A Case of Vulvar Syrigoma Clinically Suggesting Milia in a Pregnant Woman

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We report a case of vulvar syringoma suggesting milia. A 20-year-old pregnant woman visited the obstetrical department of our hospital for labor pain. On physical examination, the skin lesions were revealed accidently and showed symmetrically distributed, numerous, 2-4mm sized, yellow to skin colored papules on the vulva. The lesions developed in the fifth gestational month and were aggravated thereafter. After delivery, we performed a skin biopsy of the lesion. Histopathologic findings of the biopsy specimen showed the typical findings of syringoma and milia. (Ann Dermatol 8:(4)291~294, 1996).

Key Words : Syringoma, Milia, Pregnancy, Vulva

Syringomas are benign intraepidermal adenomas derived from the eccrine duct and develop usually on the eyelid and upper cheeks, chest, upper arms, and abdomen[1-4]. Syringoma of the vulva is a rare condition. It develops slowly and persist indefinitely without any symptoms. It may also develop at any age, and occur predominantly in women after puberty[5-7].

We found some literature reporting cases of syringoma suggesting milia appearing on the vulva [7-9]. The lesions were so numerous that the whole vulvar skin was filled with syringoma and milia giving the lesions a cobble stone-like appearance. We thought the severity of the disease was related to the pregnancy and that it could have been due to hormonal influences[5]. We tried to find a link between the syringoma and milia but we were unsuccessful. In our case, we were not certain if it was a syringoma clinically suggesting milia or syringoma with milia. We describe a case of vulvar syringoma showing numerous, symmetrically distributed papules concurrently suggesting milia during pregnancy.

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REPORT OF A CASE

A-20-year-old pregnant woman(parity; 0-0-0-0) was admitted to the Obstetrics & Gynecology department of our hospital with labor pain. On physical examination, vulvar lesions were found and the obstetrician consulted our department. Examination of the vulva revealed numerous soft skin to white or yellowish colored papules and nodules that measured 1-3mm in diameter and were located symmetrically on the labia majora; a few scattered lesions were observed on the labia minora(Fig.1). Examination of the rest of the body did not show such lesions to be present anywhere else. On history taking, the patient stated that the lesions had developed during her fifth month of gestation and were aggravated thereafter. No other member in her family was known to have a similar condition. Laboratory studies including a complete blood count and a differential count, urinalysis, serologic test for syphilis, liver function test, serum electrolytes, chest x-ray and EKG were normal.

The biopsy specimen obtained from the lesion showed numerous cystic structures containing concentric lamellae of keratin which were lined

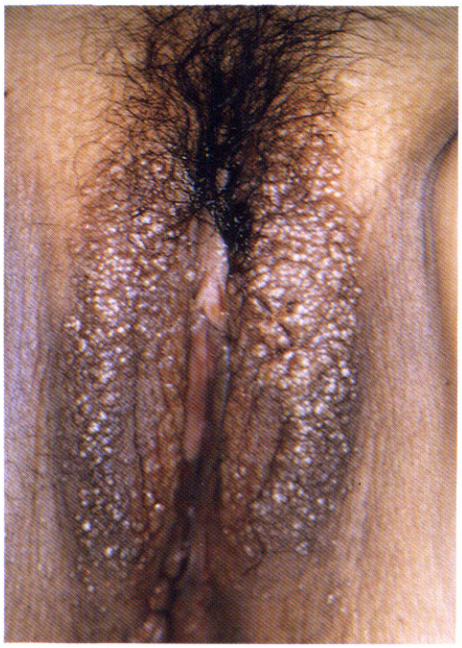

Fig. 1. Numerous soft skin to white or yellowish colored papules and nodules were located symmetrically on the labia majora; a few scattered lesions were observed on the labia minora.

by a few cell layers of stratified epithelium with a thin or single cell layer of stratum granulosum. The papillary and mid dermis contained many small ductal structures lined by two rows of epithelial cells embedded in a dense, fibrous stroma. Some ductal structures showed a comma-like tail of epithelial cells; serial sections revealed an extension of the keratin cyst towards these structures (Fig. 2).

DISCUSSION

Syringoma can develop at any age, but they frequently make their first appearance or increase greatly in number at puberty. More than half appear during the second or third decade of life, but some have developed as late as the seventh decade, but it is most frequent during the third and forth decades[1,2]. Familial occurrence has been reported, and syringomas are found frequently in patients with Down's syndrome[10]. Women are affected twice as often as men[5]. They develop slowly and persist indefinitely without symptoms, although they can cause discomfort and itching, especially during the warmer months and during mensturation. They are small translucent papules, yellow, brown or

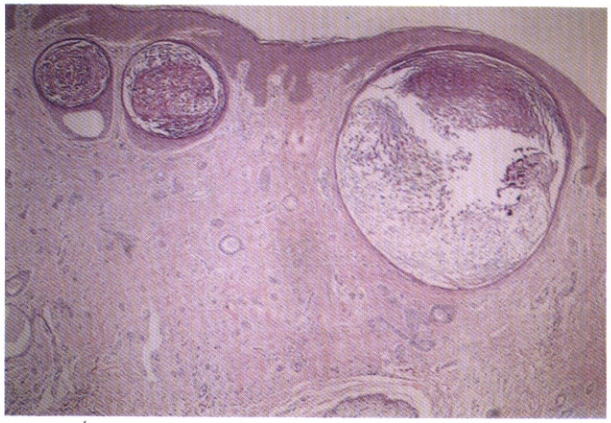

Fig. 2. Numerous cystic structures containing concentric lamellae of keratin which were lined by a few cell layers of stratified epithelium with a thin or single cell layer of stratum granulosum. The papillary and mid dermis contained many small ductal structures lined by two rows of epithelial cells embedded in a dense, fibrous stroma(H&E stain, × 40).

