Pruritic lichenoid lesions on the vulva of an 11-year-old girl

Case report

An 11-year-old premenarcheal Taiwanese girl presented with a 2-year history of itchy papules on the external genitalia. The itching was generally mild and intermittent but was aggravated by heat or during hot weather. These lesions increased gradually and had poor response to topical corticosteroid, antibiotic, or antifungal therapy. Examination revealed numerous, coalescing, 1-mm to 2-mm, slightly erythematous lichenoid papules on the labia majora. Many papules were surmounted by tiny white milia (Figure 1). There was no extragenital lesion. She denied family history, prior local trauma, or vesiculobullous disease. A skin biopsy specimen was obtained from yellow-white papules on the left labia majora. The histopathological evaluation showed numerous short cords or coma-shaped cords and ductal structures lined by cuboidal epithelial cells embedded in a sclerotic stroma in the upper and mid-dermis. In addition, many keratinous cysts were present in the superficial portion of the tumor (Figure 2). Serial sectioning revealed focal connection between the cysts and some of the underlying epithelial cords or ductal structures (arrows in Figure 2). Fontana-Masson stain failed to stain the cystic walls. Immunostaining for carcinoembryonic antigen (CEA) was positive in the epithelial cords, ductal structures, and part of the contents as well as the luminal aspect of the lower part of the larger keratinous cysts where the wall consisted of a thin layer of squamous epithelium without a granular layer (Figure 3). No CEA expression was found in the upper part of the cyst wall where the squamous epithelium was thicker with granular layer. Immunostaining for estrogen receptors (ERs) and progesterone receptor (PRs) showed negative results.

Figure 1 The patient presents with numerous, coalescing, 1-mm to 2-mm, slightly erythematous lichenoid papules on the labia majora surmounted by numerous tiny white milia.

Figure 2 Histopathology reveals a dome-shaped papule with numerous short cords or coma-shaped cords and ductal structures lined by uniform cuboidal epithelial cells embedded in a sclerotic stroma in the upper and mid-dermis. In addition, many keratinous cysts were present in the superficial portion of the tumor with focal connection with the epithelial cords or ductal structures (arrows) (hematoxylin and eosin; 200×).
Diagnosis

Vulvar syringomas with diffuse secondary milia.

Discussion

Syringomas typically manifest as asymptomatic, uniform, small, skin-colored or slightly yellowish papules on the eyelids. Vulvar syringomas are uncommon. In the largest series (18 patients) of vulvar syringomas reported by Huang et al, 33% also had periorbital syringomas; 22% had a family history of nongenital syringomas; and 72% experienced pruritus, which was exacerbated by heat or during menstruation. None of their patients presented the lesions before puberty as in our patient.

The pathogenesis of syringomas or vulvar syringomas is still not clear. Wallace and Smoller demonstrated positive PR in eight out of the nine cases of extragenital syringomas. ER and PR were demonstrated in normal vulvar epithelium in 28% and 15%, respectively. The increase in pruritus and the size of syringomas during pregnancy or menstruation and the intense nuclear and cytoplasmic staining for PR suggest that vulvar syringomas may be under hormonal regulation. However, ER and PR have not been demonstrated in vulvar syringomas by other authors and in our case.

Occasional milia may be seen in lesions of syringomas, including vulvar syringomas. Syringomas presenting as milia were rare, and only sporadic cases have been reported. Milium-like syringomas, first described in 1987, are a rare variant of syringomas with a prominent cystic component showing features of eccrine duct milia. To date, about 30 cases have been reported, mostly affecting Asians. Our case is unusual in that almost all lesions of the syringomas were literally covered by milia, which might be a secondary change because of persistent rubbing or scratching to relieve the pruritus.

The negative staining with Fontana-Masson in the wall of milia in our case and the one reported by Wang et al suggests that these cysts did not originate from epidermis or hair infundibulum. This view is further supported by the positive CEA staining of the basal half of the larger cysts, a finding in line with the incomplete-type sweat duct milia.

Treatment for vulvar syringomas include topical atropine and tretinoin, surgical excision, electrodessication with curettage, dermabrasion, and superficial CO₂ laser resurfacing. The lesions may need to be totally removed or destroyed to relieve pruritus and prevent recurrence in some patients. In our patient, we tried topical calcineurin inhibitor (tacrolimus ointment, 0.03%) twice daily with significant relief of pruritus at 5-month follow-up.

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References


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