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CASE STUDY

CHONDROID SYRINGOMA OF THE LEG: A RARE MIXED APPENDAGEAL TUMOR

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ABSTRACT: Chondroid syringoma is an exceedingly uncommon benign mixed skin tumor. The reported incidence is very low accounting for 0.01%-0.098%. This tumor is histologically analogous to pleomorphic adenoma occurring in the salivary gland. It has predilection for head and neck region of the middle-aged men. It usually presents as a slow growing, painless intradermal or subcutaneous nodule. Herein, we present an unusual case of chondroid syringoma in a 48 years old female in a relatively uncommon location. She presented with a slowly growing, painless nodule on the lower lateral aspect of right leg and a diagnosis of chondroid syringoma was rendered based on the histopathological examination of the excised nodular mass. This case describes a rare benign appendageal tumor of skin at an atypical location and also highlights the importance of histopathological examination in arriving at the final diagnosis.

KEYWORD: Chondroid syringoma, Appendageal tumour, Mixed skin tumor, Chondromyxoid, Leg, Pleomorphic adenoma of skin

INTRODUCTION :

Chondroid syringoma is an uncommon benign mixed skin appendageal tumor arising from the sweat glands. This tumor is histologically analogous to pleomorphic adenoma occurring in the salivary gland [1,2]. There are many morphological evidences to believe that they are derived from both ductal and acinar components [3]. The reported incidence of chondroid syringoma is low, ranging between 0.01% to 0.098% of all the primary skin tumors with a male to female ratio of 3-5:1 [4,5]. It exists in two forms i.e. benign and malignant. The benign type has a predilection for the head and neck region of the middle-aged men whereas, the malignant type is more commonly encountered at the extremities of the females and is characterized by rapid growth, local invasion, and distant metastasis [6,7]. We present here, an unusual case of chondroid syringoma in a 48 years old female in a relatively uncommon location, that is, lower lateral aspect of right leg. Our case report emphasizes on the pivotal role played by histopathological examination in rendering us with the final and accurate diagnosis.

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CASE REPORT:

A 48 years old female presented with a slowly-growing, painless nodule on the lower lateral aspect of right leg for 1 year. On local examination, the nodule was approximately 1.5x1cms in size and mobile. The overlying skin was normal but adherent to the nodule. However, the nodule was not fixed to the underlying structures. X-ray of the right leg showed a well-defined soft tissue mass in the lower lateral aspect of right leg without any bony involvement. All the haematological and biochemical investigations were normal. Based on the clinical history and examination, a provisional diagnosis of benign soft tissue tumour was made.

The nodule was excised along with a margin of the surrounding normal tissue and was sent for histopathological examination to the department of pathology. On gross examination, the nodular mass measured around 1.5x1.5x1 cms and was partly skin covered. The overlying skin appeared unremarkable. External surface of the mass was nodular, well-circumscribed, grey-white and firm in consistency. Cut surface was grey-white and homogenous with tiny focal cystic spaces filled with mucoid material **(Figure 1).**

Microscopic examination revealed skin tissue lined by stratified squamous epithelium, beneath which was seen a well-circumscribed but encapsulated dermal tumour. The tumour was biphasic and showed both epithelial and stromal components. Epithelial elements were seen in the form of elongated branching tubulo-alveolar structures lined by two cell layers- an inner cuboidal layer and an outer flattened cell layer. Also seen were numerous nests and islands of epithelial cells and ductal structures. The stroma showed abundant basophilic chondromyxoid, mucoid and focal hyaline material **(Figure 2A, 2B, 3A, 3B)**. Focal areas of haemorrhage were also noted. No cellular pleomorphism was noted. These histopathological findings were suggestive of a final diagnosis of benign chondroid syringoma.

The patient was asked to regularly follow-up every six months to look for local recurrence and features of malignancy.

Figure 1: Gross picture of the excised nodular mass

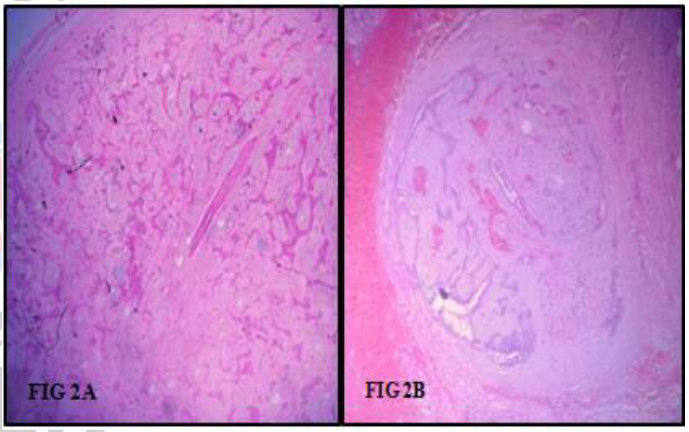

Figure 2A and 2B: Histopathology showing well-circumscribed tumor composed of both epithelial and stromal components. Compressed tubular structures are seen in a background of basophilic chondromyxoid stroma (Hematoxylin and Eosin, 40X)

