

## Chondroid Syringoma: Report of a Case with Uncommon Location

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### Introduction

Chondroid Syringoma or pleomorphic adenoma is a rare skin adnexal tumour which can have either a benign or a malignant behaviour.

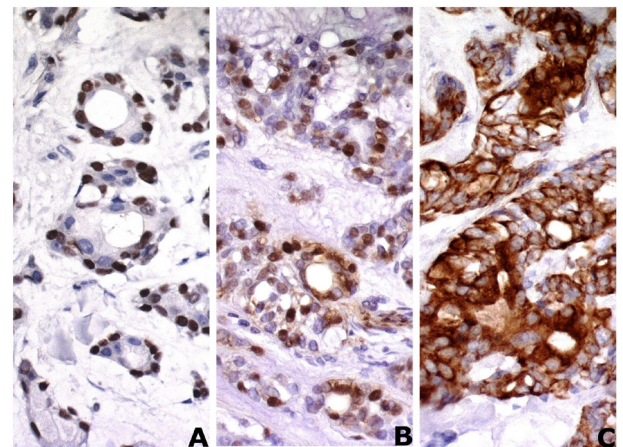
“The mixed tumor” of the skin was originally defined by Billroth in 1859 as an entity having the same histopathologic properties of the mixed tumors of the salivary glands [1]. The term ‘chondroid syringoma’ was first used by Hirsch and Helwig in 1961 to describe this sweat gland tumor, because of the presence of sweat gland elements which are set in a cartilaginous stroma [2]. The reported incidence of CS among primary skin tumor is low, ranging between 0.01- 0.098% [3]. This uncommon eccrine sweat gland tumor clinically presents as a slow-growing, painless, subcutaneous or intracutaneous nodule located usually in the head and neck region, and it affects middle-aged or older men [3,4]. Less commonly, this tumor can develop in the axillary region, penis, vulva, and scalp. We report a rare case of a chondroid syringoma with an atypical location on the back.

### Case Report

A 45 years old man was referred to the Unit of General Surgery and Surgical Oncology of the University of Siena, with a small nodular mass of 1.5 cm located at left side of the back. The lesion was light-brown with hard-elastic consistency, mobile on the subcutaneous plane. The patient reported a history of recurrent episodes of scratching and subsequent rupture of the mass, with emission of white-brown secretions and reduction of the diameter of the lesion.

A surgical excision of the mass was performed under local anesthesia.

The histological examination (Figures 1 and 2) showed a dermal and subcutaneous, well-circumscribed, multilobulated neoplasm characterized by proliferation of clusters of epithelial cells distributed in nests or in tubular structures, lined by two rows of cells, with round to ovoid nuclei. The stroma had myxoid and hyaline degeneration with areas of chondroid metaplasia. Most of the lesion showed a low

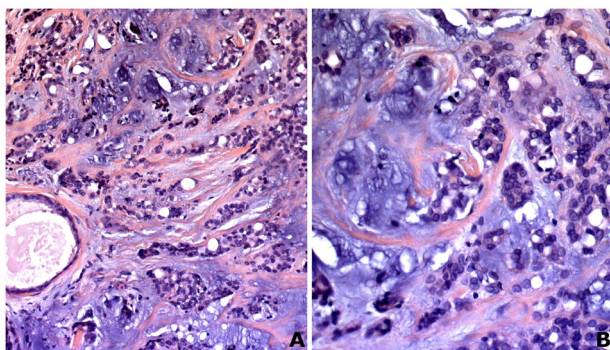


**Figure 2:** Immunohistochemistry: Areas with myoepithelial differentiation showing nuclear (A) Positivity and cytoplasmic (B) Positivity for p63 stain and S-100 stain antibodies, respectively; (C) Cytokeratin 7 decorates the inner cell layer (immunohistochemical staining, 200x).

mitotic activity. However, a focal area with nuclear pleomorphism and a mitotic index (Ki-67) of 20% was present. Immunohistochemistry was performed and the tumor cells displayed widespread and strong positivity for cytokeratin 7. In the areas with myoepithelial differentiation, p63, a myoepithelial marker, and focally S-100, were also positive. The margins of excision were positive and hence we carried out a wide surgical re-excision of the previous scar with a 2 cm margin. Histological examination of the re-excised specimen was free of tumor. At two years follow-up, the patient has no local recurrence.

### Discussion

CS is an uncommon eccrine sweat gland tumor that may originate from both secretory and ductal elements of the sweat gland. The tumor clinically presents as a slow-growing, asymptomatic, solitary, painless, nonulcerated subcutaneous or intracutaneous nodule. The lesion is commonly mobile and distinct from the surrounding tissues. CS lesions are usually not clinically distinctive, and the diagnosis is made on excisional biopsy and microscopic examination [3,4]. The differential diagnosis of CS is made with other tumors of skin adnexa,



**Figure 1:** HE: (A) Epithelial cells arranged in nests and tubular structures with a myxoid and hyalinized stroma (haematoxylin-eosin, 100X); (B) Detail of stromal chondroid metaplasia (haematoxylin-eosin, 200X).

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such as dermoid or sebaceous cysts, neurofibroma, dermatofibroma, pilomatricoma, histiocytoma, basal cell carcinomas, pilomatricomas and seborrheic keratosis.

