Hong Kong J. Dermatol. Venereol. (2005) **13**, 214-216

Case Report

Multiple piloleiomyomata cutis in a Hong Kong Chinese

多發性皮膚毛平滑肌瘤—香港華人病例一宗

YH Chan 陳耀海, WYM Tang 鄧旭明, WY Lam 林永賢

A 60-year-old Chinese gentleman presented with extensive erythematous plaques and nodules over the upper part of his left trunk for 20 years. Skin biopsy of the lesion showed piloleiomyomata cutis. The patient was currently managed symptomatically with periodic therapeutic surgical excision and oral doxazosin.

60歲華人男患者，20年來於其左側軀幹上部出現多發性紅色丘疹及結節。皮膚活組織檢查顯示皮膚毛平滑肌瘤。患者目前接受對症處理，包括定期的治療性切除術及口服多沙唑嗪。

Keywords: Chinese, multiple piloleiomyoma cutis

關鍵詞：華人，多發性皮膚毛平滑肌瘤

Introduction

Piloleiomyomata cutis (PC) is an uncommon by benign skin tumours arising from the arrector pili muscle of pilosebaceous unit. They exhibited characteristic paroxysmal pain which can be disabling to patients and are often sensitive to cold or touch. Histologically, PC appears as uncapsulated growth composed of well differentiated eosinophilic spindle cells with blunted nuclei interlacing with variable amounts of collagen. We report a 60-year-old Chinese gentleman with extensive PC over the upper part of his left trunk.

Social Hygiene Service, Department of Health, Hong Kong

YH Chan, MBBS, MRCP(UK)
WYM Tang, FHKAM(Med), FRCP(Edin)

Department of Clinical Pathology, Tuen Mun Hospital, Hong Kong

WY Lam, FHKAM(Pathology)

Correspondence to: Dr. YH Chan

Case report

A 60-year-old Chinese gentleman presented with extensive erythematous plaques and nodules over the upper part of his left trunk for 20 years (Figures 1 & 2). Besides mild hypertension on thiazide diuretics, he enjoyed good past health. The lesions started as small nodules over his left shoulder and was largely asymptomatic initially. There was an increase in number and size of these nodules and plaques with time and the patient began to be bordered by lesional paroxysmal pain. Such pain was occasionally precipitated by scratching. None

of his family members were affected by similar problem.

On examination, there were numerous skin-coloured to red dermal nodules and plaques over his left upper girdle. These lesions measured 0.5-2 cm in diameter with smooth surface and nontender. Pseudo-Darier's sign could not be elicited. There were no other cutaneous or mucosal lesions detected. Systemic examination was unremarkable. A skin biopsy was performed and showed dermal growth composed of whorled smooth muscle fibres with only mild nuclear pleomorphism without evidence of malignancy (Figures 3 & 4). These findings confirmed the diagnosis of PC.

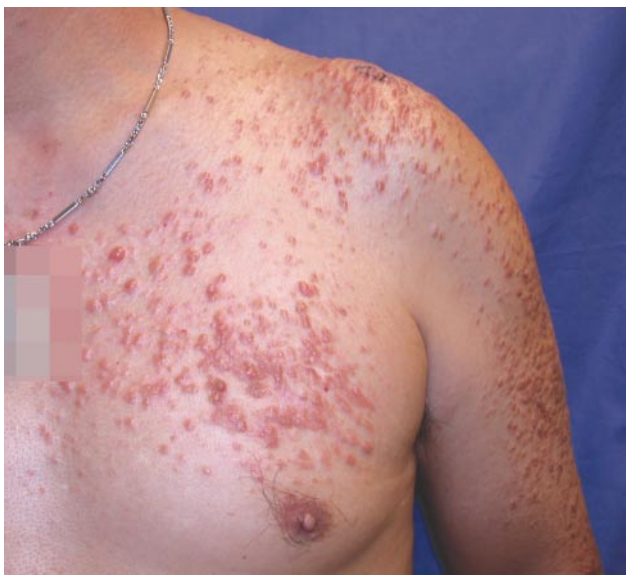

Figure 1. Extensive skin-coloured to red nodules and plaques over left chest wall.

