

THE PATTISON PRIZE ESSAY FOR SURGERY 1987

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INTRODUCTION

"The last part of surgery, namely operations, is a reflection on the healing art; it is a tacit acknowledgement of the insufficiency of surgery. It is like an armed savage who attempts to get that by force which a civilised man would by a strategem."

John Hunter (1728 - 1793), Surgeon, St George's Hospital, London.

"To study the phenomenon of disease without books is to sail an uncharted sea while to study books without patients is not to go to sea at all."

Sir William Osler (1849 - 1919), Professor of Medicine, Oxford.

(Rains and Ritchie 1984)

Many would argue that John Hunter's words are as true today as they were in the eighteenth century. Although vast advances have been made in all areas of medicine in recent decades, it remains true to say that the aetiology and pathophysiology of many of the conditions we treat are imperfectly understood. In the face of the amazing complexity and subtlety of the human organism many of the techniques of modern medicine and surgery appear crude and simplistic. However, as in John Hunter's time, clinicians today must try to do the best for their patients with the methods available to them.

With this in mind, I hope that the six cases which follow will illustrate that, although surgery often does not influence the underlying process of disease, it can, if not always cure, at least improve the quality of life of those to whom it is offered. Indeed, in a number of situations prompt surgical intervention is life-saving.

The first group of three cases have been used as backgrounds against which to consider some current information on aspects of the aetiology, pathophysiology, diagnosis and treatment of adenocarcinoma of the rectum, inflammatory bowel disease and lesions of the cerebellopontine angle respectively. The second, shorter, group of three cases addresses more specifically some examples of the problems in diagnosis which surgeons can be faced with.

Sir William Osler's words are as applicable to surgeons as they are to physicians. The ability of the clinician to sensibly utilise the ever-growing mass of information available from books and journals for the benefit of his individual patients remains one of his most important skills.

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CASE 1 : Principal Diagnosis : Adenocarcinoma of the Rectum
Operation : Abdominoperineal Resection of Rectum and Sigmoid
Colon

Mr. W.M., a 62 year old retired engineer, was referred by his general practitioner because of bleeding per rectum. This was attributed at the time of his referral to recurring trouble with haemorrhoids from which W.M. had suffered for many years. However, when seen in the outpatient clinic it was felt that the bleeding and altered bowel habit of which W.M. was also complaining probably had a more sinister aetiology. He was therefore put on the waiting list for elective admission for further investigation and possible treatment of a rectal lesion.

At the time of admission, W.M.'s main complaint was of watery diarrhoea, occasionally containing fresh blood on the surface of the stools, which had started suddenly six weeks previously. Since then he had been having two to three episodes of diarrhoea per day and had also been complaining of tenseness and a feeling of incomplete evacuation following defaecation. He had noted over the month prior to admission that his clothing was becoming loose but did not complain of any change in appetite. W.M. also admitted to having had problems with haemorrhoids with occasional bleeding per rectum for six years. Systematic enquiry revealed nothing further of note.

In the past W.M. had had varicose veins stripped from his right leg and a right medial menisectomy. Since 1967 he had suffered from Parkinson's disease which was well controlled with Sinemet and on admission his only medication was Sinemet 100/25 II q.i.d.

W.M. was a pipe smoker, living in his own house with his wife who was largely responsible for looking after him.

On examination he was noted to have the expressionless facies, monotonous speech and poverty of movement characteristic of Parkinson's disease. Varicose veins and gravitational eczema were seen in both legs. The remainder of the cardiovascular system and the respiratory system were normal to examination.

Examination of the abdomen revealed 3rd degree haemorrhoids and on rectal examination an indurated, ulcerated lesion was felt on the anterior wall of the rectum approximately seven centimetres from the anal margin. Sigmoidoscopy confirmed this finding and a small biopsy of the lesion was taken for histology. The liver was not palpable.

Examination of the nervous system showed a pill-rolling type tremor, festinant gait and minimal cog-wheel rigidity in both arms - all consistent with W.M.'s known Parkinson's disease.

A provisional diagnosis of rectal carcinoma was made and this was confirmed as adeno-carcinoma by the results of the biopsy.

Operative Procedure

At laparotomy the presence of a bulky pelvic tumour arising from the rectum was confirmed. In addition four two centimetre metastases were felt on the liver.

After ligation of the inferior mesenteric vessels at the pelvic brim, dissection of the tumour in the pelvis was attempted. The right and left ureters were identified and followed down into the pelvis. The sigmoid colon and rectum were mobilised with relative ease posteriorly but were densely adherent anteriorly. This, combined with a narrow pelvis, made this part of the dissection difficult. Perineal dissection was then undertaken, the bleeding which arose being controlled by diathermy. The rectum and sigmoid colon were then removed and a pack soaked in Eusol placed in the perineum. The colostomy was brought out through a pre-arranged site in the left iliac fossa and the abdomen was closed in layers.

Histology

Histological examination of the specimen showed a moderately differentiated adenocarcinoma of the rectum with full thickness penetration to perirectal fatty tissue and evidence of local lymph node spread. There were

multiple polyps adjacent to the tumour.

Postoperative Progress

W.M.'s postoperative course was eventful. Three days following surgery he had an episode of sharp left-sided chest pain radiating through to the back and exacerbated by inspiration and sitting up. On examination he was not dyspnoeic and there was no tachycardia. His jugular venous pressure was however elevated. Examination of the respiratory system was normal except for diminished air entry at both bases. There was no calf tenderness, swelling or redness. A chest X-ray and electrocardiogram were carried out but neither revealed any evidence of pulmonary embolism. By the afternoon of the same day he had recovered.

The following day W.M. suffered a transient loss of consciousness just after rising from his bed. This was accompanied by a bradycardia which gradually recovered. A further ECG performed at this time was normal. A tentative diagnosis of syncope secondary to postural hypotension was made and a postural drop in blood pressure was confirmed by measurements of erect and supine blood pressure. It was felt that blood loss during the operation combined with the effects of his medication may have been sufficient to decompensate an already precarious cerebral perfusion and consequently all anti-Parkinsonian medication was stopped.

W.M. also had recurrent urinary tract problems. These started when the balloon of his Foley catheter failed to deflate post-operatively. Before W.M. could be taken to theatre to have the balloon burst, the catheter blocked completely and he was unable to pass any urine at all. Drainage was temporarily effected by passing a 10 gauge nasogastric tube alongside the catheter. The following morning the balloon of the Foley catheter was burst in theatre and the catheter removed. The next day W.M. was again unable to pass urine and had to be re-catheterised. Three days later W.M. was complaining of discharge around the catheter and culture of a urine specimen showed

significant growth of *Pseudomonas aeruginosa*. A further trial without the catheter was undertaken but W.M. was again unable to pass urine and was re-catheterised for a second time.

A urological opinion was sought at this stage and it was suggested that one final trial without the catheter should be undertaken, this time combined with distigmine bromide (Ubretid) 0.5 mg i.m. and diazepam 2 mg p.o. If voiding occurred he was to continue on Ubretid 5 mg p.o. b.i.d. In actual fact it proved necessary to re-catheterise W.M. and he was therefore prepared for a transurethral resection of the prostate which was carried out uneventfully two days later. Following removal of the catheter after this procedure W.M. was able to pass urine without any problems.

Over the same period W.M. was being instructed in stoma care. He was finding it difficult to change the colostomy bags without assistance and this was felt in large part to be due to the movement disabilities resulting from his Parkinson's disease.

The combination of W.M.'s urinary problems, his poor stoma care and difficulties stabilising his anti-Parkinsonian therapy resulted in a longer hospital stay than originally envisaged. He was eventually discharged eight weeks after his original admission. He was reviewed one month following discharge and appeared to be managing his colostomy fairly well. Both his abdominal and perineal wounds were well healed.

Commentary

The differential diagnosis of conditions presenting with rectal bleeding includes some conditions which are minor and self-limiting and other conditions which, unless recognised and treated promptly may be life threatening. A further important point to bear in mind is that the presence of one possible cause of rectal bleeding does not exclude other co-existent pathologies.

Thus it is important to make as accurate a diagnosis as possible on

of carcinoma. Thus, in this case, attention to the history and the simple procedure of digital rectal examination allowed a diagnosis to be made with a fairly high degree of certainty and emphasises the importance of rectal examination in all cases of bleeding per rectum. Seventy per cent of adenocarcinomas occur in the sigmoid colon and rectum (Cutler et al 1985a) and ninety per cent of rectal carcinomas are palpable on digital rectal examination (Browse 1978).

However, in other cases, the cause of lower gastrointestinal bleeding may not be so apparent and more specialised investigations may be required. There are two particular situations in which problems of diagnosis arise:-

- (i) The patient with faecal-occult blood positive stools but no physiologic evidence of blood loss
- (ii) The patient with "significant" blood loss per rectum.

