cystic, and common bile ducts were described as "solid cords with a very small possible lumen, from which a tiny drop of bile was obtained. The ducts could not be dilated and the common bile duct appeared to end in a faint cord. The gall-bladder was smooth and shrunken." No further procedure was undertaken and his recovery was satisfactory, though he developed a biliary fistula, which closed spontaneously.

Subsequently the patient had an extensive medical history. He underwent mastoidectomy after otitis media and later developed maxillary osteomyelitis, which resolved with antibiotic treatment. When aged 3 his right eye was destroyed by an inflammatory mass. At 11 he developed inflammatory changes in the left eye. Despite treatment his sight was lost. He then fractured his skull in a road accident and underwent craniotomy for an extradural haematoma. In his early teens he suffered from spinal osteochondritis (Scheuermann's disease).

Though he maintained normal growth and development, he had persistent hepatosplenomegaly and abnormal liver function values. Itching became a great problem, which was not relieved by phenobarbitone prescribed for grand-mal convulsions after his craniotomy. In 1972 he developed xanthelasmata. His plasma cholesterol was 8.5 mmol/l (328 mg/100 ml) and triglyceride 0.72 mmol/l (64 mg/100 ml), and prominent alpha and beta bands were found on electrophoresis. He began treatment with cholestyramine with improvement of the xanthelasmata, itching, and blood lipid concentrations.

In 1975 he was complaining bitterly of itching and was unable to tolerate cholestyramine regularly. He had xanthelasmata and hepatosplenomegaly. Liver function values were: total bilirubin 22 μ mol/l (1.3 mg/100 ml), aspartate transaminase 78 U/l, alkaline phosphatase 31 KAU, albumin 44 g/l, globulin 34 g/l, and cholesterol 5.5 mmol/l (212 mg/100 ml). His serum was negative for hepatitis B antigen and gave negative or insignificant titres against routine antiviral, toxoplasma, and toxocaral antibodies. Intravenous cholangiography showed a good concentration of dye in the gall bladder.

In view of this and the pruritus a second laparotomy was performed. The operative cholangiogram (see figure) showed uniform narrowing of the ducts. Cholecystojejunostomy was performed. The liver was cirrhotic.

Operative cholangiogram showing uniformly narrowed common bile duct.

His progress was excellent, with complete loss of itching and a substantial fall in total bilirubin. Four months after operation the total bilirubin was 14 μ mol/l (0.8 mg/100 ml), aspartate transaminase 81 U/l, alkaline phosphatase 26 KAU, cholesterol 6.5 mmol/l (250 mg/100 ml), and triglyceride 0.70 mmol/l (62 mg/100 ml). He remained symptom free one year after operation.

Comment

Prolonged survival of patients with biliary atresia is unusual, though spontaneous reversal may occur.^{2,3} Krovetz³ described a patient surviving to 16 years, and she too had suffered pruritus and xanthelasmata. Recent views⁴ of biliary atresia as an inflammatory event, which may resolve like neonatal hepatitis, may explain such survival. The relation of biliary atresia to our patient's other illnesses is not clear.

The disappearance of itching after operation was gratifying. The fall in postoperative bilirubin levels suggests that, though there had been no complete obstruction, impedance to bile flow was much reduced. Neither cholestyramine nor phenobarbitone had completely relieved his pruritus. Though cirrhotic changes may regress after operation in infancy,⁵ our patient's prognosis must be guarded.

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Hypernephroma in two brothers

Hypernephroma (Grawitz tumour or adenocarcinoma of the kidney) is a relatively common tumour. Unlike tumours of the breast, stomach, or endometrium—which commonly show a familial pattern—hypernephroma has been reported in close relations only infrequently. This report concerns two brothers who came under our care.

Case reports

Case 1—A 53-year-old man was first seen in March 1974 with bilateral inguinal herniae and rectal bleeding. The findings on sigmoidoscopy and barium enema were normal. His piles were injected and the herniae repaired. He returned with further rectal bleeding in December 1974 and routine examination showed a large tumour mass in the region of the left kidney. Intravenous pyelogram showed expansion of the lower pole of the left kidney with distortion of the middle and lower calyces, consistent with a diagnosis of a renal tumour. At operation an 11-cm diameter hypernephroma was removed and the patient made an uneventful recovery. Postoperatively he complained of backache and pain down the left leg—symptoms which he had had for many years.

In September 1975, x-ray films of the lumbar spine and left leg were normal. Two months later he was injured in a road traffic accident and sustained a subcapital fracture of the left femur. Nevertheless, an x-ray film showed that the fracture had taken place through a metastasis and hip replacement was performed. At operation the upper end of the femur was infiltrated with tumour, which proved to be secondary hypernephroma. Pain in the region persisted postoperatively and he was given radiotherapy to the full level of tissue tolerance, 4125 rads in a 3-week period. His condition gradually deteriorated, and he died on 27 July 1976, 24 hours after his brother.

Case 2—A 59-year-old man first presented in November 1975 with rectal bleeding. Sigmoidoscopy and proctoscopy showed piles which were treated by injection. Two months later he developed severe pain in the area of the left kidney and a mass was felt in the left hypochondrium. Intravenous pyelogram showed an extensive tumour affecting the upper pole of the left kidney. Chest x-ray examination showed a secondary deposit in the right lung, and suspicious areas were noted in the ribs and lumbar spine on bone scan. At operation in February 1976 the mass was found to be infiltrating extensively into the surrounding structure, making removal impossible. Biopsy disclosed a hypernephroma. He was subsequently referred for radiotherapy and received 4125 rads over a period of three weeks. He gradually deteriorated and died on 26 July 1976.

