REPORT AND DISCUSSION
OF FIVE CASES IN GENERAL SURGERY
by
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(Phase III, year 4)

Submitted in fulfilment of the requirements of
'The Pattison Prize in Clinical Surgery'

May 1987
The following five cases were encountered during my general surgical attachment to Wards 9 and 10 of the Royal Infirmary of Edinburgh from 9th March 1987 to 1st May 1987.
INTRODUCTION

I have chosen each of the following five cases, to present and discuss, primarily because they have captured my interest, and thus prompted my research. However, I think they should be of interest to all, not least because of their rarity.

In each case I have had personal contact with the patient and been closely involved in their diagnosis and management, in clinics, wards and theatre. As such, I have found this a very valuable exercise for learning some of the diagnostic and therapeutic skills required in the practise of clinical surgery. It has also introduced, and helped me to develop a methodical approach to clinical research.

After presenting each case, I have chosen to discuss a particular subject which has arisen from each, and which has stimulated my interest.
CONTENTS

CASE ONE
Spontaneous retroperitoneal bile leakage and "biliscrotum".

CASE TWO
Polycystic liver disease - a case for transplantation?

CASE THREE
Myelofibrosis and post splenectomy infection.

CASE FOUR
The hazards of surgery - iatrogenic ureteric damage.

CASE FIVE
Inflammatory bowel disease and faecal peritonitis.
MR. J.B. (07.05.99)

Mr. J.B., an 87 years old retired plumber, presented to the physicians as an out patient on 3rd March, 1987 with a one week history of painless jaundice, pale stools and dark urine associated with some anorexia and a minor degree of weight loss. He also had some difficulty passing urine.

An outpatient ultrasound scan showed gallstones in the gall bladder and common bile duct dilation. The liver, kidneys and spleen appeared normal. However, a pancreatic lesion could not be fully excluded.

Mr. J.B. was due to have an ERCP as an outpatient. However, on 5th March 1987 he was admitted to the physicians as an emergency with acute onset of right sided testicular pain. This was diagnosed clinically as acute epididymo-orchitis.

Mr. J.B.'s past medical history includes transvesical prostatectomy in 1977 and insertion of a permanent pacemaker in 1986 for symptomatic episodic 2nd degree heart block.

At this admission, systematic enquiry was unrevealing, Mr. J.B. was taking no drugs or medications, had no known allergies and neither smoked nor drank alcohol.

On examination he was obviously icteric but with no stigmata of chronic liver disease.

**CVS** Pulse rate - 80/min - regular RTFV

**B.P.** 120/70

**JVP** - not elevated
Cardiovascular system was otherwise unremarkable.

**Chest** - clear

**Abdomen** - no ascites or organomegaly could be elicited.

**Rectal examination** - revealed clay-coloured stools, negative for faecal occult blood.

The scrotum was markedly swollen, erythematous and tender, especially on the right side.

**Investigations:**

<table>
<thead>
<tr>
<th>Haematology</th>
<th>Hb. 14.1, MCV 101, MCH 34.4, WCC 11.1</th>
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</thead>
<tbody>
<tr>
<td>Clinical chemistry</td>
<td>Urea 10.4, Na 139, K 3.4, Total CO₂ 26</td>
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<tr>
<td></td>
<td>Bilirubin 151, ALT 127, gamma GT 25</td>
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<td></td>
<td>Glucose 5.4</td>
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<tr>
<td>Clotting</td>
<td>PTR 1.7</td>
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<tr>
<td>Hep B -</td>
<td>negative</td>
</tr>
<tr>
<td>Urinalysis</td>
<td>Trace of blood. Bilirubinuria +++</td>
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<td></td>
<td>Trace of protein.</td>
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**Progress and management:**

Mr. J.B.'s epididymo-orchitis was treated conservatively with ampicillin - 500mg q.d.s. and analgesia - DF 118 PRN. He was also provided with a scrotal support.

On 12th March 1987, ERCP was carried out:–

A normal pancreatic duct was found but the common bile duct could not be cannulated due to obstruction by stones.

A second ERCP was booked for two weeks hence.

On 16th March the testis appeared less tender and swollen but on
the following day the scrotal swelling suddenly increased greatly in size, Mr. J.B. vomited several times and his bowels were sluggish. This was thought to be evidence of acute obstruction and a surgical opinion was sought. The presumptive diagnosis at this stage being a right incarcerated hernia.

When seen by the surgeons, he appeared very jaundiced. Pulse 92/min regular RTFV.
BP 130/90
Chest clear
Abdominal examination revealed a distended and tympanic abdomen with some suprapublic discomfort and tenderness. No organomegaly could be elicited. Bowels sounds - present. The scrotum was extremely swollen and hard mainly on the right side. It was non-tender.

Investigations:-

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Results</th>
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<tbody>
<tr>
<td>Haematology</td>
<td>Hb. 12.1, WCC 17.9, MCV 101, Platelets 461</td>
</tr>
<tr>
<td>Clinical chemistry</td>
<td>Urea 22.0, Na 138, K 4.1, CO2 24</td>
</tr>
<tr>
<td>Clotting screen</td>
<td>PTR 20 : 14, PTTK 43 : 30, Fibrinogen 6</td>
</tr>
<tr>
<td>CXR</td>
<td>NAD</td>
</tr>
<tr>
<td>AXR</td>
<td>NAD</td>
</tr>
<tr>
<td>ECG</td>
<td>NAD</td>
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Mr. J.B. was catheterized - 450mls of dark, pungent urine was collected. 50mg potassium was given intravenously to reduce the PTR and Mr. J.B. was taken to theatre.

Operative findings and procedure:-

Procedure I
A skin crease incision was made in the right groin and extended
through to the external oblique aponeurosis. In the subcutaneous tissues and under the external oblique the tissues were deeply stained green-brown, initially raising the suspicion of a severe necrotizing infection. However, it was odourless and no actual pus could be seen. The inguinal canal was opened and the cord examined. There was no obvious hernia. There was a large amount of brown-stained fluid welling up from the scrotum. The testis and epididymis were therefore evaginated and brought out of the scrotum. Approximately 200mls of brown-stained fluid came with the cord and testis and the cremasteric region appeared necrotic. Cord and testis were therefore removed. Bile stained fluid was then seen issuing extraperitoneally, along the track next to the site of ligation of the deep inguinal ring. This area was opened further with blunt dissection and copious amounts of brown bile-stained fluid issued forth. It then became obvious that the whole problem was originating from the biliary tree.

A large bore chest drain was placed in the retroperitoneal tissues and brought out through the right iliac fossa. A redivac drain was placed in the scrotum.

The patient was redraped for laparotomy.

Procedure II
A right subcostal incision was made. The whole right retroperitoneal area was boggy, oedematous and deeply bile stained. The gall bladder wall was thickened and stones were palpable. Cholecystectomy was performed. The common bile duct was opened and several small calculi removed. The liver was normal. No further abnormality was found in the abdomen. A 16-guage T-tube was placed in the common bile duct and the whole area was irrigated with tetracycline 1g/litre. The
duodenum was not mobilized nor the site of leakage identified as it was not wished to disturb the retroperitoneal biliary leak which was now draining.

A redivac drain was placed in the gall bladder bed and the wound was closed.

**Post-operative progress**

Condition stable.

Prophylactic antibiotics were used - a short course of Metronidazole - 500mg and cefotaxime 1g.

