肢端汗管瘤

—病例報告—

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Acral Syringoma

—A Case Report—

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Although syringoma is a common adnexal tumor, acral presentation is very rare. We report a case of multiple acral syringomas that appeared as symmetrical grouped, slightly pigmented papules limited to the dorsum of both hands in a young woman. This clinical presentation should be included in the differential diagnosis of papular lesions on the hands. (Dermatol Sinica 20：214-217, 2002)

Key words: Acral syringoma

汗管瘤雖然爲常見皮膚附屬器腫瘤,但其於肢端之表現卻是相當罕見。我們報告一例年輕女性以雙手手背表現對稱群集丘疹之多發性肢端汗管瘤，此臨床表徵應與其他手部丘疹樣病變做爲鑑別。(中華皮誌20：214-217, 2002)

CASE REPORT

A 24-year-old woman visited our clinic because of a ten-year history of acquired asymptomatic eruptions on both hands. Her past health was well and has no mongolism. No lesions were present elsewhere. No family history was applicable. Physical examination revealed discrete papular lesions scattered over bilateral dorsal surfaces of hands. Close-up view of the right hand showed multiple grouped, slightly pigmented, 2 to 4 mm, flat-topped papules over the dorsal aspects of all proximal phalanges (Fig. 1); some of the grouping papules formed nearly confluent lesions. An incisional biopsy was obtained from the dorsal aspect of finger and stained with hematoxylin-eosin (H & E stain). The epidermis was slightly acanthotic, hyperkeratotic, and hyperpigmentated. The dermis contained many solid strands of basophilic cells and small cystic ducts with

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tadpole formation. The lining of the ducts was composed of one or two rows of flat epithelial cells. Some ducts demonstrated an intraluminal deposition of colloidal material. Gross cystic disease fluid protein(GCDFP-15) discloses negative finding. It showed typical histopathologic findings for syringoma (Fig. 2). Serial sections revealed neither a connection to underlying sweat glands nor to the epidermis.

DISCUSSION

Syringomas were first reported under the name "lymphangioma tuberosum multiplex" in 1872. Darier subsequently recognized two types of syringoma: one called hidradenoma of the lower eyelid; the other, eruptive hidradenoma.[1]

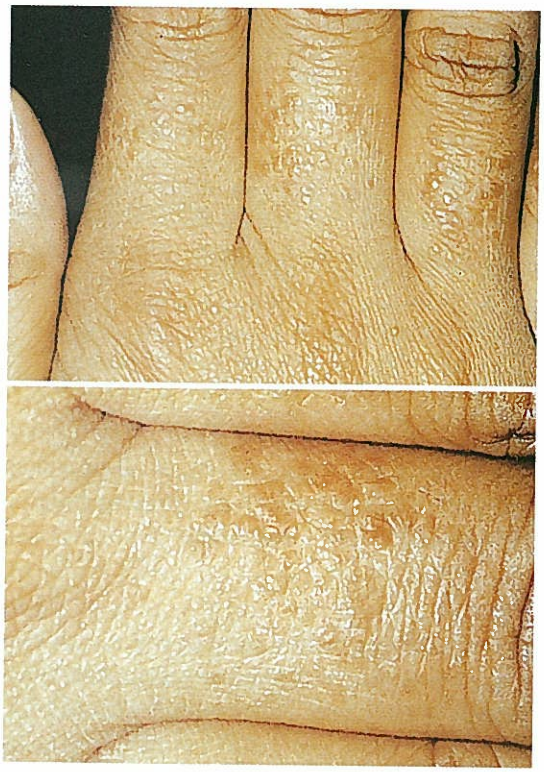

Fig. 1
Upper
Multiple grouped, slightly pigmented, 2 to 4 mm, flat-topped papules over all the dorsal aspects of proximal phalanges.
Lower
Some of them formed nearly confluent lesions.

Syringomas are common adnexal tumors most probably derived from intraepidermal eccrine ducts based on histochemical and electron microscopic examinations.[2] Usually, they present as numerous small papules on the lower eyelids and cheeks of women. (Female/male ratio of 2:1) Eighteen percent of individuals with Down's syndrome develop syringoma, a predilection that is not observed in other chromosomal abnormality syndromes.[3] Other variants are solitary,[4] milia-like,[5] and lichen planus-like lesions.[6] Unusual locations include the vulva,[7] penis,[8] ankle,[4] upper extremities,[9] forehead[10] and scalp.[11] Acral,[12] unilateral,[13] linear[14] or bathing trunk distributions[15] have also been documented. Eruptive and disseminated forms,[16] some of which are familial,[17] and even urticaria pigmentosa-like presentations[18] have been described. (Table I)

Acral syringomas should be considered in the differential diagnosis of papular lesions arising on the hands, including lichen planus, lichen nitidus, lichenoid contact dermatitis, verruca plana, molluscum contagiosum, and acrokeratosis verruciformis. The four P's-purple, polygonal, pruritic, papule-and the Wickham stirae of lichen planus is typical and sufficient to make the correct diagnosis. Lichen nitidus is composed of multiple pinpoint-sized, glistening papules. The papules are smaller and they do not become confluent. Lichenoid contact dermatitis could be diagnosed by specific contact history. Verruca plana often shows linear configuration due to autoinoculation. The lesions of molluscum contagiosum are dome-shaped papules, often with central umbilication.

