An Atlas of Lumps and Bumps: Part 20

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Syringomas

The word "syringoma" is derived from the Greek word "syrinx" meaning tube or pipe.[1] Syringomas are common, benign, sweat gland tumors that originate from the straight intradermal portion of the eccrine sweat duct.[2,3] Histologically, multiple small and dilated eccrine ducts within a dense fibrous stroma are characteristic features.[4]

Syringomas usually present during puberty or the third and fourth decades of life.[3,5] The reported prevalence ranges from 0.6% to 1% of the population.[1,6] The prevalence is higher in Asians and Africans.[7] The female-to-male ratio is approximately 2:1 in most studies.[3] Most cases occur sporadically, although familial cases have been described.[8] Familial cases may have an autosomal-dominant mode of inheritance.[8,9] The condition is more common in patients with Down syndrome, Ehlers-Danlos syndrome, Costello syndrome, Marfan syndrome, Nicolau-Balus (syringomas, milia, and atrophodermia vermiculata) syndrome, alopecia (cicatricial alopecia and rarely alopecia areata), and type 2 diabetes.[10,11]

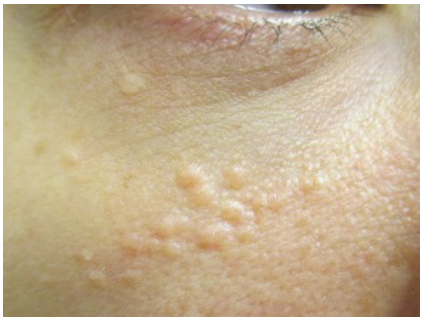

Figure 1. Syringomas present as small, soft, skin-colored to slightly yellowish papules.

Typically, syringomas present as small, soft, skin-colored to slightly yellowish papules **(Figures 1 and 2)**.[5,12] The papules are usually 1 to 3 mm in diameter, asymptomatic, and symmetrically distributed.[3] 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 are typically observed in the periorbital region, especially infraorbitally **(Figures 1 and 2)**.[13,14] Generalized syringomas are found mainly on the neck and trunk, followed by the forearms **(Figures 3 and 4)**.[5] However, syringomas may appear on other body areas such as the penis,

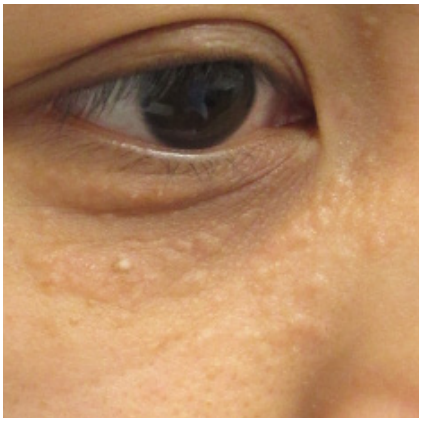

Figure 2. Syringomas are usually 1 to 3 mm in diameter, asymptomatic, and symmetrically distributed.

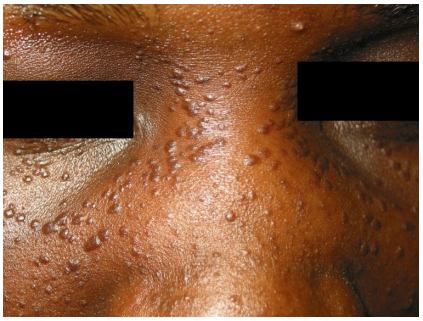

Figure 3. Generalized syringomas can present on the face but are more common on the neck and trunk.

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EDITOR'S NOTE:
This article is part of a series describing and differentiating dermatologic lumps and bumps. To access previously published articles in the series, visit https://www.consultant360.com/resource-center/atlas-lumps-and-bumps.

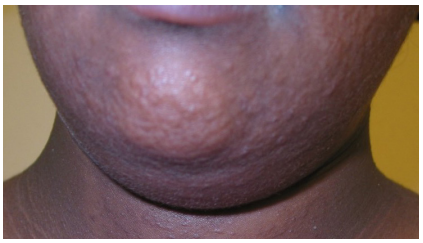

Figure 4. Generalized syringomas are found mainly on the neck and trunk, followed by the forearms.

vulva, axillae, and buttocks.[13,15-17] Generally, patients with generalized syringomas are younger than those with localized syringomas.[5]

Eruptive syringoma is a rare variant that typically presents as multiple skin-colored, pink, or yellow-brown papules and appear in successive crops on the anterior parts of the neck, chest, abdomen, axillae, upper extremities, and genital areas in addition to the usual site on the face.[2,5,14,18,19] In contrast to classic syringomas, eruptive syringomas occur most frequently before or around puberty, especially after an inflammatory condition, and may be associated with pruritus.[2,6,18] Some authors categorize eruptive syringomas as a hyperplastic response to an inflammatory reaction rather than a true adnexal neoplasm.[20] Other rare variants include plaque-like, urticaria pigmentosa-like, lichen planus-like, milia-like, and unilateral linear nevoidal-like syringomas.[11,21,22]

Milia

Milia are benign, transient, superficial keratinous cysts.[23] They present as asymptomatic, small (generally less than 3 mm), firm, white to yellow, smooth, dome-shaped papules.[24] Histologically, they appear as small infundibular cysts that are lined with stratified squamous epithelium with a granular cell layer.[25] The cyst contains laminated layers of keratin. Milia may arise spontaneously without a known cause (primary milia) or secondary to various processes (secondary milia). Primary milia may be congenital (congenital primary milia) or have an onset later in life (benign primary milia of children and adults).

