Eccrine Syringofibroadenoma (Mascaró)

An Acrosyringeal Hamartoma

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• **Two patients with eccrine syringofibroadenomas were studied. The first patient had a 3-year history of asymptomatic, unilateral, linear, moist mosaic plaques of the lower part of the left leg and foot; the second patient had a solitary papule on the back. Histologically, both tumors exhibited superficial elongated strands of eccrine ductal epithelium containing occasional lumina lined by an eosinophilic cuticle. The stroma was loose, myxoid, and fibrovascular. The secretory portions of the eccrine glands were ectatic in the first patient but were not seen in the second patient. We believe these lesions represent acrosyringeal hamartomas (nevi), although some authors interpret them as adenomas. The literature seems to indicate that lesions with similar histologic features may be solitary, multiple, or combined with other neoplasms; in other cases, the lesions may possibly represent an unusual morphologic expression of hidrotic ectodermal dysplasia.**

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Eccrine syringofibroadenoma (ESFA) is an uncommon cutaneous adnexal lesion. Histologically, it stimulates the fibroepithelioma (Pinkus) but has eccrine ductal differentiation resembling the acrosyringium. It has been compared with the intracanalicular fibroadenomas of the breast.[1]

We studied two patients with ESFA whose cases, in our opinion, represent the clinical spectrum of the lesions described in the literature but are less histologically florid than some of the previously reported cases.[2] From the assessment of our own experience compared with the literature, we propose that proliferations of the acrosyringium may occur clinically as solitary tumors or as multiple "nevoid" lesions that may be associated with other eccrine tumors or ectodermal dysplasia syndromes. Histologically, the changes are hamartomatous and seem to be more homogeneous from case to case than are the clinical features.

REPORT OF CASES

Case 1.—A 58-year-old, hypertensive, diabetic black man had had portions of the toes from his left foot amputated secondary to peripheral vascular disease several years previously. His chief complaint was a 3-year history of painless lesions initially noted on the lateral aspect of his left foot. These slowly progressed proximally onto the lower part of the left leg. There was no family history of similar lesions, and no evidence of ectodermal dysplasia or other eccrine tumors was noted. On clinical examination, the lesions were pink, opalescent papules and plaques that were moist and spongy to palpation and located on the left foot and leg (Fig 1). The plaques measured from a few millimeters to several centimeters (up to 25 cm) in diameter; they were linearly distributed approximately in the L-5 dermatome (Fig 1, top). The lesions were also present on the dorsa of the first, second, and third toes. There was no surface ulceration or scale. Close inspection revealed a mosaic or "tapioca pudding-like" appearance on the surface of the plaques (Fig 1, bottom). A trophic ulcer, separate from the other lesions, was present on the plantar surface of the left foot. No progression was noted over the course of 1 year.

Case 2.—A 48-year-old white man presented with a solitary 10 × 7-mm papule on the right lumbar region. Follow-up for progression has been unremarkable after 1 year.

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HISTOPATHOLOGIC FINDINGS

Biopsy specimens from both patients were submitted in 4% neutral buffered formaldehyde solution and processed by routine procedures to hematoxylin-eosin slides. In addition, in case one, colloidal iron, reticulin, and elastic stains were obtained.

