series of 21 cases with congenital pulmonary stenosis with cardiovascular malformations, neonatal liver disease with obstructive jaundice resembling biliary atresia or hepatitis and various minor congenital anomalies including odd facies. However, xanthomas were reported in only one case (4).

Alagille reported his series in the French literature in 1969, subsequently in English in 1975. He has studied and followed up 30 cases over the period of 15 years. The details outlined by him are chronic cholestasis, characteristic facies, mesosystolic murmur, vertebral arch defects, physical and mental retardation with hypogonadism. He also stressed the rarity of xanthomas and their pathognomony when distributed on extensors, body creases and palms. He did not state the number of patients with xanthomatosis in his series (5).

Other features of the syndrome include, in the eye, posterior embryotoxon, retinal pigmentary changes and anterior chamber anomalies, butterfly vertebrae, ataxia and areflexia (3).

Our patient had a few interesting unusual features, viz. macular and nebular corneal opacities and generalized osteoporosis, which have not been reported previously.

REFERENCES

Ulceration of the Palms and Soles
An Unusual Feature of Cutaneous T-cell Lymphoma

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Two patients, one with Sézary syndrome and one with mycosis fungoides are described, in whom lesions on the palms and soles were associated with extensive ulceration and gave rise to diagnostic difficulty. Extensive ulceration of the palms and soles is uncommon; its presence should alert clinicians to the possibility of cutaneous T-cell lymphoma.

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Cutaneous T-cell lymphomas can display a variety of clinical appearances (1). In the early stages, both mycosis fungoides and Sézary syndrome may have non-specific clinical and histological features. In the advanced stages, more characteristic appearances are usually seen, but atypical presentations with hyperkeratotic, papillomatous, hypopigmented, bullous, or aceneiform lesions have been described (1). We present 2 patients, one with mycosis fungoides and the other with Sézary syndrome, in whom atypical lesions with extensive ulceration on the palms and soles gave rise to diagnostic difficulty.

CASE REPORTS

Case 1

A 63-year-old Caucasian female presented in 1987 with a widespread scaly eruption which had the clinical appearance of guttate psoriasis and responded to coal tar paste and UVB therapy. The patient was re-referred in 1989 with a further widespread eruption which had the features of a
pre-reticulotic type of psoriasis. Skin biopsy specimens confirmed this. In the following years the patient developed lesions morphologically similar to lymphomatoid pa-
pulosis and pityriasis lichenoides chronica. Further biopsies from these lesions revealed a dense lymphoid infiltrate in the upper dermis with cerebriform nuclei and Pautrier's microabscesses and indicated a diagnosis of mycosis fungoides. Good improvement was achieved with psoralen photochemotherapy. Several more indurated plaques responded rapidly to conventional superficial X-ray therapy at a dose of 1-2 Gy.

After a full clinical remission lasting several years, a solitary ulcer developed in 1986 on the left heel and rapidly increased in size (Fig. 1). There was a slightly raised macerated margin with granulation tissue at the base. No other lesions were present and general examination was normal. The appearance of the ulcer was non-specific and the diagnosis uncertain. A biopsy from the margin of this lesion showed characteristic features of mycosis fungoides. Treatment again with conventional superficial X-ray therapy at a dose of 4 Gy resulted in complete healing over several weeks.

In May 1988, the patient developed a further lesion on the left foot, which proceeded to ulceration involving the whole of the third, fourth and fifth toes, extending onto the underlying sole. A biopsy again confirmed mycosis fungoides. Treatment with subcutaneous alpha interferon 3 Megunits daily was commenced. The therapy was continued for 3 months during which time further more typical lesions appeared over the trunk, limbs, interdigital areas, scalp and face. In view of the deterioration, interferon was
discontinued and treatment commenced with prednisolone
30 mg daily, chlorambucil 5 mg daily and etretinate 25 mg
daily, with some clinical improvement. Arrangements were
also made for generalized electron beam therapy which
produced some further improvement. However, she sub-
sequently deteriorated despite addition of pulse CHOP
chemotherapy (chlorambucil, cyclophosphamide, vincris-
tine and prednisolone) and died.