There are numerous causes of faecal occult blood positive stools in an asymptomatic patient including haemorrhoids, diverticulitis, angiodysplasia, duodenal or gastric ulcer and carcinoma (Gregor 1971). Carcinoma is obviously the diagnosis it is most important to exclude. Standard teaching suggests that sixty per cent of colonic cancers are within reach of the sigmoidoscope. More recent data shows that only one third of colon cancers are within reach of the rigid sigmoidoscope and one third of lesions are in the caecum and ascending colon (Abrams and Reines 1979; Rhodes et al 1977). Nevertheless, rigid sigmoidoscopy remains the initial investigation of choice. If this is negative, the next step should be a high quality double contrast barium enema. If this investigation reveals no pathology and the stools are consistently faecal occult blood positive, it is important to rule out upper gastrointestinal pathology with a barium swallow and follow through. This series of investigations should diagnose the site of bleeding in ninety per cent of cases (Cutler et al 1965b). If bleeding still persists then colonoscopy should be

carried out. This procedure will reveal a carcinoma in ten per cent of such patients (Brand et al 1980).

In the second situation outlined above, with significant blood loss per rectum, the clinician must not only identify the bleeding site, but control it as well. Again an upper gastro-intestinal source of blood loss must be ruled out and this can be most effectively achieved by passing a naso-gastric tube and checking for the presence of blood in the stomach. Eighty per cent of patients with lower gastro-intestinal bleeding stop spontaneously, but if the patient requires more than 2 units of blood and becomes hypotensive, he or she should be considered for angiographic determination of the site of bleeding. This will be successful in one half to two thirds of all patients with lower gastro-intestinal bleeding and in eighty to ninety per cent of patients with continuous bleeding (Athanasoulis 1980). This procedure should be preceded by rigid or flexible sigmoidoscopy to detect a low lying source of the bleeding. The increasing use of this technique has shown that in many cases of lower gastro-intestinal haemorrhage previously attributed to diverticulitis, angiodysplasia is in fact the cause (Boley et al 1979) and thus at least partly explains the high incidence of re-bleeding following blind left hemicolectomy for lower gastro-intestinal bleeding presumed to be secondary to diverticular disease. The increased pre-operative diagnostic accuracy which angiography allows has resulted in a reduced operative mortality and morbidity (Wright et al 1980).

The treatment of rectal carcinoma is primarily surgical. Although there have been improvements in diagnosis and treatment, five year survival rates have not improved significantly over the last three or four decades (Corman et al 1979), ranging from eighty to ninety per cent five year survival for Duke's stage A tumours to twenty to twenty-five per cent in stage C lesions. This lack of progress in the treatment of colorectal carcinoma has prompted a search for a program of earlier diagnosis and screening for colorectal carcinoma. A

variety of different tests have been proposed for presymptomatic diagnosis including radiography, colonoscopy, rigid and flexible sigmoidoscopy, colonic washings and cytologic study, immunosurveillance and testing for occult blood in the stool (Cutler et al 1985a). The only test which currently shows promise is the test for occult blood and even with this test there are problems of patient compliance and false positive and false negative results (Bolt 1980).

Another important consideration in the screening of asymptomatic populations for colorectal carcinoma is the identification of high risk groups. Studies of the epidemiology of the disease have allowed identification of some such groups (reviewed by Lipkin et al 1980):-

- (i) Dietary habits appear to be of considerable importance and a high correlation has been reported between the incidence of colorectal cancer and dietary excesses of fat and red meat.
- (ii) Genetic factors are also important. Individuals with inherited polyposis coli syndrome are predisposed to the development of colorectal cancer. First degree relatives of patients with colorectal cancer have a four-fold increase in incidence.
- (iii) There is also much controversial evidence concerning whether colorectal polyps are antecedent lesions in the development of colorectal carcinoma (e.g. Turrell 1983). Fifteen to twenty per cent of adenomas are greater than two centimetres in diameter and these have a fifty per cent chance of being malignant. Villous adenomas have an incidence of carcinoma ten times greater than that of tubular adenomas. In this respect it is interesting to note that the specimen resected from W.M. contained multiple polyps adjacent to the tumour.

(iv) Patients already treated for colorectal cancer have one hundred times the risk of the general population of developing a second (metachronous) cancer.

(v) Chronic ulcerative colitis carries an increased risk of the development of colonic cancer of between four and ten per cent.

Thus, it is possible to identify a number of groups which should be carefully followed and screened because of their increased risk of developing colorectal carcinoma.

The development of surgical techniques for the treatment of colorectal cancer has led to the identification of a number of principles which help to reduce post-operative mortality and morbidity.

Bowel preparation prior to surgery is of considerable importance and good preparation significantly decreases both post-operative morbidity and mortality. Mechanical cleaning and either preoperative oral antibiotics or perioperative parenteral antibiotics have been shown to be effective in this regard (Bell et al 1983; Coppa et al 1983).

Good surgical technique depends on consideration of the modes of spread of colorectal cancer and particularly of spread via the lymphatic system which proceeds in an orderly fashion from paracolic to intermediate to primary nodes before systemic dissemination occurs. Thus surgery for colorectal cancer is, as Moynihan described in 1908, "not surgery of organs: it is the anatomy of the lymphatic system". Based on the knowledge of modes of spread of cancer of the colon and rectum, general principles of surgical management have evolved to try and prevent tumour cell dissemination. These include minimal manipulation of the tumour mass at the time of surgical exploration, ligation of vascular trunks before dissection around the tumour, ligatures above and below the tumour, discarding of gloves and instruments following tumour dissection and the covering of wound edges (reviewed by Enker and De Cosse 1987).

In 1967 Turnbull described his "no touch isolation" technique and reported impressive survival figures, especially for Duke's Stage C lesions. He attributed this to early vascular isolation of the tumour. Later studies suggested that the improved survival was more likely a consequence of the wide anastomotic resection of bowel and mesentery which Turnbull undertook (reviewed by Enker and De Cosse 1987 who suggested that a minimum margin of five centimetres of normal bowel beyond the tumour should be resected.)

Miles in 1908 was the first surgeon to report on abdominoperineal excision of the rectum with permanent colostomy as a treatment for carcinoma of the rectum, and this procedure remains the standard against which other procedures for rectal lesions are compared for mortality, morbidity and long term results. In recent years, however, considerable changes have occurred in the operations performed for carcinoma in the upper two thirds of the rectum and abdominoperineal resection has been gradually replaced by low anterior resection and various sphincter saving operations (eg Corman et al 1982). Survival rates for patients with lesions between six and eleven centimetres from the anal verge are now at least equal to those of patients treated by abdominoperineal resection. Lesions in the most distal rectum (up to five centimetres from the anal verge) are still best treated by abdominoperineal resection. The management of mid-rectal lesions remains the most controversial area. Radical pelvic dissection has not been shown to improve survival and abdominoperineal resection does not seem to benefit these patients (McDermott et al 1982). Some surgeons would argue that intestinal continuity should therefore be restored wherever possible and a wide variety of techniques have been developed to this end (Weakley 1983). Others would argue that obtaining a safe margin below the tumour is not possible in such cases (Kratzer and Alia 1980).

W.M.'s lesion fell within this controversial mid-rectal zone. Surgery in his case was not likely to be curative since there was evidence of disseminated

disease at laparotomy. It could be argued that he would have been better served by a sphincter preserving operation, particularly in view of his subsequent urinary tract complications and his problem with colostomy management. Pre-operative knowledge of the presence of liver metastases, such as could have been provided by ultrasound or radioisotope scan, might have altered management in this case, shifting the emphasis more to palliation than cure. Between fifteen and twenty per cent of resections for colorectal carcinoma are done in patients with locally advanced or disseminated disease (Cutler et al 1985a). However, it remains important to balance the mortality and morbidity of the surgical procedure itself against the potential benefit of symptomatic relief and possible increase in life expectancy when deciding whether or not to operate in cases of disseminated disease. Adverse prognostic factors in such cases include multiple liver metastases, age greater than seventy-five years and a history of cardiovascular disease (Joffe and Gordon 1981). In one series operative mortality was ten per cent and post-operative morbidity fifty per cent; the mean survival was 11.4 months (Joffe and Gordon 1967).

The major causes of death following colorectal surgery are cardiovascular complications and pulmonary embolism. W.M. had symptoms suggestive of pulmonary embolism but fortunately no objective findings to confirm the diagnosis. Urinary tract problems complicate abdominoperineal resections in twenty-seven per cent of patients as a result of trauma to the bladder and urethra and their associated blood supply (Abrams 1980). W.M. had protracted problems with his urinary tract following surgery and required prolonged catheterisation. Sepsis is a further recognised cause of post-operative complications following colorectal surgery. The incidence is higher following low anterior resection than abdominoperineal resections (Cutler et al 1985).

