Multiple Syringomas on the Abdomen, Thighs, and Groin

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GOAL

To provide information on the clinical characteristics, epidemiologic features, and pathologic findings in multiple syringomas.

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Identify the predilection and incidence of multiple syringomas and their variants.
2. Describe the clinical presentation of syringomas.
3. Outline the histochemical and electron microscope findings associated with syringomas.

CME Test on page 269.

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Syringomas are benign adnexal tumors that occur most commonly in women. They typically present as soft, flesh-colored to slightly yellow papules on the lower eyelids. We present an unusual case of a healthy 33-year-old male with multiple, reddish brown syringomas located on the lower abdomen, thighs, and groin. Although these lesions can result in significant cosmetic disfigurement, treatment options are limited and generally disappointing.

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Syringomas are benign adnexal tumors derived from the intraepidermal portion of eccrine sweat ducts. Classically, they present as soft, flesh-colored to slightly yellow dermal papules on the lower eyelids of healthy individuals.[1] Syringomas can also affect other areas, including the cheeks and neck; they appear less often on the forehead, hands, upper extremities, ankle, axillae, trunk, genitals, and scalp and may result in cicatricial alopecia.[2-9] Presentation on the abdomen, thighs, and groin with a reddish brown color, as in our patient, is rare.

Case Report

A 33-year-old, healthy, white male presented with a 5-year history of eruptions that began as a few papules on the lower abdomen and gradually spread to a larger area on his lower body. The lesions were asymptomatic

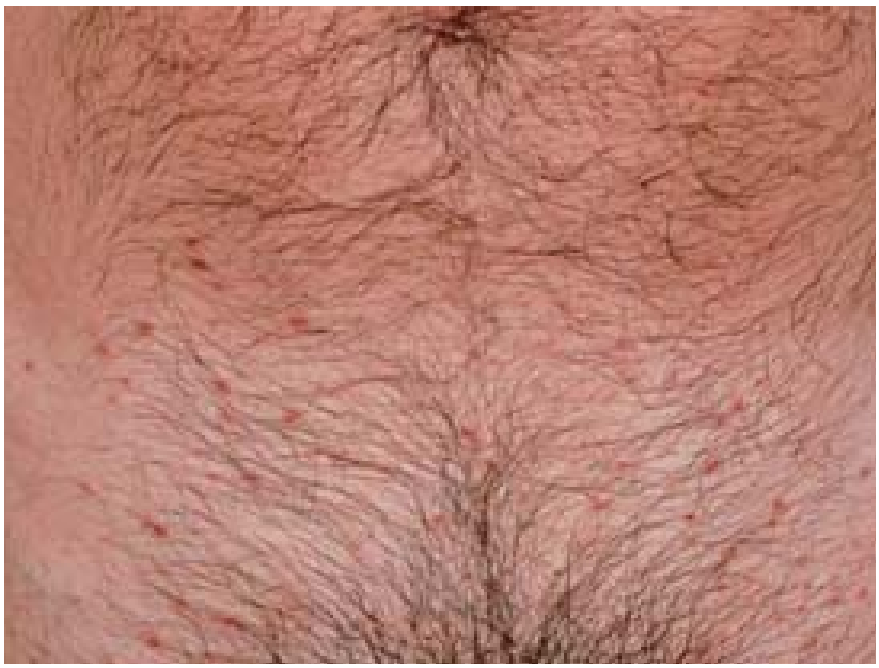

FIGURE 1. Multiple reddish brown papules on the groin.

