previous subarachnoid hæmorrhage nor previous meningitis can be incriminated as the obstructing agent. Another puzzling problem is the mechanism of hydrocephalus in the presence of apparently normal CSF pressure: a solution has been postulated by Hakim & Adams (1965) who, after consideration of Pascal's law, believe that ventricular size should always be considered in evaluating the significance of CSF pressure.

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Multifocal Fibrosclerosis

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I W, aged 49. Housewife

History: First seen at Hammersmith Hospital in December 1968, complaining of a painful tender goitre. A diagnosis of thyroiditis was made but despite a trial of cortisone (in low dosage) the gland continued to enlarge and subsequently it became painless and extremely hard. In April 1969 she became hypothyroid and required thyroxine replacement therapy.

Three months later, there was a new development – painless obstructive jaundice with pale stools, dark urine and pruritus which persisted until a laparotomy was carried out in August 1969.

Laparotomy findings: The obstruction lay at the junction of the left and right hepatic ducts which were surrounded by hard fibrous tissue that was also found in the gall-bladder bed. An operative cholangiogram showed normal gall-bladder, cystic and common bile ducts, with dye draining freely into the duodenum, but the right intrahepatic tree was markedly dilated behind an obstruction at the origin of the common hepatic duct. There was no filling of the left hepatic duct system (Fig 1). Biopsy of the obstructive lesion showed fibrous inflammatory tissue consistent with sclerosing cholangitis. Following surgery, jaundice persisted and in November 1969 she was readmitted for further assessment.

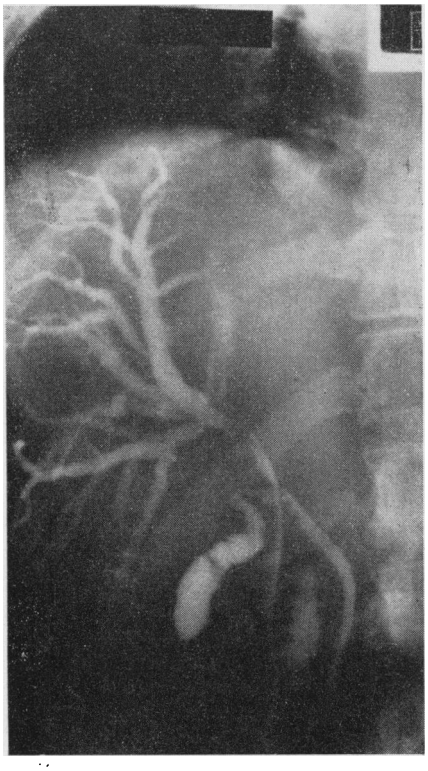

Fig 1 *Operative cholangiogram showing markedly dilated right intrahepatic tree with obstruction at origin of common hepatic duct*

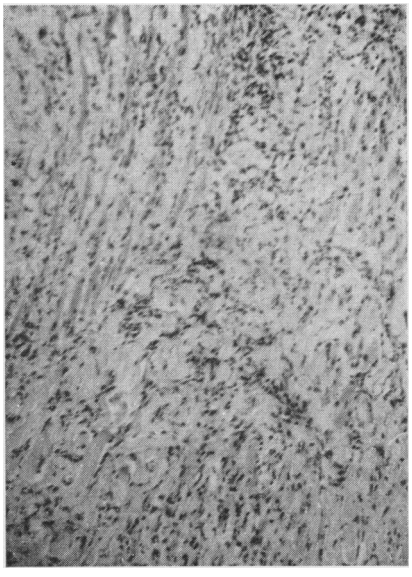

Fig 2 *Drill biopsy of thyroid showing replacement of all normal thyroid tissue by fibrous and inflammatory tissue which also shows invasion of surrounding strap muscles in the neck*

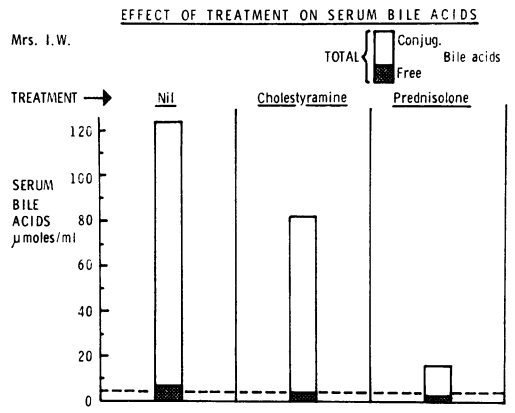

Fig 3 *Serum bile acids: values before treatment, after cholestyramine and after prednisolone therapy. Broken line indicates upper limit of normal for total serum bile acids*

