Chondroid syringoma

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ABSTRACT

Pleomorphic adenoma, or chondroid syringoma, (CS) is a rare, benign, skin appendageal tumour. Because of the unremarkable clinical presentation of this rare tumour, the diagnosis is made on microscopic examination. The usual presentation is of an asymptomatic, slowly-growing mass, typically located in the head and neck region. We present a case of a CS located over the forehead of a 32-year-old man. In the evaluation of a middle-aged male patient with a small cutaneous nodule in the head and neck region, chondroid syringoma should also be considered in the differential diagnosis. For such a lesion, excisional biopsy, without destroying the aesthetic and functional structures, is the preferred diagnostic, as well as therapeutic approach.

Keywords: chondroid syringoma, pleomorphic adenoma, skin tumour

Singapore Med J 2008;49(2):e33-e34

INTRODUCTION

Pleomorphic adenoma, or chondroid syringoma (CS), is a rare, benign, skin appendageal tumour.[1] Because of the unremarkable clinical presentation of this rare tumour, the diagnosis is often made after microscopic examination.[2] We report a case of CS on the forehead, and review the relevant literature.

CASE REPORT

A 32-year-old man presented with a progressively increasing swelling on the region of the forehead and superior-medial aspect of the left eye during the past two years. Physical examination showed a firm, painless, mobile, 5 mm × 5mm nodule, covered by normal skin. The nodule was excised and sent for histopathology. Gross examination showed a well-circumscribed, whitish firm tumour, surrounded with capsule-like tissue. Histological examination revealed abundant chondroid stroma with fibrous areas containing epithelial and myoepithelial cells, arranged as small aggregates and ducts. The epithelial cells were cuboidal with an eosinophilic cytoplasm and regular oval nuclei (Fig. 1). The mucoid stroma gave a positive reaction with Alcian blue. Based on histopathological findings, a diagnosis of CS was made. The patient was doing well and there was no recurrence at follow-up.

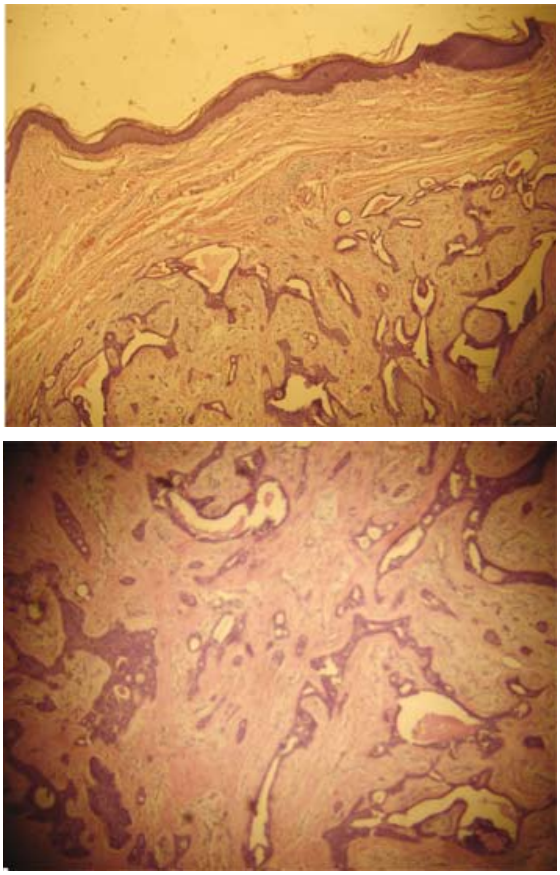

Fig. 1 Photomicrograph of the cord and duct structures in the chondromyxoid stroma shows a characteristic homogeneous basophilic chondroid stain (Haematoxylin and eosin, × 40).

DISCUSSION

CS is rare among primary skin tumours; the reported incidence is < 0.098%.[2] CS usually affects middle-aged or older male patients.[2,3] The site of predilection for CS is the head and neck region; less commonly, these tumours can involve the hand, foot, axillary region, abdomen, penis, vulva, and scrotum.[2,4-9] Typical clinical presentation of these tumours is a slow-growing, painless, firm, non-ulcerated cutaneous or intracutaneous nodule (0.5–3 cm in size).[2-4] Hirsch and Helwig used the term, chondroid syringoma, in place of pleomorphic adenoma of the skin, as the tumour is epithelial in nature, with associated secondary changes in the stroma.[10,11] Histologically, CS contains an admixture of epithelial-myoepithelial structures within a chondromyxoid and fibrous stroma arranged in cords and forming tubules.[2,11] Differentiation towards various skin adnexal structures (including hair matrix, hair follicle, apocrine, and sebaceous

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glands) is rare.[12,13] CS may be confused clinically with various skin lesions, including benign tumours of epidermal or mesenchymatous appendages, such as dermoid or sebaceous cyst, and neurofibroma.[2] Various treatment options have been proposed for CS, including electrodesiccation, dermabrasion, and vaporisation with argon or CO_2 laser.

Because of the risk of malignancy, the first-line treatment is total excision of the tumour without destroying the aesthetic and functional structures, as is in the present case.[2] This should be followed by regular follow-up to look for local recurrence and any feature of malignancy.[14] The recurrent lesion can be treated by surgical re-excision.[15] Malignant CS is one of the rarest subtype, and appears to behave in an aggressive manner.[16,17] Malignancy in CS is rare, with reported cases occurring in young female patients in the extremities and torso.[16,18] Tumours greater than 3 cm in size have a greater likelihood of malignancy.[16,19] Histological features that suggest malignant transformation in a CS include cytologic atypia, infiltrative margins, satellite tumour nodules, tumour necrosis, and involvement of deep structures.[11,20] For malignant lesions, the initial treatment modality is aggressive surgery. Adjuvant radiotherapy, with or without chemotherapy, may be recommended.[16,17]

In conclusion, in the evaluation of a middle-aged male patient with a small cutaneous nodule in the head and neck region, CS should be considered in the differential diagnosis. For such a lesion, excisional biopsy without destroying aesthetic and functional structures is the preferred diagnostic, as well as therapeutic, approach.

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