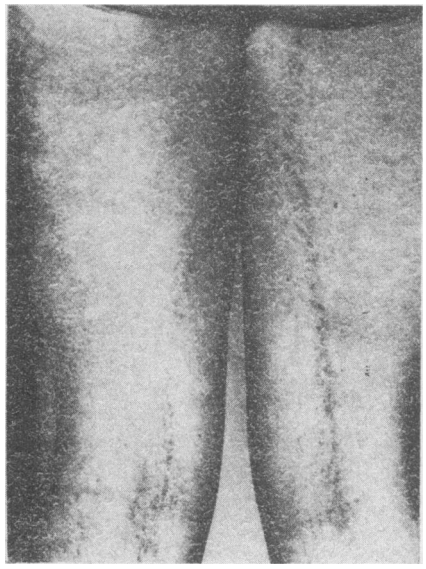

Fig 2 *Symmetrical linear telangiectasia on legs*

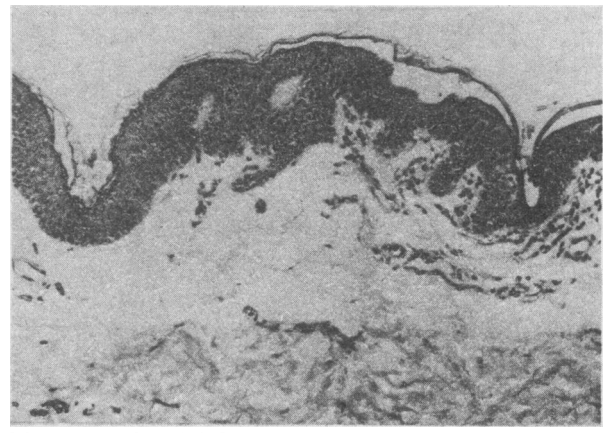

Fig 3 *Biopsy from left flank. H. & E.*

two component features are related. The histological findings suggest a primary developmental abnormality limited to the papillary dermis and associated with capillary ectasia, lymphocytic infiltration and epidermal changes. The absence of dermal atrophy and of associated skeletal defects seems to rule out the focal dermal hypoplasia syndrome, and a primary vascular abnormality such as angioma serpiginosum seems equally unlikely in view of the associated atrophy and pseudo-scarring.

Professor E Wilson Jones: The streaked zosteriform lesions with telangiectasia in this patient recall the changes seen in focal dermal hypoplasia (Goltz's syndrome). Has she any cryptic dental, ophthalmic or bony abnormalities?

Although the biopsy did not show fatty tissue close to the epidermis, this does not rule out the diagnosis as the histological changes are variable and depend on the type of lesion removed.

Dr Emmerson (replying to Professor Wilson Jones): Skeletal studies are entirely normal, and in particular do not show the changes of striate osteopathy described in the focal dermal hypoplasia syndrome. Her teeth and eyes are also unaffected.

Dr M M Black: At present we have a 31-year-old man attending St Thomas' Hospital with similar, although less striking, physical signs to the case shown by Dr Emmerson. He has a long history of discoid eczema and a rather troublesome intertrigo in the groin. In November 1973 it was noticed that he had reticulate atrophic areas mainly on the right upper trunk and also to a lesser extent on the arms and behind the knees. Close inspection revealed that there was telangiectasia around their margins. The patient stated that these lesions had been present since birth. He has not been applying topical corticosteroids to the affected areas, and they have not been the site of previous discoid eczema. The biopsy showed some œdema and mild inflammatory changes in the papillary body but there was no abnormality demonstrated on the special connective tissue stains. The condition certainly seems to be quite distinct from focal dermal hypoplasia (Goltz's syndrome).

Diffuse Cutaneous Mastocytosis in Mother and Daughter

Michael Klaber MB MRCP
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Case 1 S W, woman aged 25. Housewife

History: At birth had appearance of a macerated fetus with wrinkling and desquamation of the skin; this settled, but by the age of 6 months she was developing large blisters on minimal trauma. Dermographism was marked. Liver palpable 3.5 cm, spleen 2 cm. Diagnosis at that time was ichthyosis bullosa, biopsy having shown an infiltrate of cells of dubious type. Subsequently, she developed a bleeding tendency with cuts which oozed for some time. No coagulation abnormality was found.

On examination: Skin still leathery, especially in axillæ (Fig 1A). Dermographism.

Investigations: Prothrombin ratio, partial thromboplastin time, thrombin time and reptilase time all normal. Anti-factor 10a assay awaited. Biopsy of skin from leg: 1950, heavy infiltrate of mast cells in the dermis (Fig 1B); and again in 1971, diffuse mastocytosis (Professor E Wilson Jones).

