Eruptive syringoma in a patient with trisomy 21

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ABSTRACT

Syringomas are benign adnexal neoplasia that are classified into four main types, according to their clinical features and associations. Syringoma associated with trisomy 21 typically presents with a periorbital cutaneous involvement. We report an 18-year-old Chinese girl with trisomy 21, who presented with eruptive syringoma, an unusual generalised form involving her trunk and limbs. A histological examination was performed to confirm the diagnosis.

Keywords: adenexal tumour, Down syndrome, eccrine tumour, eruptive syringoma, trisomy 21

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INTRODUCTION

Syringomas are benign adnexal neoplasia derived from eccrine ductal elements. The presentation of syringoma in patients with trisomy 21 typically involves the periorbital region. Less commonly, syringoma can have an extensive involvement, affecting the trunk and limbs of the patient. A skin biopsy is required to confirm the diagnosis and to exclude other rare cutaneous manifestations found in trisomy 21.

CASE REPORT

We report an otherwise healthy 18-year-old Chinese girl who presented with a sudden onset of papules over her trunk and limbs over the past one month. The papules appeared in crops but were otherwise asymptomatic. The patient had a history of trisomy 21. The physical examination revealed generalised scattered skin-coloured to reddish brown papules over the face, as well as the extensor aspects of both the upper and lower limbs (Figs. 1 & 2). There was no associated hypohidrosis. The patient's nails, hair and teeth were normal. A skin biopsy of a papule was obtained.

The histological examination showed a dense fibrous stroma. There was a proliferation of eccrine-type ducts lined by rows of cuboidal cells, forming dermal strands and cords. Some of the cells were in a tadpole-like configuration, with a normal connective tissue stroma. The lumen was filled with eosinophilic, amorphous debris (Fig. 3). A diagnosis of eruptive syringoma was made.

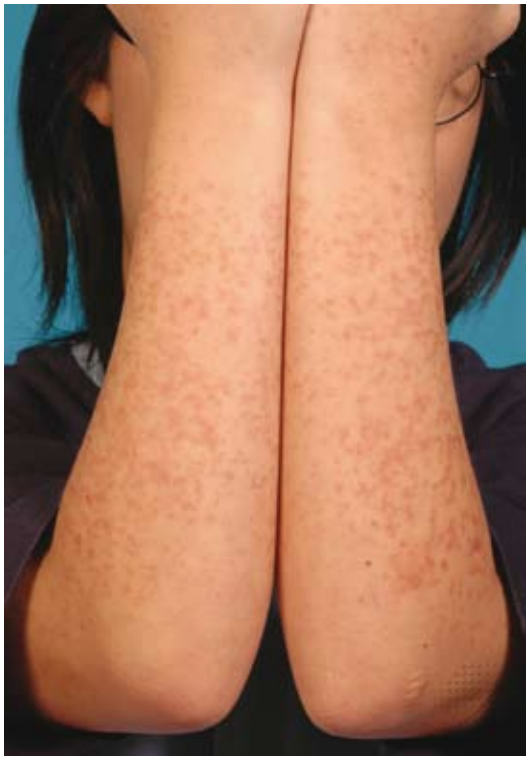

Fig. 1 Photograph shows skin-coloured and reddish brown papules on the extensor surfaces of the forearms.

DISCUSSION

Syringomas are benign adnexal neoplasia and are traditionally believed to be derived from eccrine ductal elements. Clinically, they present as small skin-coloured dermal papules and are usually smaller than 3 mm in diameter. The usual sites include the upper part of the cheek and the lower eyelids. Syringomas are fairly common, with a female preponderance. They are usually incidental although some cases may be familial. They usually arise at the onset of puberty, with additional lesions developing later. They are usually symmetrical and appear in clusters. Although facial syringomas are largely of cosmetic significance, the lesions must be distinguished from trichoepitheliomas and basal cell carcinoma. Syringomas have been classified into four groups, according to their clinical features and associations, namely localised form, generalised/

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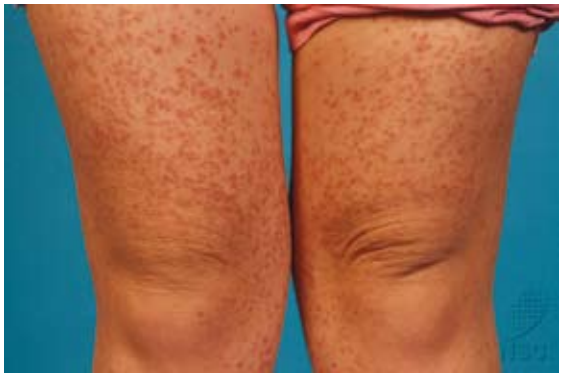

Fig. 2 Photograph shows similar papules on the thighs.

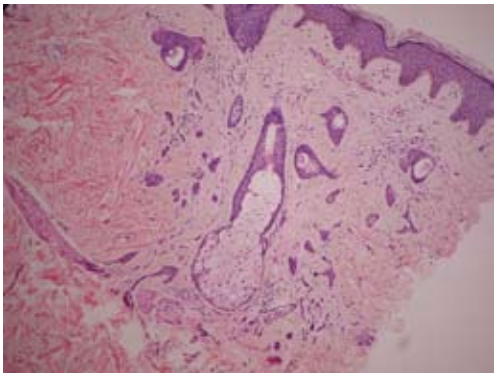

Fig. 3 Photomicrograph shows multiple strands of epithelial cords and ductular structures with tadpole appearance are seen in the upper to mid-dermis (Haematoxylin & eosin, × 100).

eruptive form, a form associated with trisomy 21 and familial form.[1]

Syringomas that are associated with trisomy 21 typically present with periorbital syringoma. Although sporadic cases of an eruptive variant have been reported, as in the case of our patient, it has yet to be established whether a higher incidence of the eruptive form is seen in patients with trisomy 21. Other rare forms of cutaneous manifestations associated with trisomy 21 include milia-like idiopathic calcinosis cutis (MICC)[2,3] and elastosis perforans serpiginosa (EPS). MICC is a micronodular, whitish acral calcified lesion of unknown pathogenesis, while EPS is a rare granulomatous inflammation, in which the abnormal elastic tissue fibres and cellular debris are expelled from the papillary dermis through the epidermis.

Eruptive syringomas, in particular, can be found in the axillae, chest and abdomen, in addition to the usual site on the face. They also tend to appear simultaneously. The incidence of syringomas appears to be higher in Asians and African-Americans. The diagnosis may be established by performing a skin biopsy, particularly for facial lesions, to exclude the abovementioned differential diagnoses. The typical histological features include multiple small ducts lined by rows of flattened epithelial cells with epithelial strands within the dermis, giving rise to the characteristic tadpole appearance.

Cosmesis is the main reason for treatment, and there is no long-term significant morbidity associated with syringomas. Facial lesions, particularly over the eyelids and cheeks, may result in significant distress in patients. The main aim of the treatment is to minimise scarring and to prevent recurrence. There have been no comparative studies to suggest definitive treatment modalities, although the possibilities include surgical excision, electrocautery, cryotherapy, dermabrasion, trichloroacetic acid and carbon dioxide laser ablation. In this case, the patient was treated with topical tretinoin without any significant improvement.

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