Case report

Vulvar syringoma exacerbated during pregnancy

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Abstract

Although syringoma of the eyelids and the cheeks are well known and defined, vulvar syringoma is a rare clinical variant of this benign eccrine tumor. A case of syringoma of the vulva exacerbated during pregnancies with regression in the periods in between is presented. Histopathological examination showed typical features of syringomas. Only 20 patients with vulvar syringoma have been previously reported in the literature.

Keywords: Syringoma; Vulva; Pregnancy

1. Introduction

Syringoma is a benign epithelial tumor commonly occurring in women especially after puberty as small translucent papules on the face that are usually asymptomatic and permanent [1]. Vulvar involvement of syringoma is rare and the majority of patients with this unusual clinical variant of syringoma are also asymptomatic [2]. In the literature only 20 cases with vulvar syringoma have been reported [3–7]. Therefore, this is the 21st reported case of vulvar syringoma in the literature to our knowledge.

2. Case report

The patient, a 25-year-old gravida 3, para 2, was referred from a family planning clinic to a gynecologic oncology out-patient clinic. She was 9 weeks pregnant and wanted her pregnancy to be terminated. Examination revealed multiple papular lesions 3–5 mm in diameter on her vulva, and a nevus 2–3 mm in diameter on her right labium minus. She had no extragenital lesion and no family history. It was learned from her history that the papular lesions had first appeared during the first pregnancy, exacerbated during subsequent pregnancies and regressed in size and number after termination of the pregnancies. Following the vulvar biopsy, she underwent a dilatation and evacuation. Histopathological examination revealed typical features of syringomas and intra-dermal nevus (Fig. 1). The patient was not given any further treatment and it was observed that these papular lesions regressed almost completely 10 weeks later.

3. Discussion

Syringoma is a benign tumor of the eccrine sweat gland ducts [2]. The tumor usually affects young women, generally appearing first at adolescence as small translucent flesh-colored to yellowish, globoid papules 2–5 mm in diameter [1]. Rarely, older women or pre-pubertal girls are also affected [3,4]. The distribution of papules is bilateral and symmetrical over eyelids, cheeks, chest and neck. Rarely, thighs, knees and legs have been reported as being involved [4]. Papules develop slowly and usually persist without symptoms. Although this is not a hereditary condition, occasion-
Fig. 1. The dermis contains two lesions, one consisting of nevus cells, the other one made up of several small ducts embedded in a fibrous stroma (H.E. x 40).

ally familial traits and increased incidences of such lesions in cases with Down’s syndrome and oriental females have been reported [2,4].

Vulvar syringoma is an unusual clinical variant of syringoma and a rarely reported lesion of the vulva [3]. However, some authors think that syringomas are probably more common than are reported, due to the fact that many are asymptomatic and unrecognized both by the patient and the clinician [1,4]. Although these vulvar lesions can cause discomfort and pruritus especially during the warmer months and during menstruation, they are usually asymptomatic [2,4].

To our knowledge, the present case is the 21st reported one and the first one that exacerbated during pregnancy and regressed after termination of pregnancy. This finding suggests a role of changing hormonal milieu. Therefore, it may be of interest to investigate the steroid receptor content. In addition, the usual appearance of syringomas first at puberty may support this finding.

Histopathological examination of this case also revealed a concomitant intra-dermal nevus. We think that the vulvar intra-dermal nevus in the present case is a coincidental finding.

Differential diagnosis of vulvar syringoma includes epidermal cyst, cherry hemangioma, angiokeratoma, comedo, soft fibroma, Fox-Fordyce disease, steatocystoma multiplex and lymphangioma circumscriptum [8]. Of these vulvar lesions, mainly Fox-Fordyce disease and epidermal cysts may create a diagnostic problem [1]. Fox-Fordyce disease, at first glance, can suggest syringoma of the vulva but the lesions are smaller, more conical and are periodically very pruritic. Small epidermal cysts, when multiple, could be confused with syringoma. They are usually fewer in number, much more yellow in appearance and often show one or more lesions with characteristic central punctum.

Although the macroscopic appearance of vulvar syringoma is not pathognomonic, it has a diagnostic histologic appearance [1]. Microscopic findings of syringoma reveal a 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 to produce a distinctive picture, resembling tadpoles. Strands of epithelial cells may occur independently of the ducts. Usually, two rows of epithelial cells line the duct walls. Recently, electron microscopic and histochemical studies have supported the view that syringoma is an adenoma of the eccrine sweat gland with differentiation towards sweat ducts [1].

Treatment of vulvar syringomas is generally not required except for cosmetic reasons. Excision or fulguration of each lesion could be performed, if needed [4,8]. Incomplete removal is often followed by recurrence [4].

References