

Giant Chondroid Syringoma - Case Report of Rare Entity at Unusual Site – Cytohistological Features

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Abstract: Chondroid Syringomas are rare benign mixed tumors of skin adnexal origin and are composed of both epithelial and mesenchymal components. These tumors usually effects middle aged and older men with commonest involvement of head and neck region. There are few case reports with cytohistological correlation. We present a rare case of giant chondroid syringoma over the thigh in 44 years male with gradually increasing size. FNAC was done and aspirated material is thick and mucoid. Microscopic examination revealed chondromyxoid background with sheets and clusters of epithelial and myoepithelial cells. Based on these findings the diagnosis of Chondriod Syringoma was made. After surgical excision we received a nodular mass of 4.5 x 3.5 x 2 cm. On histopathological examination it was found that there were epithelial and myoepithelial cells with characteristic chondromyxoid stroma. Adipose tissue is also seen. Based on these findings the diagnosis of giant chondroid syringoma was made. FNAC is useful to determine the pathology before excision; however histopathological examination of the excised tissue is more reliable for definitive diagnosis.

Keywords: Adnexal tumor, Giant chondroid syringoma, Thigh region, FNAC, Histopathology

I. Introduction

Chondroid syringoma is a rare benign skin Adnexal tumor, which was first described by Biloeth in 1859, which have both benign and malignant forms. It is also known as mixed tumor of the skin as it is composed of both epithelial and mesenchymal components. The term chondroid syringoma was introduced by Hirsch and Helwig in 1961 because of the presence of sweat gland elements (syringoma) and cartilaginous matrix (chondroid).^[1] The reported incidence of the tumor is <0.098%.^[2] It is usually located on the head and neck of adults. Most are small and less than 3cm.^[3] This case is reported because of unusual site of occurrence, large size and there are only few case reports showing the characteristic FNAC features. The diagnosis must be confirmed by histopathology.

II. Case Report

A 44 years old male presented to the surgical outpatient department with painless swelling over the medial aspect of left thigh which had been gradually increasing in size for 1year. Clinical examination revealed a non-tender swelling of 4 x 3 cm. with nodular surface and is freely mobile. Clinical diagnosis of lipoma was made and sent for FNAC.

III. Cytology

FNAC was done with the help of 22G needle and 10CC syringe. Smears were prepared and fixed in 95% isopropyl alcohol and stained with hematoxylin and eosin stain. Grossly the aspirate is thick and mucoid. Microscopic examination revealed chondromyxoid ground substance with epithelial and myoepithelial cells. The epithelial cells have round monomorphic nuclei having dispersed chromatin with moderate to abundant clear cytoplasm. Adipocytes are also seen. Based on these features the diagnosis of chondroid syringoma was made.

IV. Histopathology Findings

2.2.1 Gross

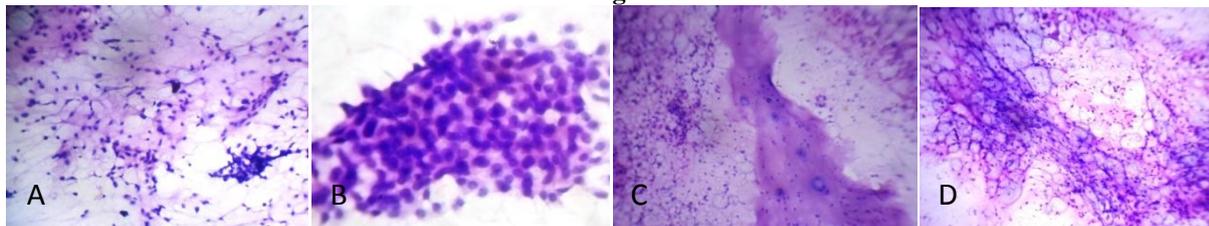
After surgical excision we received a tan colored mass of 4.5 x 3.5 x 2 cm. with nodular surface and attached skin. Cut section is lobulated, tan coloured with grey white to myxoid areas.

V. Microscopy

The histopathological examination revealed circumscribed lolulated mass composed of chondroid, fibromyxoid and osteoid areas with epithelial and myoepithelial elements. The epithelial elements are arranged in sheets, cords, tubuloalveolar pattern and duct like structures with eosinophilic material. These ducts are lined

by single layer of cells. Areas of lipomatous differentiation are observed. Based on these features the diagnosis of chondroid syringoma was made.

Fig 1



Microscopic examination of smears

Fig 1 A- epithelial and myoepithelial cells in myxoid stroma (200X H&E)

Fig 1 B- epithelial and myoepithelial cells in higher magnification (400X H&E)

Fig 1 C- Chondromyxoid stroma(200X H&E)

Fig 1 D- adipose tissue (200X H&E)

Fig 2

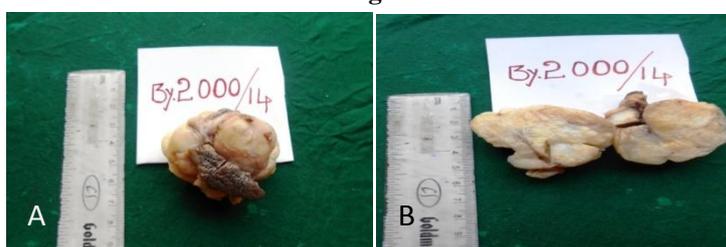


Fig. Gross: 2- A –Nodular mass of 4.5 X 3.5 X 2 cm.

