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Case Report

Syringocystadenoma papilliferum presenting as linearly arranged erythematous papules over medial thigh

乳頭狀汗管囊腺瘤表現為大腿內側線狀分佈的紅色丘疹

AYK Chan 陳綺琪, NM Luk 陸乃明, WYM Tang 鄧旭明, WY Lam 林偉業

A 3-year-old Chinese girl had three lumps over left inner thigh since birth. Histopathological examination confirmed syringocystadenoma papilliferum.

3 歲華人女孩，於其左側大腿內出現三處隆起病損，經組織學診斷為乳頭狀汗管囊腺瘤。

Keywords: Syringocystadenoma papilliferum

關鍵詞：乳頭狀汗管囊腺瘤

Introduction

Syringocystadenoma papilliferum (SP) is an uncommon cutaneous appendage tumour. Most dermatologists consider it a benign neoplasm while others categorise it as a hamartoma exhibiting apocrine differentiation.[1] We report a 3-year-old Chinese girl with syringocystadenoma papilliferum manifesting as three lumps over left inner thigh since birth.

Social Hygiene Service, Department of Health, Hong Kong

AYK Chan, MRCP(UK), FHKAM(Medicine)
NM Luk, MRCP(UK), FHKAM(Medicine)
WYM Tang, FHKAM(Medicine), FRCP(Edin)

Histopathology and Cytology Laboratory, Public Health Laboratory Centre, Hong Kong

WY Lam, MBChB, MHKCPath

Correspondence to: Dr. AYK Chan

Cheung Sha Wan Dermatology Clinic, 3/F West Kowloon Health Centre, 303 Cheung Sha Wan Road, Kowloon, Hong Kong

Case report

A 3-year-old Chinese girl was born in Hong Kong with unremarkable birth history. She developed three lumps over left inner thigh since birth. According to parent, the lumps slowly increased in size. They were non-itchy and non-painful. No systemic complaints were ever reported. No significant family history was noted. Systemic review and developmental milestones were normal. Physical examination revealed three discrete, linearly-spaced pinkish papules (Figure 1). Surface scaling, firm consistency and

hyperpigmentation were evidenced. There was no hindrance of limbs growth. Examination of skin elsewhere, hair, teeth and nails was unremarkable. Excision biopsy showed several well-demarcated cystic invaginations that communicate with the surface epidermis. There were papillae lined by a double layers of epithelium. The outer layer is low cuboidal cells while the inner layer is tall columnar cells with focal apical decapitation. The nuclei are bland-looking. The cores of the papilla contain abundant plasma cells (Figures 2 & 3). It is consistent with syringocystadenoma papilliferum.

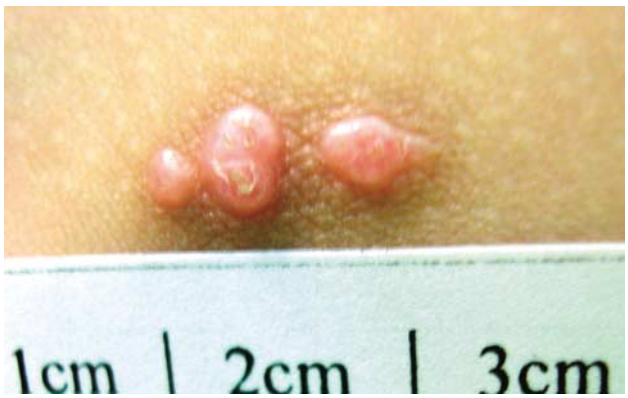

Figure 1. Three discrete, linearly-spaced pinkish papules.