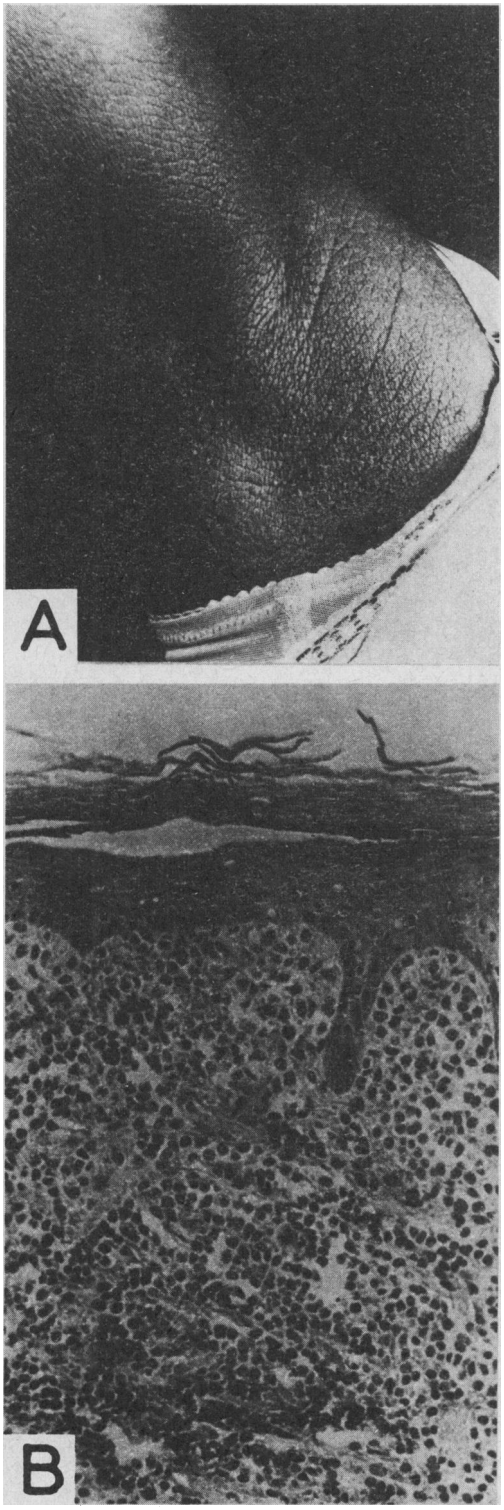

Fig 1 **Case 1** A, *axillary skin.* B, *heavy infiltrate of mast cells in dermis (stained with thionin)*

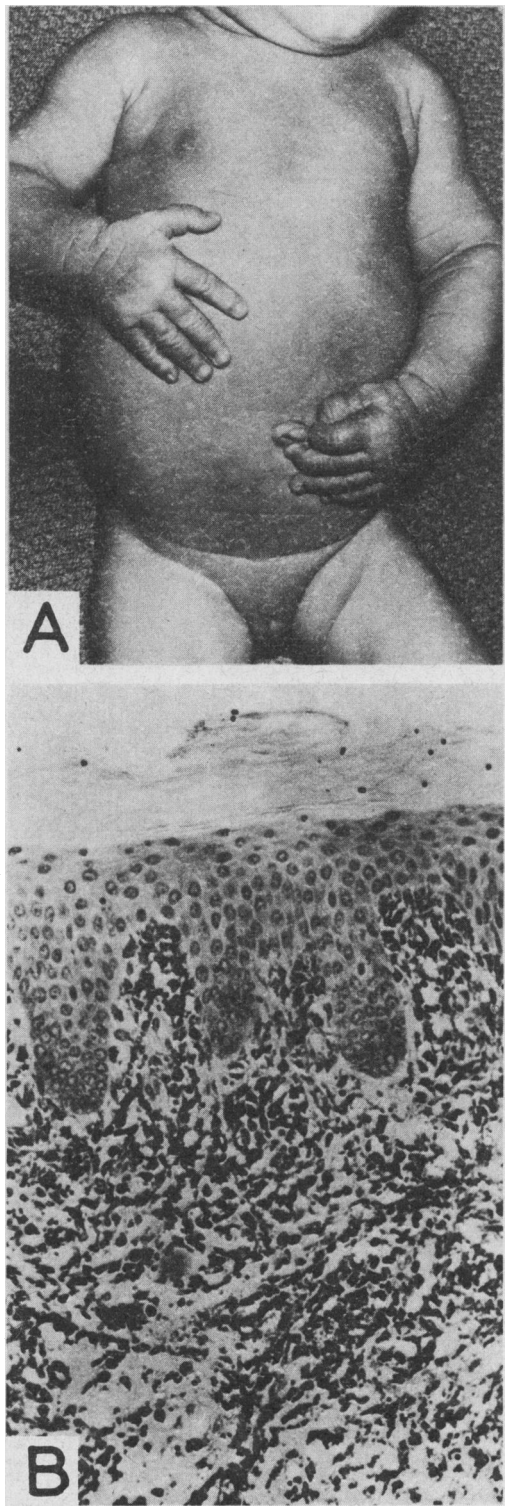

Fig 2 **Case 2** A, *at age 3 months.* B, *mast cells densely infiltrating outer dermis (thionin)*