Rare cases of malignant CS have also been reported. These malignant forms occur more commonly in younger female patients, and have a predilection for the extremities; tumors are often larger than 3 cm and locally invasive, lymph node spread is common (48%) and metastasis to bones and lung is very rare [5,6].

Histopathology of CS consists of both epithelial and mesenchymal elements; hence, the tumor has been termed mixed. The epithelial cells are arranged in cords, or in tubules with a myoepithelial layer, set in a myxoid or chondroid stroma [3]. In 1961, Headington classified chondroid syringoma into two histologic variants: the eccrine type with smaller lumens lined by a single row of cuboidal epithelial cells and the apocrine variant, which is more common, with tubular and cystic branching lumina lined by two rows of epithelial cells [7]. Some tumors may exhibit both apocrine and eccrine features. As reported in the literature, in our case there was evidence of epithelial as well as myoepithelial findings. The inner cell layer of the tubuloglandular components of chondroid syringoma expressed epithelial markers, including Cytokeratin (CK), Epithelial Membrane Antigen (EMA), and Carcinoembryonic Antigen (CEA). The outer cell layer is negative for CK, EMA, and CEA, but stained positive with mesenchymal markers, including S-100 protein. Negativity to p15, which is considered an apocrine marker, suggested an eccrine differentiation in our case. The stromal cells have immunohistochemical staining features similar to those of the outer cell layer, suggesting that the stromal components may be produced from the myoepithelial elements [8].

Differentiation between benign and malignant forms is sometimes difficult because of similarities of histological features. The atypical histological findings such as cytologic atypia, increased mitotic figures, infiltrative margin, satellite tumor nodules, and tumor necrosis are considered markers of malignant transformation and involvement of deep structures [3,9-11]. The immunohistochemical study doesn't help in distinguishing between benign and malignant lesions.

Despite the presence of a small proliferative area, showing several nuclear atypia, the lesion pursued a benign clinical course: neither recurrences nor metastases were in fact evident after 24 months.

This tumor may be treated with various methods, including electrodesiccation, dermabrasion, and vaporization with argon or CO<sub>2</sub> lasers. The treatment of choice is complete excision with a cuff of normal tissue in order to examine the histopathologic features [7]. Histologically malignant and large tumors (greater than 3 cm in size) may require a wide excision with a minimum 1-cm margin to prevent local recurrence. Because of malignant potential, a wide excision of CS must be done and patient should be followed carefully for both local recurrence and metastasis. Regional lymph node resection is indicated in the presence of clinically suspicious or palpable lymph node metastases. Few authors recommended an aggressive surgery and adjuvant radiotherapy, with or without chemotherapy as initial treatment modality for malignant tumors [12,13].

## References

1. Sheikh SS1, Pennanen M, Montgomery E (2000) Benign chondroid syringoma: report of a case clinically mimicking a malignant neoplasm. *J Surg Oncol* 73: 228-230.
2. Hirsch P, Helwig EB (1961) Chondroid syringoma. Mixed tumor of skin, salivary gland type. *Arch Dermatol* 84: 835-847.
3. Yavuzer R, Başterzi Y, Sari A, Bir F, Sezer C (2003) Chondroid syringoma: a diagnosis more frequent than expected. *Dermatol Surg* 29: 179-181.
4. Sungur N, Uysal A, Gümüş M, Koçer U (2003) An unusual chondroid syringoma. *Dermatol Surg* 29: 977-979.
5. Watson JA, Walker MM, Smith NP, Hunt DM (1991) Malignant chondroid syringoma--a rare cause of secondary bone tumour. *Clin Exp Dermatol* 16: 306-307.
6. Kiely JL, Dunne B, McCabe M, McNicholas WT (1997) Malignant chondroid syringoma presenting as multiple pulmonary nodules. *Thorax* 52: 395-396.
7. Chen AH, Moreano EH, Houston B, Funk GF (1996) Chondroid syringoma of the head and neck: clinical management and literature review. *Ear Nose Throat J* 75: 104-108.
8. Argenyi ZB, Balogh K, Goeken JA (1988) Immunohistochemical characterization of chondroid syringomas. *Am J Clin Pathol* 90: 662-669.
9. Sheikh SS, Pennanen M, Montgomery E (2000) Benign chondroid syringoma: report of a case clinically mimicking a malignant neoplasm. *J Surg Oncol* 73: 228-230.
10. Bates AW, Baithun SI (1998) Atypical mixed tumor of the skin: histologic, immunohistochemical, and ultrastructural features in three cases and a review of the criteria for malignancy. *Am J Dermatopathol*; 20: 35-40.
11. Borman H, Özcan G (1998) Chondroid syringoma at the fingertip: an unusual localization. *Eur J Plast Surg* 21: 311-313.
12. Barnett MD, Wallack MK, Zuretti A, Mesia L, Emery RS, et al. (2000) Recurrent malignant chondroid syringoma of the foot: a case report and review of the literature. *Am J Clin Oncol* 23: 227-232.
13. Hong JJ, Elmore JF, Drachenberg CI, Jacobs MC, Salazar OM (1995) Role of radiation therapy in the management of malignant chondroid syringoma. *Dermatol Surg* 21: 781-785.