CASE REPORT

Vulvar Syringoma, Report of a Case and Review of the Literature

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Syringomas are common intraepidermal sweat gland tumors most often found in women around the time of adolescence. Frequent sites of involvement include the lower eyelids and malar areas, however vulvar involvement is relatively rare. These lesions often present as small, multiple, skin-colored-to-yellowish papules and are often associated with increased vulvar discomfort and itching. We present a case of a 29-year-old female who presented to her gynecologist complaining of vulvar itching and burning. A small condylomatous-type wart observed on her vulva was biopsied and found to be a syringoma. Because of their clinical presentation and associated symptoms, vulvar syringomas should be considered in the differential diagnosis of any multicentric papular lesion of the vulva, vulvar pain syndrome, and pruritis vulvae.

INTRODUCTION

Syringomas are common appendageal tumors of the intraepidermal eccrine sweat gland ducts. Their occurrence is twice as common in women than men, with adolescence being the most common time of onset [1]. However, further lesions can develop later in life, and reported cases range between the first and sixth decades of life [2-7]. Clinically, syringomas appear as small, multiple, firm, skin-colored-to-yellowish papules, 1 to 3 mm in diameter, localized most commonly to the lower eyelids and malar areas, but can also occur in the axillae, neck, chest, upper arms, and abdomen. The lesions usually are bilateral and symmetrically distributed [4]. Vulvar syringoma is a relatively rare occurrence, with only a few cases reported in the literature to date.

CASE

A 29-year-old G0 P0 female presented in June to her gynecologist for evaluation of vulvar itching and burning of two weeks duration. b She denied any significant past medical or gynecologic history, or recent oral contraceptive use. She also denied any family history of hereditary diseases. Pregnancy test was negative.

On physical examination, a small condylomatous-type wart was observed on the right vulva. Biopsy performed at the time revealed histologic changes most consistent
with syringoma, however because the lesion extended to the base of the resection, the possibility of a microcystic eccrine carcinoma could not be excluded. A subsequent excision of the right inferior vulva with skin confirmed the diagnosis of vulvar syringoma.

Since the time of final diagnosis, the patient reports no further occurrence of syringoma, either on her vulva or any other part of her body.

**DISCUSSION**

Syringoma was first described in 1872 by Kaposi and Biesiadeki as Lymphangioma Tuberosum Multiplex [8, 9]. Since then, monoclonal antikeratin antibody tests, electron microscopy, and histochemistry have all confirmed the intraepidermal eccrine sweat gland nature of this disease [10]. These tumors are most common in females, present during adolescence, and most commonly involve the face [11]. Syringomas are typically non-regressing and asymptomatic, although malignancy has been reported [12]. There is an increased frequency of this disease among oriental females and patients suffering from Down’s syndrome [13, 14]. Although numerous reports exist in the literature describing families with a history of this disease [10, 11, 15, 16], the hereditary transmission has not been elucidated.

Biopsy with microscopic examination is key to establishing the diagnosis and to ruling out other diseases. Histologically, the epidermis is normal. However, further examination of the upper dermis and mid-dermis reveals a plethora of small colloid-material-containing cystic ducts and solid epithelial strands contained within the surrounding fibrous stroma of these two layers [7]. Two rows of flat epithelial cells with clear cytoplasm line the walls of these ducts [17, 18], although cuboidal cells may sometimes be found lining the lumen as well [17]. At the end of the ducts are often found comma shaped “tadpole” tails comprised of bulgings formed by weakly organized ductal structures.

Friedman and Butler have suggested four principle classifications of syringomas based on morphological features and associations [19]. The first is localized disease, occurring either as solitary or multiple lesions. The second is a generalized, multifocal pattern of disease. The third group of cases is associated with Down’s syndrome, and the fourth are familial cases of syringoma.

The first patients with vulvar syringoma were reported by Carneiro et al. in 1971 [5]. The prevalence of this tumor may be greater than previously reported due to the fact that many are asymptomatic and are unrecognized by both patient and clinician [5]. Because vulvar syringomas have commonly been described in association with extragenital lesions [5, 11, 20] examination of the rest of the body, especially the eyelids and malar areas, is essential when a suspected syringoma is found in the vulvar region. Likewise, examination of the vulvar skin is mandatory when this lesion is found outside the genital area.

Although the diagnostic and treatment modalities for vulvar syringomas remains the same as at each location throughout the body, the vulvar area presents a special diagnostic challenge to the physician trying to make a diagnosis. Syringoma should be seriously considered in the differential diagnosis of any multicentric papular lesion of the vulva, vulvar pain syndrome, and pruritis vulvae. Clinically, some patients with vulvar syringomas may complain of increasing discomfort and itching, especially during warmer months or during menstruation [10, 11]. The differential diagnosis for vulvar syringoma is broad, and biopsy with histologic examination may exclude such diseases as epidermal cyst, sebacocystoma multiplex, lymphangioma circumscriptum, lichen simplex chronicus, Fox-Fordyce angiookeratoma disease, senile angioma, condyloma acumi-
natum, candidiasis, scabies, pediculosis, allergic and irritant contact dermatitis, psoriasis, and lichen sclerosis et atrophicus [6, 21, 22].

It has been suggested that syringomas are hormonally responsive based on observations that they increase in size during pregnancy, the premenstrual period, with the use of oral contraceptives, and are known to occur most commonly in women and during puberty [1, 23, 24]. In a study of nine non-vulvar syringomas from both males and females, Wallace et al. demonstrated estrogen in one case and progesterone in the remaining eight syringomas by immunohistochemical staining [1]. In a recent case report by Yorganci et al., immunohistochemistry of a vulvar syringoma was shown to display progesterone positivity [25]. Both studies supplement clinical observations that syringomas may contain hormonally active receptors that influence their progression during hormonally active states.

Treatment for syringomas is usually not necessary, and is often performed at visible areas of the body for cosmetic reasons, or if it is symptomatic. Syringomas commonly do not improve following medical therapies with topical steroids or ointments [7, 11, 17, 26], and thus treatment must be surgical. Partial removal, either by excision, electrodessication, and carbon dioxide laser treatment can be performed with satisfactory results [3, 4, 11, 21]. Although pruritis may resolve, tumor recurrence and scar formation following treatment are common. In one case report, Belardi et al. favored the use of cryotherapy with nitrogen oxide because of its minimal aggressiveness. The patient tolerated the therapy and the lesions healed quickly without scarring [7].

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