

activity of PPP. This is in accordance with most investigators (4, 5).

To minimize the risk of ionizing radiation and for economic reasons, unnecessary radiographic examinations should be avoided. It is our opinion that routine radiographic examinations in patients with PPP are of little value, possibly with the exception of examination of the teeth. The high percentages of asymptomatic root abscesses in patients with PPP as well as in clinically healthy subjects seem to be a mainly odontologic problem.

Most investigators suggest that regular radiographic examinations of the teeth in adult persons should be carried out by the public health service (3, 9).

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Chequered Localized Pseudoxanthoma Elasticum: A Variety of Christensen's Exogenous Pseudoxanthoma Elasticum?

A. Dupre, J. L. Bonafe and B. Christol

Service de Dermatologie, Hôpital La Grave, Toulouse, France

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Abstract. We report a very curious case of a condition which has never been described before. Its features are comparable but not quite identical with those of Christensen's saltpetre-induced PXE which the author considers to be an exogenous variety of pseudoxanthoma elasticum. Since our case appeared spontaneously without any accidental episode we prefer to designate it under the name 'localized PXE'. The chequered appearance of the lesions seems to be the characteristic feature of the disease.

Key word: Pseudoxanthoma elasticum

We report a case which paradoxically cannot be distinguished clinically and histopathologically from pseudoxanthoma elasticum (P.X.E.) but which, nevertheless, is distinct from classical P.X.E. This case could be a variant of the features recently described by Christensen (1) concerning saltpetre-induced P.X.E.

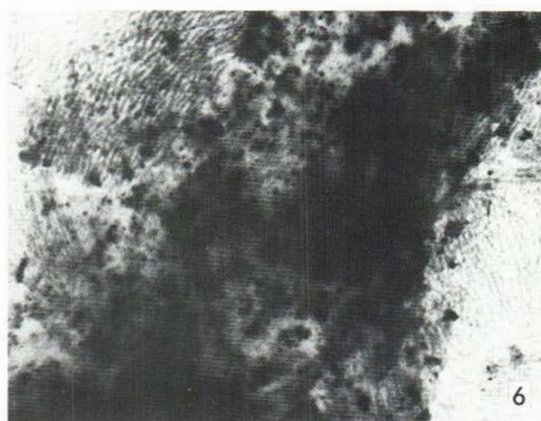
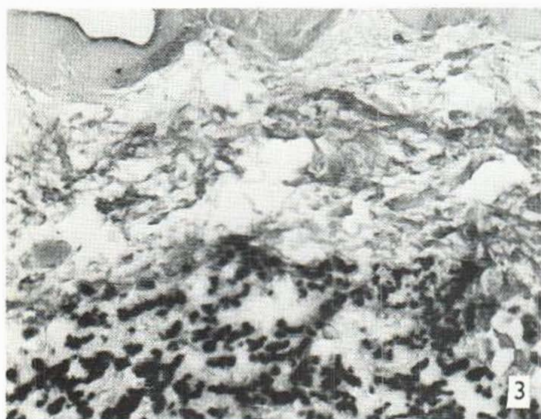
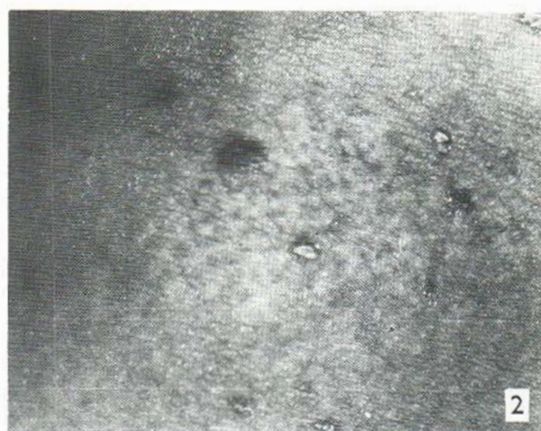
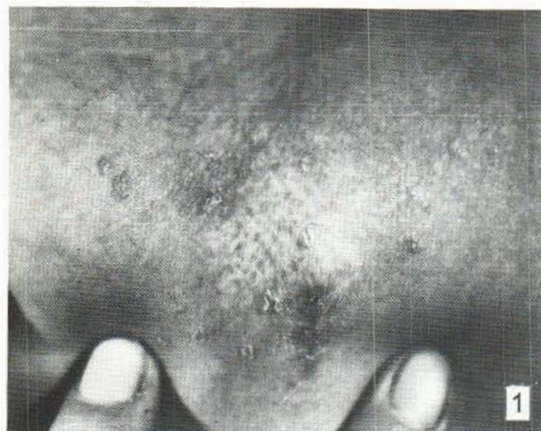
CASE REPORT

The patient, a 40-year-old charwoman, was initially seen for evaluation of a lichen planus. The eruption of lichen planus was located on the wrists and the arms. When examining the patient we were surprised to discover lesions in the right cubital fold which were quite P.X.E.-like. According to the patient no particular cause was connected with the onset of the skin lesions. They had appeared about 10 years previously and had remained stable since then, showing no tendency to disappear spontaneously.

They consisted of isolated yellow-white plaques measuring 3.5–5 cm². The plaques were not surrounded by a thread-like margin and showed a quite particular reticulate and chequered pattern (Figs. 1 and 2).

There was no actinic elastosis of the face and neck, no P.X.E.-like lesions in the flexural folds, no angioid streaks, no cardiovascular abnormalities, no cutaneous extensibility, nor any family history of P.X.E. The lesion did not cause any symptoms. The appearance was quite similar to P.X.E. or flat xanthoma plaques, except that the yellowish coalescing papules were curiously distributed and formed a reticulate and chequered pattern.

A biopsy specimen of a yellow papule examined under optic light was consistent with P.X.E. and showed the



Figs. 1 and 2. View of the right cubital fold showing: (1) some lichen planus papules; (2) a chequered P.X.E.

Fig. 3. Von Kossa's method. Calcium inhibition in the middle and lower thirds of the dermis.

Fig. 4. Verhoeff stain. Strongly positive reaction in the periphery of the elastic fibres.

Fig. 5. Electron microscopy. Twisting of collagen fibrils.

Fig. 6. Electron microscopy showing heavy calcium deposits in the ground substance and in the collagen bundles.

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 angiod 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 angiod 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 Dcgoz (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.