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CASE REPORT

Vulvar Syringoma Aggravated by Pregnancy

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Syringoma is a benign tumors of eccrine sweat gland. They appear as multiple, tiny, firm, skin-colored papules. Vulvar involvement of syringoma is rare. Only 24 cases with vulvar syringoma have been previously reported in the literature. The majority of patients with vulvar syringomas are asymptomatic. A case of syringoma of the vulva exacerbated during pregnancy is presented. The case appears remarkable for the experienced aggravated pruritic symptoms of the patient during her pregnancy. (Pathology Oncology Research Vol 9, No 3, 196–197, 2003)

Keywords: pregnancy, syringoma, vulva

Introduction

Syringomas are benign tumors of eccrine sweat gland derivation that occur more frequently in women. These neoplasms usually develop at puberty. Clinically, they appear as multiple, tiny, firm, skin-colored papules. The sites of predilection are the eyelids, malar regions, neck, and chest.[1-8] Syringoma limited to the vulva is rare. Most syringomas on the face are symptomatic and permanent, whereas most vulvar syringomas are asymptomatic and therefore overlooked. Uncommonly, vulvar syringomas may itch. Vulvar syringoma should be included in the differential diagnosis for itchy and papular lesions of vulvae.[1-3,6] We present the case of a 24-year-old woman with vulvar syringoma that was exacerbated during pregnancy.

Case Report

The patient was a 24-year-old gravida 1, para 1 female who presented with the complaint of genital itching. She had experienced intermittent vulvar pruritus for 12 years, and had noted no change in her symptoms during menstruation. She had no history of eczema or contact hypersensitivity.

Physical examination revealed multiple, soft, yellowish-to-skin-colored, 1-2 mm-diameter papules on the vulva. No similar lesions were detected elsewhere on the body. The patient reported that the papular lesions intensified during her pregnancy, and that they were reduced in size and number after that pregnancy was terminated. The results of laboratory tests were normal.

A biopsy of the lesion was performed after pregnancy was terminated and microscopic examination revealed the typical features of syringoma. There were numerous tubular structures embedded in fibrous stroma in the papillary and reticular dermis (*Figure 1*). Each of these ductal structures had a central lumen that was lined by two rows of epithelial cells. The ductal lumina contained amorphous debris (*Figure 2*). Based on these findings, the patient was diagnosed with vulvar syringoma. Immunohistochemical studies revealed no estrogen and progesterone receptors in the tumor cells.

Discussion

Syringomas are fairly common tumors of eccrine sweat ducts, but genital syringoma is rare. The first genital case of syringoma to be published was a penile tumor. Corneino et al. were the first to report a case of vulvar syringoma.[1-3] Most of these neoplasms are asymptomatic, and are detected on routine gynecologic examination.[1] Clinically, the lesions appear as multiple, tiny, firm, skin-colored papules, each approximately 1-3 mm in diameter. Some are yellowish in color. In typical vulvar syringomas, the papules are bilateral and are symmetrically distributed.[1-5] Vulvar syringoma is an unusual clinical variant of syringoma. It is unusual for the vulvar form to present as pruritus vulvae;

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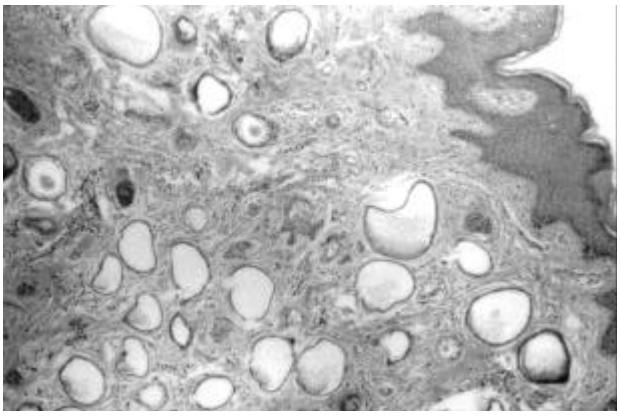