Systematic follow up is important in patients who have had resection of colorectal carcinoma. Eighty per cent of recurrences occur within two years. They are associated with symptoms in eighty per cent of cases, although these are frequently non-specific in nature (Hoto and Koyama 1982). Physical examination and basic laboratory and radiological tests should detect the majority of recurrences. Serial measurements of postoperative carcino-embryonic antigen levels have also proved an effective means of predicting recurrence in many patients (Cohen and Wood 1979). Early recognition of recurrence allows consideration to be given to further treatment - cure has been reported in thirty-seven per cent of patients following resection of local recurrences (Welch and Donaldson 1978). Repeat resections for rectal carcinomas usually involve extensive operative procedures such as abdominosacral resection and total pelvic exenteration.

More widely disseminated metastatic disease or recurrence requires systemic therapy. No effective chemotherapy has yet been developed. Some favourable results have been reported with radiotherapy and some studies have suggested that combining pre- and postoperative radiotherapy with standard surgical management may improve survival (Romsdahl and Withers 1978).

W.M. is known to have hepatic metastatic disease. The question arises as to whether treatment of these metastases will prolong his survival or improve his quality of life. There is now considerable interest in the resection of hepatic metastases sparked by the observations that there are some long term survivors after resection, that there are no long term survivors without resection and that survival after hepatic resection is better than that following resection of some visceral primaries such as stomach and pancreas. One fourth of patients with hepatic metastases from colorectal carcinoma have resectable lesions. Wedge and segmental resections result in five year survival rates of twenty-four per cent and twenty-one per cent respectively (Thomson 1983; Logan et al 1982). Hepatic lobectomy carries a thirteen per

cent five year survival (Adson and Van Heerden 1980). All these figures are considerable improvements on the figures for untreated hepatic metastases (median survival seventy-five days) (Jaffe et al 1968). However, it is important to note that only a limited number of patients with circumscribed intrahepatic disease and no evidence of extrahepatic metastasis are likely to benefit from such procedures.

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CASE 2: Principal Diagnosis : Ulcerative Colitis
Operation : Proctocolectomy

Ms A.B., a twenty-two year old lady was first admitted as an emergency with a thirteen day history of diarrhoea, passing liquid stools between four and seven times per day. For the six days prior to admission, she had noticed blood in her stools. At about the same time she had begun vomiting and this had been getting progressively worse until the time of admission.

For four days prior to admission, A.B. had also been complaining of abdominal pain, mainly in the left side of the abdomen and which was present constantly but exacerbated by passing motions.

A.B. admitted to a similar attack two months prior to the attack precipitating admission, lasting approximately two weeks and associated with bloody diarrhoea, which settled spontaneously.

In the three weeks prior to admission, she had lost 1 stone in weight.

A.B.'s past medical history was insignificant apart from minor manic episodes and depressive periods requiring short term treatment with lithium. There was no family history of bowel disease.

On examination, she looked tired and mildly dehydrated. There was no evidence of pallor or jaundice. Her temperature was 37.4° Celsius. Abdominal examination revealed generalised tenderness over the whole abdomen but most marked in the left flank and left iliac fossa. No organomegaly, masses or hernias were detected. Bowel sounds were present and slightly hyperactive. Digital rectal examination showed an empty rectum with some tenderness on the left side.

A provisional diagnosis of colitis was made and A.B. was started on intravenous fluids and cyclizine. Investigations on admission showed an elevated white blood cell count ($14.3 \times 10^3/\text{mm}^3$) and mild anaemia with a haemoglobin of 13.1g/dl. Her urea and electrolytes were within normal limits. Plain abdominal X-rays showed a smooth descending colon suggesting colitis and this

was confirmed at sigmoidoscopy at which time a biopsy was taken.

A.B. was treated with intravenous steroids and her condition slowly began to settle. Two weeks following admission her haemoglobin was 8.9 g/dl and she required a transfusion of three units of packed red cells. A barium enema done at this time showed ulcerative colitis affecting the transverse and descending colon and the rectum and this diagnosis was confirmed by histological examination of the biopsy specimen taken at the time of sigmoidoscopy.

In view of the acuteness and severity of A.B.'s attacks, it was felt that surgical intervention would be required. The possibility of proctocolectomy was discussed with A.B. She was allowed home at this time on high dose oral steroid therapy and coliform enemas to consider the implications of surgery and whilst at home was visited by an ileostomist who further discussed these with her.

A week after discharge, A.B. was re-admitted complaining of generalised malaise, left sided abdominal pain and diarrhoea. Her medication on admission was prednisolone 40mg per day, coliform enemas and temazepam 10mg nocte. On examination she looked flushed and toxic and had a pyrexia of 39° Celsius. Her abdomen was distended and the lower abdomen was tender with palpable colon in the right iliac fossa, left iliac fossa and suprapubically. Bowel sounds were present. On admission her haemoglobin was 11.4 g/dl and her white cell count $8.4 \times 10^3/\text{mm}^3$. Urea and electrolytes were again within normal limits. She was started on prophylactic cefuroxime and metromidazole and prepared for proctocolectomy.

Operative Procedure

At laparotomy there was obvious inflammation of the rectum, sigmoid colon, descending colon, splenic flexure and transverse colon. The small bowel appeared normal. During mobilisation of the splenic flexure an incipient perforation was found presumably accounting for A.B.'s pyrexia. Mobilisation of the descending colon was achieved uneventfully and both ureters were

identified and preserved. During perineal dissection of the rectum a hole in the anterior wall of the rectum and posterior wall of the vagina was made. This was repaired with catgut and the bowel delivered through the pelvis. The peritoneum was lavaged with three litres of saline containing 1 mg/ml tetracycline and two vaginal packs were placed in the pelvis. The abdomen was closed without drainage and the ileostomy brought out through the pre-arranged site to the right of and below the umbilicus.

Histology

This confirmed severe acute ulcerative colitis with the inflammatory reaction penetrating up to half the thickness of the muscle coats in several areas and suggesting imminent toxic megacolon.

Post-Operative Course

A.B.'s recovery was complicated initially by a low haemoglobin (7.9 g/dl) which required transfusion with three units of packed red cells. Liver function tests showed that her albumen (20 g/l) and total protein (38 g/l) were both low as a consequence of her poor nutrition and malabsorption pre-operatively. Her serum potassium levels were also found to be low on several occasions and this was attributed to a combination of her high dose steroid therapy and her chronic diarrhoea. All these factors contributed to making A.B.'s recovery rather slow. However, active physiotherapy, the discontinuing of her steroid therapy under Synacthen cover and improving appetite brought about a gradual improvement in her condition. Her perineal wound healed satisfactorily and seven weeks following her second admission, she was discharged home.

On review at the out-patient department two months later, A.B. was continuing to improve physically and at the stoma clinic she was noted to be managing well with her ileostomy.

Commentary

Together, ulcerative colitis and Crohn's disease comprise the idiopathic group of inflammatory bowel diseases. Both have an annual incidence of 5/100,000 in Western populations and are commoner in women than men. The incidence of Crohn's disease has been increasing over recent years whilst that of ulcerative colitis has stayed fairly constant (Watt and Spence 1986). Both conditions are more common in Europe and the United States than in Africa or India (Kirsner and Shorter 1980). The peak incidence of both conditions occurs in early adult life. Close relatives of patients with ulcerative colitis and Crohn's disease have an increased incidence of the disease. In addition, there is a cross-over risk, in that relatives of patients with Crohn's disease are more likely to develop ulcerative colitis and vice versa (Watt and Spence 1986). The aetiology and pathogenesis of these conditions remains incompletely understood. No infections or transmissible agent has been identified, although Strickland and McLaren (1981) showed that both ulcerative colitis and Crohn's disease tissue had a cytopathic effect on tissue culture monocell layers suggesting a viral infection. Others have been unable to replicate these findings (Philpotts et al 1979) and the evidence for a transmissible agent in inflammatory bowel disease remains controversial.

Immunological factors are also involved in the pathogenesis of inflammatory bowel disease, although it remains unclear whether they are secondary phenomena or whether there is an underlying immunological abnormality (reviewed by Rhodes 1981; Jewell 1980). One popular immunological theory of the pathogenesis of inflammatory bowel disease has been that type II hypersensitivity reactions are involved and high levels of anti-colonic epithelium antibodies have been found in both ulcerative colitis and Crohn's disease. These have been shown to cross-react with a lipopolysaccharide present in *E. Coli* and it has been suggested that inflammatory bowel disease may follow an initial immunological reaction against *E. Coli* (Kirsner and

Shorter 1980). There is also evidence that patients with inflammatory bowel disease have defects in their immune mechanisms and particularly impaired cellular immunity and abnormalities of suppressor T cells (Kirsner and Shorter 1980).

The differential diagnosis of diarrhoea and bleeding per rectum is wide. Causes of diarrhoea include bacterial and other infections (e.g. viruses, amoebiasis), metabolic and endocrine derangements (e.g. thyrotoxicosis), neuropathies (e.g. diabetic autonomic neuropathy) various forms of malabsorption and inflammatory bowel disease. Other causes of bleeding per rectum such as diverticulitis, polyps, haemorrhoids, carcinoma, radiation proctitis and trauma also need to be considered in the differential diagnosis. A large number of these possible causes can be eliminated in A.B.'s case on the basis of the history and examination.

