anti-thyroperoxidase antibody in addition to, or, less likely, instead of MPO-ANCA cannot be excluded.

Vesiculo-bullous SLE has been reported to respond to dapsone (15). However, in our patient, an early aggressive treatment with steroid pulse therapy and plasmapheresis was mandatory because of her life-threatening clinical condition. The contributory factors, such as an environmental trigger or an immunological factor, for the presence of a serious illness in this patient remain to be elucidated. The mechanism by which methimazole induces SLE-like reactions is unclear.

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Angiokeratoma of the Scrotum (Fordyce Type) Associated with Angiokeratoma of the Oral Cavity

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Sir,

Angiokeratoma is a rare cutaneous vascular disorder of the papillary dermis characterized by vascular ectasia associated with overlying hyperkeratosis. Five types of angiokeratoma are generally recognized: (i) the usually solitary papular angiokeratoma, (ii) the bilateral angiokeratoma that occurs on the dorsa of the fingers and toes (Mibelli type), (iii) the localized angiokeratoma of the scrotum or vulva (Fordyce type), (iv) the congenital form, angiokeratoma circumscriptum (naeviforme), and (v) the generalized systemic form, angiokeratoma corporis diffusum (Fabry's disease, fucosidosis). The types differ from each other by location and clinical manifestation, but share similar histopathological features (1). Some patients may have lesions of more than one of these types. We report on a patient with angiokeratoma of the scrotum (Fordyce type) associated with angiokeratoma of the oral cavity.

CASE REPORT

A 72-year-old Caucasian man presented with papules on the scrotum and tongue. The lesions had been present for the past two years. He had no other complaints, and, in particular, there had been no bleeding from the lesions. He had no preceding history of local trauma and his medical history was unremarkable. No other members of the family presented this condition.

Physical examination revealed multiple discrete, red or purple, well-defined, dome-shaped papules, some with keratotic overlying skin, varying in size from 2 to 3 mm, scattered over the scrotum (Fig. 1). Similar lesions could

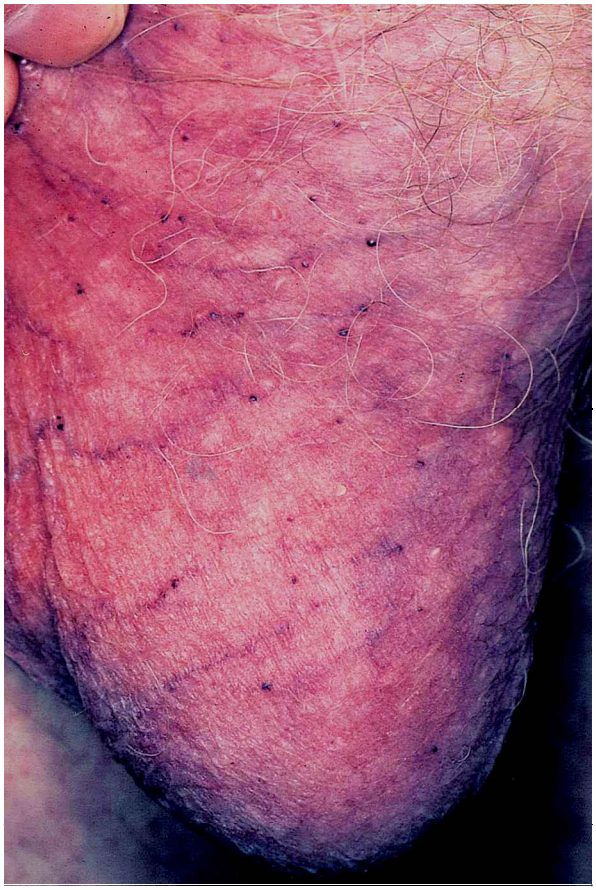

Fig. 1. Angiokeratoma of the scrotum.

be observed on the lateral aspect of the tongue (Fig. 2). There was no evidence of varicocele, tumour of the testis, or inguinal hernia.

Histopathological examination of a representative lesion on the scrotum showed an acanthotic epidermis with cystic spaces containing erythrocytes and organizing thrombi (Fig. 3). Another biopsy taken from a representative lesion on the tongue was also consistent with a diagnosis of angiokeratoma.

As the lesions were asymptomatic, the patient requested no further treatment.

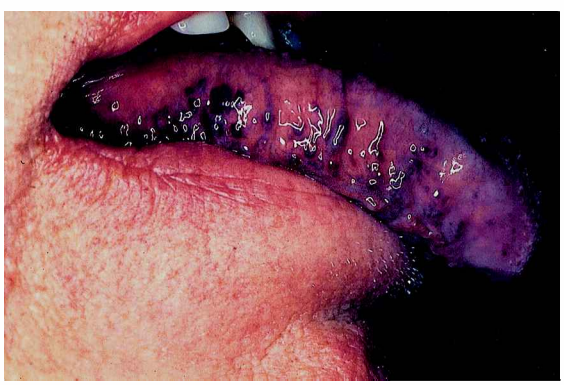

Fig. 2. Angiokeratoma at the lateral aspect of the tongue.