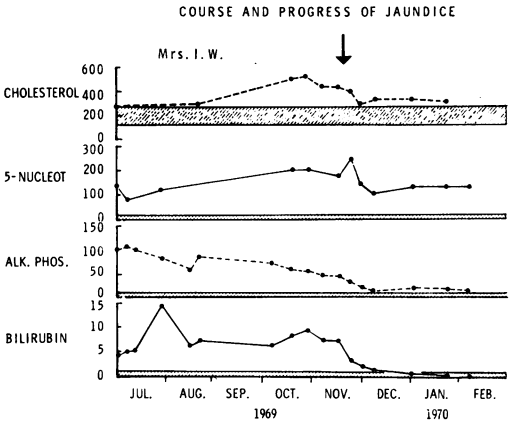

Fig 4 *Effect of prednisolone (40 mg/day) on liver function tests in patient I W. Start of corticosteroid treatment is indicated by heavy arrow*

On examination: Marked icterus with scratch marks due to pruritus. The thyroid remained diffusely enlarged and unusually hard. Respiratory, cardiovascular and central nervous systems – normal. Firm enlarged liver palpable 3 cm below costal margin. Spleen and kidneys not enlarged by palpation.

Investigations: Hæmoglobin, white cell count, blood urea, serum electrolytes, calcium and phosphate – all normal.

Liver function: Serum albumin 3·1, globulin 3·8 g/100 ml. Serum bilirubin 8·8 mg/100 ml. Serum alkaline phosphatase 59 K-A units. Serum cholesterol 530 mg/100 ml. Serum 5 nucleotidase 204 i.u. (normal 2–17). Serum lactic dehydrogenase 200 i.u. Glutamic oxaloacetic transaminase 46 i.u. Serum bile acids (Dr G Neale and Dr D Panvelliwalla) 124 μmol/litre. Urine: bilirubin present, no urobilinogen.

Drill biopsy of thyroid, two occasions (Professor Russell Fraser): Normal thyroid completely replaced by fibrous inflammatory tissue with invasion of surrounding strap muscles of the neck by inflammatory cells – the histological features of Riedel's thyroiditis (Fig 2).

IVP: Congenital rotation of right kidney, which also showed hydronephrosis and hydroureter.

Retrograde pyelogram showed that the hydroureter was due to obstruction in the lower third. No calculus demonstrated. Appearances were consistent with retroperitoneal fibrosis.

Treatment: Cholestyramine, 12 g/day by mouth reduced pruritus minimally – serum bile acids 82 μmol/litre. Oral prednisolone, 40 mg/day was started in November 1969, with rapid resolution of jaundice and pruritus. Serum bilirubin returned to normal with marked improvement in serum alkaline phosphatase, cholesterol and serum bile acids, although three months later serum 5 nucleotidase was still elevated (Figs 3 & 4). However, at this stage a repeat intravenous cholangiogram showed a normal biliary tree, and IVP showed disappearance of hydronephrotic changes in the right kidney.

Discussion

The findings in this case of Riedel's thyroiditis, sclerosing cholangitis and retroperitoneal fibrosis constitute a syndrome which was first reported by Bartholomew *et al.* in 1963. Comings *et al.* (1967), describing two further such cases, drew attention to the widespread distribution of fibrous tissue in this condition which also may include pseudotumour of the orbit and mediastinal fibrosis: they called the syndrome 'multifocal fibrosclerosis'.

The etiology of this condition is unknown. In the present case, in contrast to those described by Comings *et al.* (1967), there was no family history of similar illnesses. There had been no previous ingestion of methysergide. In the two previous accounts of this syndrome, improvement was noted with corticosteroid therapy which has also been suggested as a treatment for the early inflammatory stage of retroperitoneal fibrosis (Haché *et al.* 1962).

Summary: A further example of multifocal fibrosclerosis is reported, with inflammatory fibrous tissue diffusely affecting the thyroid, biliary tree and retroperitoneal tissue. In a four-month follow-up period, the patient has responded well to treatment with corticosteroids.