On the sixth postoperative day Mr. J.B. was given a methylene blue swallow - no obvious leak was seen. On the ninth postoperative day a T-tube cholangiogram was done - this showed no leakage. Three days later Mr. J.B. developed a pyrexia and so he was commenced on augmentin and metronidazole and a chest x-ray was carried out which subsequently proved to be normal.

On 30th March, two weeks post-operatively, a gastrograffin swallow was done - this demonstrated no leakage.

On 2nd April the T-tube cholangiogram was repeated - this demonstrated two stones in the common bile duct. It was not possible to remove the stones via the T-tube and so an ERCP and papillotomy was done on 9th April.

A stone was removed at ERCP but a second stone above the T-tube could
not be removed but a generous papillotomy was performed, and it was thought that the stone would subsequently be passed spontaneously.

Later that day the wound around the T-tube and suture point became red and inflamed - the patient spiked a temperature of 38.5°C, pulse 120/min bp. 130/90. His chest was clear. He was recommenced on metronidazole. The following day he appeared well and was afebrile.

On 14th April the T-tube was removed. Late that evening he became febrile (40°C), developed rigors and an altered mental state. He was thought to be septicaemic secondary to cholangitis and it was assumed that a stone was lodged at the lower end of the common bile duct.

Intra-venous antibiotics were commenced - gentamycin, metronidazole and amoxycillin. He was observed very closely and blood cultures, which later showed to contain pseudomonas, were taken.

Early the following morning he became hypotensive - BP 90/50, but his temperature had fallen to 37.5°C and he did not have rigors or an altered mental state. An indwelling catheter and central venous pressure line were sited to monitor carefully his fluid balance. CVP was -1cm and so fluids were increased. 10mg vitamin K was given as it was thought an emergency ERCP might be needed.

Hourly urine output, BP, pulse rate and 2 hourly CVP measurements were made.

Later that morning an emergency ultrasound scan was done, confirming
the presence of a stone at the lower end of the common bile duct.

PTR was checked - 1.6 and corrected with vitamin K and an emergency ERCP was carried out - the papillotomy was enlarged and with great difficulty the stone was removed.

Mr. J.B.'s condition gradually improved and approximately ten days later he was discharged.
DISCUSSION

Perforation of the common bile duct is a rare but well documented complication of endoscopic sphincterotomy for the treatment of common bile duct stones. Clinically apparent perforation following endoscopic sphincterotomy occurred in 1.1 per cent of cases in a recent large series (Safrany et al, 1978), although the incidence of silent perforation is probably much higher. Usually perforation leads to leakage of bile into the peritoneal cavity and occasionally bile leaks into the retroperitoneal compartment. Yet I have found only one case, reported in the literature, of such a perforation leading to a massive retroperitoneal bile leakage and thence "biliscrotum" (Neoptolemos et al, 1984). The case of "biliscrotum" which I describe was very similar in presentation and management to that described by Neoptolemos et al, the important differentiating feature being the spontaneity of the case described above.

Spontaneous rupture of the common bile duct is a very rare condition and usually leads to free leakage of bile into the peritoneal cavity. Very few cases of spontaneous bile duct rupture have been reported in adults. Spira (1976) collected 12 cases of common bile duct perforation, 11 associated with gallstones. Further cases have since been described by Plehwe et al (1978) and Wig et al, (1983).

Only one case of spontaneous common bile duct rupture leading to a large retroperitoneal encapsulation of bile has been reported (Satake et al 1985) and to my knowledge a case of spontaneous retroperitoneal bile leakage leading to "biliscrotum" has not previously been described.

The pathogenesis of nontraumatic perforation of the biliary tract
system is poorly understood, but the following mechanisms are generally accepted. 1) Calculous perforation at the site of impaction or erosion without evidence of mechanical disproportion of the ductal system; 2) increased intracanalicular pressure due to obstruction by tumour, impacted stone, or spasm of the sphincter of Oddi, 3) intramural infection; 4) intramural infection due to thrombosis of the mural vessels; 5) rupture of a diverticulum or a cyst of the biliary tract; 6) regurgitation of activated pancreatic juice through a common outlet of the common bile duct and the pancreatic duct; 7) traction from neighbouring organs. A combination of factors is probably responsible for rupture of the common bile duct by a gallstone. Erosion of a calculus through the wall of the duct is usually a slow process and is mainly associated with fistula formation. Complete distal obstruction results in an elevation of the intra-ductal pressure and in most cases this will present as obstructive jaundice. However, if the wall of the bile duct has been weakened, the raised pressure may precipitate a rupture. Infection plays an important role in the aetiology of spontaneous rupture, and, combined with the presence of calculi and/or a diverticulum, is probably the important factor in causing weakness of the duct wall. Thrombosis of the intramural vessels probably follows infection, although the mechanism itself may be obscure.

Our patient had a history of painless obstructive jaundice and ultrasound examination revealed gallstones in the common bile duct and a dilated common bile duct. The presence of gallstones was confirmed by ERCP. Although we never managed to locate the exact site of the bile leakage, it is likely that the gallstones in the common bile duct led to its perforation and hence the tracking of bile through retroperitoneal fascial planes thus leading to the formation of a "biliscrotum".
Thus I have described an extremely rare, indeed unique, presentation of common bile duct rupture.

The scrotum acts as a receptacle for a variety of rare presentations of abdominal pathology. In 1986 Noel et al described a case of urine extravasation into the scrotum following renal transplantation. Ureteral fistula with urine extravasation is a well known and serious complication of renal transplantation and may be the result of a deficit in ureteral blood supply (resulting in ureteral necrosis), a deficit in surgical anastomosis, or involvement of the ureter as well as the kidney with rejection. The occurrence of this complication in reported cases varies from 1 to 30% (Spigos et al 1977 and Texter et al 1976). However, urine extravasation into the scrotum is very rare.

Regarding the pathway of extravasated urine into the scrotum, the fluid most likely dissects through the inguinal canal along the spermatic cord.

Similarly a case of delayed rupture of the spleen presenting as a scrotal haematoma has been described (Sujka et al 1986). It was postulated in the case described by Sujka et al that the blood from the peritoneum, which originated from the delayed rupture of the spleen, dissected along the cord and tissue planes, accumulating in the scrotum. The most likely route of dissection was along the processus vaginalis.

Dissection into the scrotum has thus been postulated on several occasions in the past and the case I describe further supports this and in addition emphasises that when considering the differential
diagnosis of a scrotal swelling, abdominal pathology must be considered.

REFERENCES

CASE TWO

...
Mrs. E.G. (25.03.43)

Mrs. E.G., a 43 year old teacher, first presented to her G.P. in July 1986 complaining of nausea, eructation and a feeling of satiety even after eating very small meals. This had been gradually getting worse over the previous few weeks. She also complained of occasional epigastric pain radiating round to the back which was neither relieved nor aggravated by anything. She felt she had lost some weight over the last few weeks. Her appetite was unchanged, she had no dysphagia and her bowel habit was unremarkable. On examination she was found to have hepatomegaly. She was referred for a surgical opinion.

Gastroscopy and gamma scan of liver and spleen revealed no abnormality. Ultrasound examination revealed polycystic disease of the liver.

On 20th October 1986 she was re-examined by ultrasound scan which confirmed polycystic disease of the liver - spleen and kidneys appearing normal. The pancreas could not be visualized. Two large cysts were aspirated under ultrasound guidance and clear yellowish fluid was collected.

