Verapamil and polythiazide were discontinued without benefit. There was a poor response to bed-rest, topical steroids and antihistamines. The eruption persisted for 9 months before resolution, during which time there were acute exacerbations with marked pruritis, confluent erythema and papules.

Discussion

The 5 cases previously reported were all males aged 57-77 years. Four out of the 5 had eosinophilia and 4 lymphopenia (2 marked and 2 borderline). The patient without eosinophilia had a normal lymphocyte count. Only one patient had a past history and a family history of atopy. All the patients had similar clinical features and skin histology to the patient we now describe.

We believe that this dermatosis is a separate disease entity. The episodic nature and striking sparing of the skin creases tempt us to think that it may be due to an as yet unidentified circulating factor.

References

1 Orfuji S, Furukawa F, Miyachi Y, Ohno S. Papuloerythroderma. *Dermatologica* 1984;169:125-30
2 Farthing CF, Staughton RCD, Harper JI, Rowland Payne CME, Copeman PWM. Papuloerythroderma - A further case with the 'deck-chair sign'. *Dermatologica* 1986;172:65-6

(Accepted 1 October 1986. Correspondence to Dr R C D Staughton, Westminster Hospital, London SW1P 2AP)

Phrynoderma and perforating folliculitis due to vitamin A deficiency in a diabetic

S M Neill MRCP **A C Pembroke** MRCP
A W P du Vivier MD FRCP **J R Salisbury** BSc MB
Departments of Dermatology and Morbid Anatomy, King's College Hospital, London SE5

Keywords: perforating folliculitis, phrynoderma, vitamin A deficiency, pancreatic insufficiency

A case is reported of an insulin-dependent diabetic with longstanding malabsorption due to pancreatic insufficiency who developed ichthyosis and asymptomatic hyperkeratotic papules and nodules. Low levels of vitamins A and E were found. The skin lesions responded following the correction of the vitamin A deficiency.

Case report

A 56-year-old man was referred to the dermatology department with a 6-month history of asymptomatic papules and nodules on his limbs. He had been an insulin-dependent diabetic for 25 years and had suffered many of the complications of this disorder, including proliferative retinopathy, peripheral vascular disease, autonomic neuropathy, peripheral neuropathy with trophic ulcers of the feet and osteomyelitis of the tarsal bones. He eventually required a left mid-thigh amputation for gangrene.

In 1965 he developed intermittent nocturnal diarrhoea which was investigated in 1966 and again in 1974. Steatorrhoea was documented, but all other investigations, which included xylose absorption, pancreatic exocrine function studies, barium meal and follow through and jejunal biopsy, were normal. There was no symptomatic improvement on a gluten-free diet and finally his diarrhoea was attributed to diabetic autonomic neuropathy. He was treated with oxytetracycline with some improvement in his symptoms and has continued on this for the last 10 years. In 1975 he developed biochemical osteomalacia and was treated with calciferol. Over the last 12 months the bouts of nocturnal diarrhoea had become more frequent and severe, resulting in considerable weight loss and distress.

Case presented to Section of Dermatology, 17 October 1985

At the time of presentation to the dermatology department, examination of the skin revealed generalized hyperpigmentation and xerosis. Large dark brown ichthyotic scales covered the extensor surfaces of the forearms and the right shin. Pigmented hyperkeratotic papules were observed over the elbows and extensor surfaces of the forearms, the buttocks and the lateral aspect of the right thigh. Many of the lesions were clustered around bony prominences. The lesions over the lower limbs, however, were nodular with erythematous bases and a central crater filled with keratinous material (Figure 1). Several forearm papules were biopsied and histological examination showed moderate hyperkeratosis with distension and distortion of the upper part of the hair follicles by a large plug of keratinous debris. In some sections there

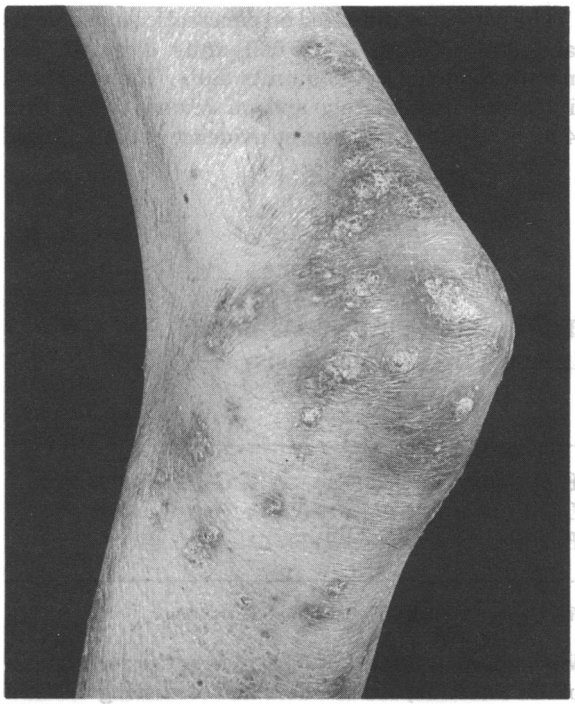

Figure 1. Hyperkeratotic nodules and papules around right knee

0141-0768/88/030171-02/$02.00/0

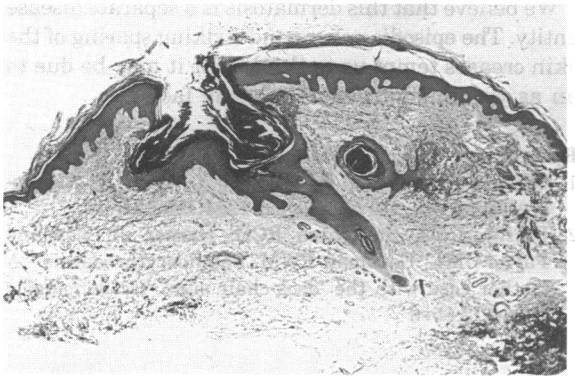

Figure 2. Widely dilated hair follicle filled with keratin. There is a breach in the infundibular wall (×25)

was a breach in the infundibular wall (Figure 2). There were no pigment-laden macrophages and elastic stains were normal.