Peete and Sabiston (1972) have reviewed the incidence of the principal symptoms of ulcerative colitis. These included diarrhoea (present in 79% of cases), abdominal pain (71%), rectal bleeding (55%), weight loss (18%), vomiting (14%) and fever (11%). It can be seen that A.B. exhibited all these common symptoms of ulcerative colitis.

Both ulcerative colitis and Crohn's disease can be associated with extra-intestinal manifestations (Watt and Spence 1986). Mild derangements of liver function tests are common. Seventy-five per cent of patients with sclerosing cholangitis have underlying ulcerative colitis and cirrhosis develops in between one and five per cent of ulcerative colitis patients. Erythema nodosum and pyoderma gangrenosum are both skin manifestations which are seen more commonly in ulcerative colitis than Crohn's disease. Episcleritis and uveitis are signs occurring more commonly in Crohn's disease. A polyarthrititis may complicate both Crohn's disease and ulcerative colitis and is more common during exacerbations of the disease. Between two and six per cent of cases of

both ulcerative colitis and Crohn's disease are associated with ankylosing spondylitis. Urinary calculi are also more common in patients with inflammatory bowel disease, occurring in up to ten per cent of patients.

Rains and Ritchie (1984) have divided the clinical features of the disease into two categories, associated with either a fulminating or a chronic type of presentation. The fulminating presentation is associated with pyrexia and incessant diarrhoea containing blood, mucus and pus; the patient looking and feeling very ill. There may be abdominal distension due to toxic dilatation of the colon. This condition should be distinguished from dysentery and typhoid. The chronic type of presentation, which is much more common, is less severe with exacerbation occurring at intervals ranging from weeks to months. With progression the patient becomes wasted and severely anaemic. During exacerbations there may be ten to twenty stools per day, often associated with tenesmus. A.B.'s presentation was obviously much closer to the fulminant than the chronic type. Her temperature was only slightly elevated on her first admission, but on her second admission, she had a considerable pyrexia and abdominal distension. The specimen removed at laparotomy was suggestive of imminent toxic megacolon which is a complication in two to four per cent of all ulcerative colitis patients and represents a surgical emergency when it occurs.

Investigation of a patient with suspected inflammatory bowel disease is directed at confirming the diagnosis and defining the extent of the disease. If amoebiasis or typhoid is suspected, stool cultures should be taken. In a fulminant type presentation plain abdominal X-ray may show free air under the diaphragm if perforation has occurred and may reveal characteristic gross dilatation of the transverse colon in cases of toxic megacolon.

Barium studies are useful in defining the extent of the disease and distinguishing between ulcerative colitis and Crohn's disease. They are contra-indicated in suspected cases of toxic megacolon. The earliest sign visible on barium enema in cases of ulcerative colitis is loss of haustral

markings, especially in the distal colon. Established disease produces a narrow, contracted colon, alteration in the mucosal outline and pseudopolyposis in fifteen per cent of cases. In A.B.'s case there was loss of haustral markings and narrowing of the transverse and descending colon. No pseudopolyps were seen. In contrast Crohn's disease is segmented with irregular involvement and frequent fistula formation (Peete and Sabiston 1972).

Proctosigmoidoscopy is a particularly useful investigation in ulcerative colitis since ninety to ninety-five per cent of cases involve the distal colon (Rains and Ritchie 1984). This will reveal an erythematous granular mucosa with contact bleeding and allows one or more mucosal biopsies to be taken. Characteristically the abnormal mucosa affects the rectum and extends proximally in a continuous fashion involving the entire circumference of the lumen.

Histological examination of biopsy specimens is the most reliable way of making a diagnosis of ulcerative colitis and distinguishing it from Crohn's disease. Characteristic histological findings in an acute attack of ulcerative colitis are an inflammatory reaction limited to the mucosa and superficial submucosa with a chronic inflammatory infiltrate throughout the lamina propria. Polymorphs invade the bases of glandular crypts to form crypt abscesses. Rupture of these abscesses may lead to mucosal ulceration. Mucosa at the margin of these ulcerated lesions is undermined and raised above the surrounding tissue to form inflammatory polyps (for a further description and comparison with the histological changes seen in Crohn's disease see Watt and Spence 1986).

The most difficult step in making a diagnosis of ulcerative colitis is often in distinguishing it from granulomatous disease of the colon. Meyer and Sleisenger (1973) conducted a study which examined the incidence of various clinical features in these two conditions. They found that pseudopolyposis,

disease limited to the recto-sigmoid area of bowel and free colonic perforation were pathognomonic for ulcerative colitis whereas associated small bowel disease, skip areas of colitis and enteric fistulae were pathognomonic for granulomatous colitis. Other features such as the passage of bloody diarrhoea, rectal involvement, perianal disease, colonic stricture, toxic megacolon and colonic carcinoma were found to be less effective discriminants.

One third of patients with ulcerative colitis eventually come to surgery compared to seventy-five to ninety per cent of those with Crohn's disease (Cutler et al 1985). Removal of the rectum and colon in a patient with ulcerative colitis will effect a cure and diminish extraintestinal manifestations of the disease. In contrast, recurrence is likely to follow resection for Crohn's disease and therefore surgery is more conservative in these cases.

The first person to perform a documented surgical operation for the treatment of ulcerative colitis was Pennel in 1850 who carried out a sigmoid colostomy for the condition (see Goligher et al (1968) for a review of the historical background to the treatment of this condition). By 1913 a completely diverting ileostomy with piecemeal resection of the colon over the ensuing years had become popular. During the late 50s and early 60s double and single stage operations became more popular and in 1959 Miller became the first surgeon to perform a single stage ileostomy and colectomy.

Currently the operation of choice is proctocolectomy with ileostomy, although some would advocate colectomy and ileo-rectal anastomosis in those cases without rectal involvement (Rains and Ritchie 1984; Binder et al 1975; Cutler et al 1985). Ileostomy alone is now only considered for gravely ill patients with fulminant disease (Rains and Ritchie 1984).

Urgent surgical intervention is indicated for the various complications which may arise from ulcerative colitis. Massive haemorrhage, toxic megacolon with impending or frank perforation, fulminant ulcerative colitis unresponsive

to steroid therapy and obstruction secondary to stricture formation all fall into this category (Peete and Sabiston 1972; Binder et al 1975). To these urgent indications, many surgeons would also add the suspicion or demonstration of colonic carcinoma and, in a younger person, failure to grow and develop normally. There is a time-related incidence of colonic carcinoma in patients with ulcerative colitis which increases at a rate of two per cent per annum after ten years (Cutler et al 1985) and patients with pancolitis of more than ten years' standing should have periodic colonoscopy and biopsy. The incidence of surgical intervention in less acute situations appears to relate to the availability and policy of surgeons in different areas ranging from ten to fifty per cent in different studies (Goligher et al 1968; Gilat et al 1976). Obviously A.B. with her acute presentation and fulminant disease course fell into the category of patients requiring urgent operation.

Surgery on patients with acute presentations of ulcerative colitis or complications arising from the disease may pose further problems. Severe, prolonged diarrhoea may upset fluid and electrolyte balance and haemorrhage may result in anaemia. Localised or systemic sepsis may further complicate management of such patients. These factors help to explain the higher mortality associated with emergency surgery (10-15%) compared to elective operations (3%) (Moody 1977). A.B. was anaemic on her second admission and required transfusion post-operatively. She was also pyrexial and toxic and this was attributed to the large colic abscess found at laparotomy.

The effects of long term high dose steroid therapy must also be considered by the surgeon. Of particular relevance is the increased risk of massive haemorrhage and free perforation as well as the increased susceptibility to pyogenic infection. Steroids may also encourage disintegration of the colon wall and encourage formation of adhesions with parietes and adjacent viscera making the procedure technically more difficult (Rains and Ritchie 1984). In

A.B.'s case the hypokalaemia and proximal myopathy which prolonged her post-operative recovery were thought to have been related at least in part to her steroid therapy.

According to Binder et al (1975) the operation of choice in urgent or emergency situation is a one stage proctocolectomy and ileostomy. They reported a series of eighty operations one stage total proctocolectomy being performed in 37 patients with a 9.1% mortality and ileostomy with sub-total colectomy being performed in 43 with a 7.5% mortality. Post-operative morbidity after total proctocolectomy did not differ substantially from that after subtotal colectomy. Seventy-five per cent of patients surviving subtotal colectomy required subsequent abdominoperineal resection of the rectal remnant and no patient had successful subsequent ileorectal anastomosis. These authors therefore recommended that a one stage proctocolectomy should be adopted as the surgical procedure of choice in emergency or urgent operations for ulcerative colitis.