Acknowledgment: We gratefully acknowledge the co-operation and assistance of the Departments of Radiology and Pathology, Royal Postgraduate Medical School.

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Mr R W Raven said that this was a very instructive case presentation showing fibrosclerosis in multiple tissues. It highlighted the advisability of investigating the biliary and renal systems in patients with Riedel's thyroiditis, for perhaps more cases would then be shown with these correlations.

Mr G Blackburn said that the uniform and tender enlargement of the thyroid initially, with the subsequent development of myxœdema, did not suggest Riedel's thyroiditis. Furthermore, resection of tne isthmus would have provided better histological evidence than a needle biopsy.

Mr G Qvist said that the cause of the obstructed hepatic ducts must have been difficult to determine at operation and Mr Selwyn Taylor was to be congratulated on avoiding an extensive operation to relieve the obstruction, relying rather on simple drainage with subsequent resolution of obstruction by drug therapy.

Jaundice due to Hepatic Artery Aneurysm

Lionel Gracey FRCS
(Royal Free Hospital, London WC1)

Mrs M R, aged 59

Previous history: Severe rheumatoid arthritis since 1945 affecting hands, feet and knees. Between 1954 and 1961, synovectomy of left knee-joint and amputation of toes. 1964, incision of abscess swelling, left knee-joint. On steroids up to present. 1956, total hysterectomy for fibroids. 1960, thyroidectomy for thyrotoxicosis. 1964, tarsorrhaphy for persistent exophthalmos.

History: In May 1969 she began to suffer a colicky pain in right hypochondrium radiating round to back, and suggestive of biliary colic. After a week of this pain she became anorexic and jaundiced, with pale stools and dark urine. Shortly before admission on 1.6.69 she began to vomit.

On examination: Deeply jaundiced, ill-looking patient. Obvious rheumatoid deformity of hands and knees, brawny œdema of legs and feet. BP 200/110. Liver enlarged. Gall-bladder area markedly tender.

A diagnosis of probable gall-bladder empyema was made, and on day of admission the abdomen was explored through a right upper paramedian incision. Free bile in the peritoneal cavity and gross distension of gall-bladder, which contained purulent bile but no stones. Head of pancreas normal, distal common bile duct collapsed; but in the porta hepatis was a pulsatile mass, an aneurysm of the hepatic artery. Cholecystostomy was performed with a Malecot catheter left in the gall-bladder, and the wound was closed with drainage.

Progress: A cœliac-axis arteriogram was performed on 3.6.69 and revealed an aneurysm of the right hepatic artery. Later that day the patient collapsed with signs of severe hæmorrhage, 250 ml blood was aspirated from the stomach, and rupture of the aneurysm into the bile duct was diagnosed. Her condition was improved with intravenous transfusion of 6 pints (3,400 ml) of blood before definitive surgery for the aneurysm.

Operation (5.6.69): The aneurysm completely eroded the wall of the common hepatic duct. Proximal and distal ligature of the aneurysm produced no colour changes in the liver. Aneurysm therefore resected together with common hepatic duct. Cholecystectomy performed and distal stump of common bile duct ligated. Roux loop of jejunum anastomosed to right and left hepatic ducts in porta hepatis.

A transient biliary fistula developed, but the patient then made a good recovery and went home, well, on 23.8.69. All liver function tests normal up to present.

Comment

Clinical diagnosis of the condition of hepatic artery aneurysm is difficult, but the combination of obstructive jaundice and hæmobilia in the absence of trauma is suggestive. Arteriography is a most valuable aid to diagnosis, and especially precision of diagnosis.

Excision of the aneurysm is the treatment of choice. Graft replacement is not needed if there is good back bleeding and no apparent colour changes in the liver on ligation of the hepatic artery; colour changes are unlikely, as the aneurysm takes months to develop and collaterals will have formed, and particularly unlikely if the aneurysm affects the main hepatic artery proximal to the origin of the gastroduodenal. The bile duct may form part of the aneurysm wall and may require resection with the aneurysm. Replacement of the bile duct by the classical Roux loop technique is then effective.

Professor Harold Ellis said that the much commoner splenic aneurysms were frequently calcified and were thus visible on plain abdominal X-rays. He wondered if this was the case in hepatic aneurysms.

Mr Gracey replied that the condition was rare but plain X-ray had not revealed the aneurysm in reported cases.