2- B –On cut section showing tan lobulated mass with grey white and myxoid areas

Fig 3

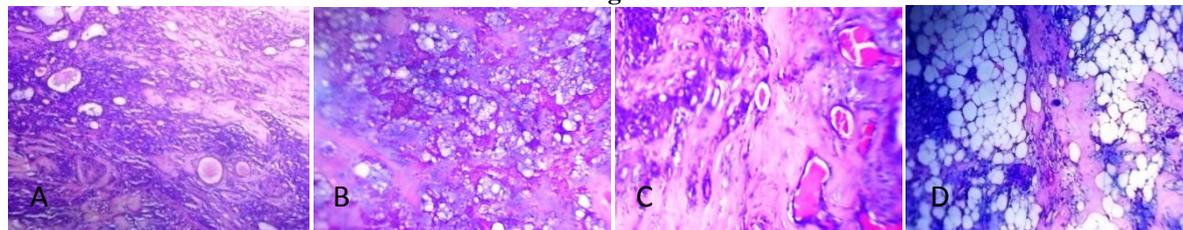


Fig 3 A and C- Tubuloalveolar and duct like pattern (100X H&E) ,(200X H&E).

Fig 3 B and D- Chondromyxoid and lipomatous differentiation, osteoid differentiation.(200X H&E).

VI. Discussion

Chondroid syringoma is a rare benign skin Adnexal tumor, which was first described by Bilioth in 1859, which have both benign and malignant forms. The term chondroid syringoma was introduced by Hirsch and Helwig in 1961 because of the presence of sweat gland elements (syringoma) and cartilagenous matrix (chondroid).^[1] The reported incidence of chondroid syringoma is < 0.098 % and effects middle aged and older men.^[2,4] The most common sites are head and neck region, hand, foot, the axillary region, abdomen, penis, vulva and scrotum. Giant chondroid syringomas occurring at unusual sites such as axilla, arm and shoulder have been reported.^[2, 5] It is non ulcerating, slow growing, subcutaneous or cutaneous nodule. Most are small and less than 3 cm. If it is more than 3 cm. indicates giant form.^[3] Virchow and Minssen referred them as mixed tumours with both epithelial and mesenchymal elements.^[1,2]

Fine needle aspiration yields thick mucoid and gelatinous material. Cytology smears show epithelial and myoepithelial cells and stromal components. Epithelial cells are round to polygonal cells with monomorphic nuclei and finely dispersed chromatin and moderate to abundant cytoplasm. Some cells show eccentrically placed nuclei. Stromal component is chondromyxoid or fibromyxoid.^[6,7]

Histologically consists of epithelial cells arranged in cords and tubules with a myoepithelial layer. Headington described two histological variants of this tumor, the eccrine type with uniform small round tubules, lined by single row of epithelial cells and the apocrine variant with tubular and cystic branching lumina, lined by two rows of epithelial cells.^[8] The tubuloalveolar components of Chondroid Syringomas are composed of two layers of cells with different immunophenotypes. The inner layer expresses epithelial marker cytokeratin. The

outer layer expresses mesenchymal markers such as vimentin, S-100 protein, neuron specific enolase (NSE) and glial fibrillary acidic protein (GFAP).^[6,7,9]

The stroma of Chondroid syringomas is mucoid, myxoid, chondroid, adipose and rarely osteoid. Stromal mixtures are common in these lesions. Cartilaginous matrix produced by myoepithelial cells is true cartilage.^[9] Differential diagnosis of this tumor includes benign tumors of epidermal or mesenchymatous appendages such as dermoid or sebaceous cyst, neurofibroma, dermatofibroma, basal cell carcinoma, pilomatricoma, histiocytoma and seborrheic keratosis.

Because of risk of recurrence and malignant transformation the first line of treatment is total excision with margin of normal tissue.^[8,10] This should be followed by regular follow up to look for local recurrence and features of malignancy. The recurrent lesions can be treated by surgical re-excision.^[2] Malignancy in chondroid syringoma is rare with reported cases occurring in young female patients in the extremities.^[11]

Tumors of greater than 3 cm. in size have greater likelihood of malignancy. Histological features that suggest malignancy includes cytological atypia, satellite nodules, infiltrative margins, tumor necrosis and involvement of deeper structures.^[8,12]

For malignant lesions the initial treatment modality is aggressive surgery. Adjuvant radio therapy with or without chemotherapy may be recommended.^[7,11]

VII. Conclusion

In the case of patients with cutaneous or subcutaneous nodules on thigh the differential diagnosis of chondroid syringoma should be considered. FNAC has been used for diagnostic purpose and may prove to be useful to determine the pathology before excision. However excisional biopsy is most reliable in establishing a definitive diagnosis and as well as therapeutic approach. If the lesion is more than 3 cm. It is giant chondroid syringoma to which follow up is essential to look for recurrence and malignant transformation.

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