Weber–Christian disease producing splenic vein occlusion and bleeding gastric varices: successful treatment with sclerotherapy

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Summary: A 48 year old woman with intra-abdominal Weber–Christian disease presented with bleeding gastric varices and evidence of splenic vein occlusion. We describe the problems encountered in making this diagnosis and subsequent treatment.

Introduction

Upper gastrointestinal haemorrhage secondary to isolated gastric varices is an unusual cause of haematemesis. Diagnosis may be difficult in the absence of oesophageal varices and where there is no evidence of liver disease. In such cases the most likely diagnosis is splenic vein occlusion, a collateral circulation being established through the short gastric veins. In the majority of cases splenic vein occlusion is secondary to pancreatitis or pancreatic carcinoma but occasionally other causes are found. We describe a case which was eventually diagnosed as being due to an intra-abdominal manifestation of Weber-Christian disease.

Case report

A 48 year old woman was admitted to a nearby hospital in March 1984 following a large haematemesis. She had been well until 1979 when she underwent a cholecystectomy for gallstones. Post-operatively she developed jaundice, but at a further laparotomy no stones were found in the common bile duct, liver biopsy was normal, as were markers for viral hepatitis. The episode of jaundice was thought to be due to a post-transfusion hepatitis and she made a full recovery.

Two years later (1981) she was noted to be hypertensive and commenced on bendrofluazide and metoprolol, which she took up until January 1984 when she was admitted to hospital with a 4-week history of a systemic lupus erythematosus (SLE)-like illness characterized by fever, widespread macular rash, pericardial rub and pleural effusions. Extensive investigations at this time did not confirm this diagnosis (normal immunoglobulins, normal platelet count, negative antinuclear antibody, negative DNA binding). Renal function was found to be impaired (creatinine 201 μmol/l compared to 110 μmol/l two years previously). Intravenous and retrograde urograms showed a normal right kidney and ureteric tract but a non-functioning left kidney and blind ending ureter. ESR was raised (80 mm in first hour). Liver function tests were abnormal (alkaline phosphatase over 500 IU/l, normal 20–120 IU/l and aspartate transaminase 120 IU/l, normal <45 IU/l). However, the metoprolol was stopped and the patient rapidly recovered on a combination of loop diuretics and vasodilators. Liver function tests returned to normal.

During her recovery she had a small haematemesis which was not thought to be significant. The haematemesis in March 1984 was therefore her second bleed, and at this time a mass in the left hypochondrium thought to be the spleen was palpable, although it appeared to be of normal size on isotope liver scan, which was also otherwise normal. Gastroscopy at the same time was reported as normal.

Twelve months later, in May 1985, she again presented with a further large haematemesis followed by two more bleeds within the next two months. Three gastroscopies, all within 24 hours, failed to reveal any lesion, and it was only when a barium meal was requested that the diagnosis of gastric varices became apparent (Figure 1).
On transfer to the medical unit at Middlesbrough General Hospital the diagnosis was confirmed by gastroscopy, using both forward and side-viewing endoscopes (F2, DUO-XL2, Fuginon). Prominent gastric folds within the fundus were noted to have 'cherry red spots' similar to those seen on the surface of oesophageal varices. The oesophagus was normal. Despite commencing injection sclerotherapy, three further haematemeses ensued. Investigation by ultrasound scan showed occlusion of the splenic vein with a patent portal vein which was confirmed by venous phase coeliac angiography (Figure 2). CT scan was normal. Repeat liver biopsy was normal. The patient was referred for splenectomy but at operation in June 1985 the whole of the upper abdomen was encased in a thick fibrous pannus covering the liver, spleen, stomach and small intestine. It proved impossible to mobilise the stomach without serious risk of perforation or uncontrollable bleeding and therefore splenectomy was not performed.

Progress

Following laparotomy the patient made an uneventful recovery. Further sclerotherapy was carried out until the varices appeared obliterated. The patient has subsequently had no further haematemeses and repeat gastroscopies up until May 1988 have not shown any evidence of recurrence. Unfortunately, she has required further admission to hospital for worsening renal function, and repeat retrograde urogram showed right
ureteric obstruction confirmed as being due to retroperitoneal fibrosis at an exploratory operation. Nephrostomy was unable to preserve renal function and the patient is now managed by regular haemodialysis. It is now 3 years since the patient last had a haematemesis.