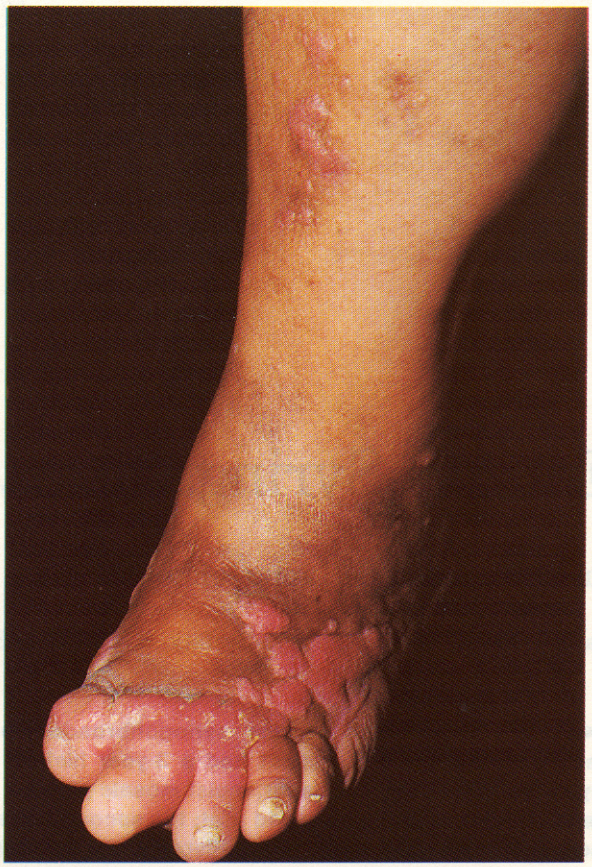
Histologically, lesions from both patients were similar. Sections from the dorsal part of the left foot (patient 1) revealed thin cords of epithelial cells, 2 to 4 cells thick, extending from the epidermis slightly into the reticular dermis. The cords were architecturally parallel or reticular. Cytologically, a monomorphous population of cells, similar to eccrine ductal cells, with areas of duct formation were seen (Fig 2, left). In addition to these epithelial changes, the fibrovascular stroma was rich in acid mucin and reticulin fibers, but devoid of elastic tissue. The eccrine secretory lobule, just deep to the lesion, showed ectatic acini and a mucinous stroma (Fig 2, right).

The specimen from patient 2 showed a multifocal lesion with an epithelial-stromal architecture similar to the "claws of a crab" or a "ball within a claw" (Fig 3, top left and bottom right). One portion showed an ectatic lumen merging with the epidermis (Fig 3, bottom left). Small ducts could be seen within some of the elongated strands. The stroma was angiomatous and myxoid.

COMMENT

A profile of patients with ESFA-like lesions described in the literature are listed with our cases (Table 1).[1-14] Several noteworthy features of the previously reported cases are listed in Table 2.[1-15]

We believe that the two patients described herein are clinicopathologically similar to some of the patients previously discussed in the literature. Both conformed to the established profile for age and time of onset of the lesions. Our first patient clinically resembled the patient of Ogino,[5] manifesting a dermatomal distribution of the lesions on the lower limb, albeit in different dermatomes. Our first patient also seems clinically unique with regard to the extensive plaquelike confluence of the lesions as well as their warmth and moistness. The striking clinical mosaic or "tapioca pudding-like" surface also seemed to be a prominent feature in the lesions of our first patient and, in our opinion, may be an important clue to the clinical diagnosis in lesions that have not developed secondary changes. The ectatic eccrine lobular changes below the acrosyringeal proliferation, seen in our first case, have not been described previously. In Weedon and Lewis' case,[6] several eccrine lobules are seen below the tumor in their Fig 1, but the acini appear to be relatively normal. However, we personally reviewed the histologic features of the lesions of patient 2 in the article by Nordin et al[12] and found that ectatic eccrine acini *were* present within the lobules below the acrosyringeal proliferations in all tumor locations. We view these changes as secondary, most likely related to physiological changes in the affected areas. Although the acrosyringeal proliferations from both of our patients are similar to those in the descriptions of some patients,[4,5,13] the extensive tumorous reticular growth pattern described in other lesions[1,2,6,9] was lacking in our cases. The clinical and histologic differential diagnoses of ESFA are listed in Table 3.[16-26]



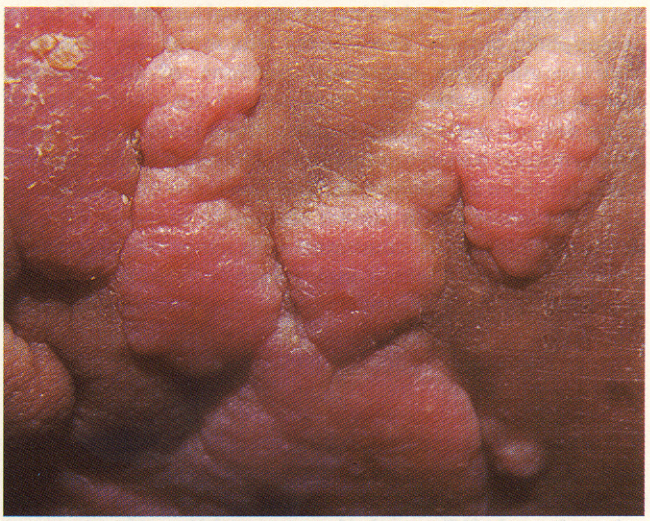


Fig 1.—Top, Patient 1. The lesions are present on the anterior lower part of the limb and dorsal part of the foot. Their distribution approximates the L-5 dermatome. Bottom, Macroscopic view of the plaques shows a mosaic or "tapioca pudding-like" surface. These were warm, moist, and spongy to palpation.

