Exogenous Pseudoxanthoma Elasticum: A New Case in an Old Farmer

Sir,

Pseudoxanthoma elasticum (PXE) is a generalized disorder of the connective tissue, characterized by an involvement of the cardiovascular system, skin and eyes. Five types of PXE have been classified on the basis of the mode of transmission (1) and the clinical picture. The typical histological pattern is characterized by fragmentation, degeneration and swelling of elastic fibres in the mid-dermis and by calcium deposition. Similar cutaneous lesions have also occasionally been reported due to penicillamine (pseudo-pseudoxanthoma elasticum) or to fertilizer contact in elderly farmers (exogenous pseudoxanthoma elasticum) (2, 3).

We report a 60-year-old man with PXE-like lesions localized on his left arm.

CASE REPORT

A crop of asymptomatic papules, 2–3 mm in diameter, was casually observed on the proximal left forearm of a 60-year-old man under treatment for a bullous pemphigoid. The patient stated that these lesions had developed at the age of 9, following burning and redness of the skin near the cubital fold, which had been injured by continuous contact with the metal handle of a bucket containing fertilizer. The patient was a farmer's son who used to keep fertilizer in a bucket supported by his left forearm whilst spreading it with his right hand. He attributed his lesions to the penetration of the fertilizer. We consulted the agronomists of the Institute of Agrarian Chemistry of our university and, on the basis of the period and of the area in which our patient lived, it was supposed that the fertilizer contained calcium nitrate and calcium ciannamidium.

Physical examination of the lateral surface of the left upper limb near the cubital fold revealed a group of annular yellow papules, 2–3 mm in diameter, with a narrow raised border and a depressed centre (Fig. 1). These lesions suggested PXE, but no other lesions were seen.

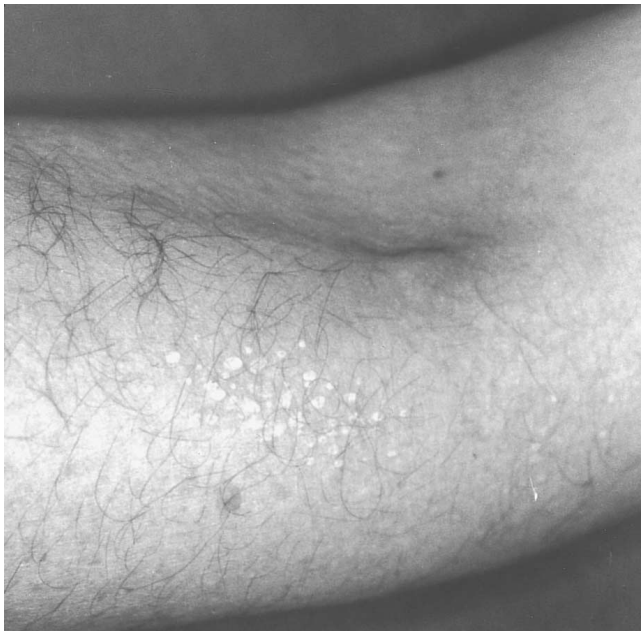

Fig 1. Crop of papules, 2–3 mm in diameter, with a narrow raised border and a depressed centre, on the proximal left forearm.

Moreover, typical bullous lesions on erythematous skin were observed on his upper limbs. Mucosae were not involved. Family history was negative for PXE. Ophthalmological examinations and echocardiogram were normal and routine laboratory tests, including serum calcium and phosphorus, urinary calcium and parathyroid hormone, were within normal limits.

Two lesional biopsy specimens were taken from the proximal left forearm. Staining with hematoxylin-eosin, Von Kossa and Weigert was performed. Histopathology revealed short, enlarged, elastic fibres in the mid-dermis and pigmented particles in the dermis (Von Kossa stain).

Electron microscopy showed important fibril dysplasia of the collagen bundles in the dermis with a "flower-like" pattern. Moreover, the elastic fibres showed notable signs of calcification.

DISCUSSION

In the previously reported 9 male patients the cutaneous lesions were due to exposure to salpeter containing a mixture of various nitrates and appeared as four or five yellow-white plaques with a reticulated surface, a slight central atrophy and a thread-like margin and also as pinhead-sized papules similar to PXE or flat xanthoma plaques. A thread-like margin was also observed in our patient; this is an important morphological feature typical of the exogenous variety of PXE. However Duprè et al. in 1979 (4) described a variant of the exogenous variety of PXE in a woman who was not a farmer, whose lesions had an insidious onset, with no symptoms and without a thread-like margin around the plaque.

Another acquired non-inflammatory skin disorder of the neck and supraclavicular regions closely resembling PXE has recently been described by Rongioletti & Rebora (5) in elderly females. The authors named it PXE-like papillary dermal elastolysis because histological examination showed the complete loss or a marked reduction of the elastic fibres only in the papillary dermis. This disorder is probably due to age-related changes in the dermal fibres. In our case previous damage to elastic fibres due to sunlight exposure or old age can be excluded because of the early onset of the cutaneous lesions.

