Case Report

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Solitary Eccrine Syringofibroadenoma (or Eccrine Syringofibroadenomatous Hyperplasia?) and Diabetic Polyneuropathy

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Key Words

Eccrine syringofibroadenoma
Diabetes
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Abstract

A 70-year-old diabetic woman with sensory polyneuropathy presented with osteonecrosis of the toes and a plaque-like lesion on the dorsum of the ipsilateral foot. Histological diagnosis of eccrine syringofibroadenoma (ES) was made. A review of the literature reveals several cases of solitary ES of the foot in diabetic patients with peripheral neuropathy. This variant of ES seems to be an eccrine sweat duct hyperplasia during the restoration of skin structures damaged by traumas in a situation of peripheral neuropathy. Diabetes and polyneuropathy should be searched for in patients with ES, particularly in acral locations.

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Eccrine syringofibroadenoma (ES) is a distinctive histological entity [1] with polymorphous clinical presentation [2]. Mainly because of such a polymorphism, its nosography is controversial.

The patient we describe provides some clues to discuss the histogenesis and nosography of ES.

Case Report

A 70-year-old woman with diabetes mellitus had had partial osteonecrosis of her toes years before a lesion developed on the dorsum of her foot which slowly enlarged. There was no family history of similar lesions and no evidence of other tumors nor of ectodermal dysplasia. The patient complained of prickling and burning sensations of her legs and feet.

On physical examination, a flesh-colored, verrucous, 8 × 20 cm wide plaque was seen on the dorsum of her right foot extending to the lateral aspect (fig. 1). A chronic ulcer of the sole was also noted. Neurological examination of the lower limbs showed a decreased or absent sensation for touch, pinprick and heat-cold due to a diabetic sensory polyneuropathy, confirmed by nerve conduction studies.

The whole lesion was surgically removed. Histopathology showed an epithelial proliferation with thin, anastomosing strands

of small cuboidal cells extending into the underlying dermis (fig. 2). The epithelial cords contained cystic spaces and ductal structures lined by a single layer of flattened cells and by a

distinct cuticle (fig. 3). The stroma showed an increased vascularity and fibrosis with an inflammatory infiltrate of few lymphocytes, plasmacytes and an exceedingly high number of mast cells, confirmed by to-luidine blue stain. The luminal ductal cells expressed carcinoembryonic antigen.

Discussion

ES was described by Mascarò in 1963 in two 63-year-old patients. They had a solitary nodule on the upper lip and a solitary plaque on the leg, respectively [1]. Both lesions showed a proliferation of anastomosing cords and strands of epithelial cells, ductal aspects within the latter and a fibrovascular inflammatory stroma. About 31 cases have been reported since [2, 3].

Although all these cases have the same peculiar histological pattern, they are clinically polymorphous. They range from a solitary papule, nodule or plaque to multiple lesions with a linear and symmetric arrangement or with a palmoplantar distribution. Even a mucous presentation has been described [4]. The lesions may also involve sites as different as the face, back, abdomen or upper and lower

extremities. ES occurs preferentially in patients between the 5th and 8th decades and evolves within 6 months to 20 years.

The present classification includes solitary ES, multiple ES, multiple ES with hy-drotic ectodermal dysplasia and ES associated with other tumors [5]. Nosography is controversial, however. Solitary lesions of short duration have been considered a neoplasm while multiple symmetric, linear, long-standing lesions have been regarded as hamartoma [6, 7]. Finally, ES has also been considered a nonspecific histological pattern of proliferation reactive to various skin disorders [8, 9].

Accordingly, the solitary plaque-like lesion of a relatively short duration in our patient should be considered a neoplasm. The same clinical presentation was found by Hurt et al. [5] and Sueki et al. [10]. Their 3 patients were middle-aged diabetics who had had osteonecrosis and partial amputation of toes years before they saw a plaque-like ES developing on their dorsal foot.

Altogether, the association of ES with insulin-dependent diabetes has been reported in 5 out of 31 cases in the literature [4, 5, 10, 11, present case]. All patients had their foot involved. A peripheral neuropathy was also observed in another patient affected by lepromatous leprosy [12]. He had a nodular lesion with histological features of ES at the edge of an ulcer in an anesthetic foot [12]. The plaque-like solitary ES of

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Fig.
1. A

solitary, flesh-colored, verrucous plaque on the dorsum of the foot. The toes were involved in previous osteonecrosis.

Fig. 2. An epithelial proliferation of anastomosing strands in the dermis.

Fig. 3. Typical ductal structures within the epithelial cords.

the foot, therefore, may be a reactive hyperplasia of the distal portion of the eccrine sweat apparatus. Such a process may result from the regrowth and restoration of skin structures damaged by traumas in a situation of sensorial neuropathy. Accordingly, diabetes and polyneuropathy should be searched for in ES, particularly in its acral locations.

In addition to our observation, mast cell hyperplasia was signaled only by Sueki et al. [10] (incidentally, their patient had the same clinical presentation as ours). A well-documented phenomenon in wound healing [13],

mast cell hyperplasia may also contribute to form the angiofibrotic stroma which supports the reactive epithelial and eccrine hyperplasia of ES [10]. In case our findings will be confirmed in future, mast cell hyperplasia could help to distinguish between a neoplastic and a reactive hyperplastic ES.

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