We believe that ESFA is, histologically, a hamartoma (nevus) as opposed to an adenoma. It is a ham-

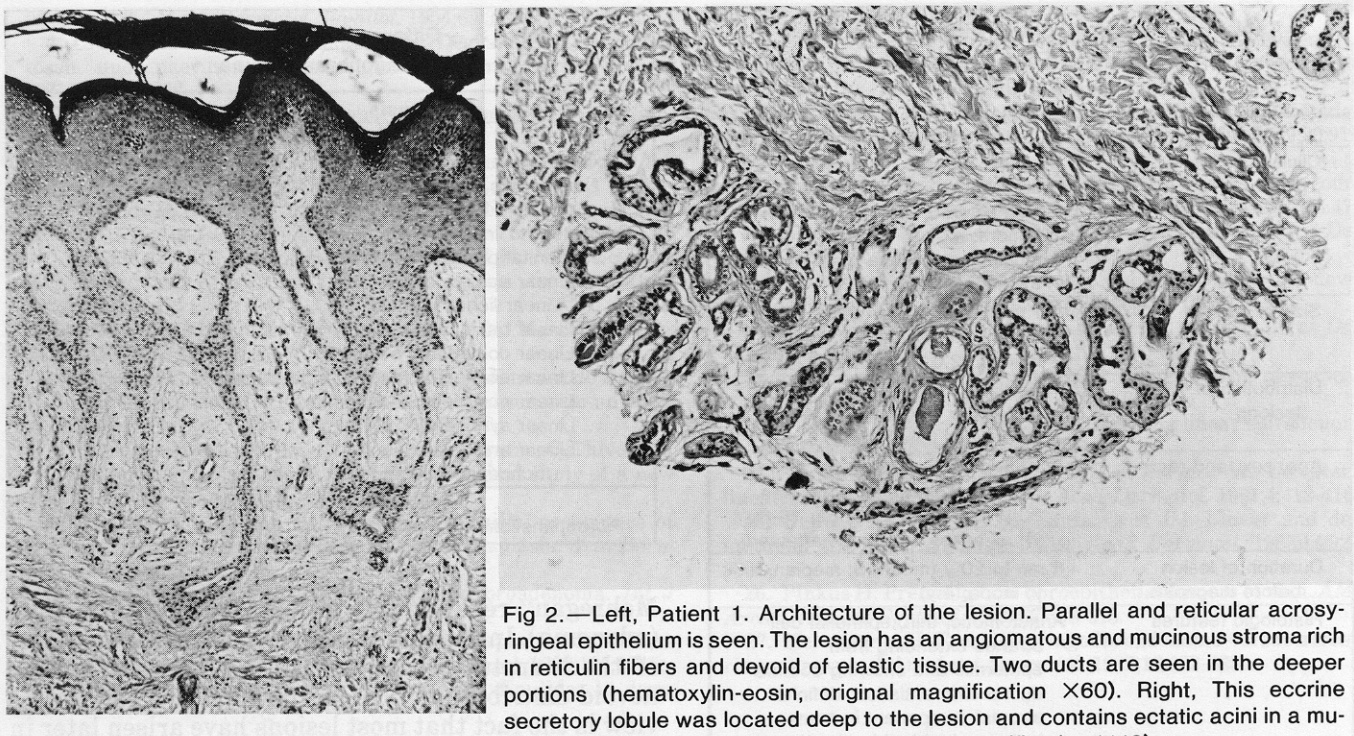


Fig 2.—Left, Patient 1. Architecture of the lesion. Parallel and reticular acrosyringeal epithelium is seen. The lesion has an angiomatous and mucinous stroma rich in reticulin fibers and devoid of elastic tissue. Two ducts are seen in the deeper portions (hematoxylin-eosin, original magnification ×60). Right, This eccrine secretory lobule was located deep to the lesion and contains ectatic acini in a mucinous stroma (hematoxylin-eosin, original magnification ×40).