The following day she complained of severe abdominal pain. On examination she was apyrexial but looked very ill. She displayed rebound tenderness, abdominal rigidity and guarding. Bowel sounds were absent. Pulse rate was 80/min; BP 130/80.

A diagnosis of peritonitis secondary to bile leakage due to aspiration was made. She was commenced on amoxycillin, gentamycin, metronidazole and analgesics. An intravenous infusion was started and a nasogastric tube passed.
Investigations

Haematology
Hb. 13.4, WCC 18.3, PTR 1:1

Clinical chemistry
Urea 4.9, Na 138, K 4.0, Amylase 169

CXR – AXR – unremarkable except for demonstrating the enlarged liver.

Ultrasound scan – showed free peritoneal fluid.

Over the next two days she became gradually better while being managed conservatively. She soon returned to a normal diet.

On 28th October, in view of her original complaint of epigastric discomfort, nausea and easy satiety, a barium follow-through was performed as the recent gastroscopy had not visualized the duodenum and hence a duodenal lesion could not be excluded. However, these symptoms were thought to be due to simple extrinsic compression of the stomach by the enlarged liver. The result of this test revealed persistent indentation of the lesser curve aspect of body and antral areas of stomach. This was consistent with extrinsic pressure on the stomach from the enlarged left lobe of liver.

The barium meal and follow-through was otherwise normal and gastric emptying was said not to be impeded, transit time from the stomach being quite rapid.

A few days later Mrs. E.G. was discharged from hospital.

On 3rd February 1987 Mrs. E.G. presented again to her G.P. complaining of worsening epigastric pain, inability to eat adequate quantities of food, severe eructation and upper abdominal swelling.
A decision was made to deroof some of the cysts rather than reaspirate, and so she was admitted to hospital on 18th March 1987.

Her past medical history included a laparotomy, D & C and other minor gynaecological procedures in 1971.

Mrs. E.G. seemed to think that her mother might have had liver cysts - ? She drank alcohol occasionally and smoked about 8 cigarettes/day.

Systematic enquiry was unremarkable.

On examination she was undistressed and apyrexial. She was anicteric, had no lymphadenopathy or finger clubbing. There were no stigmata of chronic liver disease.

Pulse 72/min BP 145/90 - CVS otherwise NAD

Chest - clear

Abdomen - obviously distended - upper abdomen
  soft, non-tender
  Hepatomegaly - 5 finger breadths below xiphisternum
    - irregular outline
  No KKS
  Bowel sounds - present
  No ascites

P.R. Rectum empty. No pain. No masses. FOB negative

CNS Grossly intact

Investigations

| Haematology | WCC 7.2, Hb 13.0, platelets 221, PTR 1:1 |
| Clinical chemistry | Urea 3.4, Na 142, K 4.0, Total CO₂ 25 |
|                | Bilirubin 7.0, Alkaline phosphatase 69 |
Clinical chemistry (cont)  ALT 22, gamma GT 76
Albumin 42, Ca 2.27, Glucose 4.5

On the 19th March 1987 Mrs. E.G. was taken to theatre.

Operative procedure and findings

The abdomen was opened through a right subcostal incision. There appeared to be very little normal tissue in the right lobe of the liver, which contained myriads of cysts (Figure 2.1). Cystic changes extended across into the medial part of the left lobe of liver, left of the falciform ligament. The only sizeable piece of functioning liver tissue appeared to be the lateral part of the left lobe which was hypertrophied and also contained a small number of cysts. The size of the liver as a whole was such that simple compression of the stomach could certainly have been responsible for Mrs. E.G.'s symptoms.

Brief discussion as to the possibility of major resection of liver tissue was undertaken - but this would have meant a trisegmentectomy which was inappropriate due to cysts in the "good" part of the liver.

Therefore, a large number of the larger cysts were marsupialized and a central segment of the liver was excised (Figure 2.2).

This appeared to relieve pressure on the stomach. A redivac drain was sited and the wound closed.
Fig. 2.1a) Appearance of polycystic liver at laparotomy. Diaphragmatic surface.

Fig. 2.1b) Appearance of polycystic liver at laparotomy. Visceral surface.
Fig. 2.2 Appearance of polycystic liver after marsupialization of some of the larger cysts.
Post-operative progress

Per-operative blood loss totalled 880ml and so Mrs. E.G. was given two units of red cell concentrate.

Her immediate post-operative condition was stable, but the following day she developed a mild pyrexia of 38°C.

By 21st March 1987, a swinging pyrexia had developed. Mrs. E.G. complained of breathlessness on minimal exertion. On examination there was found to be decreased air entry at the right lung base and localized rhonchi could be heard. Chest x-ray revealed a small area of consolidation at the right lung base. Mrs. E.G. was treated conservatively with antibiotics and she made a rapid recovery.

Mrs. E.G. experienced severe abdominal discomfort throughout the following month which was treated with analgesics and gradually resolved. Bile continued to discharge from the abdominal drain for approximately two weeks. Once drainage ceased, the drain was removed.

Mrs. E.G. returned home on 19th April 1987.
DISCUSSION

Polycystic disease of the liver in association with polycystic disease of the kidneys is rare. Polycystic disease of the liver in isolation, as in the case described above, is very rare.

Cystic disease in the liver occurs more often in females than males, the ratio being 4 or 5 to one.

Although many liver cysts are congenital (i.e. non parasitic), they usually do not become clinically expressed until the patient is between 30 - 50 years of age. Indeed 75% of patients with polycystic liver disease present between 40 and 70 years of age.

Many theories have been postulated as to the aetiology of polycystic liver disease.

Moschowitz in 1906 suggested that liver cysts arise as an embryological abnormality of bile duct development - this is supported by the fact that the cysts of polycystic liver disease are usually lined by bile duct epithelium and indeed this is the theory which still holds today.

An alternative suggestion was proposed by Norris and Tyson (1947), i.e. over production of intra hepatic bile ducts giving rise to cysts.

Polycystic disease of the liver and kidneys has been shown to be inherited in both autosomal recessive and autosomal dominant Mendelian manner, and as such there appears to be at least two specific types of adult polycystic disease - not to mention the paediatric polycystic conditions.
The inheritability of polycystic liver disease without any kidney involvement has not yet been elucidated satisfactorily.

Polycystic disease may also affect other organs: pancreas, spleen, lungs, ovaries, uterus, seminal vesicles etc. and is associated with a variety of congenital abnormalities, the commonest being cerebro-vascular aneurysms.

The most common clinical presentation of adult polycystic liver disease is distension in the right upper abdomen first noticed as a lump in about 20% of patients.

Episodes of severe abdominal pain are also common. Should there be associated polycystic disease of the kidneys, and especially if renal function is impaired, hypertension, cardiac failure and cerebrovascular accident may be prominent clinical features.

Laboratory findings are usually normal even when the liver is large in size; hence liver function is rarely impaired - (Comfort et al 1952). When kidneys are also involved by polycystic disease albuminuria and microscopic haematuria may be detected.

Complications of liver cysts include torsion, strangulation, haemorrhage, rupture and abscess. The symptoms and signs of these conditions are essentially pain and those of peritoneal irritation.

The vast majority of patients with non-communicating (cysts do not communicate with bile ducts) polycystic disease do not need treatment of cysts. Patients with obstructive jaundice or severe pain may benefit from deroofing of as many cysts as possible. The small minority
of patients who develop portal hypertension may be managed by portacaval shunting.

