

following characteristic features: massive accumulation of swollen and clumped fibres in the middle and lower thirds of the derma, frankly positive reactions with Orcein and Verhoeff stains (Fig. 4), as well as Von Kossa's method (Fig. 3). The presence in large amounts of calcium deposits allowed us to confirm the diagnosis of P.X.E.

Ultrastructural examination of the lesions revealed most of the classical findings: twisting of the collagen fibrils (Fig. 5) with wire-like figures, masses of granulofilamentous material, altered fibroblasts with signs of hyperactivity, pseudoclastic transformation of the collagen fibres and heavy calcium deposits (Fig. 6).

COMMENT

The prominent clinical and histological features of our patient are very suggestive of the cases recently described by Christensen under the title "an exogenous variety of pseudoxanthoma elasticum in old farmers". This author observed the following features: 1) onset exclusively in male patients in Denmark and Sweden; 2) elderly farmers 52–87 years old; 3) who suffered burns 30–50 years ago while spreading Norwegian hydrous saltpetre; 4) the P.X.E. had followed a superficial ulcer which had healed 2–3 weeks later; 5) the lesion was asymptomatic and was discovered accidentally; 6) there was neither P.X.E. in the other folds nor angioid streaks; 7) the histological, histochemical and electron-microscopical features (2) allowed one to state that the microscopic structures were quite similar to those of classical P.X.E.

Many of the symptoms described by Christensen were found in our patient, i.e., localization in the cubital fold, reticulate pattern of the yellow plaque, absence of classical P.X.E. in the flexural folds and of angioid streaks, accidental discovery during a routine dermatologic examination, histologic features quite similar to those of classical P.X.E., absence of a family history of P.X.E.

However, some features of Christensen's disease such as history of accidental or professional contact with saltpetre and thread-like margin around the plaque, were not found in our patient. 1) Our patient was a woman. 2) She was not a farmer, but a charwoman. 3) The lesion appeared insidiously without any previous trauma or burning or ulcer. 4) There was no history of accidental or professional contact with saltpetre. 5) no thread-like margin around the plaque.

Therefore we can conclude that our patient is a very unusual case of P.X.E. We believe it could

be a variant of Christensen's exogenous type of P.X.E., whose etiology is unknown.

We think that besides the systemic hereditary endogenous P.X.E., different varieties of non-hereditary localized exogenous P.X.E. can be described, just as concerning ochronosis, both endogenous and the exogenous types can be distinguished.

Diffuse Cutaneous Mastocytosis: A Report of Neonatal Onset

P. V. Harrison,¹ L. J. Cook,² H. J. Lake³
and S. Shuster²

¹Department of Dermatology, The General Infirmary, Leeds, ²Department of Dermatology, Royal Victoria Infirmary, Newcastle upon Tyne, and ³Department of Physiology, University of Newcastle, Newcastle upon Tyne, England

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Abstract. A child developed diffuse cutaneous mastocytosis when 20 days old. His prognosis appears good, compared with other reports of similar neonatal onset.

Key words: Diffuse cutaneous mastocytosis; Neonatal onset

Diffuse cutaneous mastocytosis (DCM) is included in the spectrum of mastocytosis disorders. Nettle-ship (6) first described urticaria pigmentosa in 1869 and the first description of the rare DCM is usually attributed to Dęgos (3) in 1952, although Dowling (4) had described a child who probably had the disease. DCM is characterized by generalized skin involvement and absence of the classical pigmented lesions of urticaria pigmentosa (7).

CASE REPORT

When 20 days old, a male child developed blisters on his hands and erythematous skin elsewhere. He later developed more blisters on the trunk and limbs and it became apparent that he had DCM.

On examination, the skin was diffusely thickened, which in some areas gave it a leathery appearance. No pigmented macules were present and skin folds and creases were exaggerated, particularly on the palms.

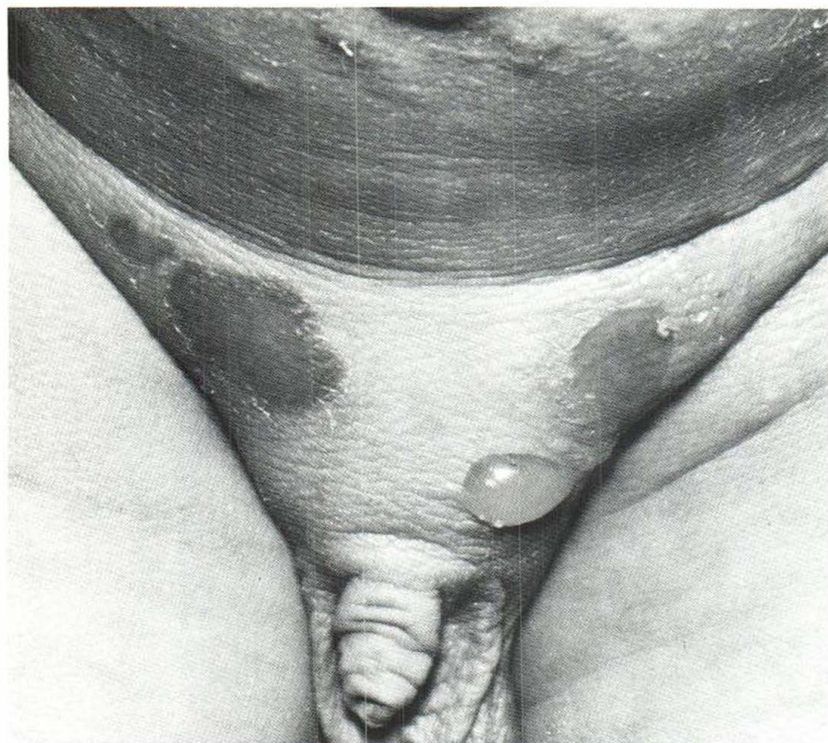


Fig. 1. Trunk, age 10 months, showing bullous lesion.