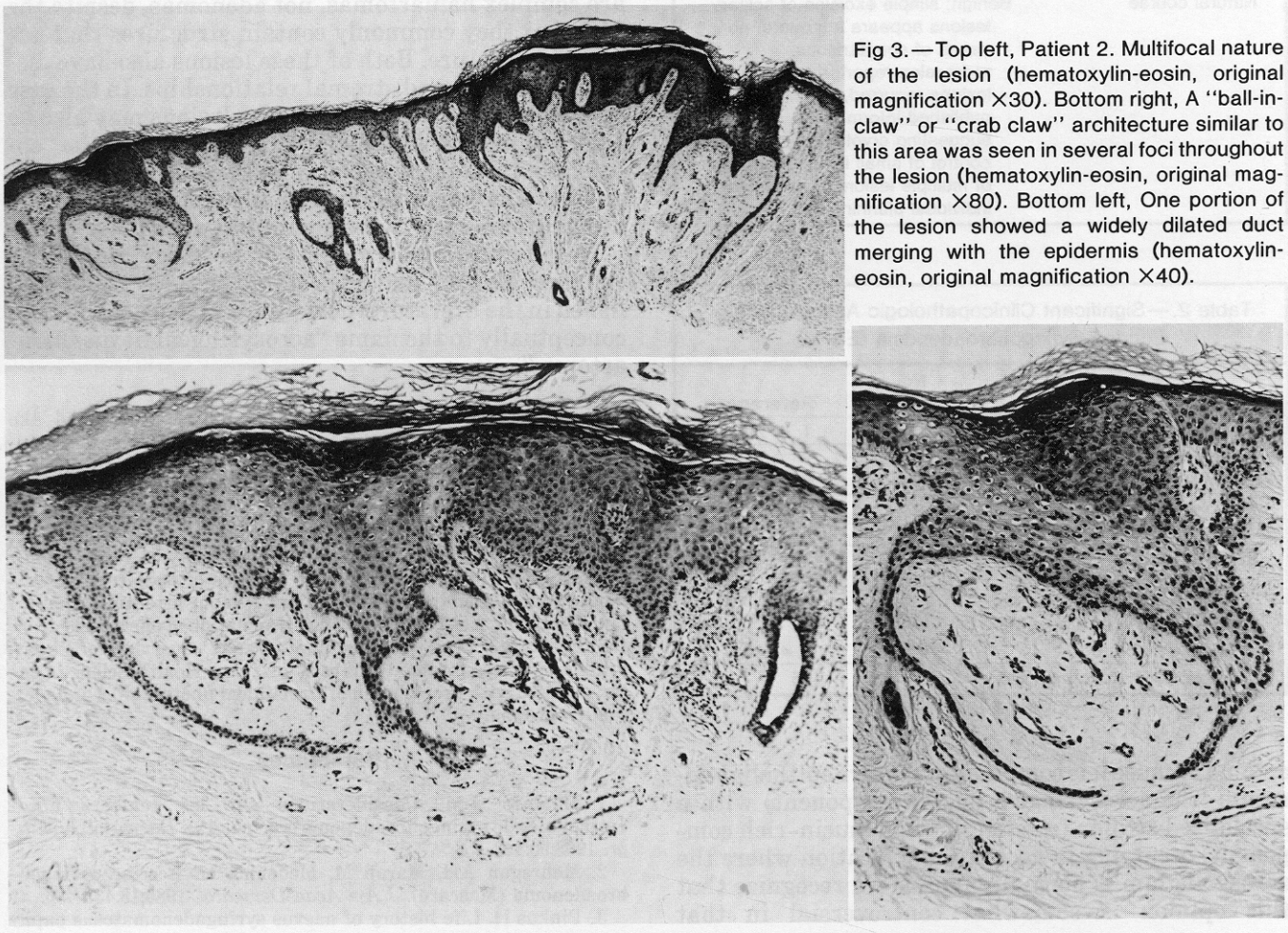


Fig 3.—Top left, Patient 2. Multifocal nature of the lesion (hematoxylin-eosin, original magnification ×30). Bottom right, A "ball-in-claw" or "crab claw" architecture similar to this area was seen in several foci throughout the lesion (hematoxylin-eosin, original magnification ×80). Bottom left, One portion of the lesion showed a widely dilated duct merging with the epidermis (hematoxylin-eosin, original magnification ×40).