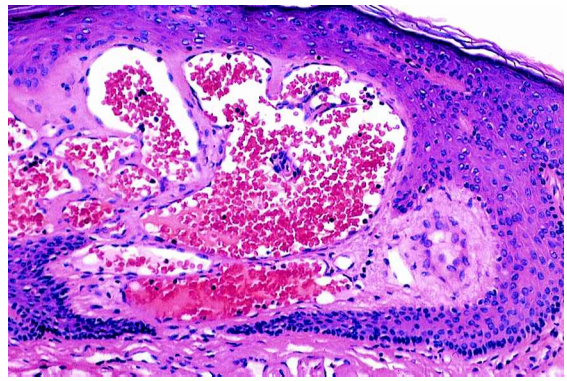

Fig. 3. Histopathology showing an acanthotic epidermis with cystic spaces containing erythrocytes and organizing thrombi. H & E stain.

DISCUSSION

Angiokeratoma of the scrotum was first reported in 1896 by Fordyce (2) in a 60-year-old man. The lesions are usually multiple in number and located mainly on the scrotum, but are occasionally found on the shaft or glans of the penis, the upper part of the thighs, the abdomen, and the buttocks. They tend to be small (1–5 mm) and can range in colour from red to blue or black. The young lesions are soft and compressible while the older lesions may be firm and non-compressible, often keratotic and scaly, sometimes warty. Angiokeratoma of the scrotum usually develops in late adulthood or in elderly persons, although it may sometimes manifest in childhood or adolescence. The condition is frequently asymptomatic, but irritation, itching, and episodes of bleeding from the lesions may occasionally occur (3). Lesions entirely analogous to angiokeratoma of the scrotum may occur on the labia majora in older women (4).

So far, only a few cases of angiokeratoma of the scrotum associated with angiokeratoma of the oral cavity have been reported in the literature (5, 6). Recently, Karthikeyan et al. reported a similar case (5). In our patient, the presentation was different from that of the three cases reported by Rappaport & Shiffman (6) who had simultaneous involvement of the jejunum, oral mucosa, tongue, and scrotum. Mucosal involvement is otherwise uncommon in other types of angiokeratomas (7, 8). Fabry's disease (9) and fucosidosis (10) have mucosal involvement as a part of their generalized involvement.

Treatment is generally not necessary, except for bleeding, discomfort, and cosmetic reasons. Individual lesions can be destroyed with simple excision, cryosurgery, electrodesiccation, or lasers. Evidence of associated disorders capable of increasing the scrotal venous pressure should be sought, particularly varicocele or inguinal hernia, the treatment of which may lead to regression of angiokeratoma (11). In our patient, however, there

was no evidence of a local factor that could have lead to the condition.

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Ulcerated Giant Juvenile Xanthogranuloma Accompanied by Hyperlipidaemia

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Sir,
Juvenile xanthogranuloma (JXG) is a tumour frequently seen in childhood and is one of the most common non-Langerhans' cell histiocytoses. It is believed that JXG occurs without any metabolic disorder such as hyperlipidaemia. The macronodular form of JXG has a high tendency to ulcerate. We recently studied a rare case of ulcerated giant JXG accompanied by hypercholesterolaemia.

CASE REPORT

A 4-month-old female infant was taken in for examination of three, gradually enlarging nodules on her scalp and umbilicus. Two months earlier, two pea-sized dome-shaped nodules with yellow to pale red hues were initially noticed on the left temporal and umbilicus regions. One month later, a new dome-shaped nodule, yellow in colour, was found on the left frontal region of the scalp. Two nodules on the temporal and umbilicus became eroded and bled. The baby was delivered normally at term, and physical and mental development was normal. No particular diseases, such as diabetes mellitus, hypertension, hyperlipidaemia or malignant tumour, were found in the patient's or the family past medical history.

Physical examination revealed that this baby suffered from three discrete nodules on the left temporal, left frontal region of the scalp and umbilicus. Each nodule showed different clinical features. The largest one was on the umbilicus and showed a sessile, exophytic, round tumour, measuring $15 \times 15 \times 8$ mm, with a red–brown hue and partial ulceration. The temporal nodule was an ulcerated, dome-shaped tumour, measuring 12 mm in diameter, with a central excoriation covered by a black crust and a brownish yellow halo. The frontal one was a yellowish dome-shaped tumour measuring 8 mm in diameter. The mucous membranes were unaffected. Ophthalmologic examination was normal. The infant had no adenopathy or organomegaly.

The laboratory investigations revealed hyperlipidaemia: serum cholesterol 368 mg/dl (normal upper limit 220 mg/dl), serum triglyceride 136 mg/dl (normal upper limit 160 mg/dl), serum HDL-C 51 mg/dl (normal 40–70 mg/dl), estimated serum LDL-C 344 mg/dl (normal upper limit 130 mg/dl), serum ß-lipoprotein 884 mg/dl (normal upper limit 628 mg/dl). Endocrine functions (GH, ACTH, TSH, cortisol, free T_3 and T_4), erythrocyte sedimentation rate, full blood count, liver and renal function tests were all within the normal range. Chest and abdominal X-rays were normal. Abdominal ultrasound, computed tomography and magnetic resonance imaging showed no internal lesions.

A skin biopsy was taken from the eroded larger nodule on the temple. Hematoxylin and eosin-stained sections revealed a dense, diffuse infiltration composed of monotonous histiocytes intermixed with lymphocytes throughout the dermis. The histiocytes were distributed more compactly from the middle to deep dermis. Many discrete adipocytes were found in this lesion. The majority of the histiocytes had slightly lobulated nuclei and vacuolated cytoplasm. Neither giant cells nor foam cells