Congenital primary milia are present in

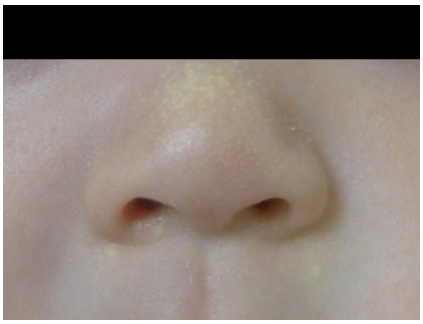

Figure 5. Congenital primary milia favor the nose.

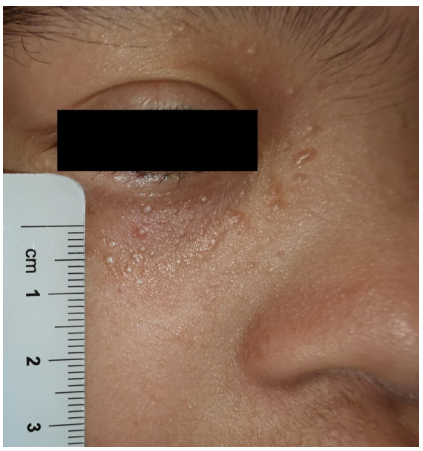

Figure 6. Benign primary milia of children favor the eyelids.

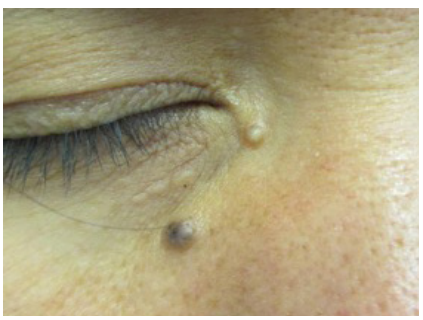

Figure 7. Benign primary milia of adults favor the eyelids and periocular areas.

40 to 50% of newborn infants with no sex predilection.[26] The condition is less common in premature infants.[23,25] Its onset in premature infants could be delayed by several weeks.[23] While congenital primary milia favor the nose **(Figure 5)**, benign primary milia of children and adults favor the eyelids, periocular areas, cheeks, and forehead **(Figures 6 to 8)**.[23,24] Other unusual sites of involvement include the nasal crease, trunk, extremities, vulva, glans

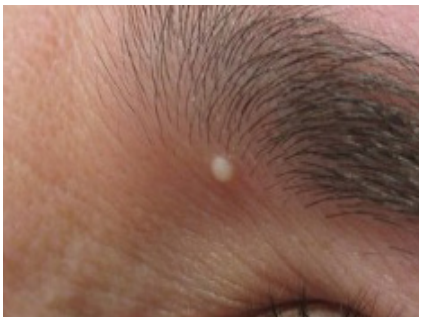

Figure 8. Benign primary milia of adults favor the eyelids and periocular areas.

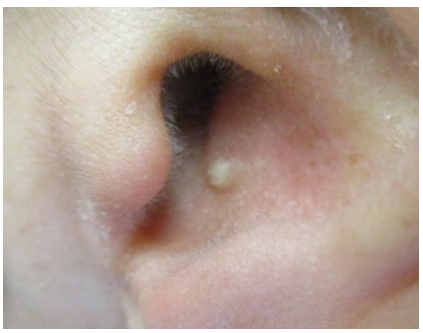

Figure 9. Benign primary milia of adults favor the eyelids and periocular areas.

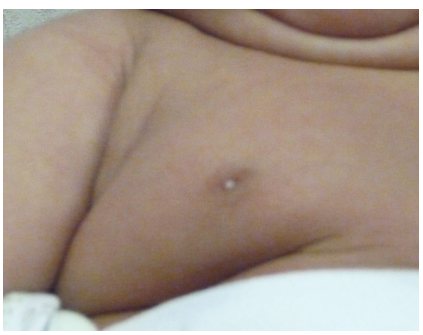

Figure 10. Unusual sites of involvement include the areola.

penis, auricle, and areola **(Figures 9 and 10)**.[23-25,27,28]

Primary milia may be a feature of trisomy 13, Gorlin-Goltz syndrome, Rombo syndrome, basal cell nevus syndrome, Bazex-Dupre-Christol syndrome, Basan syndrome, Brooke-Spiegler syndrome, Marie-Unna hypotrichosis (congenital hereditary trichodysplasia), Nicolau-Balus syndrome, pachyonychia congenita type 2 and oro-facial-digital syndrome type 1.[23,25,29,30]

Multple eruptive milia and milia en plaque are considered clinical variants of milia.[31,32] Multiple eruptive milia are characterized by a sudden onset of multiple asymptomatic milia over a period of weeks to months, mainly on the head, neck, and trunk.[31] Milia en plaque is characterized by numerous tiny milia on an erythematous plaque.[32-34] The condition is asymptomatic and is most common in middle-aged individuals with a female predominance and no predilection for race.[32] The periocular area is most commonly affected.[33] Other sites of involvement include the postauricular area, nasal bridge, cheek, submandibular area, supraclavicular area, trunk, arms, and lower limbs.[32]

Secondary milia usually occur in older children.[23] They may occur in association with systemic disease (eg, porphyria cutanea tarda, epidermolysis bullosa, lichen planus, discoid lupus erythematosus, bullous pemphigoid,), medication use (eg, topical corticosteroids, oral cyclosporine, oral 5-fluorouracil, oral isotretinoin, oral dovitinib, oral vemurafenib, oral penicillamine), photodynamic therapy use, radiotherapy use, or trauma (chemical peels, dermabrasion, tattoos, burns, skin grafts).[35-47]

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