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Case Report

Pathology Section

Chondroid Syringoma–An Unusual Presentation

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ABSTRACT

Chondroid syringoma is a rare mixed tumour of the skin. It was first described by Hirsch and Helwig. The tumour is composed of a proliferation of epithelial cells present in a myxoid and chondroid matrix. Although chondroid syringoma was predominantly benign, sweat-gland tumour, the usual presentation is of an asymptomatic, slowly growing mass, typically located on the head and neck region. The present case was located in right leg, lateral aspect and rare.

Keywords: Chondroid syringoma, Mixed tumour, Sweat-gland

CASE REPORT

A 27-year-old female presented with a swelling in the lateral aspect of right leg since 8 years. The swelling had started as small in size and had grown slowly and attained the present size. The local examination revealed a firm to hard mass measuring 4.5 cm x 4.3cm situated over the lateral aspect of right leg. It was mobile and neither attached to the skin nor to the underlying structure.

Patient was not a known diabetic, hypertensive, or having tuberculosis. Her general condition was fair.

A provisional diagnosis of calcified lipoma was entertained.

Routine investigations

Hemoglobin – 10.5gm/dl, Total WBC count – 8600 cells/cumm, Differential count – N 68%, L 23%, E 4%, M 5%

Ultra sonogram: A well-defined heter ogeneous mass of 47 x 45 x 37mm size in the right leg. The mass had mixed echo texture on the lateral aspect. No obvious calcification was present.

The tumour was excised and sent for histopathological examination.

Grossly, the excised tumour mass with attached elliptical piece of skin measured 4.5x4.3x3.5 cm. Its external surface was smooth and nodular. Cut section was solid and showed islands of cartilage [Table/Fig-1].

Microscopic features [Table/Fig-2,3,4]

Sections showed a well-circumscribed tumour. The tumour was composed of cuboidal to polygonal cells with abundant cytoplasm. The tumour cells were arranged in the form of cords, sheets and intercommunicating tubuloalveolar structures. Some of the tubules contained eosinophilic material. In some areas small cysts were lined by squamous epithelium. The stroma was abundant and predominantly composed of myxoid tissue. In areas, they appeared like cartilage. Few lobules of adipose tissue were also present. No mitosis was present. The surgical margin was very near to the tumour.

A definite diagnos is of chondroid syringoma was made and a follow up was advised.

DISCUSSION

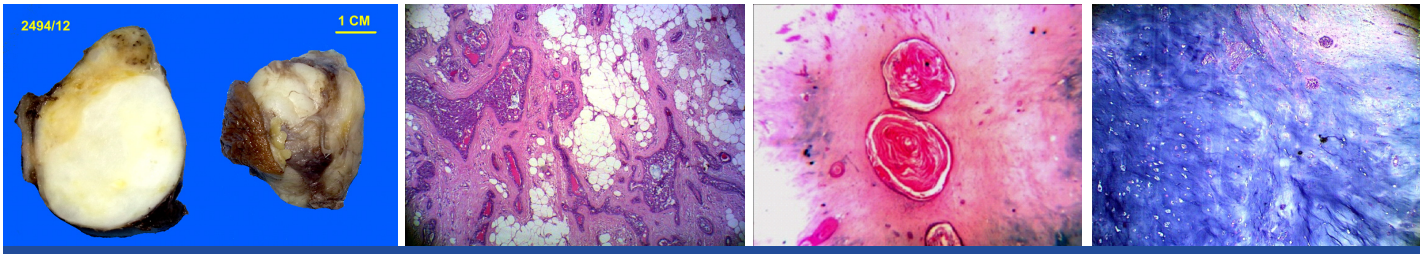
Chondroid syringoma is also known as mixed tumour of the skin. The tumour is composed of both epithelial and mesenchymal components. Hirsch and Helwig named it chondroid syringoma because of the presence of sweat gland elements in a cartilaginous stroma [1] and the histological criteria for diagnosis.

Histological features

(1) Nests of cuboidal or polygonal cells; (2) intercommunicating tubuloalveolar structures lined with two or more rows of cuboidal cells; (3) ductal structures composed of one or two rows of cuboidal cells; (4) occasional keratinous cysts; (5) a matrix of varying composition. Chondroid syringomas may have all five characteristics or manifest only some [1].

In our case all above features are present.

Chondroid syringomas share similarities with pleomorphic adenomas, which are mixed tumours that arise from the salivary glands [2]. In contrast to pleomorphic adenomas, chondroid syringomas are thought to arise from sweat glands.



[Table/Fig-1]: The excised tumor mass with attached elliptical piece of skin measured 4.5x4.3x3.5 cm. The external surface was smooth and nodular. Cut section was solid and showed islands of cartilage
[Table/Fig-2]: Sections showed a well-circumscribed tumor composed of cuboidal to polygonal cells with abundant cytoplasm. The tumor cells were arranged in the form of cords, sheets and intercommunicating tubuloalveolar structures. Some of the tubules contained eosinophilic material. Few lobules of adipose tissue were also present. No mitosis was present
[Table/Fig-3]: Section showed keratin cysts, **[Table/Fig-4]:** Section showed abundant myxoid stroma and in areas the stroma appeared like cartilage

Incidence: (Age, Sex, Site)

The incidence of chondroid syringoma is low 0.01-0.098 percent [3]. The middle-aged men were affected more than women [3, 4].

The lesions are typically located in the head and neck region.

In the present case the lesion was located on the extremity (right leg, lateral aspect) and the affected person was female.

Biological behaviour

They are slow-growing, and present as subcutaneous or dermal nodules [3] however, Sungur et al., reported a benign case where rapid growth, ulceration, and necrosis was evident at the tumour site [5]. The malignant form occurs predominantly in females, has no age-related predilection, and is observed more commonly on the extremities [4, 6, 7].

Excessive amounts of mucoid matrix and poorly differentiated chondroid components are important indicators of the tumour's malignancy and metastatic potential [8].

Varela-Duran et al., determined that the chondroid area of the tumour shows ultrastructural features identical to myoepithelial cells, a component of the outer layer of cells. They suggest that the chondroid regions are pseudo cartilaginous areas that reflect the pluripotentiality of the neoplastic myoepithelial cell [9]. However, subsequent immunohistochemical evaluation of the stroma suggested that myoepithelial cells may not play a major role in the formation of stromal cells [10]. Other studies concluded that the cartilaginous matrix is true cartilage produced by ultra-structurally typical chondrocytes and not pseudo cartilage produced by myoepithelial cells [11]. Mills suggests that mixed tumours are monoclonal neoplasms with replicating cells that have the ability to differentiate toward epithelium or mesenchyme, and may account for the histologic variability of mixed tumours of the skin [11].

Treatment

The optimal treatment of benign chondroid syringomas is surgical excision. Because of the lobulated nature of the tumour, it is important to include a margin of normal tissue with the excision of complete removal of the tumour. Otherwise, local recurrence may occur.

Note: Advised a close follow-up as (i) tumour site is in the extremity; (ii) the patient is female; (iii) it contains large amount of myxoid matrix; and (iv) tumour was very close to surgical margin.

CONCLUSION

We presented this case because chondroid syringoma is a rare benign skin adnexal tumour. The lesion was located on extremities, right lateral aspect of leg and the affected person is female, large amount of myxoid matrix, and the tumour was very close to surgical margin.

We gave advice to surgeons to follow-up the case.

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