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. Brinton[2] 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 μmol/l (0·5 mg/100 ml; normal range 7–17 μmol/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 μmol/l (1·1 mg/100 ml; normal 53–124 μmol/l (0·6–1·4 mg/100 ml)), creatinine clearance 67 1/24h. 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.

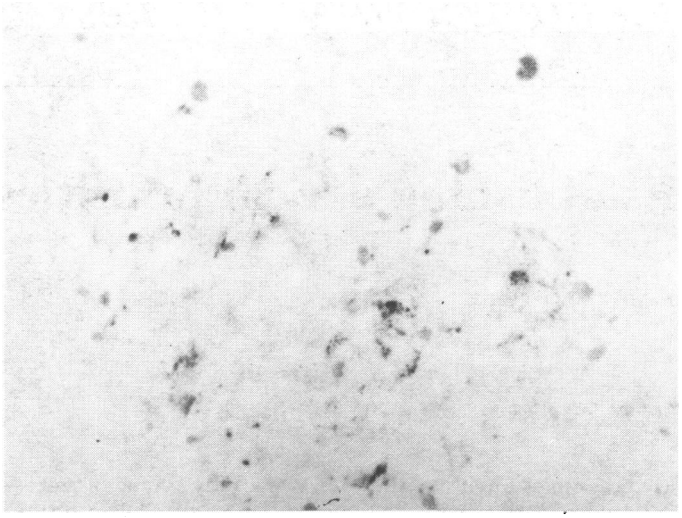
Liver histology—There was evidence of bile duct destruction, marked lymphocytic infiltration with portal tract expansion, and scattered piecemeal necrosis compatible with stage I PBC. Immunofluorescence was not performed.

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_2 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 (×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 diseases[5] 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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