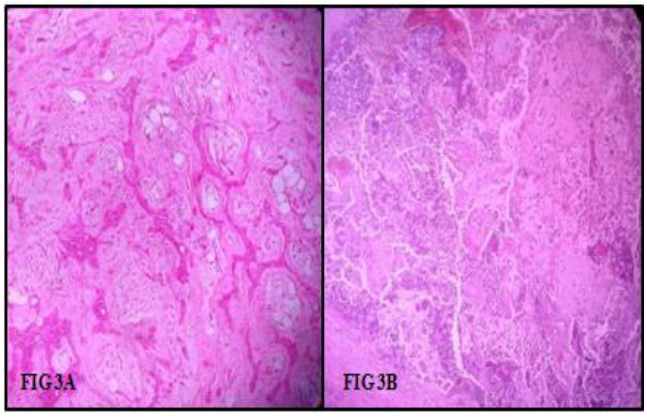

Figure 3A and 3B: Histopathology showing nests of epithelial cells and few tubular structures in a chondromyxoid stroma (Hematoxylin and Eosin, 100X)

DISCUSSION:

The term 'chondroid syringoma' was first introduced by Hirsch and Helwig in 1961 to describe this neoplasm because of the presence of sweat gland elements set in a cartilaginous stroma [8]. The term, mixed tumor, was coined by Virchow and Minssen for tumors with microscopic features that indicate both epithelial and mesenchymal origin. The reported incidence of chondroid syringoma is low, ranging between 0.01% to 0.098% of all the primary skin tumors with a male to female ratio of 3-5:1. Chondroid syringoma usually has a predilection for the head and neck region affecting nose, cheek, upper lip, eyelid, scalp, forehead except for rare cases involving the trunk, back, hand, foot, abdomen, pelvis, vulva, and scrotum [4,5,9,10]. The malignant type is more commonly encountered at the extremities of the females and is characterized by rapid growth, local invasion, and distant metastasis [6,7].

Table 1 shows a comparison of the age, sex, location and duration of tumor in our case with that of other case reports.

Table 1: Comparison table showing the age, sex, location and duration of tumor in our case with that of others.

SL. NO	STUDY	AGE (in years) / SEX	LOCATION OF THE TUMOR	DURATION
1.	Abi-Falah R et al. [11]	51 / Male	Right parietal scalp	1 year
2.	Shilpa K et al. [12]	48/ Male	Right upper lip	1 year
3.	Kumar V et al. [13]	35/ Male	Right medial canthus	6 months
4.	Shidlingappa S et al. [14]	34/ Male	Philtral dimple	6 months
5.	JY Hong et al. [15]	54/ Female	Right lower leg	5 years
6.	Sharma S et al. [16]	40/ Female	Left foot	6 months
7.	Madi K et al. [17]	47/ Female	Left foot	20 years
8.	Present case report	48/ Female	Right lower leg	1 year

The tumor usually presents as a slowly-growing, painless, solitary, solid, non-ulcerating, well-circumscribed subcutaneous/ intradermal nodule ranging in size from 0.5-3.0 cm [7]. In our case too, the patient presented with a slow-growing, painless nodule over the right lower lateral aspect of leg measuring approximately 1.5x1 cms in size. Clinically, the tumor may be confused with an epidermal cyst, pillar cyst, calcifying epithelioma, or a solitary trichoepithelioma [12,14]. Therefore, histopathological examination is mandatory for arriving at an accurate diagnosis of the tumor and also for an optimal management.

Five histological criteria were described by Hirsch and Helwig for the diagnosis of chondroid syringoma: (1) Nests of cuboidal or polygonal cells; (2) interconnected tubuloalveolar structures lined by two or more rows of cuboidal epithelial cells; (3) ductal structures lined by one or two rows of cuboidal cells; (4) occasional keratinous cysts; and (5) a matrix of mixed chondroid and myxoid material. Chondroid syringoma may exhibit all five characteristics or manifest only some, with the most common feature being the nests of cuboidal or polygonal cells [11,14]. In our case, microscopic examination of the excised nodular mass revealed a biphasic intradermal tumor showing both epithelial and stromal components. Epithelial elements were seen in the form of elongated branching tubulo-alveolar structures lined by two cell layers. Numerous nests of epithelial cells and ductal structures were noted. The stroma showed abundant basophilic chondromyxoid, mucoid and focal hyaline material. A final diagnosis of benign chondroid syringoma was conferred upon.

The treatment of choice for chondroid syringoma is surgical excision. An inadequate resection of the tumor can lead to its local recurrence. One of the major post-operative concerns of chondroid syringoma is the potential risk of malignant transformation. Therefore, excision techniques which allow pathological evaluation of the entire specimen should be always considered as the treatment of choice [18]. Periodic follow-up is important to look for local recurrences and also to determine the malignant potential.

Our case report is unique because of the uncommon location of this benign tumor on the leg and that too in a female patient. Chondroid syringomas are common in the head and neck region of the middle-aged males and the malignant type are more common at the extremities of the females. Very few cases of benign chondroid syringoma arising from the leg have been documented in the literature till date. Our case report will definitely add on to the existing literature.

CONCLUSION:

Chondroid syringoma of the leg is a rare benign skin appendageal tumor about which the pathologists and clinicians should be aware of while dealing with the lesions on leg. Histopathological examination is mandatory for arriving at an accurate diagnosis of the tumor and also for deciding the treatment modality. Histopathology is still considered as a cornerstone tool in the diagnosis of chondroid syringoma. Regular follow-up of the patient is mandatory to look for local recurrences and the potential risk of malignant transformation.

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