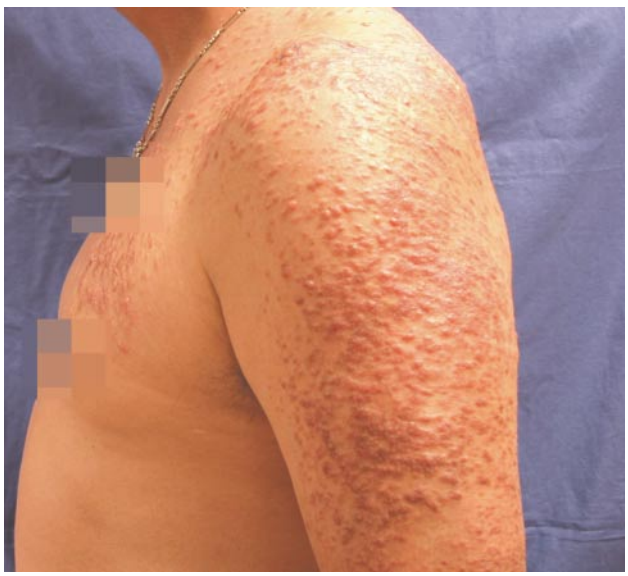

Figure 2. Extensive nodules and plaques over left upper girdle.

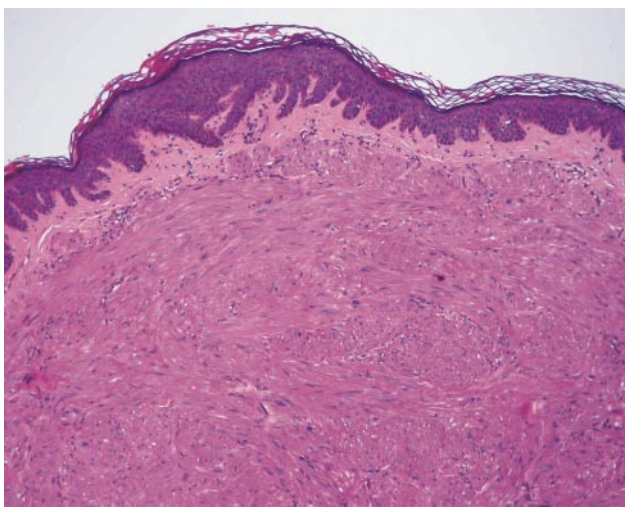

Figure 3. Unencapsulated tumour growth composed of fascicles of smooth muscles and intercalating collagen in the dermis and subcutis. (H & E, Original magnification X 4)

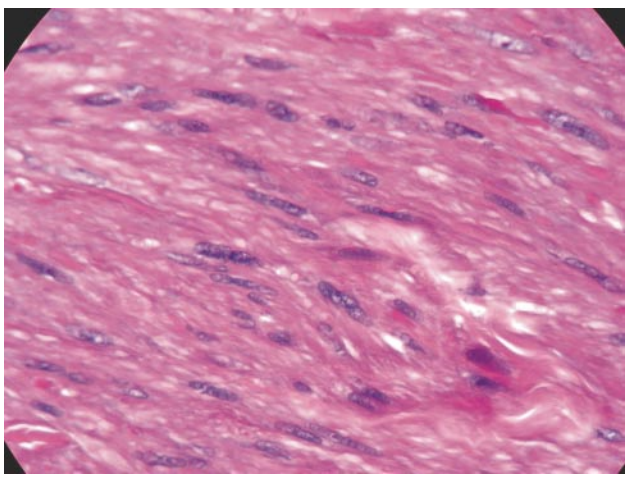

Figure 4. Eosinophilic spindle cells with elongated nuclei, abundant cytoplasm and no mitoses. (H & E, Original magnification X 40).