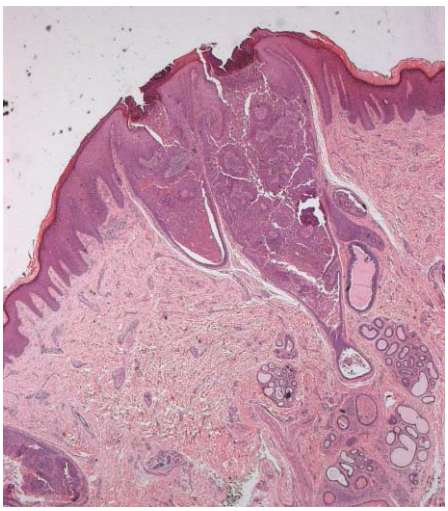

Figure 2. Several well-demarcated cystic invaginations communicates with the surface epidermis (H&E, Original magnification x 20).

Discussion

SP is usually seen at birth, infancy or early childhood. It develops either independently or arises in association with a nevus sebaceus, in one third of cases.[1] Three clinical types of SP have been described. The plaque type presents most commonly as yellowish or brownish papules with central ostium and typically devoid of hair over the scalp. It is often associated with sebaceus nevus of Jadassohn or arises de novo. [2] The linear type is characterised by multiple pink, red or brown papules or nodules. They show predilection over neck or face but may sometimes be over the extremities. The solitary nodular type usually arises at puberty, affecting shoulder, axilla, genito-crural and trunk. It is commonly associated with hamartomas of follicular or sebaceous gland origin. It may increase in size at puberty, becoming papillomatous and crusted.[3] Most of the lesions occur on head and neck region, while the remaining 20% on trunk and 5% on the extremities (mainly on legs). Possible affected areas include eyelid, external auditory canal, vulva and scrotum.[4-8] In literatures, there was only few reported cases of malignant transformation in syringocystadenoma papilliferum (for instance,

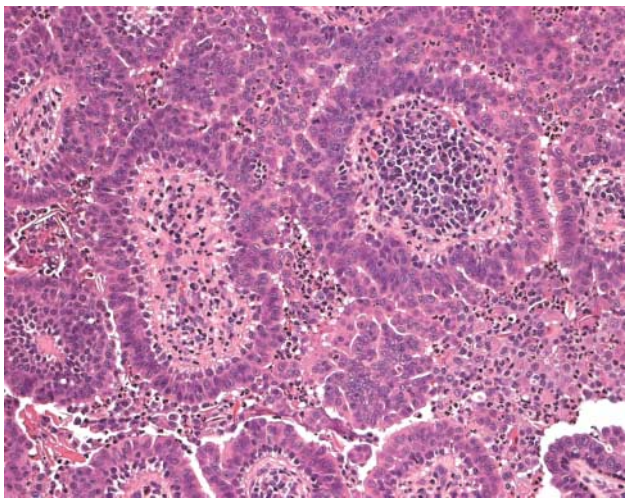

Figure 3. The cores of the papilla contain abundant plasma cells (H&E, Original magnification x 200).

syringocystadenocarcinoma papilliferum in situ).[9] Our case is a rare presentation of SP with multiple papules arranged linearly over the lower limbs. A similar case of linear SP on thigh in a 14-year-old girl without associated nevus sebaceous or epidermal naevus has been reported by Patterson et al.[3] Similar cases of linear array of SP occurring over the chest, arm, neck and lower extremity were noted.[3, 7] Linear epidermal naevus, common wart and linear porokeratosis can mimic this condition. Linear epidermal naevus is a hamartomatous proliferation from embryonic ectoderm. Most lesions occur at birth or in infancy, characterised by crops of brown verrucous papules sometimes coalescing to form well demarcated papillomatous plaques. Linear configuration is common and appears to follow the line of Blaschko. Common wart is a common benign epithelial hyperplasia secondary to human papilloma virus infection. It may develop in a linear fashion secondary to köebner phenomenon. Linear porokeratosis is an uncommon benign epidermal proliferation. It is firm, keratotic papule, with characteristically raised hyperkeratotic border. It mainly affects infancy or childhood and may follow the line of Blaschko. Histopathological study can reliably distinguish between the aforementioned conditions.

Surgical excision is the treatment of choice. CO_2 laser may be an alternative treatment for anatomic sites unfavourable to surgical excision. It produces infrared radiation that is highly absorbed by water. A rapid increase in skin temperature results in tissue vaporisation.

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