In any case there is probably a relationship between the clinical picture here described and the traumatic exposure to fertilizer containing calcium salts. Traumatic cutaneous calcium deposition has been reported as the result of minor trauma and/or prolonged exposure to calcium salts. It was first described by Oppenheim in 1935 in an ice-cream maker (6). It was also reported in coal-miners (7), in oil-field workers (8) and due to prolonged contact with electroencephalogram electrode paste (9, 10). Calcium chloride or calcium nitrate were responsible for the lesions occurring in all these cases. Clinically they were very different from PXE and appeared as yellow-white plaques, papules or nodules and, only in one patient, with a follicular pattern. In most cases histology revealed a diffuse deposition of calcium in the dermal connective tissues and not in the elastic fibres.

It is likely that the peculiar clinical aspect of exogenous PXE reported here is due to the chemical qualities of the fertilizer, which is thought to contain calcium nitrate and calcium ciannamidium, similar to the salpeter used by the Scandinavian farmers, but the exact way in which they induce

specific damage to the elastic fibres with calcium deposition is unknown. Moreover, the specific genetic biochemical defect that allows for the calcification of elastic fibres is also unknown (11).

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Congenital Multiple Annular Glomus Tumors

Sir,

The glomus is a specialized arteriovenous shunt, involved in thermal regulation. It is made up of an efferent arteriole, which extends into thick-walled arteriolar segments called Suquet-Hoyer canals, and a venule. Glomus cells are found in layers in the walls of the Suquet-Hoyer canals – the site of A-V anastomosis. Glomus bodies are normally concentrated in fingertips and nail beds. Glomus tumors, first described by Masson (1) in 1924, are rare benign proliferations of glomus bodies. We describe the first case of congenital multiple glomus tumors arranged in an annular pattern and review the literature on congenital multiple glomus tumors.

CASE REPORT

A 3-year-old Hispanic male was evaluated for multiple asymptomatic skin lesions, present since birth, which were slowly increasing in size and number. Past medical history was negative, as was family history of similar lesions. Physical examination revealed multiple bluish-purple macules, 2–3 mm in diameter, in an annular configuration on the right upper (Fig. 1) and lower back and buttock. In addition, small individual bluish macules were scattered bilaterally across the trunk and shoulders, with a total of about 20 lesions. The macules were non-tender and did not blanch on diascopy. Laboratory values, including a CBC, SMA-20, and PT/PTT, were normal. A biopsy of an annular lesion revealed within the superficial and deep dermis, extending to the deep margin of the specimen, irregularly shaped interconnected cystic spaces that in areas had jagged outlines and dissected in between collagen bundles (Fig. 2a). These spaces were empty and were lined by cuboidal cells having round, small and uniform nuclei. The cells were arranged as a single row or multiple layers. In some areas one could appreciate widely spaced slender spindle-shaped cells on top of the cuboidal cells, separating these cells from cystic lumina (Fig. 2b). Immunohistochemical studies demonstrated the cuboidal cells to be vimentin-positive, desmin-negative. These findings were diagnostic of multiple glomus tumors.

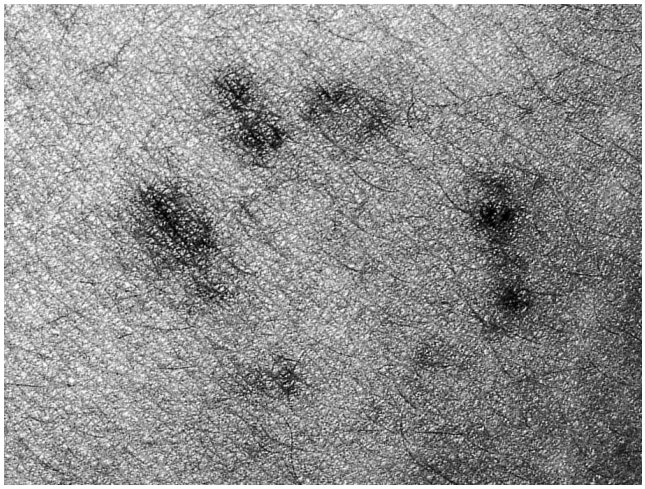

Fig. 1. Annular glomus tumors on the back.

DISCUSSION

Glomus tumors, also known as glomangiomas, are uncommon neoplasms that occur as a solitary tumor or as the rare multiple type. Solitary glomus tumors are usually small bluish painful nodules, situated in the nail bed or on an extremity. The pain can be exacerbated by changes in temperature, pressure and trauma. Subungual solitary tumors are more common in females and develop at an average age of 40, whereas solitary tumors at other sites occur equally in both sexes and occur earlier, at an average age of 25 (2).

In 1937, Weidman & Wise (3) described multiple glomus tumors. Unlike the solitary type, they are often inherited as an autosomal dominant trait and are more common in childhood, with a male predominance. They are generally asymptomatic and not situated in the nail bed. Multiple glomus tumors