Acrokeratosis verruciformis may resemble acral syringoma clinically; however, punctate keratoses on the palms and the nail plates changes are usually present in the former. Conclusively, the differential diagnosis on clinical grounds alone may be difficult, and histological examination is frequently required.

Syringomas rarely affect the extremities. Usually, they appear in a form of eruptive syringomas. To the best of our knowledge, symmetrical syringomas located on the distal

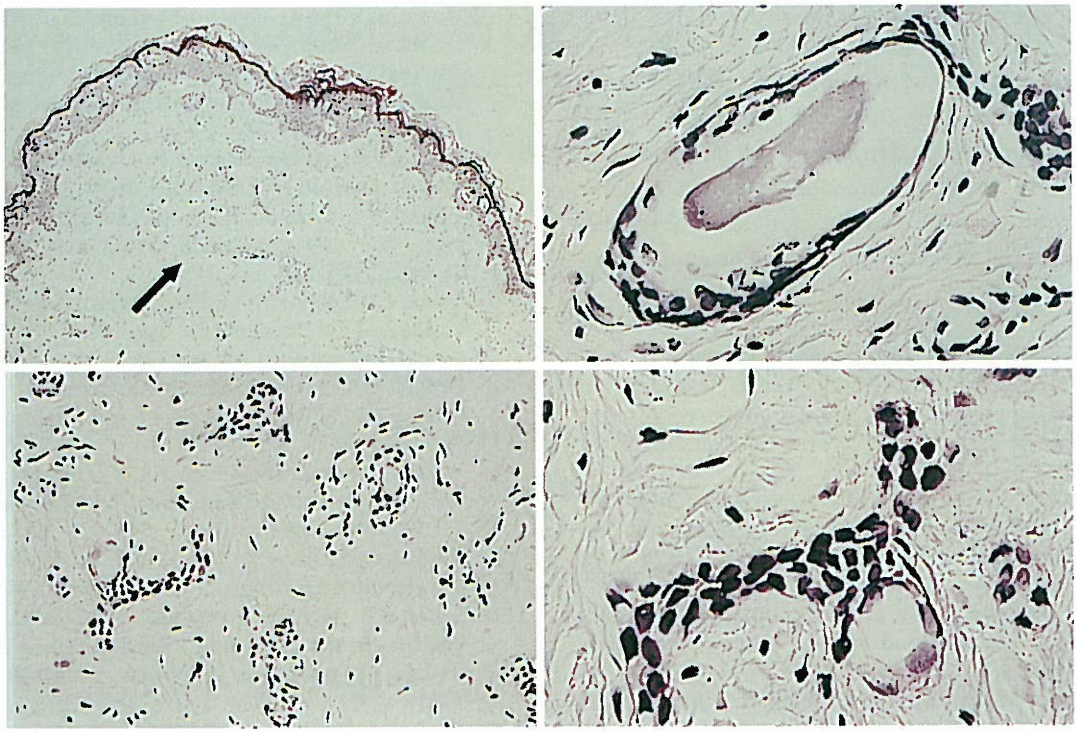

Fig. 2

Upper left: Epidermis was slightly acanthotic, hyperkeratotic, and hyperpigmentated. The arrow pointed out where the tumor located. (H & E stain 40x)

Lower left: The tumor was composed of many solid strands of basophilic cells and small cystic ducts. (H & E stain 100x)

Upper right: The walls of the ducts were lined by one or two rows of flat epithelial cells. It demonstrated an intraluminal deposition of colloidal material.(H & E stain 400x)

Lower right: The duct had comma-like tail, giving it the appearance of tadpole. (H & E stain 400x)

Table I. Clinical variants of syringoma (Friedman and Butler classification)

Solitary	Multiple	
Papulonodular[4]	Localized	Disseminated
Alopecia: scalp[11]	*Papular*	*Papular*
	Infraorbital	Eruptive[16]
	Genital[5,6]	Lichen planus-like[6]
	Acral[12]	Urticaria pigmentosa-like[18]
	Forehead[10]	Milia-like[16]
	Upper extremities[9]	Familial[17]
	Bathing trunk[15]	In Down's syndrome[22]
	Unilateral[13]	
	Linear[14]	
	Milia-like[5]	
	In Down's syndrome[21]	
	Plaque-like	
	Unilateral	

aspects of the upper extremities have been described in 4 cases as an isolated findings[9,12] or in association with typical eruption around eyelids.[1,19] Besides, there was one report of acral syringomas on the dorsal aspect of the hands and feet.[20] Our case presented a distinctive clinical picture and should be considered in the differential diagnosis of papular lesions of the hands.

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