The attraction of subtotal colectomy in patients without rectal involvement is obviously the possibility of performing an ileo-rectal anastomosis and avoiding the need for an ileostomy (Farnell et al 1980). However, the high incidence of disease recurrence and carcinoma in the rectal stump means that between twenty and eighty per cent of patients eventually require proctectomy (Cutler et al 1985; Binder et al 1976). Consequently much current interest is centred on the effectiveness of alternatives to conventional ileostomy such as the continent ileostomy (Koch pouch) and ileoanal reservoir (Parks pouch) (Cranley 1983; Goligher 1980; Rotherberger 1987) and these may represent a psychologically and socially more acceptable alternative for these patients in the future.

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CASE 3: Principal Diagnosis : Acoustic Neuroma
Operation : Right Suboccipital Craniectomy for excision of
acoustic neuroma

Mrs A.A., a 59 year old factory worker, was admitted with a ten to fifteen year history of progressive deafness in the right ear, associated more recently with dryness of the right eye and mild ataxia. She had attributed her progressive hearing loss to working in a noisy factory. A routine hearing test carried out at her work had shown that there was no hearing in her right ear at all. A computed tomography scan carried out subsequently showed an enhancing three by three by two centimetre mass in the posterior fossa extending into the right internal auditory canal and causing displacement of the fourth ventricle and early hydrocephalus.

On admission she was otherwise well. There was a past medical history of cholecystectomy in 1965 and hysterectomy in 1982. She was on no medication at the time of admission.

Family history, social history and systematic enquiry revealed nothing of note.

Examination showed an obese woman who was noted to have a large diffuse bilateral goitre. This had been investigated on a prior admission and classified as benign. She was also found to be hypertensive with a blood pressure of 200/110 millimetres of mercury. The remainder of the cardiovascular system, the respiratory system and the gastrointestinal system were normal to examination.

Testing of the cranial nerves showed bilateral horizontal nystagmus on lateral gaze. Rinne's test showed bone conduction to be better than air conduction on the right and Weber's test lateralised to the left. There was no objective evidence of a facial palsy other than reduced tear production in the right eye. An increased jaw-jerk and hyper-reflexic gag reflex were also noted. Further tests of cerebellar function revealed minimal dysdiadochokinesis and minimal gait ataxia. The remainder of the nervous

system was normal to examination.

X-rays of the chest and neck at this time showed evidence of a large soft tissue swelling in the region of the thyroid gland with some associated constriction of the trachea. Consequently pulmonary function tests were carried out. These were found to be within normal limits. Routine blood investigations including thyroid function tests were all found to be normal.

Operative Procedure

A right sub-occipital craniectomy was performed during which venous haemorrhage from a large venous sinus was encountered. This was controlled by tamponade. A "Y" shaped deural incision revealed a somewhat distended cerebellar hemisphere. The cisterna magna was opened releasing a considerable amount of cerebrospinal fluid and allowing retraction of the right cerebellar hemisphere to expose the tumour in the cerebello-pontine angle. Frozen section histology of a biopsy of the tumour confirmed the presence of an acoustic neuroma. The tumour was internally decompressed using an ultrasonic probe. This was accompanied by some bleeding which was controlled by intermittent tamponade.

Dissection of the tumour capsule from the cerebellar hemisphere and brainstem proved difficult because of a densely adherent arachnoid cap surrounding the tumour.

Drilling off of the lateral aspect of the internal auditory meatus exposed the intracanalicular portion of the tumour which was dissected free of the facial nerve. The facial nerve was then followed towards the brainstem until it was obscured by thickened arachnoid. The inferior pole of the tumour was noted to be separate from cranial nerves IX, X and XI.

The tumour extended superiorly and anteriorly towards the tentorial hiatus where it indented the antero-lateral aspect of the pons. This part of the tumour was mobilised and removed.

At this stage it was felt that the majority of the tumour had been removed although a thick, scarred layer of arachnoid remained adherent to the brainstem and lateral cerebellum. It was thought that dissection of this layer was likely to result in considerable damage to the underlying brain and consequently, once haemostasis had been secured, the dura, galea, sub-occipital musculature and skin were closed in layers.

Histology

Microscopical examination of the tumour removed at operation showed a vascular tumour whose appearance was consistent with that of an acoustic schwannoma showing both Antoni A and Antoni B types of tissue.

Post-operative Progress

Mrs A.A. made a very satisfactory and uneventful post-operative recovery, awaking quickly and with a full level of consciousness. Bilateral horizontal nystagmus as on lateral gaze and a mild right-sided upper motor neurom facial palsy were noted. The facial palsy did not progress.

One week following the operation Mrs A.A. was discharged on a reducing dose of steroids, to be followed up after six weeks in the out-patient department.

Commentary

Acoustic neuroma was first described as a post-mortem finding by Sandifort in 1777 (Hoogland 1984). These tumours are derived from Schwann cells forming the nerve sheath of the VIIIth nerve. They characteristically arise from the vestibular component of the nerve at the neuroglial-neuroleminal junction just inside the internal auditory meatus (Nager 1985). They comprise between seven and ten per cent of all intracranial neoplasms and seventy-five per cent of space occupying lesions within the cerebello-pontine angle (Gonzales-Resilla 1947). Four per cent of acoustic tumours are bilateral. Most of these cases are an expression of the central form of von Recklinghausen's neurofibromatosis. Acoustic neuromas occur most frequently in the middle

decades of life with a peak age incidence between thirty-five and forty years (Nager 1985) and are found twice as commonly in females as in males. Although A.A. was older than the age of peak incidence when she presented, it is worth noting that her history of progressive deafness went back over a period of ten to fifteen years.

Because of their origin within the internal auditory canal, acoustic neuromas can be considered as having two components - an intracanalicular portion and an extratemporal portion in the cerebello-pontine angle (Nagler 1985). Acoustic neuromas are extremely slow growing tumours and it has been suggested that some tumours may cease to grow entirely and remain asymptomatic for long periods of time (Olivecrona 1950). Their slow growth is an important factor in the development of the clinical manifestations of these tumours.

Macroscopically they are usually well demarcated encapsulated neoplasms of pale colour and firm consistency and on larger tumours arachnoid cysts are frequently found around the caudal pole. Histologically the tumours consist of compact groups of elongated spindle-shaped cells with indistinct cell membranes (Nager 1985). On the basis of the spatial arrangement and configuration of the tumour cells, two types of tissue can be distinguished - the dense cellular fibrillary structure of Antoni type A and the loose less cellular reticular structure of Antoni type B (Nager 1985). It has been suggested that the type B tissue may represent a degenerate form of type A (Zulch 1957). Histological examination of A.A.'s tumour showed a mixture of both types of tissue.

As mentioned above, the slow insidious development of the clinical manifestations of this tumour can be attributed to its slow growth. The clinical signs and symptoms themselves can be related to the location of the tumour in the cerebello-pontine angle. In a study in 1984, Verhagen and Schulte found that the time from the appearance of the first symptom to diagnosis was between six months and twenty years, the widest range and longest

time period of any of the space occupying lesions of the cerebello-pontine angle which they considered. A.A. had had symptoms of progressive hearing loss for fifteen years at the time of diagnosis. According to Verhagen and Schulte (1984) the commonest first symptom in the history was hearing loss, with tinnitus, dizziness, facial numbness or headache being much less common initial symptoms. The commonest predominant symptom at admission was also hearing loss but with dizziness, headache and tinnitus being relatively much more common.

The commonest sign found on clinical examination was a hearing deficit (Verhagen and Schulte 1984) - most sensitively detected by audiometry - and a routine hearing test provided the information which allowed the diagnosis to be made in A.A.'s case. Ataxia was a further common sign with trigeminal nerve impairment (especially a diminished corneal reflex), nystagmus, facial nerve impairment, pyramidal tract lesions, optic nerve impairment and chronic hydrocephalus being further less common findings on clinical examination. Obviously the particular constellation of clinical signs seen will depend on both the size and location of the tumour although the only sign which Verhager and Schulte found to be strongly associated with tumour size was nystagmus. Fisch (1978) divided acoustic nerve tumours into three groups according to their size (groups A and B with a maximal diameter from the fundus of the internal auditory meatus of less than twenty-five millimetres and group C with a diameter of greater than twenty-five millimetres). Groups A and B present with otological symptoms related to the acoustic nerve only; whereas patients with tumours of Group C have neurological symptoms attributable to the trigeminal or facial nerves, the brainstem or the cerebellum. According to these clinical criteria, A.A.'s tumour fell into category C.

Having made a provisional diagnosis on clinical grounds it remains to confirm the diagnosis and determine as accurately as possible the size and extent of the tumour. Seventy-five per cent of neuromas cause a considerable increase in cerebrospinal fluid protein (to levels in excess of 400 mg/100 ml)

and this can help to distinguish them from other space occupying pathology in the cerebello-pontine angle (Jennett and Galbraith 1983). Further investigation of these tumours depends on various imaging procedures and has been reviewed by Thijssen and Merx (1984). According to these authors the radiological diagnosis of cerebello-pontine angle pathology can be divided into three stages - imaging of the configuration of the internal auditory meatus, imaging of the contents of the cerebello-pontine angle cistern and internal auditory meatus and judgement of the exact location of large tumours in relation to the brainstem and cerebellum. All of these objectives can be achieved using computed tomography scanning with software for high resolution and reconstruction zoom magnification with or without contrast enhancement and cisternography or equally accurately using more traditional but also more invasive techniques (Thijssen and Merx 1984). In the case presented above the diagnosis was made using CT scanning with contrast enhancement and confirmed at operation by frozen section histology. The other common pathologies which have to be considered in the differential diagnosis of space occupying lesions in the cerebello-pontine angle are meningiomas (six to seven per cent of cerebello-pontine angle tumours), epidermoids (6.7%) and gliomas (6%) (Nager 1985).