The development of obstructive jaundice is very rare, but may occur as a result of cystic pressure on the common bile duct. However, cysts grow so slowly that adjustment by the common bile duct is gradually made to the rising pressure.

Congenital cysts very rarely progress to malignancy: Willis R.A. in 1943 described carcinoma arising from the epithelial lining of multiple developmental cysts of the liver. Manes et al 1977 reported occurrence of liver cell carcinoma in a patient with adult polycystic disease.

A presenting feature is very rarely massive haematemeses from oesophageal varices (Cambell et al, 1958).

As already stated most patients with congenital polycystic disease of the liver do not require treatment. However, the surgical management of polycystic liver disease is directed at those cysts causing most symptoms. If the multiple cysts are located only in one lobe of the liver, a lobectomy or partial hepatectomy may be feasible (Grime et al, 1959). Occasionally, multiple punctures of cystic areas in polycystic liver disease have been successful. Lin et al, 1968, described a fenestration procedure for polycystic livers. Multiple cysts may be marsupialized to the peritoneum, to the gall bladder when they are in the right lobe of the liver and to the intestine by a Roux-en-Y procedure when in the left lobe of liver. Medical decompression of the cysts using sodium restriction and diuretic therapy has also been reported to control the symptoms of abdominal pressure.
Portocaval anastomosis may become necessary to reduce secondary portal hypertension and hypersplenism.

Generally the prognosis for isolated polycystic liver disease is very good. When in combination with polycystic kidneys the outcome is usually worse.

In Mrs. E.G.'s case the severity of her polycystic liver disease is such that it is not inconceivable that the remaining cysts will grow and cause recurrence of her symptoms, thus requiring further surgical intervention.

Brief discussion was undertaken by the surgeons and physicians as to the possible need for liver transplantation at a future date.

However, the five year survival of patients after liver transplantation is still very poor (McMaster et al 1985) and so long as Mrs. E.G. has some functioning liver tissue remaining, capable of regeneration (the liver has a remarkable capacity for regeneration) transplantation would be unacceptable.

As we know from previous studies on polycystic liver disease (Comfort et al, 1952; Melnick et al, 1955) liver function is rarely compromised to any great extent and I could not find a single case of liver transplantation for polycystic liver disease in the literature. So it is likely that Mrs. E.G.'s future management will involve further surgical deroofing, marsupialization or fenestration of cysts, should symptoms recur.
REFERENCES


Textbooks

Diseases of the gastrointestinal tract and liver – Shearman and Finlayson, 1st edition.
CASE THREE

Mrs. W., a 38-year-old female, presented to the clinic on the 6th
February 1997 with a four-day history of severe headache. She had
also noticed a lump on the left side of her abdomen since November
1996.

She was referred to the medical oncologist department and there
presented to the surgical unit on 19th February 1997.

Her past medical history included appendectomy in 1985, subtotal
colectomy in 1990, and an ovarian cystectomy in 1993.

She had been doing relatively occasionally but was not on any
medications. She had no history of surgery, and her daily
activities were not restricted. She did not have any appetite
problems.

On physical examination she was found to have a mass in the left
upper abdomen. The mass was firm and movable, and there was
no pain.

Rectal examination was normal.

Investigations:

Total leukocyte count: 11.2 x 10^3/μl

Hemoglobin: 12.5 g/dl

Chest X-ray: normal

Ultrasound: a mass was seen in the left upper abdomen.

CASE THREE
MRS. G.H. (13.12.47)

Mrs. G.H., a 39 year old law cashier, presented to her G.P. on 4th February 1987 with a four day history of severe heartburn. She had also noticed a lump in the left side of her abdomen since November '86.

She was referred to the medical out patient department and thence admitted to the medical wards on 19th February 1987.

Her appetite was good, her weight being stable at 7st 12 lbs. She had no other gastrointestinal symptoms.


She had been taking antacids occasionally but was on no other medications. She has no known allergies and neither smokes nor drinks alcohol. She is a single lady with no family history of illness.

Systematic enquiry was unrevealing.

On Examination she was cachexic with prominent ribs and muscle wasting. Slight cervical lymphadenopathy was present.

CVS

- Pulse 100/min regular RTFV
- BP 120/60
- JVP not elevated
- Heart sounds I + II + 0 + 3/6 systolic murmur LSE

Chest

- Clear

Abdomen

- Visibly distended
- Hepatosplenomegaly to the level of the iliac crests.
Spleen - very tender No ascites.
Bowel sounds present.
Grossly intact

Investigations

Haematology

Hb 10.8, WBC 9.7, RBC 4.18 Hct 0.31
MCV 75, MCH 25.8 MCHC 34.4
platelets 243
Reticulocytes 9.8%
Differential:- Myelocytes 9%, Metamyelocytes 2%
Neutrophils 68%, lymphocytes 17%
Monocytes 1%, Eosinophils 1%
Nucleated red blood cells 2%

Blood film:- platelets - normal,
red blood cells - anisocytic,
normochromic, some hypochromic cells,
some poly-chromasia, occasional "tear-
drop" poikilocytes, occasional spherocytes.

Clotting

PTR 1 : 1.5, PTTK 32 : 30, Fibrinogen 5.4

Clinical chemistry

Urea 2.6, Na 138, K 3.9
Bilirubin 7, Alkaline phosphatase 334
ALT 24, gamma GT 84.

Serum electrophoresis

NAD

CXR

NAD

ECG

NAD

Bone marrow aspiration was attempted - yielded a "dry" tap so two
trephine biopsies were taken. They revealed an almost totally aplastic
bone marrow in which only occasional small aggregates of erythroid cells
and megakaryocytes were seen. Focally the marrow showed replacement by loose fibrous tissue but this was not a striking feature.

A 99m Tc scan of liver and spleen showed marked enlargement of liver and spleen which extended well below the costal margins to the level of the iliac crest. There was no evidence of focal defects within either organ and no reversal of normal liver : spleen ratio. The appearance was consistent with a diagnosis of myelofibrosis.

Radiographs of the long bones showed a generalized increase in bone density and a prominent trabecular pattern; consistent with myelofibrosis.

Iron studies showed decreased sacral uptake and increased splenic uptake.

On 5th March the bone marrow trephine biopsy was repeated. This revealed a hypoplastic picture with immature megakaryocytes, fibroblasts, scanty lymphoid cells and plasma cells. Almost no normal haemopoietic tissue was present. There was also increased reticulin deposition.

Mrs. G.H. was transferred on 1st April for splenectomy on 2nd April.

Operative findings and procedure
A mid line incision with oblique extension to the left costal margin was made.

The spleen was grossly enlarged (3.5 kg) and there were two splenunculi (Fig 3.1, 3.2). There was also considerable hepatomegaly but the
surface appearance and consistency was normal. Approximately two litres of free fluid was present in the peritoneal cavity but there were no features of portal hypertension.

Haemostasis was ensured and a large bore redivac drain was sited.

Pathology
The spleen was grossly enlarged measuring 28cm x 18cm x 11cm and weighing 3.5kg (Fig 3.3).

It had a smooth capsular surface.