Figure 1. Tubular structures embedded in fibrous stroma in the papillary and reticular dermis (Hematoxylin-eosin, x 100)

however, these neoplasms do occasionally cause discomfort and pruritus, especially during the warmer months or during menstruation.[3] Although the macroscopic appearance of vulvar syringoma is not pathognomonic, the tumor's histologic appearance is diagnostic.[3] The microscopic findings of syringoma include normal epidermis, and dilated cystic sweat ducts embedded in a fibrous stroma in the dermis. Some of these dilated ducts have small comma-like tails, which produce a distinctive picture that resembles tadpoles. In addition, there may be strands of epithelial cells independent from the ducts. Typically, two rows of epithelial cells line the duct walls. Recent electron microscopic and histochemical findings have supported the theory that syringoma is an adenoma of the eccrine sweat gland with differentiation towards sweat ducts.[1-7]

Enlargement of syringomas during pregnancy has been reported previously, and, interestingly, immunohistochemical studies have detected intralesional progesterone and estrogen hormone receptors in these neoplasms. This suggests that cyclical hormonal changes are likely responsible for periodic exacerbation of genital pruritus during menstrual periods or pregnancy.[1] Our patient only experienced aggravated pruritic symptoms during pregnancy, not during menstruation.

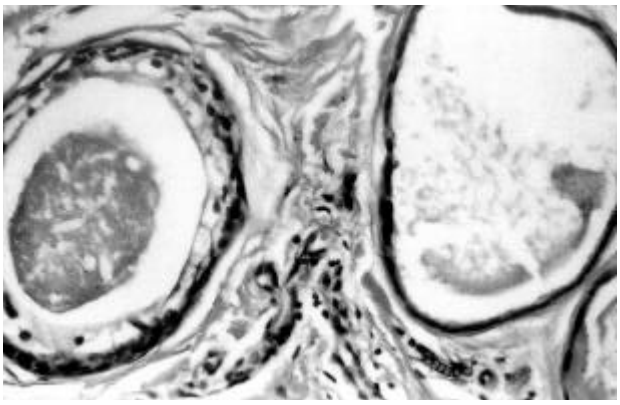

Figure 2. Ductal structures were lined by two rows of epithelial cells and amorphous debris in ductal lumina. (Hematoxylin-eosin, x 400)

The differential diagnosis for vulvar syringoma includes itchy lesions of vulvae such as candidiasis, lichen sclerosus and atrophicus and popular lesions of the vulvae such as Fox-Fordyce disease, epidermal cyst, senile angioma, cherry hemangioma, lichen simplex chronicus, steatocystoma multiplex, lymphangioma circumscriptum, and condylomata acuminate.[1-3,6] With respect to histolopathological features, syringoma must be distinguished from several conditions. Desmoplastic trichoepithelioma typically features numerous keratocysts and solid strands of basophilic epithelial cells. Although there is obvious histological overlap with eccrine epithelioma, the clinical features of this neoplasm are quite different from those of syringoma.[8] The solid strands of basophilic epithelial cells embedded in the fibrous stroma in some cases of syringoma may appear similar to strands seen in fibrosing basal cell carcinoma. Syringoma can be distinguished from microcystic adnexal carcinoma by its smaller size, its greater symmetry, lack of prominent horn cyst formation, and infrequent single-file strand formation.[5] Histological examination is essential in all cases of syringoma, as this is the only way to establish a definitive diagnosis and rule out malignancy.[1] Treatment of syringoma is usually not necessary unless there are cosmetic issues. Electrodesiccation and carbon dioxide laser treatment can be performed with satisfactory results, though the lesions may recur.[2]

In conclusion, it is important to always include syringoma in the differential diagnosis for papular lesions of the vulva. This case of vulvar syringoma is of particular interest because the patient's pruritic symptoms were aggravated by pregnancy, which is evidence for hormonal etiology.

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