Distinction between these pathologies can usually be made on the basis of cerebrospinal fluid and radiographic criteria, but in cases of doubt frozen section histology at the time of surgery allows a definitive diagnosis to be made.

The management of this condition is surgical. The first sub-occipital removal of an acoustic neuroma was performed by Balance in 1894. However the high (80%) operative mortality of this approach prompted Quix to attempt traslabrynthine removal of the tumours in 1911 and Cushing to advocate intracapsular subtotal removal in 1918. Dandy in the 1920's returned to the

original aim of total tumour removal and in the 1960's Hanse and Doyle combined the skills of neurosurgeon and ENT specialist to further improve the results of operative treatment (Hoogland 1984). There is still discussion concerning the relative merits of the total and intracapsular subtotal removals, but continuing improvements in neurosurgical technique and patient monitoring make total removal of the tumour a more attractive approach (Jennett and Galbraith 1983; Harner and Ebersold 1985). The most common complications to follow a total tumour removal are facial palsy, neuroparalytic keratitis, temporary bulbar palsy, ataxia, cerebrospinal fluid otorrhoea, meningitis and haemorrhage (Jennett and Galbraith 1983; Harner and Ebersold 1985). The advantage of the subtotal intracapsular removal is the avoidance of most of these complications, but at the risk of tumour recurrence at a later date and this operation is only recommended in the elderly or those with intercurrent disease. A second operation carries a higher risk than a total initial removal (Jennett and Galbraith 1983). In a recent study (Harner and Ebersold 1985) employing a retrosigmoid suboccipital craniectomy and the combined expertise of neurosurgeon and otological surgeon, total tumour removal was accomplished in ninety-eight per cent of cases. Facial nerve function was preserved in eighty-one per cent of cases with the aid of peri-operative nerve function monitoring. The most common complication occurring in this study was CSF otorrhoea which was present in twelve per cent of the patients of whom approximately half required a subsequent procedure. In one hundred and sixty-two cases considered there were two recurrences in a follow-up period ranging from three months to five years.

Thus, it would appear that even though A.A.'s operation was technically difficult because of the thickened adherent capsule of the tumour, the prospects of a cure or at least a long term remission are very good.

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CASE 4 : Principal Diagnosis : Rectus Sheath Haematoma
Operation : Appendectomy

Mrs D.W., an eighty-three year old nursing home resident was admitted as an emergency with abdominal pain. On admission her mental status questionnaire score was five out of ten and she was a poor historian. She was complaining of lower abdominal pain mainly on the right. She was unable to accurately describe its nature, duration or mode of onset. There appeared to be no radiation. She did not admit to any vomiting, change in bowel habit or other gastrointestinal system related symptoms. According to the letter which accompanied her to hospital the pain had come on gradually over a period of three to four hours that evening.

Mrs D.W.'s past medical history was unremarkable apart from a cholecystectomy in 1960. There was no family history of note.

On admission her only medication was temazepam 10 mg nocte. She had no known allergies.

D.W. was a widowed lady living in Part IV accommodation. She had no living relatives other than a daughter now residing in England. She was a non-smoker and did not take alcohol. Systematic enquiry was unrevealing.

On examination she was noted to be an obese lady in some distress. She was disorientated in time and place. At the time of admission she was pale and restless. She was afebrile but had a tachycardia of 115 beats per minute and a blood pressure of 140/80 millimetres of mercury. There was no jaundice. Examination of her abdomen was made difficult by her obesity. There were no visible signs of abdominal pathology. An old cholecystectomy scar was noted. No masses, hernias or organomegaly could be detected on palpation. There was tenderness to palpation in the right lower quadrant with moderate guarding. There was minimal rebound tenderness. Bowel sounds were present and normal in character. Digital rectal examination revealed some generalised tenderness and a rectum containing a few soft faeces. These were faecal occult blood

negative. The remainder of the physical examination revealed no other abnormalities.

A full blood count, urea and electrolytes, serum amylase, a chest X-ray, erect and supine abdominal x-rays, an electrocardiogram and urinalysis were ordered. Her urea, electrolytes and amylase were all normal as were the results of her full blood count although the white cell count was at the upper limit of normal. Her abdominal X-rays and urinalysis were also normal. The chest X-ray and electrocardiogram were within normal limits.

A provisional diagnosis of appendicitis was made on the basis of the clinical findings and D.W. was prepared for laparotomy. Division of the rectus muscles revealed a large haematoma within the rectus sheath. The appendix and associated bowel appeared normal. The appendix was removed to prevent possible confusion at a later date. The haematoma was evacuated, haemostasis secured and the abdomen closed.

Mrs D.W. made an uneventful recovery. Post-operatively she remained rather confused. One week following admission she was discharged back to her nursing home.

Commentary

Rectus sheath haematomas are uncommon and only of interest to the surgeon in that they may mimic acute intra-abdominal disease. The haematoma is situated beneath the navel in ninety per cent of cases and lies to the right of the midline twice as often as it does to the left (Cullen 1937). Their frequent occurrence below the semicircular line of Douglas means that these haematomas can cause peritoneal irritation and hence mimic intra-abdominal disease (Sabiston 1977). Haemorrhage usually occurs from the inferior epigastric vessels and is more commonly from the vein than the artery.

Aetiologically these haematomas may occur as a result of trauma, infectious and debilitating disease (often precipitated by a bout of coughing in such cases), degenerative vascular disease, blood dyscrasias and

coagulopathies, postoperatively as a result of trauma to the rectum muscle during surgery or without any obvious precipitating cause. In D.W.'s case there was no evidence of widespread disease, cardiovascular problems or trauma, although in view of her poor historical ability a fall prior to the onset of the pain could not be ruled out.

The most pertinent question in this case is whether the diagnosis of rectus sheath haematoma could have been made without resorting to laparotomy with the associated risks this carries for an elderly lady. The symptomatology associated with a rectus sheath haematoma is usually characteristic with a sudden onset of abdominal pain localised to the area of rupture which after a period of increasing severity, slowly improves. Nausea and vomiting are relatively common. On examination a hard tender mass may be palpable in one or other rectus (Murray and Burger 1954). Fothergill (1926) has described a test which allows such masses within the recti to be distinguished from intraperitoneal masses. When the recti are contracted a mass in the wall may still be palpated and is fixed whereas an intraperitoneal mass becomes harder to palpate. Contraction of the recti may also exacerbate the abdominal tenderness. An additional late sign is the appearance of echymosis around the umbilicus.

In the case considered here D.W. was unable to give a detailed history of the onset and progression of her pain. Although there was tenderness and guarding in the right iliac fossa there was no palpable mass and no visible echymosis. Thus the only aspect of physical examination which might have been revealing and which wasn't carried out was getting D.W. to contract her recti and determine if this exacerbated the pain.

In the circumstances therefore the provisional diagnosis of appendicitis and the decision to proceed to laparotomy were reasonable. The differential diagnosis of pain in the right iliac fossa is wide, especially in women, and

includes appendicitis, ovarian cyst, mesenteric lymphadenitis, mittelschmerz, pelvic inflammatory disease, gastroenteritis, obstruction, pyelonephritis and caecal diverticulitis (Cutler 1985). In D.W.'s case a number of these causes can be ruled out in view of her age and loss of reproductive capacity. Her normal urinalysis and lack of fever argued against pathology in the renal tract. Potentially the most serious cause of her pain was acute appendicitis. The presentation of this condition in the elderly is often late and atypical and the threshold for operative intervention correspondingly lower than in younger patients. Perforation occurs in seventy-five per cent of elderly patients and death occurs in as many as twenty-four per cent of patients over seventy-five (Owens and Hamit 1978). The diagnosis of acute appendicitis is recognised as being harder to make in the elderly since twenty to twenty-five per cent do not have an elevation in temperature or white cell count (Lewis et al 1975). Consequently a more aggressive approach to abdominal pain is justified in this group.

It seemed therefore that laparotomy was the correct course to follow in D.W.'s case. However the question arises as to whether, once the diagnosis of rectus sheath haematoma has been made and a normal appendix seen, an appendectomy should have been performed. Some would argue that such an operation prevents possible future problems and avoids confusion when the abdomen is examined at a later date by other practitioners. The alternative argument (Nockerts et al 1980) suggests that the increased morbidity and mortality associated with the removal of a normal appendix does not justify routine incidental appendectomy in the elderly.