Microscopically the spleen showed diffuse infiltration by a mixed population of cells. Erythroid and myeloid precursors were present, indicating extensive extramedullary haemopoesis. A population of large cells with pleomorphic enlarged nuclei was present representing atypical megakaryocytes. Some large nucleated cells with granular chromatin staining were present, probably representing early myeloid precursors. Appearances were consistent with splenic enlargement secondary to myelofibrosis.
Fig. 3.1  Gross hepatosplenomegaly seen at laparotomy

Fig. 3.2. Grossly enlarged spleen after splenectomy for myelofibrosis
Fig. 3.3 Grossly enlarged spleen measuring 28cm x 18cm x 11cm and weighing 3.5 kg.
Post-operative progress

Intraoperatively Mrs. G.H. was given 11 units of platelets and postoperatively 20 units of platelets.

The following day she appeared unwell, developed a pyrexia of 38.5°C and a pulse rate of 120/minute. She also developed a cough, unproductive of sputum. On auscultation of the chest coarse expiratory crackles could be heard at the right lung base.

Blood cultures were performed and a catheter specimen of urine was sent to bacteriology - both of which subsequently showed no growth.

She was commenced on penicillin and given physiotherapy. By the 7th April Mrs. G.H. was eating, drinking and mobilizing but her pyrexia had risen to 39°C.

On examination Shivering and generally unwell
pulse 120/min
JVP raised 3cm
Heart sounds I, II and III i.e. gallop rhythm.
An ejection systolic murmur was heard over the aortic area. Grade 3/6.
Ankle and sacral oedema - present

On percussion of the chest there was bilateral basal dullness and on auscultation the breath sounds were absent at both lung bases and aegophony was heard at the left midzone.

Chest x-ray revealed bilateral basal consolidation and collapse.
Sputum grew commensals only.
Further blood cultures were taken and more samples of sputum were sent to bacteriology for culture.

Her antibiotic therapy was changed from penicillin to augmentin. Physiotherapy was continued.

The presumptive diagnoses at this stage being chest infection and high output state due to pyrexia.

The following day her clinical condition was unchanged but staphylococcus aureus was isolated from her sputum and a repeat chest x-ray revealed left basal consolidation and pleural effusion, the right lung base being relatively clear.

So she was thought to have a staphylococcal pneumonia with some degree of fluid overload, JVP now being raised by 5cm.

Augmentin was ceased and netilmicin and flucloxacillin commenced.

An oral dose of 40mg frusemide was also given.

By the 9th April Mrs. G.H. was miserable. She had a cough productive of sputum and had also developed haemoptysis. Her chest x-ray showed widespread bronchopneumonia. Mrs. G.H.'s wound was also inflamed, as were her venous access sites. Her WCC was 11.7 and her haemoglobin 7.6. Antibiotics were continued and two units of red cell concentrate were given, 20mg frusemide with each.

On 12th April her condition had improved, her temperature being 38°C. A good fluid throughput had been achieved with a negative fluid balance.
Her JVP had fallen to normal and heart sounds I and II could be heard only. She was producing less sputum and there was better air entry at both lung bases. The inflammation around the wound appeared much worse with evidence of spreading cellulitis (Fig. 3.4). Intravenous netilmicin and flucloxacillin were continued. Further 20mg doses of frusemide were administered. Oral metronidazole was also added.

The following day her temperature rose to 40°C, she developed a tachypnoea of 28/min with a pulse rate of 118/min and a JVP elevated 8cm. There was still decreased air entry at both lung bases and bilateral dullness on percussion. She was given a bolus dose of 40mg frusemide, 4 hourly paracetamol and her intravenous flucloxacillin was changed to oral penicillin in view of her spreading cellulitis around her wound; the likely organism being streptococcus. Repeated wound swabs were negative.

On 16th April she was taken to theatre for debridement of the wound (Fig. 3.5).

\[
\begin{align*}
\text{Hb} & \quad 9.4 \\
\text{WCC} & \quad 18 \\
\text{platelets} & \quad 660 \\
\text{PTR} & \quad 17:14 \\
\text{U} & \quad 4.6 \\
\text{Na} & \quad 130 \\
\text{K} & \quad 4.0
\end{align*}
\]
Fig. 3.4 Splenectomy wound with surrounding spreading cellulitis.

Fig. 3.5 Wound debrided after spreading cellulitis.
After debridement of the wound it was lavaged with hydrogen peroxide solution and dressed.

Pathology

The skin specimen showed no organisms.

The appearance raised the question of pyoderma gangrenosum. Islands of haemopoetic tissue were also present in the dermis and subcutis. These were characterised by prominent megakaryocytes and early myeloid elements and were consistent with myelofibrosis.

Over the next week Mrs. G.H.'s condition improved slightly but she continued to spike temperatures from day to day. However, by the 23rd April her platelet count had risen to $771 \times 10^9/1$, other haematological indices being WCC 14.1, Hb 10.9. By 24th April, platelets $-1000 \times 10^9/1$. She was commenced on hydroxyurea in the hope that this would reduce platelet levels and so prevent any thromboembolic episodes. By 30th April her platelet count had fallen to $200 \times 10^9/1$ but her WCC had also fallen to 1.0, and so when I left the unit Mrs. G.H.'s prognosis was still very much in the balance.
DISCUSSION

Myelofibrosis is a chronic myeloproliferative disorder, characterized by (1) splenomegaly, (2) immature granulocytes and erythroblasts in the blood, (3) distorted and teardrop-shaped red cells, and (4) some degree of marrow fibrosis. The disorder was originally described by Heuck in 1879.

It is a condition of the elderly - most patients being in the sixth and seventh decades of life when the disease is diagnosed. Males are a little more frequently affected than females. Approximately one third of patients are asymptomatic at the time of diagnosis. They are either discovered by the abnormalities observed in the blood or by the finding of a large spleen on physical examination.

Most patients have symptoms and signs of anaemia, such as weakness, fatigue, exertional dyspnoea, palpitations or pallor. Patients may complain of a dragging sensation in the left upper abdomen or of early satiety resulting from the pressure of the enlarged spleen on the gastric outlet. Splenic infarction with severe left upper quadrant pain may occur in occasional patients. Petechiae or ecchymoses secondary to thrombocytopenia, qualitative platelet defects, or disseminated intravascular coagulation are often present.

Splenomegaly is present in all patients, sometimes extending into the pelvis. Splenomegaly is due to extramedullary haemopoiesis in the spleen (i.e. myeloid metaplasia). Liver and other organs also exhibit myeloid metaplasia and hepatomegaly is a common feature.

The blood picture shows anaemia, polychromasia, anisocytosis, and
poikilocytosis with "tear-drop" red cells. A proportion of patients become folate deficient, with a raised MCV and macrocytes. Nucleated red cells are often present in association with immature granulocytes (leuco-erythroblastic reaction). The number of granulocytes may be low, raised or normal and the platelet count is usually increased at the time of diagnosis. Bone marrow examination usually yields a "dry" tap.

The course of myelofibrosis is variable. The median survival is usually given as approximately five years but it is not uncommon for patients to live 10–20 years after diagnosis. The usual cause of death is progressive anaemia which becomes refractory to blood transfusion. Approximately 20 per cent of cases terminate in acute myeloblastic leukaemia. There is a sudden worsening of the anaemia and a marked increase in the number of blast cells in the peripheral blood.

There is no definitive treatment for patients with myelofibrosis. They should be carefully followed and complications such as iron or folate deficiency corrected. If they become symptomatic due to progressive anaemia, they should be started on a blood transfusion programme. Splenic irradiation and chemotherapy have been used in an attempt to reduce the size of the spleen but results have been equivocal. Alternatively, hydroxyurea can be used to reduce spleen size: starting with a low dose, e.g. 500mg and gradually increasing it keeping a careful watch on white cell and platelet counts.

