Vulvar syringoma: A rare cause of pruritus vulvae

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Abstract

Syringomas commonly occur in women over the face, neck, and chest. They are usually asymptomatic and mainly of cosmetic concern. The vulva is an uncommon site for syringomas. A 45-year-old woman had asymptomatic lesions over the face, of 28 years duration and presented with vulvar papules, associated with severe pruritus for the past 2 months. Clinical and histopathological examination confirmed them to be syringomas. Coexistent facial and vulvar syringomas are rare. Further, vulvar syringomas presenting as pruritus vulvae is still rarer. We report a case with severe pruritus vulvae causing sufficient distress to seek medical care, which is remarkably unusual.

Key words: Female, genital, lichenification, pruritus, syringomas, vulvar

Introduction

syringoma is a benign eccrine tumor, commonly seen in adult females, though they may appear in adolescence. They present as multiple, small, 1–3 mm sized, flat-topped or dome-shaped papules, occurring bilaterally symmetrically over the face, neck, and upper chest.[1]

Vulvar syringomas are rarely reported in the literature. There may be co-existing facial syringomas, or they may occur as a part of a more generalized eruptive pattern. Pruritus is more commonly reported in vulvar syringomas than over extragenital sites.[2] We report an unusual case of coexistent facial and vulvar syringomas who presented with severe pruritus vulvae. Incidentally, her sister also was noted to have facial syringomas.

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Case history

a 45-year-old married woman presented with a history of skin lesions and severe itching over genitalia for the past 2 months. Pruritus was unrelated to menstruation and treatment with oral antihistamines, topical steroid/antifungal creams in the past gave her only partial relief. She had also noted multiple asymptomatic skin lesions over the face for the past 28 years, that had gradually increased in number. She gave a history of similar facial lesions in her sister. On detailed examination, there were multiple skin colored, smooth, mostly flat-topped, and a few dome-shaped, soft papules

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ranging from 1–5 millimeters in size, predominantly over the forehead, eyelids, cheeks, and sparsely over the upper lip and chin [Figure 1]. Nasolabial creases were characteristically spared. Examination of genitalia revealed lichenification and hyperpigmentation of labia majora with multiple discrete and few confluent smooth, flat-topped papules [Figure 2], and similar to facial lesions. A per speculum examination was normal. No similar lesions were seen in the body elsewhere, and systemic examination was normal.

Routine hematological, biochemical, urine examinations and chest X-ray were within normal limits. Blood venereal disease research laboratory test and serology for Hepatitis B, Hepatitis C, Herpes Simplex Virus-I and II, and Human immunodeficiency virus were negative. Vaginal swabs for microscopy, culture for bacteria and fungi were negative. Punch biopsies taken from representative lesions over the face and vulva revealed similar histopathological features. There were many superficial and deep dermal ductal structures lined by cuboidal epithelium with bland nuclei. Dilated lumina contained amorphous debris. The characteristic tadpole-like appearance was noticed [Figures 3 and 4]. No mitoses or desmoplasia was seen. Immunohistochemistry for estrogen and progesterone receptors could not be performed for want of facility.

The patient was given antihistamines and advised carbon dioxide laser treatment. However, she did not consent for the same and was started with 70% trichloroacetic acid application, twice a week over the lesions. There was moderate relief in pruritus and mild improvement in the lesions, after 4 weeks of treatment.

Discussion

kaposi and Biesiadeki reported the first case of syringoma in 1872 as "Lymphangioma Tuberosum Multiplex." Carneiro *et al*. in 1971, reported the first case of vulval syringoma in the English literature.[3] Syringomas are most commonly seen around the eyes, and angulated borders are characteristic. Eruptive forms have been rarely described in Down's syndrome involving the trunk, vulva, and penis. Syringomas are asymptomatic and are usually of cosmetic concern.[4]

Family history may sometimes be present, although it is not a hereditary condition.[2] Our patient's sister

