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Original Research Paper

Ophthalomology

CHONDROID SYRINGOMA :A RARE UPPER EYELID TUMOR

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ABSTRACT Chondroid syringoma is a benign adnexal mixed tumor of skin characterized by sweat gland elements in a cartilaginous stroma1. It is a tumor found to be arising from the sweat glands which can be apocrine (found throughout the surface of lid) or eccrine (glands of moll) with a mean age of 50 years2. Chondroid syringoma is a rare benign tumour that accounts for 0.01% of all primary skin tumours3 and occurs rarely in the upper eyelid region. Head and neck region involvement is common (nose is the most common site).It is morphologically similar to pleomorphic adenoma or benign mixed tumour of the salivary gland4. A 18-year-old male patient presented to us with a slow growing, painless mass on right upper eyelid since 6 months. Complete surgical excision of the mass was done under local anaesthesia. This report aims to discuss about this rare benign tumor of the eyelid and to study the histopathological aspects of it.

KEYWORDS : Chondroid Syringoma,benign, eyelid mass, mixed tumor,rare

Case Report

18 year old male patient presented with a slow growing, painless swelling since 6 months, 3 mm above medial canthus which was insidious in onset,gradually progressive in nature.Swelling was not associated with fever, blurring of vision or eyeache.

There was no associated change in surface or consistency of swelling and no history of any similar swelling in body.There was no history of loss of body weight .There was also no relevant family history available.Rest of the ocular examination were within normal limits and systemic examination was normal.

The mass was situated on the right upper eye lid, measuring 12mm X 8mm(Refer to figure A and B) .Firm to hard in consistency, non-tender, not fixed to underlying tissue and overlying skin,and was having a smooth surface. The skin over the swelling was freely mobile.

All the haematological investigations were within normal limits.Xray orbit was performed (refer to figure C) which revealed no bony involvement.AfterXray and clinical evaluation it was found out that there was no deeper structure involvement so we avoided getting MRI or CT scan done.

Complete excision of mass was done(refer to figure D) under local anaesthesia and sent for histopathological examination, which revealed a circumscribed partly encapsulated tumour exhibiting histological heterogenecity in form of varied patterns of epithelial elements admixed with chondroid,osseous, hyalinized and myxoid stromal areas.

Epithelial areas show tubular,glandular,nesting patterns and keratin filled multiple horn cysts of varying size.At places sheet of myoepithelial cells also noted.There was no evidence of mitotic figures or pleomorphism.

Finally the histopathology report of excised lesion showed features consistent with chondroid syringoma (refer to figure E and F) On post operative day 1, there were no signs of any swelling or inflammation(refer to figure G).

DISCUSSION

Chondroid syringoma is exceptionally rare in the periorbital region, particularly in the upper eyelid.This tumor accounts for only 0.01% of all primary skin tumors and occur only rarely in eyelids.Till now **27 cases of chondroid syringoma** of ocular adnexa have been reported so far[3,5]

Chondroid syringoma is a benign , non- ulcerated and nodular tumor that occur largely on the face, head and neck but also on the extremities and trunk[5] . Histologically, histochemically and ultrastructuraly, their appearance can be very well comparable to that of mixed tumors of salivary gland origin[7-9] . This includes the presence of cells with an abundant hyaline cytoplasm[10,11].Sometimes eosinophilic globules having radiating fibrillary structures are found within and around the lumina. Their appearance is quite identical to that seen in collagenous spherulosis of the breast[12,13]. Immuno-histochemically the cell layers show a variability in staining for myoepithelial markers, such as vimentin , S-100 protein , actin , calponin, p63 and glial fibrillary acidic protein[14-17] .Even though most of these tumors are considered to be eccrine type,a variety of them exhibit clear evidence of apocrine differentiation . This is often admixed with the follicular and sebaceous components[18,19]. Regardless of the occasionally atypical appearance of the cartilaginous components, the vast majority of these tumors are benign.

Tumors with an appearance similar to cutaneous chondroid syringomas and myoepitheliomas can also arise in the deep soft tissue[20].The exact cause of chondroid syringoma is not clear, but it is classified into apocrine and eccrine types.Most recently they have been defined as hamartomas.

The **differential diagnosis** of chondroidsyringoma may include neurofibroma, dermoid cyst, epidermoid cyst, retention cyst, papilloma,lipoma, sebaceous cysts, dermatofibroma, salivary gland pleomorphic adenoma and basal cell carcinoma[21,2]

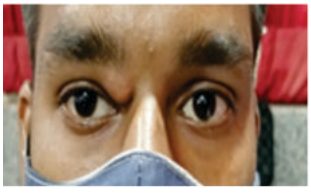
Diagnosis is confirmed with **biopsy** of the mass.The treatment is done by complete local surgical excision of the tumour along with its capsule.Additional treatment measures include electro dessication and CO_2 laser. But with these procedures the recurrence rate is much higher as compared to the surgical excision of the mass[4]

CONCLUSION

Chondroid syringoma is a rare benign mixed tumor of the exocrine glands and its incidence of occurring on the eyelid is a rare finding. Many times it goes unnoticed due to its rare occurence and its non-specific clinical presentation. These tumours are mostly benign but if the capsule is ruptured, they may become locally invasive and malignant transformation is also possible[1] The size of the mass is found to be somewhere between 0.5 cm and 3.0 cm.The risk of malignancy increases in a tumor greater than 3.0 cm in size[23] .Histopathological examination is essential to confirm the diagnosis and identify any signs of malignancy .Complete surgical excision along with its capsule is the treatment of choice[22].

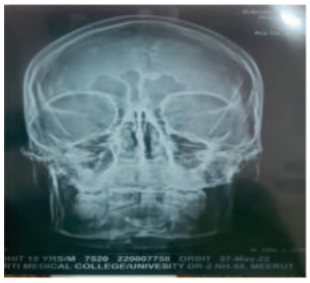
Chondroid syringoma should always be suspected in case of any small subcutaneous nodule in the head , neck or periorbital region particularly in the middle-aged patients.[24]

So it can be concluded that as pre-operatively, we cant differentiate between dermoid cyst, epidermoid cyst ,retention cyst, papilloma, lipoma, sebaceous cysts, salivary gland pleomorphic adenoma, or basal cell carcinoma from chondroid syringoma, so it is thereby recommended to excise the tumor completely along with its capsule. If complete excision is not performed, it can lead to malignant transformation.



A

B



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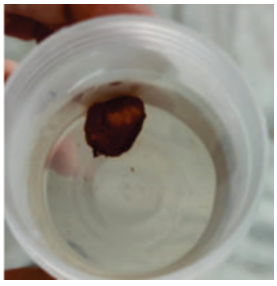

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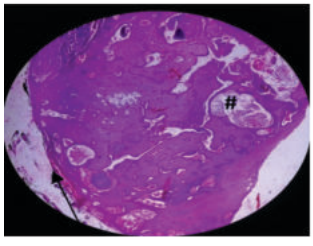

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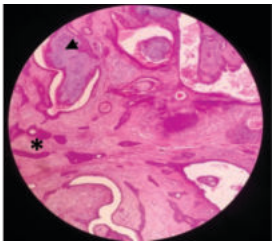

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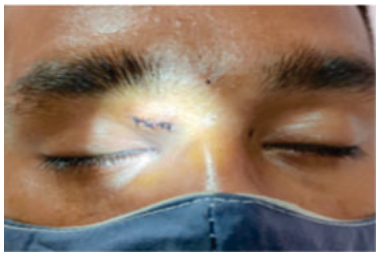

G

DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has given his consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts Of Interest

There are no conflicts of interest.

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