The place of splenectomy is controversial but it is generally accepted that it should be reserved for those cases in which there is gross splenomegaly causing severe symptoms, as in the case described above.
Clinical complications of splenectomy include post-operative bleeding, subphrenic abscess, thrombo-embolic disease and post-operative infection.

In 1952, King and Shumaker first suggested that in children, there may be an increased susceptibility to infection after splenectomy. They also asked why serious infection apparently tends to follow splenectomy performed in infancy and not in patients splenectomized at an older age. However, they noticed that there was some evidence in the literature tending to show that splenic activity varies with age. Gross, in 1919, studied 111 spleens obtained at post-mortem and from the many anatomical and pathological changes noted from birth to old age concluded that the spleen probably has its greatest functional activity very early in life.

Since the first study in 1952, many cases of post splenectomy sepsis have been reported, predominantly in children but also in adults. The syndrome of post-splenectomy sepsis is now a well-recognised clinical entity predominantly caused by streptococcus pneumoniae. In decreasing order of frequency Neisseria meningitidis, E. Coli, Haemophilus influenzae, Staphaureus, Streptococcus and Pseudomonas have been implicated. Numerous case reports have further documented that the risk of development of overwhelming sepsis is lifelong with a mortality of 50 to 90 per cent (Dickerman, 1981). Prevention of overwhelming post splenectomy infection by long-term antibiotic administration and vaccine prophylaxis, as well as by surgical preservation of the injured spleen, has been the focus of recent attention.

Splenic salvage is technically more difficult and time-consuming than splenectomy and may increase the patient morbidity and mortality.
The philosophy of aggressive splenic preservation is based on the presumption that the risk of this procedure to the patient is less than the risk of fulminant sepsis developing after splenectomy. There have been several attempts to study the incidence and estimate the relative risk of developing overwhelming postsplenectomy sepsis.

In 1962, Horan and Colebatch estimated that life threatening or fatal sepsis would develop in 5.6 to 8.7 per cent of asplenic children. In their own series of 142 paediatric patients, there were five lethal cases of overwhelming infection after splenectomy.

Only recently has postsplenectomy infection been studied in adults. In 256 asplenic adults, O'Neal and McDonald (1981) found a 2.7 per cent incidence of fatal infection after a mean follow-up of 3.8 years. This rate of lethal sepsis was estimated to be 540 times greater than that in the general population. However, in 1985, Chaikof and McCabe published a study of 776 patients who underwent splenectomy between 1962 and 1972. Follow-up information was obtained on 637 patients (82 per cent) including 584 adults and 53 children. There was a total of 4,837 person-years of follow-up with a mean observation interval of 8.4 years. Four cases of fatal overwhelming postsplenectomy infection were identified. The incidence of fatal overwhelming post-splenectomy infection in their paediatric population was 3.77 per cent: the incidence in their adult population being 0.34 per cent. They concluded that the aggressive approach to splenic preservation in the adult should be tempered.

And thus the risk of developing overwhelming post-splenectomy infection in adults is still questionable, in the light of the widely differing results of the studies carried out by O'Neal and McDonald, and Chaikof.
and McCabe. However, it is probably better to err on the side of caution and splenorrhaphy, where possible, is probably more justifiable than splenectomy. The question of splenectomy versus splenorrhaphy is probably most important after trauma to the spleen and not of great consideration in elective splenectomy for gross splenomegaly as in the case I describe above. In Mrs. G.H.'s situation, splenectomy appeared to be the only choice.

REFERENCES


Textbooks

CASE FOUR
MR. F.S. (07.08.17)

Mr. F.S., a 69 year old retired security officer, presented to his G.P. at the end of December 1986 complaining of severe stomach cramps after eating a Chinese meal. On examination the G.P. found him to be very pale and tired. He also noted a large hard left iliac fossa mass. Investigation revealed a haemoglobin of 10.6g/dl and so he was commenced on ferrous gluconate 200mg T.i.d. He was referred to the physicians for a medical opinion.

On further questioning, Mr. F.S. admitted to having had vague lower abdominal pain for the previous three months, and he had also noticed some bleeding per rectum two months previously, but as he could feel some haemorrhoids, he had dismissed the bleeding. There had been no change in his appetite or his weight recently. Further gastrointestinal symptoms were absent.

Past medical history includes a small degree of chronic obstructive air ways disease, which necessitated one admission to hospital 17 years ago, when he was commenced on a ventolin inhaler which he has used ever since.

He lives with his wife who is disabled by respiratory disease. He has three children, including a daughter who lives in Edinburgh and who is very supportive.

On examination He is a pleasant but extremely anxious man with no evidence of finger clubbing or lymphadenopathy. He is slightly pale.

Cardiovascular system

Pulse rate 100/min regular
BP 170/95
otherwise NAD
Respiratory system
chest clear

Gastrointestinal system
Examination of the abdomen revealed a 7cm firm tender mass in the left iliac fossa. Rectal examination showed a small amount of soft faeces which was negative for occult blood. A mass could not be palpated rectally.

Central nervous system
Grossly intact

Investigations:--

Haematology
Hb 10.7, WCC 10.4, RBC 4.85
Hct 0.36 MCV 74.2 MCH 22.1
MCHC 29.7 platelets 200
Blood film - microcytic hypochromic cells occasional burr cells/elliptocytes
ESR 14mm/hr

Clinical chemistry
Urea 4.5 Na 138 K 3.3
ALT 11 Bilirubin 8 Alkaline phosphatase 72 gamma GT 11 Total protein 66 Albumin 39
Revealed "a few small circular shadows in the lung fields and these are in keeping with small intrapulmonary metastases. Cardiac size normal."

ECG
NAD

Barium enema
revealed a large annular carcinoma proximal to the rectosigmoid junction.

On 17th February 1987, Mr. F.S. was seen by the surgeons and on 19th February he was taken to theatre.
Operative findings and procedure
At laparotomy a large tumour was present at the recto-sigmoid junction. It was penetrating into perirectal tissues posteriorly and was invading the posterior wall of the bladder anteriorly. The liver was clear of metastases and regardless of the chest x-ray finding of pulmonary metastases it was decided, in view of the size of the lesion, to proceed with an anterior restorative resection of the rectum, and to prevent severe local complications.

On removal of the rectum it was seen that the left ureter had been accidentally divided: the continuity was restored. Partial cystectomy was also carried out because of invasion.

Pathology
Moderately differentiated adenocarcinoma with lymph node metastases (Duke's Stage C).

Post-operative progress
On the first post-operative day Mr. F.S.'s respiratory rate fell to 5/min - a reasonable respiratory rate was restored by the administration of 0.8mg naloxone.

By the fourth post-operative day he was vomiting large amounts of bile and so a nasogastric tube was passed and an intravenous infusion started.

Two days later the NGT was removed. By this time the wound had started to leak sero-sanguinous fluid.

On 3rd March the skin sutures were removed and this resulted in dehiscence
of the wound. That day he was taken to theatre for repair of his wound at which time he was found to be in acute retention with a bladder at the level of the umbilicus. And so he was catheterised.

On the 9th March a catheter specimen of urine revealed a moderate number of pus cells and many Pseudomonas aeruginosa. This was treated with chlorhexidene bladder washouts and the catheter was removed.

By the 12th March he developed urinary retention yet again and so he was recatheterised.