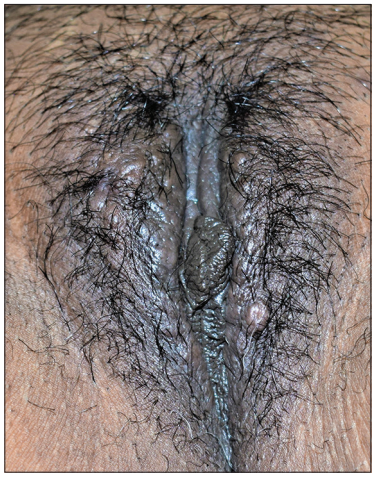

Figure 1: Bilateral vulvar syringomas with lichenification

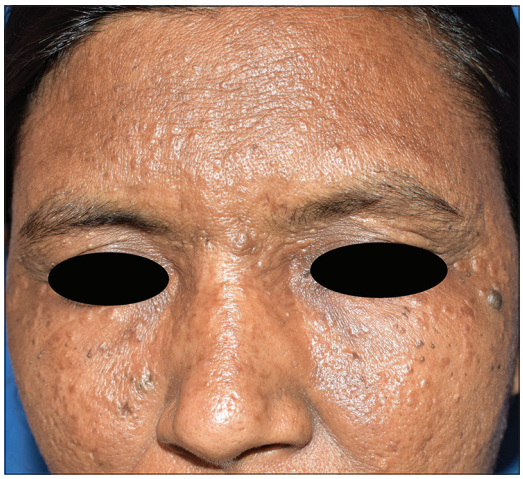

Figure 2: Facial syringomas involving the forehead, cheeks, and periorbital area

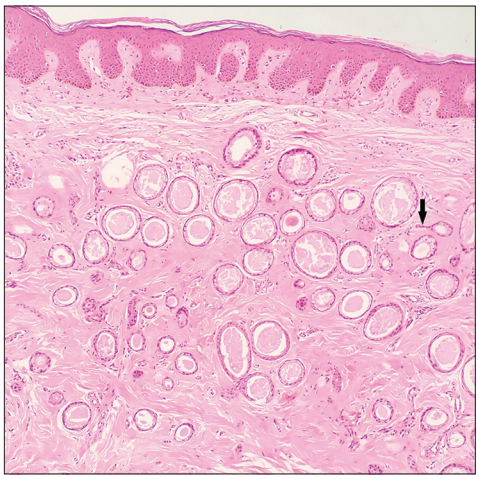

Figure 3: Histopathology (vulva): Showing multiple ductal structures with tadpole appearance (arrow) in the fibrous stroma (H and E, ×100)

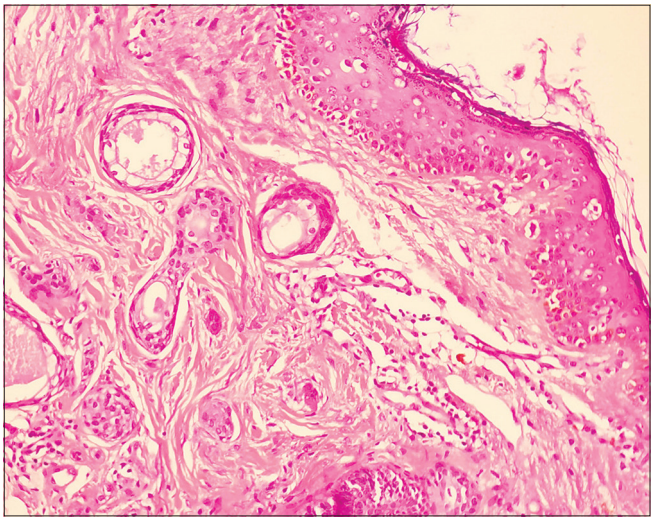

Figure 4: Histopathology (face): Showing multiple dilated ductal structures with amorphous debris (H and E, ×100)

had similar facial lesions, suggestive of familial involvement.

Vulvar syringomas are an unusual variant of syringomas. They are usually detected on routine gynecological examinations as symmetrical papules on labia majora and can be confirmed only by its diagnostic histopathological features.[2,5,6] They may be more common than is reported as most are asymptomatic.[3] They are usually revealed if symptomatic, unlike other areas where the cosmetic concern is the major problem.[5,7]

It is rare for vulval syringomas to present as pruritus vulva of a severe degree, as in our case. It is usually associated with intermittent or cyclical pruritus of moderate degree with possible exacerbations and an increase in size during pregnancy, menstruation, and summer months.[2,8] It is thought that this may be due to the hormone responsiveness of these benign tumors, supported by the demonstration of estrogen and progesterone receptors in the lesions.[8,9] However, our patient did not exhibit such exacerbations.

Syringomas may morphologically resemble various other lesions such as trichoepithelioma, milia, syringocystadenoma, condyloma acuminatum, lymphangioma circumscriptum, fox-fordyce disease, and epidermal cyst. Vulvar pruritus may be seen in conditions such as candidiasis, lichen simplex chronicus, and lichen sclerosus.[4,7,8]

Histopathology is diagnostic and shows a normal epidermis or changes related to chronic scratching and excoriation. In the dermis, multiple duct-like and cystic structures are seen embedded in a fibrous stroma. The structures are lined by a double layer of small cuboidal cells. Lumina usually contain amorphous debris. Characteristic tadpole or comma-shaped structures may be seen due to tail-like elongation of strands of the cells from one side of the ductal structures into the stroma.[4,8] In our case, the diagnosis was made based on the characteristic clinical and histopathological findings.

Treatment of syringoma is not necessary except for cosmetically disfiguring or symptomatic lesions. Various methods such as excision, electrodessication, carbon dioxide laser, argon laser, and dermabrasion have been tried successfully for localized lesions and topical tretinoin for eruptive syringomas. However, lesions can often recur.[10]

In conclusion, syringomas though rare should be considered as a cause for pruritus vulvae. Confirmation with histopathology is necessary. It is important to be aware of this condition for appropriate evaluation and management of patients with pruritus vulvae.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published, and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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