Haematological investigations showed a normochromic, normocytic anaemia of 10.1 g/dl. The serum iron was reduced at 10 mmol/l with a normal total iron-binding capacity, serum ferritin, B_{12} and red cell folate. Thyroid and liver function tests were normal. There was evidence of renal impairment with a raised blood urea (14 mmol/l; normal range 3.3-6.7) and raised serum creatinine (140 umol/l; normal 45-105). Serum calcium was low (2.15 mmol/l; normal 2.3-2.6) and alkaline phosphatase elevated (245 iu/l; normal 30-85) in keeping with osteomalacia.

Other recent investigations included a low serum albumin (27 g/l; normal 35-50), low vitamin A (0.3 μmol/l; normal 1-3.5), low vitamin E (2.83 μmol/l; normal 16-47) and serum zinc of 10.8 μmol/l (normal 11-20). A breath test confirmed fat malabsorption with 2.5×10^{-4}% of an ingested dose of ^{14}C-labelled triolein being excreted at 7 hours (normal $>50\times10^{-4}$%). A plain film of the abdomen showed extensive calcification of the pancreas. Pancreatic insufficiency was confirmed by a Lundh test, which showed very low concentrations of tryptic activity in the jejunal aspirate <2000 u/l (normal 7000-38 000).

The patient was started on pancreatic supplements and oral vitamin A: 50 000 units daily for one month reducing to 2000 units daily. The diarrhoea improved immediately and on review of his skin 4 months later there was no evidence of the acquired ichthyosis. The forearm papules had resolved leaving no residual scarring. The lesions on the knees had flattened leaving depressed scars, and there was no inflammation as previously noted. The vitamin A was within the normal range at 1.6 mmol/l, but the vitamin E was still low at 3.0 μmol/l.

Discussion

The term phrynoderma, meaning toad skin, was first used by Lucius Nicholls[1] in 1933 to describe the hyperkeratotic folliculitis he had observed in African labourers who were vitamin A deficient. However, not all those with vitamin A deficiency develop this cutaneous manifestation. Phrynoderma has recently been reported in 2 cases following intestinal bypass surgery[2,3]. In these 2 reports the patients developed cutaneous signs and night blindness 2 and 4 years respectively after bypass surgery for obesity. Both patients had low vitamin A levels which, when corrected, resulted in a dramatic improvement of the cutaneous lesions.

Initially in our patient we suspected a diagnosis of an acquired primary perforating disorder of the skin in association with diabetes mellitus. The investigation of his weight loss and diarrhoea, however, led to the subsequent discovery of pancreatic insufficiency, malabsorption and vitamin A deficiency. In view of these findings, an alternative diagnosis of phrynoderma was considered. The response to correction of the vitamin A deficiency supports this diagnosis. The vitamin E levels have remained low in our patient as they did in the case reported by Wechsler[2]. We believe, like Barr *et al.*[3], that the perforation may be an incidental finding.

Acknowledgment: We thank Dr P J Watkins and Dr D A Pyke for referring the patient and Dr P Powell-Jackson for his help in the management of this case.

References

1 Nicholls L. Phrynoderma: A condition due to vitamin deficiency. *Indian Medical Gazette* 1933;**68**:681-7
2 Wechsler HL. Vitamin A deficiency following small-bowel bypass surgery for obesity. *Arch Dermatol* 1979;**115**:73-5
3 Barr RJ, Riley RJ, Greco DJ. Bypass phrynoderma. *Arch Dermatol* 1984;**120**:919-21

(Accepted 6 November 1986. Dr Neill is now at St Stephen's Hospital, London SW10)

The treatment of obstructing intestinal endometriosis

E P Perry MSc FRCSEd **A L G Peel** MChir FRCS
Department of Surgery, North Tees General Hospital, Stockton-on-Tees

Keywords: endometriosis, intestinal obstruction

0141-0768/88/030172-02/$02.00/0

We report 3 cases of obstructing endometriosis of the intestine, one of whom had coexisting Crohn's disease. The diagnosis is more easily achieved in colonic than ileal disease. The differential diagnosis from malignant disease in women of childbearing age is important. Endocrine treatment proved ineffective in intestinal endometriosis, producing symptoms.

Case reports

Case 1: A 42-year-old woman had a 4-year history of left-sided colicky abdominal pain and diarrhoea, which had been diagnosed on barium studies as Crohn's disease and treated medically with sulphasalazine (Salazopyrin) and occasional short courses of steroids. Over the previous year the pain had become predominantly right-sided and she was treated with high-dose steroids, but without improvement.

She then proceeded to laparotomy and had a limited right hemicolectomy with resection of 35 cm of