Perforating pseudoxanthoma elasticum of the arms: a rare case report

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under the bed. The lack of definitive guidelines for diagnosis and management reflects the diagnostic difficulties for a not so rare medical condition, sometimes associated with serious complications. ■

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Perforating pseudoxanthoma elasticum of the arms: a rare case report

Pseudoxanthoma elasticum (PXE) is a rare inherited disorder of the connective tissue that causes fragmentation and mineralization of elastic fibres. The coexistence of PXE and elastosis perforans serpiginosa (EPS) is now considered "perforating PXE" (PPXE) [1]. To the best of our knowledge, PPXE is most commonly observed in the periumbilical region. Herein, we present an extremely rare case of PPXE on the arms of a middle-aged woman.

A 43-year-old obese woman presented with symmetric reddish-brown, annular, and pruritic lesions with slightly elevated borders on her upper arms which she had complained about for half a year (*figure 1A, B*). She worked as a cleaner and had no history of practicing sports, sun exposure, medication, or preceding traumas or surgeries. She had no other comorbidities, and her family history was unremarkable. A cutaneous examination revealed well-demarcated, hyperpigmented, atrophic reticulate plaques on both arms and scattered keratotic papules, surrounded by a rim of hyperpigmented skin. The surrounding skin was slack, with multiple striae. No lesions on any other body part, including the neck, abdomen or flexural areas, were identified. The clinical impression was that of tinea corporis, granuloma annular, and perforating dermatoses. A microscopic examination of the fungi on the lesions yielded negative test results, and the complete blood count, liver and renal function, and blood sugar test results were within the normal ranges. Dermoscopy revealed keratotic plugs, telangiectasias, and dotted and hairpin vessels on an erythematous background (*figure 1H*). Histopathological examination of the papule around the patch showed degenerative changes affecting the elastic fibres of the mid dermis. The elastic fibres were shortened, fragmented and curled, giving the appearance of ravelled wool (*figure 1E, F*), and a narrow zone from the dermis to the external area of the epidermis was detected, which was full of degraded elastic fibres. Granular and basophilic debris with calcium deposits were observed. Verhoeff-Van Gieson staining demonstrated that the pseudoxanthoma elasticum was perforated (*figure 1G*). No abnormalities were identified on the ophthalmological examination, chest X-ray, electrocardiograph, echocardiograph, or abdominal ultrasound. A diagnosis of PPXE was made. The patient was treated with topical corticosteroids but without improvement. Two years later, although the patient's cutaneous lesion had enlarged (*figure 1C, D*), the general physical examination did not reveal abnormalities.

PXE is a rare inherited disorder of connective tissue, characterized by the degeneration and calcification of elastic fibres in the ocular, cutaneous, and cardiovascular systems. In the past, perforating PXE was considered to represent PXE coexisting with EPS. However, in 1976, Lund and Gilbert [2] discovered that this was a separate entity. Later, Neldner and Martinez-Hernandez described this condition [3] as "localized acquired cutaneous pseudoxanthoma elasticum" due to a lack of family history and systemic manifestations. Another, more recent, viewpoint suggested that it could be considered as a perforating variant of PXE [4].

The pathogenesis of PPXE is controversial. It may be caused by cutaneous trauma, surgery, traction following ascites, or pregnancy [3]. In our patient, the perforation appeared on both upper arms, and we speculated that this manifestation had been caused by local friction and the environmental conditions of daily living, which were damp and wet.

PPXE typically presents as a well-demarcated hyperpigmented plaque around the umbilicus in middle-aged women, although cases have been reported in two-year-old children [5]. Clinically, the manifestations of PPXE and EPS are similar, but their pathology is different. In PPXE, it is common to observe short, thick, curly elastic fibres, studded with calcium salts, residing in the lower dermis. In EPS, the elastic fibres do not show calcification and are usually present in the upper reticular and papillary dermis. The histopathological features of our patient were consistent with PPXE findings. Therefore, the diagnosis of PPXE was made.

Currently, no established treatment for PPXE exists. Therefore, follow-up is necessary. To the best of our knowledge, this report is the first to describe a case of PPXE occurring bilaterally on the flexion side of the upper arm. At

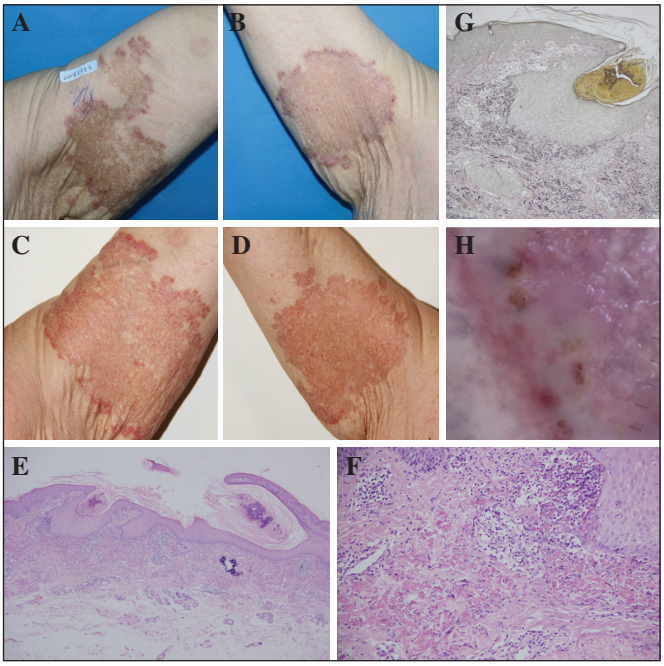

Figure 1. A, **B**) A reticulated reddish-brown plaque with peripheral keratotic papules on each arm. **C**, **D**) After two years, the cutaneous lesions were enlarged. **E**) Histopathology showing degenerative changes affecting the elastic fibres of the mid dermis and granular and basophilic debris with calcium deposits (H&E staining; x100). **F**) Histopathology showing shortened, fragmented and curled elastic fibres, giving the appearance of ravelled wool (H&E staining; x200). **G**) Curled and granular elastic fibres (Verhoeff-Van Gieson staining; x200). **H**) Dermoscopy showing keratotic plugs, telangiectasias, and dotted and hairpin vessels on an erythematous background.

present, our patient has no systemic involvement, but in an earlier case, cutaneous involvement of progressive PPXE, occurring from 12 years of age, was shown to have affected the ocular and cardiovascular functions after 24 years [6]. Patients with perforating PXE should undergo fundoscopy, an electrocardiogram examination, as well as liver and kidney function tests.

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Successful switching to brodalumab in a patient with severe psoriasis developing ixekizumab-induced eczema

A 70-year-old Japanese male with a four-year history of psoriasis vulgaris had been treated with topical maxacalcitol, calcipotriol and betamethasone dipropionate, but his disease gradually deteriorated. He had no past history of atopic diseases. Physical examination revealed sharply demarcated erythematous patches with white scales on his head, trunk and extremities (*figure 1A, B*). A skin biopsy was performed to rule out the possibility of contact dermatitis caused by topical medications. Histopathological examination of a skin biopsy from the right forearm revealed regular acanthosis with rete ridge elongation, thinning of the suprapapillary epidermis (*figure 1C*), hypogranulosis and Munro