

The present case of PXE-like PDE with characteristic skin changes and corresponding histopathology is unusual since the lesions had been observed already in the first years of life. Most patients with PXE-like PDE reported onset of disease after the fifth decade.^{2,3} Only one case in early middle age has been reported so far.⁴ Because of the late onset of the disease, some authors hypothesize PXE-PDE is related to intrinsic skin ageing and/or extrinsic ageing due to ultraviolet radiation.^{2,3} Using immunohistochemistry, loss of elastin and fibrillin-1 has been observed in PXE-like PDE.⁵ On ageing, however, there is only loss of fibrillin-1 while elastin remains normal or decreased.⁵ So far, one familial case of PXE-PDE has been documented in the literature, suggesting influence of genetic or inheritable factors.⁶ The history of the present case does not favour intrinsic or extrinsic ageing as pathogenetical factor but rather indicate that genetics may be of importance at least in certain cases of PXE-like PDE. Notably, PXE-like PDE is characterized by exclusive female predominance.^{2,3} The differential diagnosis PXE is of great importance in the present case, but one have to consider also other rare elastolytic disorders including linear focal elastosis, white fibrous papulosis of the neck, and mid-dermal elastolysis.^{2,3,7-9} PXE is a rare genetic disorder, caused by a mutation in the ABCC6 gene.^{8,9} The cutaneous lesions in PXE are very similar to lesions seen in PXE-like PDE. The predominant pathological findings of PXE, consisting of fragmentation, clumping and calcification of the elastic fibres, are located in the mid and lower dermis.⁹ As also observed in the present case, the absence or marked loss of elastic fibres in the papillary dermis and the absence of calcifications and fragmentation of elastic fibres are characteristic features of PXE-like PDE.¹⁻³ The collagen fibres are normal. Unlike PXE-like PDE, PXE usually develops during childhood, has systemic involvement, characterized by calcification of the elastic fibres of the skin, retina, and cardiovascular system, which may lead to serious complications.^{2,3} In the present case, there was no evidence for systemic involvement.

In conclusion, we observed a case of early-onset PXE-like PDE. This observation, the exclusive female predominance documented in the literature, and the report of familial PXE-like PDE indicate genetical factors in the aetiopathogenesis of this very rare disease.

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Metal implant-induced skin ulcer mimicking scrofuloderma

Editor

The use of metal implants to reposition and secure fractured bones is very common in primary practice. Wear, breakage, loosening, infection and metal allergy are known to be the major complications.¹ In contrast, non-allergic cutaneous reactions over the implant site are rare. Herein, we report a case of metal implant-induced skin ulcer on the neck clinically mimicking scrofuloderma.

A 66-year-old Japanese woman with dementia was referred to our hospital complaining of a several-year history of recurrent skin ulcers on the left neck. Physical examination revealed 5-mm-diameter and 2-mm-diameter ulcers, with purulent drainage on the left supraclavicular fossa (Fig. 1a). There was neither tenderness nor swelling around the lesion. Our clinical differential diagnosis was scrofuloderma. No bacterial, fungal or mycobacterial infection was identified in culture specimens from the ulcers. Laboratory investigations showed no abnormal haematological results. Computed tomography scan showed no evidence of pulmonary tuberculosis or lymphadenopathy. During the biopsy procedure, we detected metal implants underneath the ulcers (Fig. 1b). We covered the tips of the implants with surrounding soft tissue and sutured the wound. Her family confirmed that the patient had a history of the bone fracture of the left clavicle for which she had received metal implants 25 years earlier. A plain X-ray image revealed broken metal implants (Kirschner

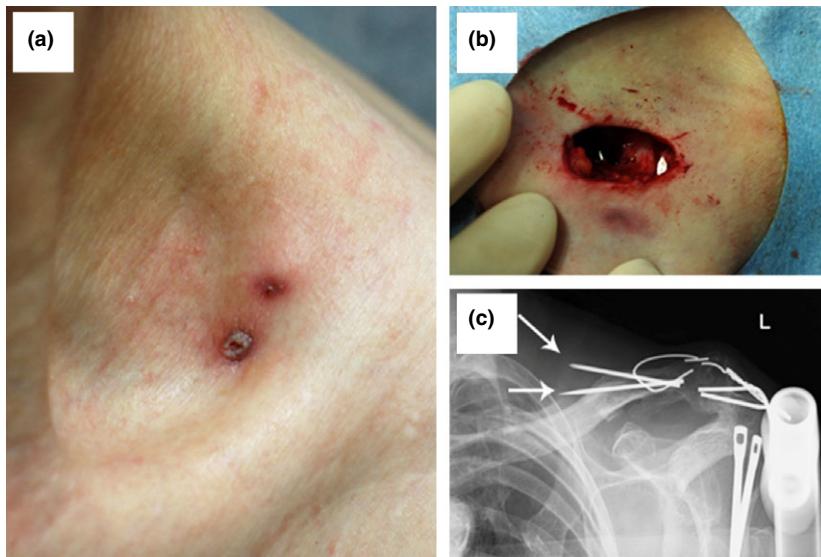


Figure 1 (a) Two ulcers, 5 mm and 2 mm in diameter, are observed on the patient's left supraclavicular fossa. (b) The tips of metal implants were found during the procedure of excisional biopsy of the ulcers. (c) A plain X-ray image revealed broken metal implants in the left clavicle. The tips of the implants were close to the skin surface (arrows).

wire) in her left clavicle. The tips of the implants were turned up toward the skin surface ([arrows], Fig. 1c). Histological specimens showed fibrosis and the infiltration of a few mononuclear cells in the dermis. No granuloma formation was seen, which ruled out both foreign body reaction and mycobacterial infection. No malignant cells were identified in the specimens. We made the diagnosis of metal implant-induced skin ulcer. The implants were removed due to recurrence of the skin ulcers. No relapse of the ulcers was seen during our follow-up in 15 months.

The most important clinical differential diagnosis of this case was cold abscess due to tuberculosis infection because scrofuloderma presents as recurrent ulcerative lesions, especially around the neck, and tuberculosis infections are not yet rare in elderly people in Asia.²

Dermatitis caused by a reaction of the skin to orthopaedic implants was first described in 1964 by Foussereau and Laugier.³ Metal implantation dermatitis has been explained as a hypersensitivity reaction against components of the metal devices.⁴ Eczema is the most frequent clinical manifestation, but urticarial and non-eczematous bullous reactions have also been reported.⁵ Although rare, skin erosions can also develop due to physical irritation from the metal implants when the tips are near the covering skin. The present case showed no signs of metal allergy, represented by erythematous skin lesions above the implants, and the plain X-ray image clearly showed the tips of the metal implants adjacent to the covering skin. Therefore, we made the conclusion of non-allergic skin erosion caused by physical irritation from the metal implants. When clinicians encounter recurrent ulcerative skin lesions around bones, they must be aware of the patient's history of metal implantation and take an X-ray image of the lesions to exclude or confirm the involvement of metal implants.

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Lichen planus pigmentosus-inversus: 5 Turkish cases

Editor

Lichen planus pigmentosus-inversus (LPP-I) is a rare clinic variant of lichen planus pigmentosus (LPP) which particularly affects the intertriginous and flexural areas and skin folds in light-skinned individuals that presents as asymptomatic to