

Answer to Dermacase *continued from page 765*

#### 4. Pseudoxanthoma elasticum–like papillary dermal elastolysis

First described in 1992, pseudoxanthoma elasticum–like papillary dermal elastolysis (PXE-like papillary dermal elastolysis) is a rare acquired skin disorder that commonly affects women in late adulthood.<sup>1-5</sup> Clinically, it is characterized by multiple, asymptomatic or slightly pruritic, small, white-to-yellow, nonfollicular papules that might coalesce into large plaques over the neck and supraclavicular areas.<sup>1-5</sup> Less commonly, it might involve the forearms, axillae, inframammary folds, and lower abdomen. The condition might remain stable or progress, with the appearance of new lesions over the course of months to years.<sup>1-5</sup> Histology shows epidermal atrophy and bandlike loss of elastic fibres (confirmed by elastic tissue stain) in the papillary dermis—features that were also present in our patient.

The pathogenesis of PXE-like papillary dermal elastolysis is still not clear. Some believe that it might result from intrinsic aging, ultraviolet radiation, or abnormal formation of elastic fibres. No specific drug or chemical exposure has been associated with this entity.<sup>1-5</sup>

#### Differential diagnosis

Clinical differential diagnosis of PXE-like papillary dermal elastolysis should include syringomas, PXE, and milia.<sup>6-8</sup> Syringomas are sporadic, benign adnexal neoplasms that present most commonly as multiple, small, smooth, firm,



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slightly yellowish or skin-coloured papules over the face, especially the lower eyelids of adult women.<sup>6</sup> Inherited PXE is a rare genodermatosis that commonly manifests in children. Although the cutaneous lesions might clinically resemble those of PXE-like papillary dermal elastolysis, there are usually associated ophthalmic (angioid streaks) or cardiovascular abnormalities (blood vessel calcification that might lead to ischemic or hemorrhagic events). Unlike PXE-like papillary dermal elastolysis, histology of cutaneous lesions of PXE reveals fragmentation and calcification of elastic fibres in the reticular dermis.<sup>7</sup> Milia are benign, small, white, superficial keratinous cysts that usually present on the eyelids, cheeks, and genitalia of children and young adults.<sup>8</sup>

#### Management

The first step in the management of this condition is to exclude PXE. This can be accomplished by demonstrating the absence of calcified elastic fibres on histology and the absence of associated systemic abnormalities. There is currently no effective treatment.<sup>1-5</sup> Topical retinoids have been tried to no avail. Our patient elected to receive no treatment and to use sunscreen. 🌿

**Ms Abdullah** was a registered nurse in the Department of Nursing at the American University of Beirut in Lebanon. **Dr Abbas** is Assistant Professor of Clinical Dermatology in the Department of Dermatology at the American University of Beirut.

#### Competing interests

None declared

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