Usually, incisors and canines were affected. All our 3 patients exhibited dental abnormalities of one or more of the types mentioned above.

ACKNOWLEDGEMENT

The author wishes to thank K.-V. Sarnäs, orthodontist, Centre for Cranio-facial Anomalies, General Hospital, Malmö, who has examined the dental status of the propositus and the propositus' mother.

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Forty Years of Diarrhoea in a Patient with Urticaria Pigmentosa

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Received September 9, 1981

Abstract. A patient with urticaria pigmentosa who gave a 40-year history of diarrhoea was found to have systemic mastocytosis with gut involvement. The radiological appearances of the gut in this disease, although not widely recognized, are specific and should be looked for carefully in patients with urticaria pigmentosa who complain of gastro-intestinal symptoms. Gastro-intestinal symptoms, due mainly to alterations in bowel motility or peptic ulceration, are said to occur in some 25–50% of cases of systemic mastocytosis (3, 6). These symptoms have usually been ascribed to generalized histamine release acting on the gut, although cases where mast cell infiltration of the bowel has occurred have also been reported (4, 5). In a review of the radiological features (2), increased gastric rugosity with or without evidence of peptic ulceration and nodular space-filling defects of the bowel mucosa were the most commonly found. Occasionally, diffuse thickening of the bowel wall was seen. It was concluded that these appearance were probably due to local release of vasoactive substances causing submucosal oedema following mast cell accumulation in the gut. Another result of such infiltration may be malabsorption (1).

CASE REPORT

A 64-year-old man had suffered from diarrhoea and a widespread skin rash for 40 years. His rash had become gradually more noticeable over this period and for the last 2 years he had in addition suffered from flushing attacks, last approximately 3 days and occurring once every 2–3 months. His diarrhoea was severe on average once a year. At these times he would produce up to 20 motions a day, sometimes blood-stained. More usually he passed four or five watery stools each day and no treatment appeared to alter this pattern. Twenty years previously he had been told he was suffering from 'colitis' after a sigmoidoscope examination, but no histological proof was obtained.

On examination he had a widespread rash consisting of discrete, pigmented telangiectatic macules consistent with the 'telangiectasia macularis eruptiva perstans' variant of urticaria pigmentosa. Physical examination revealed 3 cm hepatosplenomegaly. A skin biopsy confirmed an increased number of mast cells in the dermis, while bone marrow biopsy showed focal aggregation of mast cells. Screening for bone involvement was negative, as was the peripheral blood. Five-day faecal fat estimation was within normal limits. Carcinoid syndrome was excluded on the basis of normal urinary 5-HIAA excretion.

Sigmoidoscopy showed an oedematous granular rectal mucosa and gastroscopy revealed a mild gastritis. A barium meal revealed increased rugosity in the stomach and in the duodenum many nodular space filling defects. These 'indentations' were seen in several areas of the small bowel on follow through examination, and also in the transverse colon and hepatic flexure following a barium enema. Histological examination of the jejunal and rectal biopsies showed a mixed infiltrate of mast cells and eosinophils.

DISCUSSION

The duration of our patient's diarrhoea is rather unusual in this disease, but not without precedent. There have been several cases recorded of long-standing gastrointestinal involvement in mastocytosis, the longest of which is 25 years (5). As in our case, there was no evidence of malignant transformation.

Oral disodium cromoglycate has been shown in a doubl-blind trial to control the symptoms of the disease quite well (7). In our patient the flushing attacks and excerbations of diarrhoea have disappeared following institution of this drug, although his overall bowel frequency remains three to four stools per day.

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Suppression of Erythema nodosum by Indomethacin

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Received August 17, 1981

Abstract. Erythema nodosum (EN) can be a debilitating illness. Many treatment modalities have been suggested but none is universally effective. We describe three patients with severe EN secondary to streptococcal pharyngitis who were unresponsive to large doses of aspirin. Following the administration of indomethacin in doses of 100 to 150 mg per day, all three showed a dramatic response, with prompt resolution both of systemic symptoms and of local signs of inflammation. The lesions of EN involve the lower cutis, with inflammation of septae and fat lobules. It is likely, therefore, that the local liberation of fatty acids leads to increased prostaglandin synthesis which, in turn, is responsible for the intense inflammation. The impressive suppression of EN by indomethacin could be related to the inhibition of prostaglandin synthetase in the subcutaneous fatty tissues.

Key words: Erythema nodosum; Indomethacin

Although frequently severe and debilitating, erythema nodosum (EN), is usually self-limited, resolving within 3–6 weeks (1, 2). A number of medications have been employed, but none has been found to be universally satisfactory (1).

We have observed 3 patients with severe, debilitating EN secondary to an antecedent streptococcal pharyngitis, who responded promptly and dramatically to indomethacin. Our experiences, reported here, suggest that indomethacin be used in the treatment of this disorder and further imply a possible mechanism for the local inflammatory process of EN.

CASE REPORTS

Case 1. R. G., a 57-year-old Mexican-American female, was admitted to the hospital with an acute febrile illness of 2 days' duration. She complained of numerous painful, red, raised lesions on all extremities. Ten days prior to the onset of her acute illness, she had had a sore throat and received oral penicillin. She had taken penicillin numerous time in the past without complications. On physical examination the patient was acutely ill, with an oral temperature of 101°F and numerous tender EN lesions, most prominently over the ankles, dorsum of the feet, the wrist and the dorsum of both hands, with underlying edema, perarticular inflammation and tenosynovitis. The pertinent abnormalities from an extensive laboratory evaluation were an elevated ESR of 128 mm/h, and an ASO titer of 500 Todd units. Treatment initially consisted of aspirin 4 g/day in divided doses. On the 4th hospital day, the plasma salicylate concentration was 17.1 mg/dl. Disability due to EN with fever persisted and new lesions developed daily. On the 6th hospital day, aspirin was discontinued and indomethacin was given in a dose of 50 mg t.i.d. Within 12 hours, the patient dramatically improved. Inflammation of the skin and joints had markedly subsided. Twenty-four hours after the initial indomethacin dose, a new nodule was noted, but it was minimally inflamed and non-tender. No new lesions developed subsequently. With the onset of treatment, she became afebrile. Indocin was continued for 2 weeks. There was no recurrence of EN.

Case 2. B. S. a 34-year-old white female, had a sore throat which subsided untreated and was followed in 4 days by migratory polyarthralgias and the development of painful red nodules on the extremities. She had taken oral contraceptives containing estrogen for 3 years. On admission to the hospital, the patient had the classical dermal lesions of EN and synovitis of joints proximal to the EN lesions. Abnormal laboratory findings were a leukocytosis of 15900/mm^3, ESR 60 mm/h, positive CRP and throat culture positive for Group A streptococci. Her ASO titer was initially 166, increasing to 500 Todd units.

Initially, she was considered to have acute rheumatic fever and received benzathine penicillin and large doses of aspirin. Even though her plasma salicylate levels equalled or exceeded 30 mg/dl, joint complaints persisted and new EN lesions developed. On the 4th hospital day, aspirin was discontinued and indomethacin started in