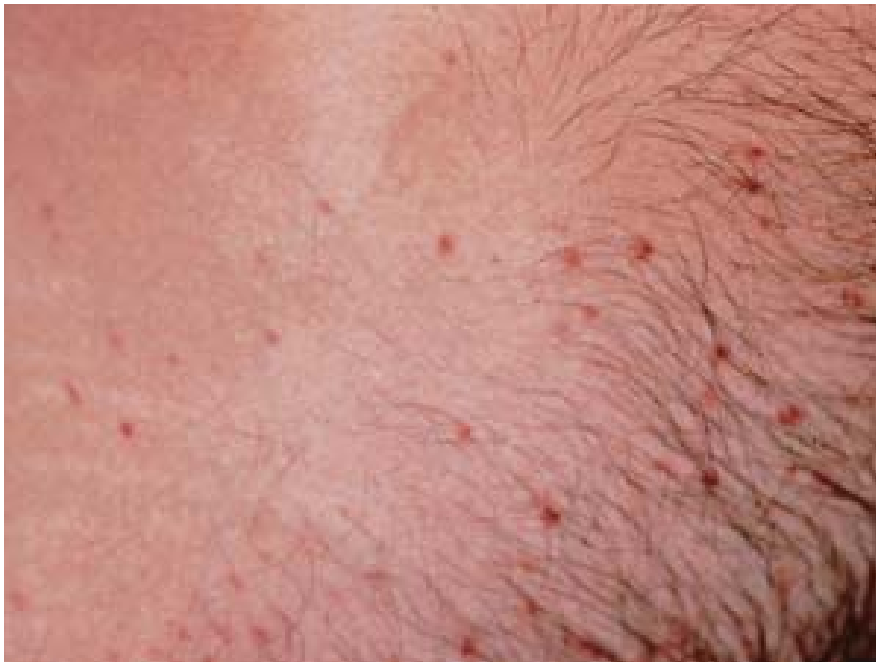

FIGURE 2. Individual papules are bilateral, symmetrical, and both follicular and nonfollicular in distribution.

and, since they first appeared, the patient's skin had never been completely clear. He was not on any medications, no family member had ever had a similar skin condition, and a review of systems was noncontributory.

Physical examination revealed multiple, reddish brown, flat-topped papules 1 to 3 mm in diameter on the lower abdomen, thighs, and groin (Figure 1). The lesions were bilateral, symmetrical, and had both a follicular and nonfollicular distribution (Figure 2). No puncta or significant surface changes were noted and Darier's sign was negative. The remainder of the physical examination was unremarkable.

A punch biopsy specimen from the right thigh revealed a normal epidermis overlying a dermis that was filled with multiple ducts embedded in a fibrous stroma. The ducts were lined by an inner layer of clear cuboidal cells and outer layers of flattened epithelial cells. Some had a tadpolelike appearance due to the presence of a commalike tail that was formed by cells projecting from one side of the duct into the stroma.