On the 17th March an intravenous urogram was performed. It revealed that the right kidney and right ureter were normal. A stone was present in the upper calyx of the left kidney. The left kidney was functioning but the pelvi-calyceal system was dilated. The urogram also demonstrated a partial obstruction of the left ureter distally (probably at the site of the surgical transection). There was some irregularity of the left upper wall of the bladder and the prostate was calcified.

On the 19th March 1987 he was transferred to the urologist for trans-urethral resection of prostate which alleviated his retention. He was later discharged.
DISCUSSION

I have chosen the case of this very unfortunate man because it highlights a few of the many complications which may occur peri- and post-operatively.

I have decided to look particularly at iatrogenic utereral damage.

The ureters are at risk in every abdominal operation, most especially in those involving the pelvic organs. Sometimes the disease process actually involves the ureter, making damage inevitable; in other cases the disease distorts the normal anatomy thus putting the ureter at risk by displacing it; and in yet other cases, peroperative difficulty such as heavy bleeding, poor light, inadequate exposure or obesity can lead to inadvertent injury. Gynaecologists, urologists, rectal surgeons and vascular surgeons all face the problem of occasionally damaging ureters.

Congenital anomalies pose a constant challenge, as they inevitably increase the possibility of damage as a result of capricious and unpredictable variations in the normal anatomy.

The ureters are particularly at risk of damage in gynaecological surgery, the incidence of such injury, when in the hands of a competent gynaecologist, and with modern techniques, probably being less than 0.5 - 1.0% of all pelvic operations (R.F. Mattingley et al 1978) possibly rising to nearer 2% with radical hysterectomy (M.A. Macasaet et al 1976). The ureters may be included in ligatures; they may be crushed in clamps and subsequently undergo necrosis; or they may simply be partially or completely divided.
In general surgery the ureters are most at risk in abdomino-perineal resection of the rectum, especially if the growth is extensive. In diverticulitis, especially if there is a pericolic abscess, and in Crohn's disease, the normal tissue planes may be obliterated and the ureter may be concealed in inflammatory reactive tissue.

If the ureter is injured during operation and the surgeon is aware of this, he should repair it at the time of operation. The choice of procedure for high injuries is end-to-end anastomosis, and for low injuries ureteric reimplantation with or without a psoas hitch or Boari flap. Early recognition and prompt repair of the injury make for a quick recovery (J.T. Flynn et al 1979).

If a low ureteric injury is found it is usually possible to reimplant the ureter into the bladder. Care should be taken to produce a submucosal course of at least 2cm to prevent reflux.

The most important point is to ensure that the ureteric reimplantation is done without tension. This is best achieved by suturing the apex of the bladder to the psoas muscle on that side (the psoas hitch).

After radiotherapy or in the presence of dense adhesions, it may not be possible to elevate the peritoneum sufficiently well to perform the ureteric reconstruction extraperitoneally. In these circumstances a transperitoneal Boari-Ockerblad flap (N.F. Ockerblad, 1947) procedure may be preferable. Indeed some surgeons prefer the Boari-Ockerblad flap procedure to the ureteric reimplantation.

With psoas hitch, for all low ureteric injury (W.G. Bowsher et al 1982). A tongue of full-thickness bladder is rolled into a tube and joined
to the lower end of the ureter over a splint. The defect in the bladder is then closed. This procedure has the advantage of using well-vascularized bladder wall to bridge the gap to the lower end of the ureter (W.F. Hendry, 1977).

Ureteric injuries may present after operation in an urgent or non-urgent manner. If the damaged ureter continues to drain urine into the extraperitoneal tissues, pelvic cellulitis may follow, if it drains into the peritoneal cavity, urinary peritonitis will be produced; or if both ureters are injured anuria may result. On the other hand, ligation of one ureter may produce little more than transient loin pain. Probably the most commonly delayed presentation is by the development of a urinary fistula, presenting usually per vaginam, or occasionally through the wound.

When there is evidence of ureteric damage after operation, the precise nature of the injury should be defined before deciding on the best approach to repair. The minimum investigations are urine culture, blood urea and electrolytes and a high-dose intravenous urogram is essential. To confirm the exact location of the complication cystoscopy and ascending ureterogram should be performed.

In some cases catheter drainage may be all that is required to allow a small fistula to heal. More often, however, some form of operative intervention is necessary. If a ureter is completely obstructed, reconstruction or reimplantation should be undertaken within three weeks if worth-while kidney function is to be preserved.

The ureter is either repaired or reimplanted, with or without a psoas hitch or Boari-Ockerblad flap. If a considerable defect has to be bridged, it may occasionally be necessary to do a high transuretero-
ureterostomy. The latter operation is seldom performed, however, because of the risk of injuring the other ureter. Good results have, of course, been obtained by some (Smith and Smith, 1975), but not always by others (Ehrlich and Skinner, 1975). Very rarely it may be necessary to interpose an isolated loop of ileum between ureter and bladder. Seldom, if ever, should the need arise for tying off a ureter, nephrectomy, or ureterocolic anastomosis.

So we can conclude that occasional injury to the ureters appears to be almost inevitable: during surgery in the presence of difficult congenital anomalies, or with distortion or involvement of the tissues by disease; when technical difficulty is encountered; or as a result of accidental or malicious wounding. If the injury is recognised at the time primary repair will give the best results. If the damage only becomes evident later, extreme care must be exercised to locate the injury or injuries accurately, and then to choose the optimum timing and surest technique for repair.

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CASE FIVE

There was no history of trauma with abortion or blood transfusion. She had been recently married and expectant.

There was no past medical history of importance. Her history, social and family history were all uneventful as was her systemic review.

On examination, she looked well but not diarrhoeic. Her tongue was furred, sticky dry and the vesiculation was 2-3. There was no

CASE FIVE
MRS. B.M. (29.01.60)

Mrs. B.M., a 27 year old nurse, was seen by the physicians, after emergency admission from her G.P. on 10th April 1987. She complained of a two week history of diarrhoea and abdominal pain. The abdominal pain preceeded the diarrhoea and was originally periumbilical. After a few days it moved down to both iliac fossae and suprapublic region. It was a constant pain made worse if she moved or coughed. There was no associated nausea or vomiting but her appetite had decreased and she had lost 16 lbs in two weeks. The diarrhoea started shortly after the abdominal pain; her bowels were initially opening 7-9 times/day and the stool was formed. By the second week of diarrhoea her bowels were opening about 4 times/day and the motion was watery and yellow/green in colour. There was no blood or slime in the motion. She also had pain on passing urine and the urine was concentrated.

There was no history of contact with animals or with infected persons. She had not recently travelled abroad.

There was no past medical history of significance. Drug history, social and family history were all unrevealing, as was systematic enquiry.

On examination she looked unwell but not distressed. Her tongue was furred, skin dry and she was pyrexial at 39.5°C. There was no finger clubbing or lymphadenopathy.

**CVS**
- Pulse 110/minute - regular
- BP 135/70
- Otherwise - NAD

**RS**
- Chest clear

**GIT**
- The abdomen was not distended and there
were no scars.  
No masses palpable  
No LKKS  
Vague generalized abdominal tenderness especially over the descending colon.  
Some guarding and rebound tenderness over the descending colon.  
Bowel sounds - few tinkling  

Mild haemorrhoids  
Non-tender. Rectum empty  
No masses  
FOB positive  

Grossly intact  
No joint, eye or skin symptoms or signs.  