Table 1.—Clinicopathologic Features of Eccrine Syringofibroadenoma From the Literature[1-14] and Present Report

Feature	Description
Clinical lesions	Nodules, papules, or fleshy, sometimes spongy, verrucous, skin-colored or erythematous tumors; may be single or multiple; may have other clinical associations (Table 2)
Size of lesions	0.1 to 25 cm in greatest dimension (mean, 4.6 cm; median, 2.0 cm; mode, 2.0 cm)
Distribution of lesions	Face, back, abdomen, buttock, and upper or lower extremity; sometimes linear
Age, sex, and race	5th-8th decade, except for 1 teenager (mean, 61 y; median, 63 y; mode, 63 y; range, 16-80 y); all sexes and races affected
Duration of lesion before diagnosis	6 mo to 20 y (mean, 6; median, 4 y; mode, 4 y)
Histologic features	Anastomotic, thin, epithelial cell strands extending from epidermis and showing eccrine ductal differentiation; definite stromal component in many lesions with "ball in claw" architecture; may be associated with other tumors in some cases (Table 2)
Natural course	Benign; simple excision of solitary lesions appears adequate; no cases of spontaneous regression reported; no excised lesions recurred, according to published information, suggesting cryotherapy for local control of small lesions; removal of multiple lesions may require individual planning

Table 2.—Significant Clinicopathologic Associations of Eccrine Syringofibroadenoma (ESFA)

Feature	No. of Patients	References
Solitary ESFA only	6	1, 2, 9-11
Multiple ESFA without known syndrome	3	5-7, 13
Multiple ESFA with hidrotic ectodermal dysplasia	4	8, 12, 14
ESFA associated with other tumors (papillary syringoadenoma, clear cell acanthoma, and verrucous eccrine poroma)	4	3, 4, 12
Possible ESFA in dog gingiva	1	15

artoma because it is composed of mature epithelial elements (the acrosyringeal-ductal component) with a specialized stroma (a reticulin-acid mucin-rich component) occurring in an anatomic location where the acrosyringium is normally found. We recognize that this opinion is somewhat controversial in that

Table 3.—Differential Diagnosis of Eccrine Syringofibroadenoma

Diagnosis	Reference
Clinical Diagnosis	
Solitary lesion	
Nonspecific (unless mosaic or "tapioca pudding-like" surface is noted)	. . .
Multiple and linear lesions (usually congenital or arise in childhood)	
Linear epidermal nevus	16
"Linear lichen planus"	17
Linear basal cell nevus syndrome	18-20
Linear comedonal nevus	21
Linear eccrine nevus with comedones	22
Linear eccrine spiradenomas	23
Linear syringomas	24
Linear and dermatomal trichoepitheliomas	25
Histologic Diagnosis	
Fibroepithelioma of Pinkus	26

Mehregan[27] regards this lesion as a true neoplasm (adenoma). In support of our view, we refer to several of the literature cases, as well as our case 1, with nevoid distribution of the lesions.[5-8,12-14,27] Against this view is the fact that most lesions have arisen later in life and that some were associated with eccrine poromas. It is widely held, however, that lesions such as nevus sebaceus (Jadassohn) and trichoepitheliomas are complex hamartomas, not adenomas, despite the fact that they commonly contain structures that are less than mature. Both of these lesions also have distinct epithelial and stromal relationships. In the case of solitary trichoepithelioma, the lesions may also be acquired in later life. Thus, we do not believe the "epithelial-stromal" or age argument necessarily distinguishes a hamartoma from an adenoma; in fact, these designations seem to be somewhat arbitrary.

Despite these controversies in nosology, we believe the term *ESFA* should be retained as it is well established in the literature; however, we have no objection conceptually to the name "acrosyringeal nevus/hamartoma."

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