Case 2 L W, girl aged 6 months. Daughter of Case 1

History: Red dry scaly skin at birth. Thick skin on legs at 2 months old, with blisters on sites of trauma from the age of 3 months.

On examination: Erythematous leathery appearance of skin especially on legs and napkin area with thickening of skin folds, most marked over knees and wrists (Fig 2A). Blistering at times on pressure areas. Dermographism. Spleen palpable 1.5 cm, liver 2.5 cm (normal).

Investigations: Simple coagulation tests negative. X-ray of legs showed nothing abnormal. Histology of skin from leg (Dr J W Landells) showed a continuous dense infiltration of mast cells in the outer dermis (Fig 2B).

Comment

Both mother and daughter seem to show identical patterns of diffuse cutaneous mastocytosis as described by Hissard *et al.* (1951) and by Degos *et al.* (1951). The mother's dizygotic twin brother is not affected. Familial mastocytosis has been described in 36 families. Bazex *et al.* (1971) and Shaw (1968), who have collected the largest series, believe it to be inherited in an autosomal dominant fashion and our family would fit in with this suggestion.

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Degos R, Lortat-Jacob, Mallarme J & Sauvan R (1951) *Bulletin de la Société française de dermatologie et de syphiligraphie* **58**, 435

Hissard R, Moncourier L & Jacquet J (1951) *Annales de médecine* **52**, 583

Shaw J (1968) *Archives of Dermatology* **97**, 137

Dr J A Savin: It may not be valid to draw inferences about the inheritance of diffuse cutaneous mastocytosis from a survey of published cases of other forms of mastocytosis. I feel that the diffuse form is clinically, and probably genetically, separate.

I have under my care a child with the diffuse form and have checked the literature. The death rate seems high. Usually the cause of death is not clear, histamine shock being suspected. Excessive bleeding from heparin release may occur. Urinary histamine estimations are difficult to interpret if the patient is not kept on a strictly controlled diet.

The dermographism differs from ordinary dermographism in that the marks last for several hours.

Meeting 17 April 1975

Cases

Tuberculous Lupus Miliaris Faciei

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Mrs L P, aged 40. Housewife

History: This West Indian woman arrived in the UK from Grenada thirteen years ago. In January 1974 an acne-like rash developed on her face. Subsequently there were small areas of lesions on abdomen and elbow flexures. Otherwise well; no relevant previous or family history.

On examination (April 1974): Acneiform eruption of cheeks. Papules set in mild lichenification in elbow flexures.

Investigations: Biopsy: conspicuous dermal infiltration with large lymphocytes and histiocytes. No tuberculous or sarcoidal granuloma. Microscopy of biopsy smears: acid-fast rods seen. Culture of biopsy material: *Mycobacterium tuberculosis* grown. Guinea-pig inoculation: generalized tuberculosis at post-mortem. Chest X-ray normal. Tuberculin test positive at 1:10 000.

Follow up (August 1974): Clinical picture unchanged. Repeat biopsies from face again grew *Mycobacterium tuberculosis* despite nontuberculous histology. Subsequent inpatient investigation has failed to reveal any evidence of systemic tuberculosis in pelvis, kidneys or elsewhere. The most recent biopsy showed a multinucleate cell granuloma centred on hair follicles (face).

Treatment: Isoniazid 300 mg daily and ethambutol 100 mg daily.

Comment

In recent years, lupus miliaris faciei has been considered to be neither tuberculous nor a tuberculide, the alternative names of acnitis or acne agminata being preferred. This patient had an acne-like eruption with banal histology which has been unequivocally shown to be tuberculous by culture of the organism.

Dr H T Calvert: The patient's condition does not resemble lupus miliaris faciei, acne agminata, or acnitis of Barthélemy. She does, however, have tuberculosis of the skin, the proof of which may lie in the response to antituberculous drugs.

Dr J R Simpson: The American literature contains many references to atypical immunological reactions amongst Negroes who present lesions which clinically and histologically appear to be sarcoidosis but which develop into frank tuberculosis. It is therefore not surprising that this interesting case should show unexpected features.

Postscript (December 1975): The lesions subsequently healed completely with prolonged antituberculous therapy. – H B

(*meeting to be continued*)