

Can You Identify the Papules Below This Woman's Eyes?

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A 25-year-old woman presented with multiple flesh-colored papules in the infraorbital areas. The papules had developed slowly over the past few years and gradually had increased in size and number. The lesions were asymptomatic but were of cosmetic concern. Her past health and family history were not contributory.

Physical examination revealed multiple, skin-colored, smooth, soft to firm papules, measuring 1 to 3 mm in diameter. The papules were symmetrically distributed in the infraorbital areas bilaterally. No similar lesion was observed elsewhere on the woman's body. The rest of the physical examination findings were normal.

What's Your Diagnosis?

- A. Milia
- B. Xanthelasma
- C. Syringomas
- D. Molluscum contagiosum

Answer: Syringomas

Syringomas are common benign sweat gland tumors that originate from the straight portion of the intradermal eccrine sweat duct.¹ The condition was first described by Kaposi in 1872.² Clinically, syringomas present as skin-colored to slightly yellowish papules, commonly found in the periorbital areas.

EPIDEMIOLOGY

Syringomas usually appear at puberty or in the third and fourth decades of life.¹ The reported prevalence is 0.6% of the population.³ The prevalence is higher in the Asian and African American populations.⁴ The female to male ratio is approximately 2 to 1 in most studies; however, a female to male ratio of 6.6 to 1 also has been reported.¹

Most cases occur sporadically, although familial cases have been described.⁵ Familial cases likely have an autosomal dominant mode of inheritance.^{5,6} The condition is more common in persons with Down syndrome, Ehlers-Danlos syndrome, Costello syndrome, Marfan syndrome, Nicolau-Balus syndrome (syringomas, milia, and atrophoderma vermiculata), and diabetes mellitus.⁷

PATHOGENESIS

The exact pathogenesis of syringomas is not known. It has been hypothesized that the condition may represent a hyperplastic response of the eccrine sweat duct to an inflammatory response.⁷ Obstruction of the upper eccrine sweat duct by keratin plugs, leading to ductal proliferation, also may play a role.⁶ A hormonal factor cannot be excluded, since the condition is more common in women and has a peak incidence during puberty.¹

HISTOPATHOLOGY

Histologically, multiple small and dilated eccrine ducts within a dense fibrous stroma are characteristic features.⁸ The eccrine ducts are lined by 2 rows of cuboidal to flattened epithelial cells, the outer layer bulging outwards and giving

rise to the characteristic comma-like tail or "tadpole" appearance.⁸ Ductal lumina are filled with an amorphous, periodic acid-Schiff-positive material.⁹

CLINICAL MANIFESTATIONS

Typically, syringomas present as small, soft to firm, skin-colored to slightly yellowish papules.^{9,10} The papules usually are 1 to 3 mm in diameter, asymptomatic, and symmetrically distributed. The lesions may be solitary or, most often, multiple. The distribution may be localized or generalized. Localized syringomas are the most common clinical variant, and the lesions usually are found in the periorbital areas as illustrated in the case presented here. Generalized syringomas are found mainly on the chest and neck, followed by the forearms.⁹ However, syringomas may appear on other body areas such as the penis, axillae, and buttocks.

The generalized form encompasses eruptive syringomas. Eruptive syringomas typically present as multiple yellow-brown papules in large numbers and in successive crops on the anterior parts of the neck, chest, abdomen, axillae, and upper extremities, in addition to the usual site on the face.⁹ In contrast to classic syringomas, eruptive syringomas occur most frequently before puberty.³ Other rare variants include lichen planus type, alopecia type, and unilateral linear nevoidal type.

DIAGNOSIS

The diagnosis usually is clinical, especially if lesions occur in the classic periorbital location. Skin biopsy occasionally is warranted if the diagnosis is in doubt and to establish a definitive diagnosis.

Differential diagnoses include milia, xanthelasma, eruptive xanthoma, trichoepithelioma, sebaceous hyperplasia, seborrheic keratoses, molluscum contagiosum, acne vulgaris, Langerhans cell histiocytosis, and folliculitis.^{7,10}

COMPLICATIONS

Syringomas can be aesthetically disturbing and socially embarrassing for patients with them. This is particularly true

of facial lesions. Syringomas generally are more bothersome for affected women. The condition has a negative impact on the quality of life and may result in psychological disturbances.

MANAGEMENT

The condition is benign and usually asymptomatic. As such, treatment is primarily for cosmesis. A number of treatment modalities are available, including surgical excision, laser surgery, electrodesiccation, dermabrasion, chemical peeling, cryotherapy, topical tretinoin, and combinations thereof.⁷ Referral to a dermatologist should be considered if the diagnosis is in doubt or if treatment is sought. ■

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