Thus, it is apparent that clinical features and investigations do not always allow an accurate diagnosis and frequently this must await surgery itself and the pathologist's report. It remains important, however, to balance the risks of surgical procedures against the potential risks and complications of inactivity in each case in order to determine the best course of action from

the patient's point of view.

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CASE 5 : Principal Diagnosis : Colloid cyst of thyroid gland
Operation : Excision of colloid cyst of right lobe of thyroid gland

Mrs P.M., a 48 year old medical receptionist, came in as an elective admission for a right partial thyroidectomy. She had first noticed a painless lump in her neck six months prior to admission and since that time it had been slowly enlarging. She had noted no change in her voice or dysphagia during that time and she complained of no symptoms of thyroid hyper- or hypofunction. There was no past history of thyroid disease or neck irradiation. Her mother had had a similar complaint thirty years previously which had not been treated and she had recently been found to be thyrotoxic.

P.M.'s past medical history consisted of a right oophorectomy for an ovarian cyst in 1976 and a total abdominal hysterectomy for menorrhagia in 1984.

Her only medication on admission was Gaviscon p.r.n. for a longstanding complaint of heartburn. Previous investigation had failed to demonstrate any evidence of hiatus hernia. There were no known allergies.

P.M. was an ex-smoker and non-drinker. Systematic enquiry was unremarkable.

On examination there was an obvious goitre to the right of the midline four centimetres in diameter and moving with swallowing. The goitre was soft and smooth to palpation. No fluctuation could be elicited. The goitre was not fixed to surrounding tissues and no cervical lymphadenopathy was present. Her pulse was eighty beats per minute and regular. There was no exophthalmos, lid lag or chemosis. There was no abnormal tremor or sweating. P.M.'s voice was normal and there was no evidence of myxoedematous faices. Tendon reflexes were symmetrical and normal. The remainder of the physical examination was normal.

An ENT consultation prior to surgery confirmed that P.M.'s vocal cords were functioning normally. Preoperative full blood count, urea and



electrolytes, chest X-ray and electrocardiogram were all normal. Thyroid function was also within normal limits.

Operative Procedure

The thyroid gland was approached through a collar incision. The left lobe of the thyroid appeared normal. There was a large lump in the right lobe of the gland which was demonstrated after the strap muscles were divided when it also became apparent that the lump was a simple cyst. The cyst was therefore excised without further dissection. Haemostasis was secured with catgut. The strap muscles were sutured and opposed to the midline with catgut and the skin closed with clips after a redivac drain had been inserted.

Histology

Histological examination of the specimen confirmed the presence of a three centimetre colloid cyst with a well-defined fibrous capsule.

Postoperative Course

P.M. made an uneventful recovery from the operation and was discharged home two days following surgery. A further ENT consultation confirmed satisfactory post-operative function of the vocal cords.

Commentary

Although this was a simple colloid cyst, the case provides a background against which to consider the differential diagnosis and investigation of solitary thyroid swellings.

The clinical importance of thyroid nodules lies in the fact that their differential diagnosis includes thyroid carcinoma. Determining the best course to follow in the diagnosis and treatment of solitary thyroid nodules is made difficult by confusion over classification and the fact that the low incidence of thyroid cancer and the small death rate limit the size of studies and require long term follow-up.

The incidence of clinical apparent solitary thyroid nodules is quite high at around seven per cent (Ashcraft and Van Herle 1981). The incidence of

thyroid carcinoma on the other hand is low at 4/100,000 population per year (Ashcraft and Van Herle 1981). However some studies have suggested that the incidence of carcinoma in a solitary nodule may be as high as forty per cent (Katz and Zager 1976).

The most important criterion determining whether or not to remove solitary thyroid nodules must therefore be the likelihood that they are malignant and a number of investigations are available to the clinician to help him select patients for surgery. However, there are also a number of other indications for surgery. The pressure effects of large nodules may impair breathing and/or swallowing. Excision may also be undertaken for primarily cosmetic reasons - as in this case.

The differential diagnosis of a solitary thyroid nodule includes the following - adenomatous goitre, adenoma, chronic thyroiditis, toxic adenoma, well differentiated carcinoma and thyroid cyst (Cutler et al 1985). The first clues to the nature of a solitary thyroid nodule come from the history and physical examination and some authors would argue that clinical features alone are the most reliable indications for operation (Hoffman et al 1972).

The length of time a nodule has been present and the presence or absence of pain are important features. A family history of multinodular goitre increases the chance of a nodule being non-malignant whilst a family history of medullary carcinoma or multiple endocrine tumours is an indication for early surgical excision (Hoffman et al 1972). A history of radiation to the neck increases the likelihood of carcinoma and in such cases the chance that a solitary hypofunctioning thyroid nodule is cancer is sixty per cent (Cutler et al 1985). Age is a further factor to be borne in mind - a nodule in a child or young adult is more likely to be malignant (Rallison et al 1975). The physical features of a nodule also have a bearing on the likelihood of malignancy. Thus a stony, hard, solitary mass fixed to surrounding structures

and associated with hoarseness or dysphagia is almost certainly malignant (Ashcraft and Van Herle 1981).

Various tests and investigations are in use to try and further evaluate solitary thyroid swellings, each with its own advantages and disadvantages. Most patients with solitary nodules are euthyroid but occasionally autonomous nodules may result in elevated T_3 and T_4 levels. Serum calcitonin levels are raised in patients with medullary carcinoma of the thyroid and serum thyroglobulin levels are usually elevated in differentiated thyroid carcinoma and in some cases of thyroiditis (Ashcraft and Van Herle 1981).

The thyroid scintiscan is widely used in the evaluation of solitary nodules (Ashcraft and Van Herle 1981b). Nodules are classified as "hot" if they take up more radionuclide than the surrounding thyroid tissue and "cold" if they take up less than the surrounding tissue or none at all. The incidence of cancer in a hot nodule is reported to be less than one per cent, although more recent reports suggest that it may be greater and that the incidence of carcinoma in a cold nodule may be less than originally accepted (O'Holleran et al 1982). In addition there are reports of a twenty-five per cent incidence of cancer in patients with normal radioiodine scans (O'Holleran et al 1982). The technical limitations of the method must also be considered. In general larger nodules near the periphery of the gland are most easily visualised and identified. Small nodules in the isthmus may not be visualised at all. A number of different radioisotopes have been used in an attempt to decrease the radiation employed and increase the specificity of the investigation (Saxe 1979). Nevertheless the technique remains limited and Hoffman (1972) has reported that if hypofunction on scintiscan were the sole criterion for operative selection, forty per cent of carcinomas in his series would have been missed.

If a nodule is found to be cold or non-functioning on scintiscan, then ultrasound can effectively differentiate solid from cystic lesions (Lees et al

1978). However this technique also has limitations as a means of selecting cases for operation. Lesions less than one centimetre in diameter cannot be resolved and although completely cystic lesions can be differentiated from mixed and solid lesions, the distinction between benign and malignant solid lesions cannot be made. Beta mode ultrasound is reported to be ninety per cent effective in differentiating cystic from solid lesions (Miskin et al 1973), but it is worth noting that the cystic nature of a lesion does not rule out carcinoma (Hammer et al 1982).

In nodules shown to be solid, coarse needle biopsy is a further technique available for evaluating thyroid nodules and is reported to have a diagnostic accuracy of over ninety per cent. However, in small lesions the sampling error is significant and reported complications include bleeding and pneumothorax (Karlán et al 1983). Malignant seeding of the needle track, however, would appear not to be a valid concern (Wang et al 1976). In an attempt to decrease limitations, eliminate complications and increase accuracy, fine needle aspiration cytology may be employed. Accuracy is reported between ninety and ninety-five per cent and the sampling error in small lesions is eliminated (Ashcraft and Van Herle 1981b). However, sophisticated cytotechnology equipment and an experienced cytologist are prime requisites for effective use of this diagnostic technique.

Lesions which appear cold on scintiscan and are shown to be solid on ultrasound and benign on biopsy may be treated by thyroid suppression. An increase in size or failure to regress is an indication for surgical intervention (Blum and Rothschild 1980). Lesions shown to be cystic can be aspirated. Malignant cells, bloody or cloudy fluid and cyst recurrence would be indications for surgery.

The systematic application of the investigations described above, with appropriate consideration of their limitations, according to an established

algorithm (eg Thomas 1981) should allow most cases of malignant disease to be diagnosed and many unnecessary operations avoided. Nevertheless, none of these investigations, either alone or in combination, are infallible and some authors argue that clinical factors alone offer the best guide to the need for surgical intervention (Hoffman et al 1972). If so, then additional investigations themselves become superfluous and in the case of invasive procedures actually add to the morbidity associated with the condition. In addition there are indications for surgical intervention other than the possibility of malignancy including the cosmetic and pressure effects of the goitre. If one of these other indications for surgery is present, then investigation aimed at determining if the nodule is malignant again become unnecessary.

