

Case Report

Plaque like syringoma presented as plaque like lesion over right vulva

斑塊狀汗管瘤於右側外陰呈斑塊狀皮損

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A 14-year-old Form 2 student presented with more than one year history of itchy slightly pigmented papules over the right vulva. These lesions wax and wane. An initial diagnosis of lichen simplex chronicus was made but she did not respond to steroid. Skin biopsy was subsequently done which showed plaque like syringoma.

14 歲中二學生，一年多來於其右側外陰出現癢性色素沉著性丘疹。病損呈復發性。早期診斷為慢性單純苔癬，但對局部類固醇的治療無反應。其後皮膚活組織檢查顯示斑塊狀汗管瘤。

Keywords: Syringoma, vulva

關鍵詞：汗管瘤、外陰

Introduction

Syringoma is a benign adnexal neoplasm which often occurs around the cheeks and the lower

eyelids as skin coloured papules measuring a few millimeters in size.¹ Plaque like syringoma is a clinical variant of syringoma. We present a case of this unusual form of syringoma over the vulva region.

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Case report

A 14-year-old Form 2 student presented with a more than one year history of itchy skin coloured papules over the right vulva. These lesions wax and wane and were aggravated during menstruation. The patient enjoyed a good past health. She had no ongoing medication taken. No family member had similar lesion. Physical examinations showed thickened, slightly

pigmented papular lesions over the right vulva (Figure 1). There was no pustule and no skin atrophy. Topical steroid was prescribed by the family doctor but there was no improvement. No other similar skin lesion could be found in other parts of the body. Differential diagnoses included vellus hair cysts, epidermoid cysts, milia en plaque, syringoma en plaque, lichen simplex chronicus, lichen nitidus, plane wart, molluscum contagiosum and adnexal carcinoma. A skin biopsy was done over the right vulva which showed milia like structures associated with comma- and tadpole-shaped tubules and cell cords in scirrhous stroma (Figure 2). The pathological diagnosis is consistent with plaque like syringoma.

Discussion

Syringoma is a benign adnexal neoplasm formed by well differentiated ductal elements. It usually presents as multiple, skin coloured, small papules over the cheeks and lower eyelids. The lesions are usually asymptomatic and they tend to first appear at puberty.² Other clinical variants include solitary, eruptive, milia-like, lichen planus like and plaque like syringoma. Other unusual sites of involvement include the penis, vulva, ankle, upper extremities and scalp.

Syringoma is usually an incidental finding. Rarely, some familial cases may occur.³ In addition, it may be associated with Marfan, Down and Ehlers-Danlos Syndrome.⁴ Friedman and Butler proposed a classification of syringoma in 1987 based on the clinical features and associated conditions.⁵ It is divided into categories of localised, generalised, familial, Down's syndrome associated syringoma. There have been around ten documented cases of plaque-type syringoma. The first of which was published in 1979.⁶ In all cases, the lesions were asymptomatic and they involved the face, arms and trunk.

Histopathology of syringoma typically shows multiple eccrine ducts, lined by two layers of cuboidal epithelium which are dispersed within a fibrous stroma in the dermis. Histopathology of plaque-like syringoma is identical to that of typical syringoma. The main histopathological diagnosis include morphoeaform basal cell carcinoma, eccrine syringocarcinoma, microcystic adnexal carcinoma and desmoplastic trichoepithelioma. The lack of subcutaneous fat involvement and the



Figure 1. Thickened, slightly pigmented and papular lesions over the right vulva.

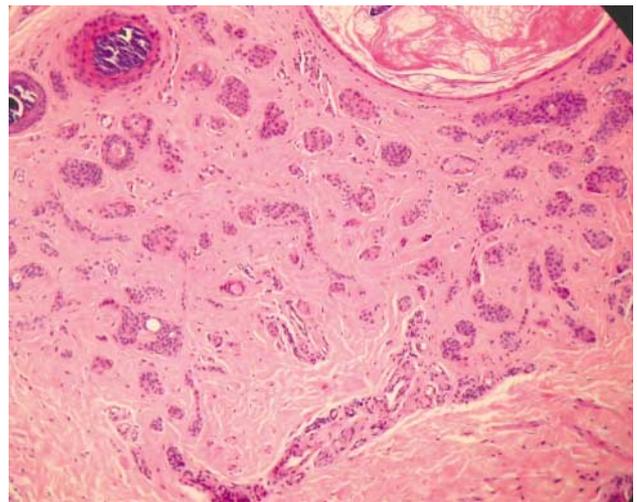


Figure 2. Milia like structures associated with comma- and tadpole- shaped tubules and cell cords in scirrhous stroma.

absence of cellular atypia help to distinguish syringoma from the other malignancies.

Syringoma requires no treatment. Cosmetic concern may prompt patients to seek treatments. The goal of the treatment should be the destruction of the lesions with minimal scarring and no recurrence. In one report, lesions have improved significantly using a topical retinoid.⁷ Syringoma can also be effectively treated by ablative methods, such as electrodesiccation,⁸ cryosurgery and CO₂ laser.⁹ In one case series in Taiwan, CO₂ laser has shown to be effective in the resolution of the lesions and symptoms.¹⁰

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