

What Is Your Diagnosis?



A 45-year-old woman presented with multiple flesh-colored to slightly hyperpigmented papules on the face, neck, axillae, chest (left), cubital fossae, medial forearms (right), anterior abdomen, and groin. The papules were approximately 3 to 4 mm in diameter and were not painful or pruritic. The papules first appeared at 13 years of age and became enlarged during her premenstrual cycle. Her medical history was otherwise unremarkable. A 4-mm skin biopsy was taken from the right forearm.

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The Diagnosis: Eruptive Syringoma

Syringoma is a benign eccrine gland tumor that appears as a single papule or cluster of papules. There are 4 types of syringomas that are categorized based on location and association: localized syringoma, which is the most common and usually presents on the lower eyelids; eruptive syringoma, a rare variant that presents with a wider and more varied distribution of papules; familial/hereditary syringoma; and trisomy 21-associated syringoma.

Eruptive syringoma was first described by Jacquet and Darier¹ in 1887. In this rare variant, successive crops of numerous, small, firm, flesh-colored to brown-pigmented papules typically appear during childhood or adolescence (Figures 1 and 2). The distribution of papules usually is bilateral and symmetrical. There is a predilection to the superior and anterior surfaces of the body, namely the flexor

aspects of the upper arms, chest, shoulders, abdomen, and eyelids. A younger age of onset is associated with involvement of the groin and axillae.² The papules tend to remain stable throughout adulthood; however, regression is a rare possibility.

The differential clinical diagnosis generally includes eruptive vellus hair cysts, steatocystoma multiplex, disseminated xanthoma, and disseminated granuloma annulare.² A skin biopsy provides valuable additional information. Multiple small and dilated eccrine ducts within a dense fibrous stroma are characteristic histologic findings of syringomas. These ducts are lined with 2 rows of elongated epithelial cells, with the outer layer in a comma-like configuration. Histologically, the appearance of syringomas is similar to desmoplastic trichoepithelioma, morpheaform basal cell carcinoma, and microcystic adnexal carcinoma. Correlation between histological and clinical features is needed to establish a definitive diagnosis. In our patient, histologic examination of the dermis revealed small comma-shaped tubules of epithelial cells, many with central lumina and luminal cuticles dispersed throughout a collagenous stroma (Figure 3). The epithelial cells were polygonal or flattened and had eosinophilic to clear cytoplasm. They demonstrated sharp circumscription and lacked nuclear atypia or infiltration. No nucleoli or mitoses were seen.



Figure 1. Multiple papules on the neck and chest wall.



Figure 2. Multiple papules on the right medial forearm.

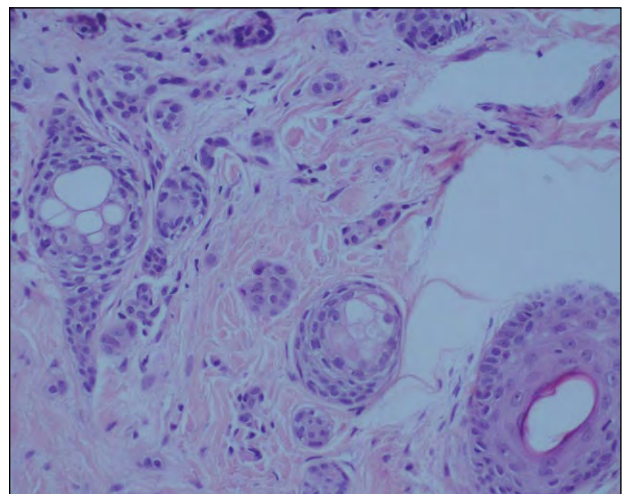


Figure 3. Biopsy of the chest wall revealed small comma-shaped tubules of epithelial cells, many with central lumina and luminal cuticles dispersed throughout a collagenous stroma (H&E, original magnification $\times 20$).

The pathogenesis of eruptive syringoma is unclear. Classically, syringomas are described as benign neoplasms involving the intraepidermal portion of the eccrine glands; however, recent observations of atypical presentations of eruptive syringomas suggest a primary inflammatory response resulting in structural eccrine duct changes as an alternative hypothesis.³ Additionally, the development of eruptive syringoma may be influenced by hormones, as progesterone receptors are highly expressed in a large proportion of syringoma biopsies.⁴ Increased frequency in females with the usual age of onset around puberty and papular enlargement during the premenstrual period also suggest hormonal involvement.

The primary treatment goal is cosmetic. Unfortunately, there currently is no intervention that yields consistent satisfactory results in patients with eruptive syringoma. Although some case reports have noted variable positive outcomes using the CO₂ laser, chemical peels, and topical tretinoin, the risk for scarring and recurrence of symptoms should be carefully considered prior to beginning treatment.^{5,6} After a discussion of the available treatment options, many patients deny intervention and are content with the reassurance that the lesions are benign.

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