Discussion

Leiomyomas were first described by Virchow in 1854 as benign neoplasm composed of smooth muscle.[1] There are three types of leiomyomata cutis, namely piloleiomyoma arising from the arrector pili muscle of hair follicle, genital leiomyoma from the scrotal and labial dartos muscle as well as erectile smooth muscle of the nipple, and angioleiomyoma from the smooth muscle of vessels. Piloleiomyomata cutis may be solitary or multiple. Solitary lesion of PC has a slight male predominance whilst multiple lesions are more common in female. PC tends to affect young adults with a mean age of onset at 30. Familial cases of PC with associated uterine leiomyomas had been reported and such cases were named familial leiomyomatosis cutis et uteri or Reed syndrome.[2,3] PC usually presents as skin coloured to red brown smooth, round and firm nodules with diameter less than 2 cm. The nodules may coalesce to form extensive plaques. Paroxysmal pain which may be precipitated by cold and pressure is characteristic and can be incapacitating to patients with multiple lesions. Differential diagnoses of PC include other painful skin tumours such as blue rubber bleb naevus, eccrine spiradenoma, glomus tumour, granular cell tumour, neurilemmoma and cutaneous endometriosis.

Histologically, PC appears as uncapsulated growth in reticular dermis composed of well differentiated eosinophilic spindle cells with blunted nuclei interlacing with variable amounts of collagen. Mitotic rate was low even if present. Special stains such as Masson trichrome stain, Von Gieson stain, and PTAH (phosphotungstic acid haematoxylin stain) can be used to distinguish smooth muscle cells from surrounding collagen. PC also stain positively for desmin and actin which are markers of smooth muscle differentiation.[4] The measurement of haemoglobin in patients with multiple PC was advocated in some literature because leiomyoma may produce erythropoietin like substance. In female patients with multiple PC and positive family history, evaluation for visceral lesions such as uterine leiomyoma may be warranted.

Treatment of PC aims at symptom relief and improved cosmesis. Camouflage with cosmetics and cold avoidance can be the only measures required in asymptomatic cases. Surgical excision is the gold standard treatment for solitary PC. Excision of multiple PC is, however, impractical in most cases and associated with high recurrence rate. Various medications such as nifedipine, phenoxybenzamine, nitroglycerin,[5] doxazocin[6] and gabapentin[7] have been tried with variable results. There medications were proposed to either relax the in-situ smooth muscle or reduce the associated neurologenic pain. Carbon dioxide laser ablation was reported to result in promising pain relief in a case of multiple PC.[8] The prognosis for solitary PC is excellent after surgical excision. PC with multiple lesions tend to progress with no spontaneous involution.

References

1. Virhow R. Ueber Makroglossie und pathologische Neubildung quergestreifter Muskelfasern. Virchows Arch (Pathol Anat) 1854;7:126-38.
2. Garman ME, Blumberg MA, Ernst R, Raimer SS. Familial leiomyomatosis: a review and discussion of pathogenesis. Dermatology 2003;207:210-3.
3. Martinez-Mir A, Gordon D, Horev L, Klapholz L, Ott J, Christiano AM, et al. Multiple cutaneous and uterine leiomyomas: refinement of the genetic locus for multiple cutaneous and uterine leiomyomas on chromosome 1q42.3-43. J Invest Dermatol 2002;118:876-80.
4. Raj S, Calonje E, Kraus M, Kavanagh G, Newman PL, Fletcher CD. Cutaneous pilar leiomyoma: clinico-pathologic analysis of 53 lesions in 45 patients. Am J Dermatopathol 1997;19:2-9.
5. Holst VA, Junkins-Hopkins JM, Elenitsas R. Cutaneous smooth muscle neoplasms: clinical features, histologic findings, and treatment options. J Am Acad Dermatol 2002;46:477-90.
6. Batchelor RJ, Lyon CC, Highet AS. Successful treatment of pain in two patients with cutaneous leiomyomata with the oral alpha-1 adrenoceptor antagonist, doxazosin. Br J Dermatol 2004;150:775-6.
7. Alam M, Rabinowitz AD, Engler DE. Gabapentin treatment of multiple piloleiomyoma-related pain. J Am Acad Dermatol 2002;46(2 Suppl Case Reports):S27-9.
8. Christenson LJ, Smith K, Arpey CJ. Treatment of multiple cutaneous leiomyomas with CO2 laser ablation. Dermatol Surg 2000;26:319-22.