Abstract: Eccrine syringofibroadenoma is a rare benign adnexal neoplasm derived from cells of the acrosyringium of eccrine sweat glands. ESFA usually manifests as a solitary nodule on the extremities of elderly patients, but it may also present as papules, nodules or plaques. Its clinical appearance is nonspecific and malignant neoplasms should be considered in the differential diagnosis. However, histopathological findings are typical. The main treatment is surgical excision. In order to illustrate a typical presentation of the tumor, we report a case of solitary eccrine syringofibroadenoma, including the surgical treatment used and its result.

Keywords: Eccrine glands; Skin neoplasms; Surgical flaps

INTRODUCTION

Eccrine syringofibroadenoma (ESFA) is a rare neoplasm considered to originate from the excretory portion of eccrine sweat glands. It was first described by Mascaro in 1963. The clinical appearance of ESFA varies considerably and may include multiple papules, erythematous plaques or a solitary nodule. ESFA is classified into five subtypes according to the clinical presentation. However, its histopathological appearance is peculiar and common to all subtypes. Solitary ESFA is the most common subtype. In order to illustrate a typical presentation of the tumor, we report here a case of solitary eccrine syringofibroadenoma, including the surgical treatment used and its result.

CASE REPORT

A 81-year-old white female patient had had a painful tumor at the lateral malleolus of the right ankle for three years. Physical examination revealed an erythematous tumor with a small central ulceration on the right lateral malleolus (Figures 1 and 2). The following hypotheses were formulated: cutaneous annnex tumor, fibrosarcoma, schwannoma, cutaneous lymphoma, basal cell carcinoma and squamous cell carcinoma. Next, a lesion biopsy was performed. Histopathology showed epidermis-derived, intertwined epithelial cords, ending in thin projections. Some of these projections had a typical ‘crab claw’ appearance. Between these cords, a rich fibrovascular stroma is present (Figure 3). Light formations of the eccrine ducts were identified within some of the cords (Figure 4).
The patient was referred to the plastic surgery service, where surgical excision and reconstruction by a reverse-flow supramalleolar flap were done (Figure 5). Three months postoperatively, the patient was completely recovered (Figure 6).

DISCUSSION
ESFA predominantly affects the elderly, between the seventh and eighth decades of life. The distribution of lesions is wide and includes face, back, abdomen, buttocks, extremities and, rarely, the nails. It is classified into five subtypes according to its clinical presentation: (i) solitary ESFA, a single, non-hereditary, verrucous nodule in elderly patients; (ii) multiple ESFA with Schopf syndrome (an anhidrotic ectodermal dysplasia with hydrocystomas of the eyelids, hypotrichosis, hypodontia and nail abnormalities); (iii) multiple ESFA without cutaneous
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findings, also called syringofibroadenomatosis; (iii) Nonfamilial unilateral linear ESFA, also referred to as nevoid ESFA, and (iii) reactive ESFA, associated with inflammatory or neoplastic dermatoses, such as erosive lichen planus, bullous pemphigoid, burn scar, squamous cell carcinoma and chronic diabetic foot ulcer. The case reported here was classified as solitary ESFA, because it was a single lesion on one of the extremities of an elderly patient.

Histopathology is characterized by anastomosing epithelial cords and columns, surrounded by abundant fibrous tissue. Some of these cords have tubular lights in their interior. The epidermis shows long, thin projections, with confluent terminations and a peculiar appearance, similar to crab claws. The tumor is usually limited to the papillary dermis.

The treatment of choice is surgical excision. Other rarely reported treatments are cryotherapy, curettage, electrodessication, laser and radiotherapy, with variable patient outcomes. In the case reported here, we opted for a reconstruction by a reverse-flow supramalleolar flap because the tumor was bulky and located in an extremity, which makes the reconstruction of the surgical defect difficult, due to poor skin mobility. Furthermore, the use of the flap promotes better coverage of the area and provides a better protection to the bone surface, given that the lateral malleolus is an area that is highly exposed to trauma. Primary closure with a graft would result in a very thin protective coating. Other flaps used for reconstruction of this region are very thick, which makes wearing of some footwear difficult. In addition, they have an unaesthetic appearance. Complications include the association with squamous cell carcinoma and malignant transformation into syringofibrocarcinoma and eccrine porocarcinoma.

The aim of this paper was to report a characteristic presentation of solitary ESFA, including the good results of surgical treatment.

Erratum


Image 2 was switched for image 3 and vice-versa. Figure 2 should show the image of a skin lesion (ankle nodule) and Figure 3 should show the image of the histopathological exam. The legends are correct.