Investigations  
Clinical chemistry  
U 4.4  Na 126  K 3.0  CO₂ 28  
Haematology  
Hb 12.7  WCC 4.6  
Urinalysis  
No pus cells, no organisms  
Stool sample + MSU  
No growth of enteric organisms  
No ova, no cysts, no parasites  
Serum virology titres  
Negative  
Blood cultures  
Negative  
Abdominal x-ray  
Erect and supine - revealed dilated transverse and descending colon with loss of haustral markings. Fluid levels in the small bowel. No free gas seen.  

She was commenced on metronidazole 500mg t.d.s. and cefotaxime 1g t.d.s. and an intravenous infusion was started.
On 12th April she was given 200mg i.v. hydrocortisone and regular maintenance doses. She was also started on salazopyrin 500mg every six hours. The abdominal x-ray was repeated, which showed the colon to have dilated to 7cm in diameter. Small bowel fluid levels were still present as was some thumb-printing. No free gas was present.

Urea - 2.6, Na - 126, K - 2.9
WCC - 6.7 - 70% neutrophils, 10% lymphocytes, 9% macrophages.

Sigmoidoscopy was attempted but this was difficult in view of the diarrhoea. It did manage to show some mucosal oedema but no haemorrhagic areas.

Later that day she was transferred to the surgeons.

On arrival she was pale, distressed and pyrexial at 39.5°C with a tachycardia of 110/minute and a BP of 120/70. She was also clinically dehydrated with loss of skin turgor. She had a distended abdomen and there was generalized tenderness, rebound tenderness and guarding. Bowel sounds were absent.

U - 3.1, Na - 127 K 3.2, CO₂ 28
Hb - 11.6, WCC 7.5, platelets 327.

She was observed carefully, given analgesia and an intravenous infusion was recommenced. She was given i.v. prednisolone 20mg t.d.s.

The following day she was apyrexial and somewhat brighter. But by the 14th April she was experiencing a lot of pain only settling when the pethidine was changed to three hourly omnipon. There was no improvement on the abdominal x-ray.
And so on the 15th April she was taken to theatre.

Operative findings and procedure
A left paramedian incision was made. There was no free fluid in the abdomen. The entire large bowel from the caecum to the junction of the upper and middle third of the sigmoid colon was markedly distended and showed obvious evidence of severe inflammatory bowel disease.

Caecum, ascending colon, transverse colon, descending colon and the proximal half of the sigmoid colon were resected together with about 10-15 cms of healthy terminal lieum. During mobilization of the grossly inflamed colon spontaneous perforation occurred at three different sites and there was gross peritoneal soiling. It appeared that a small perforation may have occurred in the region of the splenic flexure preoperatively as the colon was densely adherent to the abdominal wall in this situation and there appeared to be a localized collection of pus. A swab of this was sent for bacteriology which subsequently grew penicillin sensitive alpha-haemolytic streptococci.

The peritoneal cavity was thoroughly lavaged with 6 litres of tetracycline/saline solution. A standard ileostomy was made.

The patient was then transferred to the intensive care unit in view of the gross peritoneal faecal soiling.

Pathology
Macroskopically the colon showed mural thickening throughout most of its length and there was submucosal oedema and focal ulceration which varied from small punctate superficial ulcers to deeper, more
linear ulcers. The mucosa appeared oedematous and small inflammatory polyps were seen at the ulcer margins. These changes were widespread throughout the colon.

Microscopically the specimen showed inflammation with ulceration and focal perforation. There were also areas of ischaemia - presumably a part of the "toxic dilation". The specimen was not entirely typical of ulcerative colitis.

Post-operative progress

On 16th April she returned to the surgical ward at which time she was being monitored with a CVP line and indwelling catheter to measure urine output, in addition to regular observations of temperature, pulse and B.P. Her pain was being controlled by a morphine infusion and she was also receiving intravenous ampicillin, gentamycin and metronidazole. Her steroids were gradually being tailed off.

On 17th April her condition was greatly improved but by 20th April she complained of breathlessness. Her sputum was purulent. On examination there was decreased expansion and reduced air entry at the left lung base, which was also dull to percussion. Chest x-ray revealed some collapse and consolidation of the left lower lobe associated with a small pleural effusion. Sputum culture grew Staphylococcus aureus and so she was commenced on flucloxacillin.

By 26th April her chest was clear, she was eating and drinking and her main problem was pain. She was being treated with regular paracetamol, dihydrocodeine and diazepam but the opiates were gradually being reduced.

The next two weeks were uneventful. Her strength gradually returned,
she became pain-free and she left hospital on 10th May 1987.
DISCUSSION

Colonic dilatation during a severe acute attack of ulcerative colitis usually occurs at the initial presentation of the disease and is often referred to as toxic megacolon. Toxic dilatation of the colon may also occur in Crohn's disease and occasionally in amoebiasis, ischaemic colitis, pseudomembranous colitis, obstructing tumours and Hirschsprung's disease.

Although colonic dilatation in acute colitis may respond to medical therapy, toxic megacolon requires surgical intervention. This bowel tends to fall apart with the gentlest handling at operation and even when surgery is performed prior to perforation, there is an increased risk of postoperative peritonitis and abscess.

Perforation of the colon is not an infrequent finding when operating during severe attacks of colitis. This may be a frank opening from the bowel into the general peritoneal cavity leading to a diffuse faecal peritonitis, or alternatively it may be found that the part of the colon penetrated is firmly adherent to the anterior or lateral abdominal wall or to an adjacent viscus, so that the hole in the bowel is in effect sealed off and there is no general peritoneal contamination. With regard to open perforations, it is obvious that, whatever is done surgically at operation, post operative peritonitis will be one of the major hazards and will require management including peritoneal irrigation at the conclusion of the laparotomy and massive systemic antibiotic therapy during and after the operation. Probably the best operative plan in these patients is to press on with ileostomy and colectomy, making an effort to control further leakage of faecal material into the peritoneal cavity by the judicious use of the sucker inserted into the bowel early in the operation.
In dealing with sealed perforations, however, if a colectomy is performed, this will result in unsealing the perforation with consequent contamination of the peritoneum. This can be avoided by making an ileostomy alone or caecostomy as suggested by Klein et al (1960). Truelove et al (1965) employed a double-barrelled ileostomy combined with installation of steroid solutions into the colon in 14 emergency operations for colitis and had four failures (as indicated by operative death or by need for a subsequent emergency colectomy). Turnbull et al (1970) favoured a loop ileostomy combined with a decompressing transverse (and possibly sigmoid) colostomy. In 26 patients treated by this method for severe attacks of colitis, there was only one operative death. Of the 25 survivors, 12 patients required emergency colectomy during the 6 months after operation and 13 patients proceeded to elective colectomy.

Extensive peritonitis is still associated with significant morbidity and mortality rates. The concept of mechanically cleansing the peritoneal cavity using lavage is not new, being first advocated for use at the time of operation by Nolan in 1893. In 1957, Burnett et al reported the advantages of adding antibiotics to the operative lavage, and McKenna et al in 1970 halved the mortality rate with the use of continuous antibiotic lavage. Stephens and Loewenthal (1979) treated 27 patients by continuous postoperative peritoneal lavage with 6 deaths, and mention that in a former series of 68 patients treated by perioperative lavage alone, combined with a course of postoperative systemic antibiotics, there were 33 deaths.

Although these new techniques have been introduced peritonitis still remains a significant cause of morbidity and mortality.
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I should like to thank Mr. I.B. MacLeod for allowing me to discuss his patients.