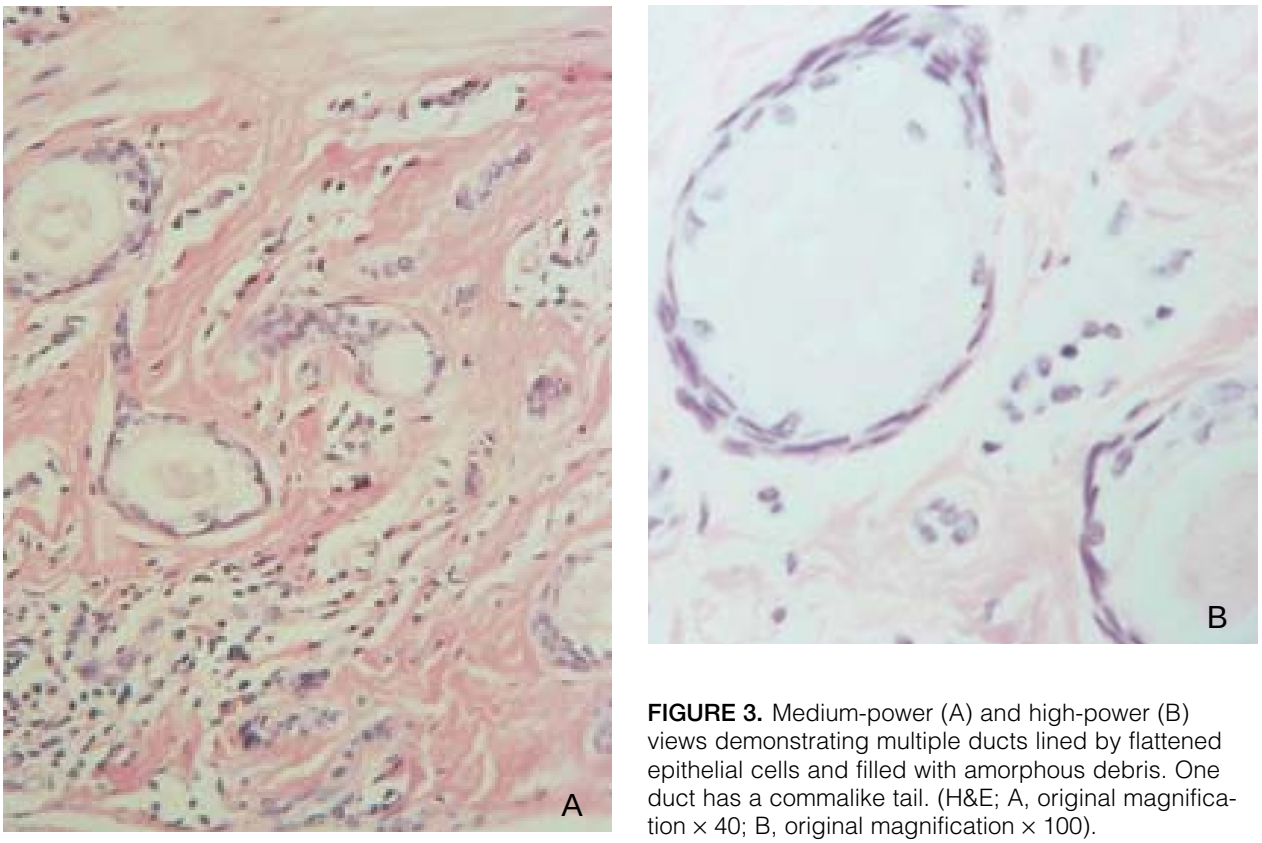

FIGURE 3. Medium-power (A) and high-power (B) views demonstrating multiple ducts lined by flattened epithelial cells and filled with amorphous debris. One duct has a commalike tail. (H&E; A, original magnification × 40; B, original magnification × 100).

Ductal lumina were filled with amorphous debris (Figure 3, A and B). These histologic features are typical for syringomas.

Comments

Typically, syringomas demonstrate a female predilection, usually with spontaneous onset in puberty or the third or fourth decade of life. They are most commonly observed in Japanese women, who have a 6-fold higher incidence of syringomas than white or African American women.[10] A few familial cases have also been reported.[11,12] Interestingly, syringomas occur in 18% of individuals with Down syndrome, leading some to propose including them among the diagnostic criteria for this disease. In these patients, a female predominance is maintained, with syringomas demonstrating a 2:1 female-to-male ratio.[13]

Several variants of syringomas exist. Eruptive syringoma is a rare variant that typically presents in children between the ages of 4 and 10 years.[14] It is characterized by numerous papules arising in successive crops on the anterior surfaces of the body. Nicolau and Balus' syndrome, which is extremely rare, is characterized by the combination of eruptive syringomas, milia, and atrophoderma. An unusual histologic variant, the clear-cell syringoma, is associated with diabetes mellitus.[15] There has also been one case reported of syringomas on the upper extremities associated with a carcinoid tumor.[16]

Syringomas typically present in a bilateral, symmetrical distribution, but there have been reports of unilateral, unilateral linear nevoid, bathing trunk, and generalized distributions.[17-21] Clinically, syringomas may be mistaken for acne vulgaris, sebaceous hyperplasia, milia, lichen planus, eruptive xanthoma, urticaria pigmentosa, or hidrocystoma. However, syringomas demonstrate distinctive histopathologic features. Examination of the dermis demonstrates numerous, small ducts lined by a double row of flattened epithelial cells. Often, the outer layer extends into the surrounding stroma, forming a commalike projection. Ductal lumina are filled with an amorphous, periodic acid-Schiff–positive material.

Histochemical and electron microscopic findings have confirmed that syringomas represent adenomas of eccrine sweat ducts. Histochemically, syringomas react strongly with S100, the carcinoembryonic antigen; epithelial membrane antigen; lysozymes; and antibodies to the breast cystic fluid proteins GCDFP-15 and GCDFP-24.[6] In addition, eccrine sweat gland enzymes, such as succinic dehydrogenase, phosphorylase, and leucine aminopeptidase, predominate in syringoma cells.

Cosmetic treatment for syringomas has included dermabrasion, various methods of excision,

cryosurgery, electrodessication, and oral and topical retinoids.[22-24] Recently, successful treatment of facial syringomas with the carbon dioxide laser has also been reported.[25] Unfortunately, there is no satisfactory treatment for widespread syringomas, as in our patient, because any method of surgical or chemical destruction carries the risk of scarring. Furthermore, there is a high rate of recurrence.

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