Dermatitis Herpetiformis, Steatorrhoea, and Malignancy

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An association is known to exist between idiopathic steatorrhoea and intra-abdominal lymphoma (Fairley and Mackie, 1937; Shreeve *et al.*, 1968; Brunt *et al.*, 1969). Malignancy of various other types also occurs often in patients with idiopathic steatorrhoea (Harris *et al.*, 1967). Structural changes of the small-intestinal mucosa resembling those in idiopathic steatorrhoea are prevalent in dermatitis herpetiformis, as first reported by Marks *et al.* (1966) and van Tongeren *et al.* (1967) and later reviewed by Lyell (1969). Apart from poor correlation between intestinal structural changes and malabsorption, the dermatitis herpetiformis enteropathy shows great similarity to that of idiopathic steatorrhoea (Shuster *et al.*, 1968).

We have recently studied two patients with dermatitis herpetiformis and steatorrhoea who also had malignancy adding a new feature to the dermatitis herpetiformis enteropathy.

CASE 1

The patient, a man aged 44, was admitted to our department in January 1969. In 1946 he had suffered a long period of diarrhoea. Dermatitis herpetiformis had been diagnosed at the University Hospital in 1951, and later he had continuous dapsone treatment. Since July 1968 he had dyspeptic upset in the upper abdomen. During the six months before admission he lost 10 kg. in weight. On examination he was a healthy looking man with no pathological findings in the heart, lungs, or abdomen.

Investigations.—Haemoglobin 80%; R.B.C. 4,460,000/cu.mm.; W.B.C. 6,400/cu.mm.; platelets 219,000/cu.mm.; E.S.R. 45 mm./hour; serum electrolytes normal; alkaline phosphatase slightly increased (61 i.u./100 ml.). Stools contained increased amounts of fat and nitrogen (12·1 and 3·7 g./day respectively). Barium meal and follow-through examination was normal. Immunoelectrophoresis of serum showed moderate general increase of immunoglobulins, with slight increase of IgG and IgM and pronounced increase of IgA. Peripheral blood film was normal. Sternal marrow showed normoblastic erythropoieses and slight increase of plasma cells. Serum vitamin B_{12} was 230 pg./ml. Pentagastrin test gave a maximal acid output of 6·2 mEq/hour. Schilling test showed that 11% of dose was excreted in urine in 24 hours. Serum iron was 30 µg./100 ml., total iron-binding capacity 520 µg./100 ml. Jejunal biopsy: normal villi with slightly increased number of leucocytes in the stroma.

On 10 February a sudden life-threatening melaena necessitated laparotomy. Twenty cm. of upper jejunum (20-40 cm. distal to the ligament of Treitz) was removed because of an ulcerating tumour measuring 4 by 5 cm. Histological examination showed a homogenous hypercellular tissue composed of large cells with eosinophilic cytoplasm. The nuclei showed some variation from pyknotic to large, with coarse granular chromatin and nucleoli. These malignant cells did not produce mucin and probably originated from the reticuloendothelial system.

Another dramatic melaena required a second laparotomy on 6 April, and a further 20 cm. of jejunum distal to the anastomosis was resected. This specimen contained ulcerating tumour tissue with a similar histological picture to that described above.

CASE 2

The patient, a man aged 44, was first admitted to our department in October 1966 because of anaemia and probable malabsorption. Dermatitis herpetiformis had been diagnosed in 1954 and he had since taken dapsone continuously. During the six months before admission he had experienced increasing asthenia, anorexia, pigmentation of the skin, and complaints of eructation and audible borborygmi and had lost 4·5 kg. in weight.

Investigations.—Haemoglobin 73%; R.B.C. 3,650,000/cu.mm.; W.B.C. 5,300/cu.mm. Immunoelectrophoresis of serum showed slightly increased concentration of IgG and IgM, with normal IgA. Vitamin A was abnormally low (295 i.u./100 ml.; normal>800 i.u.). Fat and nitrogen in stools were 19·3 and 2·9 g./day respectively. Glucose tolerance test showed a flat curve. Barium meal and follow-through examination showed rapid passage, but was otherwise normal. Histological examination of jejunal biopsy specimen showed pronounced villous atrophy compatible with idiopathic steatorrhoea.

A diagnosis of idiopathic steatorrhoea was made and a gluten-free diet led to some improvement. The clinical situation was, however, not satisfactory and the E.S.R. and serum fibrinogen remained consistently raised. Intravenous pyelography and renal arteriography showed a large left-sided kidney tumour, which on removal was found to be a renal carcinoma metastasizing into the renal veins and a regional lymph node. Postoperatively he received local *x*-ray treatment and continued with dapsone and the gluten-free diet, with pronounced increase in weight and good clinical condition. Intractable anaemia, weight loss, and intermittent fever recurred throughout 1968. He died in June 1969 from metastatic deposits.

COMMENT

Enteropathy in dermatitis herpetiformis is now so well documented that all patients with this condition should undergo small-intestinal investigations (Shuster *et al.*, 1968). The relation between skin and gut symptoms in this dermatointestinal syndrome remains unexplained. The enteropathy resembles idiopathic steatorrhoea in many respects, but the common denominator for the two lesions has not been found. Gluten-free diet may improve the malabsorption without affecting the course of the skin disease. This may, on the other hand, be successfully treated with dapsone without influencing the severity of the enteropathy.

The histological changes found in dermatitis herpetiformis enteropathy are indistinguishable from those seen in idiopathic steatorrhoea. Fry *et al.* (1967) reported a high prevalence of other abnormalities usually related to idiopathic steatorrhoea, like IgM-globulin deficiency and signs of splenic atrophy in the peripheral blood. Malignancy with dermatitis herpetiformis enteropathy as reported here strengthens the similarity between this condition and idiopathic steatorrhoea.

E. GJONE, M.D.,
A. NORDÖY.
Medical Department A,
University Hospital, Rikshospitalet,
Oslo, Norway.

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