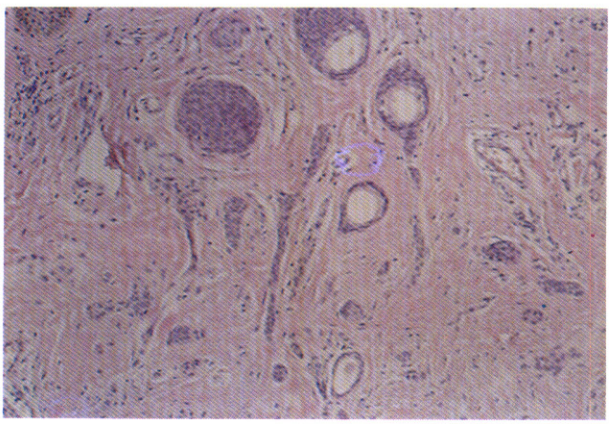

Fig. 3. Some ductal structures showed a commalike tail of epithelial cells; serial sections revealed an extension of the keratin cyst towards these structures(H&E stain, × 100).

pink in color and globoid, being 2-3mm in diameter. They usually appear on the infraocular areas and also appear in large numbers on the chest, upper arms, and abdomen. The lesions are usually multiple and tend to have bilateral symmetry in distribution, particularly so in those occuring on the vulva[8,11,12].

The pathogenesis of the syringoma is unclear, but women are affected twice as often as men and increase in size during puberty suggesting hormonal influences. Increase in size during pregnancy, premenstural enlargement and swelling after estrogen administration support the idea that the syringo-

ma is under hormonal influences[5]. Our patient also developed this condition during pregnancy and it was aggravated thereafter. In our case, the patient was pregnant and the vulvar lesion was more aggravated during pregnancy. This explains that syringoma is under hormonal influence. In fact, increase in the size of the lesion during pregnancy, premenstrual enlargement, or swelling by estrogenic hormones have been observed[5,11].

The histogenesis of the syringoma was debated for many years, but histochemical, electron microscopic[4] and monoclonal antikeratin antibody studies[13] support the theory that syringomas are of eccrine duct differentiation.

Microscopic findings are chracterized by the presence of solid strands of basaloid cells embedded in a fibrous stroma, horn cysts near the epidermis and dilated cystic sweat ducts, some of which have small comma like tails to produce a distinctive picture resembling tad poles. Cystic ductal lumina filled with keratin and lined by cells containing keratohyaline granules were noted in the papillary dermis of some of the cases[2,4]. The keratin cysts may become large and sometimes rupture, producing foreign body reaction and focal calcification. Although vulvar syringoma has a characteristic clinical appearance, microscopic examination is essential to establish the diagnosis, because its gross appearance often resembles other lesions(including epidermal cystws, cherry hemangioma, angiokeratoma, comedones, soft fibroma, Fox-Fordyce disease, steatocystoma multiplex, lymphangioma circumscriptum, etc.)[8,12].

There are many clinical varients of syringoma. Eruptive hidradenoma generally occurs on the anterior half of the body of young adults. Syringoma associated with noncicatrical alopecia[13,15] and lichen planus-like lesion of syringoma have also been described[6,13].

Milia are white keratinous cysts, 1-4 mm in diameter, appearing chiefly on the face, especially under the eyes. They may occur in great numbers, especially in middle-aged women. They arise spontaneously in the skin(primary milia) and, also develop in the setting of underlying factors such as bullous disease or trauma(secondary milia). Primary milia grow from the undifferentiated sebaceous collar and secondary milia arise from either hair follicles or eccrine ducts[7]. In our case the milia arose combined with syringoma after trauma.

The clinical and histological features of keratin-filled cystic lesions in our case were typical of milia. Friedmann et al[7] described two cases of syringoma presenting as milia. They indicated that no cases of this clinical presentation had been reported and proposed it as a new clinical varient of syringoma. But at first we thought it was not syringoma presenting as milia but syringoma combined with milia, because clinically some of the lesions looked more milia than syringoma. In addition on microscopic examination the upper dermis showed typical findings of milia, and the papillary and mid dermis show typical findings of syringoma.

Usually treatment for syringomas is not necessary although it is, sometimes carried out for cosmetic purposes. These include electrolysis, laser ablation and cryotherapy[3].

In summary, vulvar syringoma is distinctly rare and, what is more, in our case milium-like syringomas had severely infiltrated on the whole vulvar skin and were aggravated during pregnancy. The relationship between syringoma and milia is unclear and seems to be associated with pregnancy. We propose that further study about the pathogenesis of syringoma and milia would be helpful in the understanding of the relationship between syringoma and milia and hormonal influences.

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