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CASE 6 : Principal Diagnosis : Left testicular torsion
Operation : Exploration of left and right testis
Correction of torsion

Mr G.T., a 15 year old schoolboy, was admitted as an emergency with severe left-sided lower abdominal pain. The pain had come on quite suddenly at 0200 that morning, waking G.T. from sleep. The pain was severe, fairly constant in nature and with no radiation. The pain was made worse by movement and relieved by lying still. There was no relation to coughing. Although nauseated, G.T. had not vomited. There had been no change in bowel habit. There were no other abdominal symptoms and no history of trauma. There had been no dysuria, frequency, haematuria or discoloration of the urine. G.T. had not complained of any fever or rigors.

Three months prior to this admission, G.T. had had a similar episode of left abdominal pain for which he had been admitted to the Emergency Department. On that occasion it was felt most likely that the pain was of renal origin. However, urinalysis and an intravenous pyelogram at that time were both normal. The pain settled spontaneously and G.T. was discharged home.

There was no past medical history or family history of note. G.T. was a non-smoker and did not take alcohol. Systematic enquiry was unremarkable.

On examination G.T. was in some distress, lying still on his bed. He was afebrile with a pulse of ninety beats per minute and a blood pressure of 120/70 millimetres of mercury. On abdominal examination a two centimetre spherical swelling was noted four centimetres left of the pubic symphysis. This was extremely tender, firm and not fixed to the overlying skin. There was no cough impulse and the swelling was not reducible. No bowel sounds were audible in the swelling. The scrotal sac itself was non-tender and not inflamed. A normal testis was palpable on the right side. The remainder of the abdominal examination was normal. There were no other masses or areas of tenderness and no organomegaly. Bowel sounds were normal and rectal

examination was unremarkable.

The remainder of the physical examination was also normal. Urinalysis and a full blood count revealed no abnormalities.

Based on the location of the swelling, a provisional diagnosis of inguinal hernia was made despite the absence of a cough impulse and the irreducibility of the swelling. There were no signs to suggest intestinal obstruction secondary to a strangulated hernia, although a Richter's hernia remained a possibility. A urological opinion was also sought and the urologist made a diagnosis of torsion of the left testis.

G.T. was prepared for surgery and the scrotum and mass explored ten hours after the onset of pain. At operation a seven hundred and twenty degrees torsion of the left testis was found. The cord was untwisted and since the left testis itself viable, it was fixed in place. The right testis was fixed at the same time to remove the possibility of a similar event on that side.

G.T. made an uneventful recovery from the operation and was discharged from hospital two days following the operation.

Commentary

Testicular torsion is an important diagnosis to consider in the differential diagnosis of a scrotal swelling or lump in the groin since delay in making the diagnosis may result in irreversible damage or necrosis of the involved testis.

It is estimated that testicular torsion occurs three to four times per year in a large general hospital (Williams 1977). There is approximately a fifty per cent testicular loss from either atrophy or orchidectomy (Williams 1977). The two peak periods during which torsion is likely to occur are in the first year of life and at puberty (Allan and Brown 1966) although twenty-three per cent of cases occur in men over the age of eighteen years (Donohue and Utley 1978).

The initial effect of testicular torsion is obstruction of venous return. Venous thrombosis occurs, followed by arterial thrombosis. The degree of obstruction is a function of the degree of rotation. Incomplete rotation leads to the development of oedema and congestion. Complete obstruction results in the development of testicular neurosis and infarction promptly follows arterial thrombosis. Compromising the blood supply to the testis for six hours may lead to an irreversible loss of spermatogenesis and if the torsion persists for ten hours neurosis of the Leydig cells is likely to occur. Thus preservation of spermatogenic and hormonal function is only likely if the torsion can be terminated within six to ten hours and it is this fact which makes the prompt diagnosis of testicular torsion so important. Most torsions occur in testes with an underlying anatomical abnormality. Most commonly there is a large tunica vaginalis with a high insertion round the spermatic cord. This results in a redundant length of cord and the testis dangles in the scrotum - the so-called bell-clapper deformity (Donohue and Utley 1978). This anomaly is usually bilateral and thus forms the rationale for fixing both testes at operation.

The clinical features of testicular torsion are usually well defined. An early clue is provided by the fact that forty-one per cent of patients report a history of similar pain which resolved spontaneously (Donohue and Utley 1976; Parker and Robinson 1971). S.T. fell into this category, describing a previous episode of pain similar to his current complaint. The pain characteristically begins suddenly in the scrotum but may also be inguinal or lower abdominal as in the case described above. Straining at stool, lifting heavy weights and coitus are all possible precipitants (Rains and Ritchie 1984). The occurrence of torsion during sleep is also recognised (Harwood 1983) and was the case in this instance. Characteristically there is an absence of urinary symptoms.

In the straight forward case physical examination will reveal an extremely tender, swollen hard hemiscrotum. The classical signs of a high riding testis

with a transverse lie may not be detectable on the affected side. Examination of the opposite testis is often helpful and it may be noted to lie in the horizontal axis indicating the presence of an underlying bell-clapper abnormality (Williams 1977; Donohue and Utley 1978). In this case the contralateral testis was felt to be normal. Prehn's sign - the relief of scrotal pain by elevation of the testis in cases of epididymitis is no longer felt to be an accurate means of distinguishing between torsion and epididymitis (Allan and Brown 1966).

However, there are a number of situations in which the physical findings and diagnosis are not so straightforward. Torsion of an imperfectly descended testis may be impossible to distinguish from a strangulated inguinal hernia (Rains and Ritchie 1984). Torsion of a completely descended testis may be exactly mimicked by a small strangulated inguinal hernia compressing the cord and causing congestion of the pampiniform plexus.

There are a number of aids to diagnosis which may be useful to the clinician although unnecessary delay in arriving at a diagnosis may result in the loss of the testicle and it remains important to have a low threshold for surgical exploration of cases of possible testicular torsion. A urine analysis and full blood count are both useful in that they should be normal. Doppler ultrasound examination may show decreased or absent blood flow in the affected testis and this is considered diagnostic of testicular torsion (Iuchtman et al 1979). Radionuclide imaging has also been used with considerable success (Boedecker et al 1979).

The condition most commonly confused with testicular torsion is epididymitis. Features useful in distinguishing between the two conditions are that epididymitis tends to occur in an older age group (Allan and Brown 1966) and the pain is usually of more gradual onset. Seventy-five per cent of cases have urinary tract symptoms which precede the onset of the pain (Berger et al

1979). Ninety-five per cent of cases of epididymitis are associated with fever but twenty per cent of patients with torsion are also mildly febrile (Allan and Brown 1966). The presence of a leucocytosis or pyuria are further features suggestive of epididymitis.

The differential diagnosis between epididymitis, torsion and testicular tumour can also be difficult to make and epididymitis is in fact the most common incorrect diagnosis made in cases of testicular tumour with an incidence of between six and sixteen per cent (Bigley et al 1977). Usually testicular tumours are painless but sudden onset testicular pain may be secondary to haemorrhage within the tumour.

Torsion of the testicular appendages may produce a clinical picture indistinguishable from testicular torsion. As noted above, some presentations of testicular torsion may be difficult to clinically distinguish from small strangulated inguinal hernias.

The only definitive treatment of testicular torsion is surgical. Temporary relief may be obtained by manual detorsion. Most torsions occur in a medial direction. Successful normal detorsion will be accompanied by immediate relief of pain and should be done with the patient in the supine position after administering appropriate parenteral analgesia (Frazier 1975). This procedure removes the urgency but not the necessity for surgical treatment which should include fixing of both the affected and unaffected testis (Ranis and Ritchie 1984).

Thus, this case illustrates that conditions frequently do not present in the clear cut ways described in textbooks and that in some instances undue procrastination concerning the diagnosis may have serious consequences for the patient.

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CONCLUSION

I hope that the cases presented above have served to illustrate the benefits of surgical intervention in situations when other forms of treatment have failed or have nothing to offer and the importance of prompt diagnosis in situations where early surgical intervention is essential to avoid morbidity.

The brief reviews of the diagnostic and treatment options available to the contemporary surgeon highlight the vast amount of factual information at his disposal. As this information continues to increase in complexity and scope, it becomes more and more difficult for the surgeon to keep up to date and select the most effective strategy in individual cases. Increasing use of computers, expert systems and the application of decision analysis (Pauker and Kassirer 1987; Szolovitz 1982) may prove to be useful means of rationally applying such information in the future.

Future improvements in medical treatment and the advances in the screening for, and prevention of, some diseases may decrease the contribution of surgery in some areas. However, continuing technological advances are certain to open up new areas of application for the surgeon's skill and ensure that surgery remains a vital component of the wider field of clinical medicine.

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Acknowledgements

I am indebted to Professor I.D. Miller, Department of Neurosurgery, WGH, Mr. R. Buchan, Victoria Hospital, Kirkcaldy and Dr. D.S. Dettman, Emergency Department, Vancouver General Hospital for allowing me to present case histories of patients under their care.