Case 2
A 58-year-old man presented in November 1985 with a
5-year history of an intermittent erythematous scaly rash
principally affecting the palms, soles and scalp. The patient
had a long history of depression necessitating several hospi-
tal admissions. There was no other relevant past medical
history. General examination was entirely normal. A diag-
nosis of psoriasis was made on clinical grounds and a satis-
factory response was achieved with topical steroid prep-
arations. In July 1986 the rash became more widespread
and was associated with marked hyperkeratosis of the
palms and soles. Limited improvement followed treatment
with a combination of etretinate and PUVA, and sub-
sequently with hydroxyurea.

In April 1988, the patient was admitted to hospital fol-
lowing worsening of his condition over a period of several
weeks. On examination there was widespread exfoliative
dermatitis with marked hyperkeratosis of the palms and
soles. In addition there was a marked nail dystrophy and
large areas of ulceration on the palms (Fig. 2). Cervical and
axillary lymphadenopathy were noted, but the examination
was otherwise normal. A clinical diagnosis of psoriasis was
made but the ulcers on the palms were incompatible with
this and a diagnosis of ulcerated lichen planus was consid-
ered.

A skin biopsy specimen taken from the edge of one of
the palmar ulcers revealed a lymphocytic dermal infiltrate
consisting of normal small lymphocytes and large trans-
formed cells, some with nuclear indentations. In addition
the epidermis was sparsely infiltrated with small lympho-
cytes, some forming small clusters in the upper epidermis.
These findings indicated a diagnosis of cutaneous T-cell
lymphoma. Sézary cells were identified on routine blood
counts accounting for 64% of circulating leukocytes.

The palms were treated with electron beam therapy with-
out significant improvement. The patient was subsequently
treated with subcutaneous alpha-interferon in a dose of 3
Megauunits three times a week, increasing to 3 Megauunits
daily after 4 weeks. Hydroxyurea was subsequently added
in a dose of 1 g daily. The palms and soles were treated
with potent fluorinated topical steroids under occlusion.
On this regimen there was a general improvement in the
skin and a reduction in circulating Sézary cells. However,
although there was partial healing, the palmar ulcers have
persisted and new ulcerated areas have appeared on the
soles.

DISCUSSION
Chronic ulceration of the palms or soles is uncom-
mon in the absence of significant sensory neuropathy
or ischaemia. It may occur in lesions of cutaneous
vasculitis and can be seen in localized areas at the
site of lesions such as calcinosis cutis, goyti tophi or
malignant tumours such as squamous cell carcinoma
and malignant melanoma. Ulceration of mycetoma
on the sole is not uncommon in the tropics and can
be seen in the late stages of Treponemal infections.
Other causes of chronic palmoplantar ulceration are
rare. Persistent erosions of the palms and soles may
occur in epidermolysis bullosa. Ulceration of the
fingers has been reported in one patient with sarco-
diosis (2). Extensive chronic ulceration of the feet
has been described in a number of cases as an un-
usual feature of lichen planus (3).

We have described here 2 patients with cutaneous
T-cell lymphoma who developed extensive chronic
ulceration, one of the palms and the other of the
soles. In the first case the patient developed discrete
areas of ulceration on the sole of the left foot some
years after a prolonged remission of mycosis fungoi-
des. In the second case the patient developed extensive
areas of ulceration of both palms against a background
of palmar hyperkeratosis during an evolving Sézary
syndrome. Ulceration is known to be a complication of tumourous mycosis fungoides
lesions. However, we are not aware of any previous
reports of palmar or plantar ulceration in these
conditions. Hyperkeratosis of the palms and soles is a
common feature of Sézary syndrome, but ulceration
has not previously been reported.

In conclusion, extensive chronic ulceration of the
palms or soles can occur in conjunction with mycosis
fungoides and Sézary syndrome. These disorders
should be considered in the differential diagnosis of
chronic palmar or plantar ulceration.

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