**Histology**

The biopsy of the fibrous tissue was reviewed by two independent histopathologists. The reports indicated that the biopsy contained degenerating adipose tissue, fibrous septa and inflammatory cells similar to that seen in Weber–Christian disease (Figures 3 and 4).

**Discussion**

The cause of this patient’s intra-abdominal fibrosis remained undiagnosed for some time. Beta-blocker therapy seemed unlikely because of the distribution and appearance of the fibrous tissue and inability to ‘strip’ this at operation. The histological appearance was unlike sclerosing peritonitis. Although retroperitoneal fibrosis has been reported anecdotally in association with beta-blocker therapy, there is insufficient evidence to support this as cause and effect. Nevertheless, it is possible for retroperitoneal fibrosis to cause localized portal hypertension. It must also be remembered that the patient had undergone surgery in 1979 when the abdominal cavity was normal. However, the nature and extent of the intra-abdominal fibrosis was so extreme that it seemed most unlikely to be a result of previous surgery. There was also no evidence of ‘talc’ peritonitis on histological grounds. Finally a diagnosis of Weber-Christian disease was made after 2 independent histologists separately described the classical changes of fat inflammation in intra-abdominal fibrous tissue.

Weber–Christian disease is an inflammatory disease of fat first described in 1882, and extensively reviewed by Panush et al. It is predominantly a disease of middle-aged females presenting with inflamed subcutaneous nodules but may also present with fever, arthralgia and myalgia. Although a variety of abnormalities may be found on routine screening (elevated ESR, anaemia and thrombocytopenia), the diagnosis is usually made by histological examination of the skin lesions. However, a visceral form involving the fatty tissues of the gastrointestinal tract and occasionally the retroperitoneal structures may present without dermal involvement. A more diffuse ‘systemic’ form has also been reported that involves organs of the thoracic cavity and may explain the pericardial friction rub observed in the patient. Mesenteric panniculitis may cause abdominal pain and in some cases can produce bowel obstruction. Retroperitoneal involvement may occur and involve the kidneys. This may explain the renal failure secondary to increasing retroperitoneal fibrosis and raised alkaline phosphatase.

Weber–Christian disease may also be associated with other pathology, such as alpha-1-antitrypsin deficiency, scleroderma, fungal infections and amyloidosis, but none of these was present in this patient. Often no cause is found, but the patient

![Figure 3](http://pmj.bmj.com/)

**Figure 3** Histology (H and E × 40) of biopsy showing degenerative adipose tissue with fibrous tissue, forming septa.
suffers life-threatening complications such as in this case where splenic vein occlusion resulted in multiple episodes of upper gastrointestinal haemorrhage from gastric varices which required a total of 28 units of blood.

In addition to this problem there now remains the long-term problem of renal failure secondary to ureteric obstruction. Both conditions are probably related to the fibrosis associated with Weber-Christian disease and the prognosis for improvement in her renal function is poor. Long term haemodialysis seems to be inevitable.

Other aspects of the case are worthy of note. Firstly, the enthusiasm for early endoscopy has perhaps left us over-reliant on this technique as difficulty may be experienced in diagnosing upper gastrointestinal bleeding due to gastric varices. In retrospect, a barium meal should have been performed earlier. Secondly, we were puzzled by the discrepancy between the clinical finding of splenomegaly and the normal ultrasound and isotope scan which showed the spleen to be of normal size. At operation it appeared that this discrepancy was due to thick fibrous pannus covering the spleen. Thirdly, although splenectomy was the treatment of choice in this patient it is reassuring to have a second line of management in such cases, namely injection sclerotherapy. Others have also reported success with this procedure although benefits have previously not been documented for sclerotherapy of gastric varices.

In conclusion, we feel that although the presentation of this patient was not typical of Weber-Christian disease, histological evidence is sufficient to label this as the cause of the massive peritoneal and retroperitoneal fibrosis. As a consequence, the patient developed ureteric obstruction and splenic vein occlusion resulting in bleeding gastric varices which have to date been treated successfully by injection sclerotherapy. We feel these should now be added to the list of complications associated with the intra-abdominal manifestations of Weber-Christian disease.

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References


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