Blisters were present (Fig. 1) and were in various stages of evolution, some fresh with thin walls and filled with clear fluid, others healing without scarring. When damaged by trauma or scratching, secondary infection of the blisters often resulted. Itching was intense and dermatographism marked (Darier's sign). The liver was enlarged, two fingers below the right costal margin, but there was no splenomegaly.

The following investigations proved normal: full blood count (including film which revealed no mast cells), clotting screen, whole blood histamine levels, 5-hydroxyindoleacetic acid excretion in urine, skeletal survey and bone marrow examination.

Urinary histamine levels, to correlate with the clinical course, were determined fluorometrically after condensation with *o*-phthaldehyde, using an improved version (5) of the method of Shore, Burkhalter & Cohn (9). Unfortunately, the presence of drugs or their metabolites in the patient's urine rendered impossible a reliable estimation of the histamine content.

The skin histopathology showed a diffuse infiltrate of mast cells in the upper dermis and a percutaneous liver biopsy showed an excess of mast cells, located mainly in the portal tracts, without associated fibrosis.

Therapy has included conventional antihistamines, reserpine and the histidine decarboxylase inhibitor tritroqualine (Hypostamine). The child, now aged four and receiving no treatment, shows normal growth and de-

velopment. The skin has no blistering at present and is only slightly erythematous and thickened in some areas. Itching is only slight. The skin has shown a reduced mast cell infiltrate when compared with the previous histology and the hepatomegaly has regressed.

Table 1. *Onset of DCM in neonatal period (1, 2, 4, 8, 10, 11, 12)*

Reports with insufficient data, and in which the distinction between DCM and urticaria pigmentosa is not clear, are not included

Number of patients	8	
Males	6/8	
Bullous lesions	7/8	
Systemic involvement	3/8	Includes one patient who developed mast cell leukaemia and later died
Death	3/8	Causes:
		1. Severe blistering and infection, age 8 days
		2. No apparent cause, age 6 months
		3. Mast cell leukaemia and bleeding disorder, age 5 years

DISCUSSION

DCM is a rare condition. A literature review of DCM with neonatal onset is shown in Table I. This form of DCM is more frequent in males and blistering is common. The prognosis appears to be unfavourable compared with disease onset after the neonatal period, when a good prognosis is expected (7).

The present patient probably has a good prognosis, as illustrated by the clinical improvement and the regression of both cutaneous mast cell infiltration and hepatomegaly. The favourable prognosis may be related to the disease onset occurring late in the neonatal period.

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Dermatomyositis Induced by Penicillamine

N. B. Simpson¹ and J. R. Golding²

Departments of ¹Dermatology and ²Rheumatology, St. James's (University) Hospital, Leeds 9, England

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Abstract. A case of dermatomyositis is reported in a 50-year-old woman receiving D-Penicillamine therapy for rheumatoid arthritis. There was no evidence of neoplasia on full investigation. Remission of dermatomyositis occurred on withdrawal of D-Penicillamine.

Key words: D-Penicillamine; Rheumatoid arthritis; Dermatomyositis

D-Penicillamine has been used successfully in the treatment of Wilson's disease and is being used with increasing frequency in the treatment of rheumatoid arthritis and other 'collagen' diseases. Side effects are unfortunately frequent and have been well reviewed (5).

Less common side effects include the development of other auto-aggressive disorders: Systemic lupus erythematosus (3), myasthenia gravis (2), and polymyositis (1). There has been a single case report of dermatomyositis (4) and we report a second case.

CASE REPORT

A 50-year-old housewife developed a sero-positive rheumatoid arthritis (Rosewaaler 1:512) in April 1976. Treatment with D-Penicillamine was started at 250 mg/day and continued with satisfactory clinical improvement on that dosage. After 19 months of treatment the patient complained of a month's history of tenderness, redness and swelling of the fingers and knuckles of both hands and also redness and puffiness of her face and eyelids. Examination revealed a dusky violaceous discoloration of her eyelids and forehead; dilatation of nail fold capillaries on all fingers with a dusky violaceous discoloration of the skin overlying all metacarpophalangeal joints, and extending along the dorsal aspect of the fingers to overlie the first inter-phalangeal joint. There was no clinically detectable muscle weakness.

Investigations

ESR was 5 mm in 1 hr. Antinuclear factor 1:10, 24-hour urinary creatine 1920 μ mol/24 h (normal <380). Other investigations including haemoglobin, full blood count, serum transaminases, creatine phosphokinase and elec-