Discussion

Clemmesen¹ reported a remarkable family of 15 siblings of whom two had hypernephromas and six had polycystic kidneys. Brinton² treated a family in which three siblings had hypernephromas confirmed and the father had been treated for a renal tumour but this had not been confirmed histologically. Steinberg *et al*³ reported the same tumour in a mother and daughter and cited six further case reports in three pairs of siblings. A remarkable family of five siblings was reported by Franksson *et al*⁴ in which four siblings had multiple bilateral tumours and a further sibling had a solitary tumour in one kidney. Thus, if the parent in Brinton's cases is included, there have

been a total of 19 cases in seven families reported, our patients bringing the total to 21 cases in eight families.

The familial incidence of hypernephromas may be fortuitous, but if a genetic factor is present the question of screening other members of the family arises. Brinton² and Steinberg *et al*³ reported hypernephroma in two generations and it would therefore seem reasonable to carry out regular five-year investigations in the other siblings and the children of our two brothers.

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Primary biliary cirrhosis, cutaneous capillaritis, and IgM-associated membranous glomerulonephritis

Primary biliary cirrhosis (PBC) is often associated with autoimmune phenomena. These include thyroid disease, arthropathy, sicca complex, hyperglobulinaemia and non-organ-specific autoantibody formation. We describe a patient with PBC, membranous glomerulonephritis, and itching cutaneous papules in whom IgM was identified in both renal and skin lesions.

Case report

A 43-year-old woman presented with a six-week history of generalised itching. Examination showed inconspicuous 1-2-mm pink papules and vesicles scattered over the trunk, and splenomegaly. Investigations showed: serum bilirubin 8 μ mol/l (0.5 mg/100 ml; normal range 7-17 μ mol/l (0.4-1 mg/100 ml)), alkaline phosphatase 612 IU/l (normal range 20-90 IU/l), aspartate aminotransferase 29 IU/l (normal 2-21 IU/l), fasting cholesterol 8.1 mmol/l (313 mg/100 ml; normal range 3.6-7.8 mmol/l (139-301 mg/100 ml)), albumin 27 g/l (normal 36-53 g/l), urea 5.4 mmol/l (32.5 mg/100 ml; normal 2.5-7.0 mmol/l (15.1-42.2 mg/100 ml)), creatinine 96 μ mol/l (1.1 mg/100 ml; normal 53-124 μ mol/l (0.6-1.4 mg/100 ml)), creatinine clearance 67 l/24h. The Wrong-Davis renal acidification test showed a minimum urine pH of 5.9 confirming distal renal tubular acidosis. There was proteinuria (7.2 g/24h) with numerous hyaline and cellular casts. Serum immunoglobulins (normal ranges in parentheses) were: IgM 4.3 g/l (0.65-2.00 g/l), IgG 19.7 g/l (9.5-16.5 g/l), IgA 3.4 g/l (0.9-4.5 g/l). Mitochondrial antibodies were present at a titre of 1/80 but HBsAg was not detected in serum.

Liver histology—There was evidence of bile duct destruction, marked lymphocytic infiltration with portal tract expansion, and scattered piecemeal necrosis compatible with stage I PBC. Immunofluorescence was not performed.

Kidney—Uniform thickening of capillary loops was present with multiple spikes on the epithelial surface of glomerular basement membrane. There was extensive IgG deposition on basement membranes and patchy IgM, probably within the mesangium, on immunofluorescent staining by the technique of Coons *et al*.¹ There was no other important immunoglobulin deposition, and the appearances were compatible with membranous glomerulonephritis.

Skin—A localised area of upper dermal perivascular neutrophilic and lymphocytic infiltration was present. The overlying epidermis showed parakeratosis, a prominent granular layer, and some epidermal cell swelling.

Papillary vessel walls in this area contained IgM and complement (C3), which was also present as scattered granules in the surrounding dermis and especially at the dermoepidermal junction.

Discussion

The clinical, biochemical, immunological, and histological features of this patient were consistent with PBC.² Itching in this condition is usually attributed to raised bile acid concentrations, and inflammatory skin lesions to the consequent scratching. Nevertheless, inflammatory papules similar to this patient's have been described.³ Periportal deposition of IgM is recognised in PBC but deposition in other organs is hitherto unreported. About 74% of patients with PBC have raised serum IgM concentrations, as did this patient. A fall in IgM concentrations follows treatment with azathioprine and there is some evidence that this drug also relieves itching.⁴ The finding of capillaritis associated with IgM and C3 in the skin lesions of this patient suggests that immunological and possibly complement-mediated events may have contributed to the itching.

Single glomerulus stained with fluorescein-labelled anti-IgM after washing in saline for 30 minutes ($\times 221$). There are patchy deposits of IgM throughout.

The biochemical features of both renal tubular acidosis and nephrotic syndrome were present, although oedema was absent. Renal tubular acidosis is common in PBC but although glomerular damage occurs in various liver diseases⁵ it is not a recognised feature of PBC. One patient with biliary cirrhosis (type unspecified)⁵ had mild proteinuria and glomerulosclerosis on light microscopy.

Membranous glomerulonephritis associated with IgM deposition is uncommon, as is also PBC. The simultaneous presentation of liver, renal, and skin disease in this patient is unlikely to represent chance association. Possibly deposition of circulating immune complexes including IgG and IgM might account for both the renal glomerular lesion and the cutaneous capillaritis.

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