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Ophthalmology

PLEOMORPHIC ADENOMA OF LACRIMAL GLAND WITH INTRACONAL EXTENSION – A RARE CASE REPORT

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ABSTRACT Tumours of lacrimal Gland are rare in clinical practice and the most common epithelial tumour is Pleomorphic Adenoma. Here we are presenting a case of ploemorphic adenoma of lacrimal gland with intraconal extension, which is a rare occurrence. A middle aged female presented with painless eccentric proptosis of left eye. Contrast Enhanced MRI of orbit showed multilobulated enhancing intraconal mass in left orbit. Mass was surgically removed by anterior orbital approach. Histopathological examination revealed Pleomorphic Adenoma. Although intraconal extension of Pleomorphic Adenoma of Lacrimal Gland is rare, it should be considered as one of the differential diagnosis for any intaconal mass presenting with proptosis and dystopia.

KEYWORDS : Pleomorphic Adenoma, Lacrimal Gland, Intraconal Extension, Dystopia

INTRODUCTION

Lacrimal gland tumour are rare, representing only 10% of orbital space occupying lesion.[1] Epithelial tumors are the most common tumours of Lacrimal gland, consisting approximately 50-60% of all benign tumours and 40-50% of malignant tumours.[2] Pleomorphic adenoma is the most common epithelial tumour of lacrimal gland.[3], representing 3-5% of all orbital tumours, 25% of lacrimal mass lesions, and 50% of epithelial lacrimal gland tumour.[4] Pleomorphic adenoma is a neoplastic proliferation of epithelial cells that form characteristic ductal structures with surrounding myoepithelial cell. These structures gradually trail out into myxomatous mesenchyme of the lacrimal gland. [1] Pleomorphic Adenoma of Lacrimal gland does not seem to have a male or female preponderance. Because of the rarity of this tumor, the risk factors have not been elucidated.[1] The exact cause of PALG is unknown. Genetic rearrangements, such as chromosomal translocations involving 8q12 target the PLAG1 gene, may contribute to the pathophysiology of PALG.[1] Alterations in this developmentally regulated zinc finger gene can lead to deregulation of target genes, including IGF2, and is likely to play a major role in the genesis of pleomorphic adenomas.[1] The orbital lobe of the gland is most commonly involved. After developing from the orbital lobe the benign mixed tumour tends to extend farther back into the orbit and become quite large. Pleomorphic Adenoma of lacrimal gland most commonly occur in the second to fifth decades of life, and usually present with painless proptosis, downward displacement of the globe, diplopia, fullness of upper eyelid, retinal striae and a palpable eyelid mass.[4]

CASE REPORT

A 45 year old Indian female home maker by profession presented with a complain of bulging of left eye for the past 1 year which was gradually progressive and not associated with pain and diminision of vision with visual acuity of 6/18(OD) & 6/12(OS). On examination, all the findings were within normal limit except for a mild proptosis of 3mm more than the contralateral eye with significant dystopia (downward) [Fig 1(A)] and a Smooth, firm, non-tender mass of 1.5x1 cms size in lacrimal gland fossa.

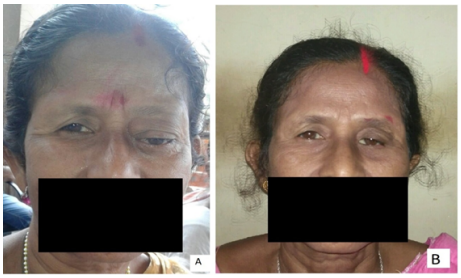

Fig. 1 A- Pre-operative Picture B- Post-Operative Picture(3 month)

Contrast Enhanced MRI finding revealed multilobulated enhancing intraconal lesion in left orbit pushing the globe forward and downward suggestive of neoplastic etiology of left lacrimal gland.[Fig 2] All other routine pre-operative investigations were done and was within normal limit as the case was planned for surgical removal of the tumour.

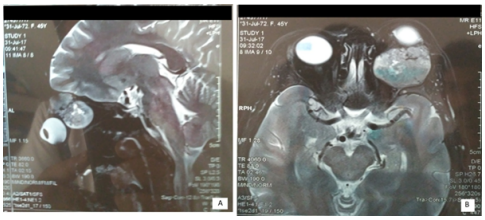

Fig. 2 Contrast enhanced MRI Image of the lesion; A- Saggital , B- Transverse

The Orbital mass was surgically removed by anterior orbitotomy with lateral extension under general anaesthesia by combined afford of Ophthalmology and Head & Neck Surgery Department. The mass was excised in piecemeal as it could not be excised en masse and sent for Histopathological examination.[Fig 3]

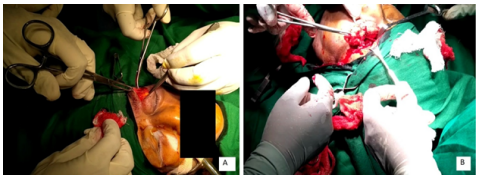

Fig. 3 Intra-operative picture showing A- Orbitotomy, B- Removal of tumour mass

On Histopathological examination, there was a multiple irregular tissues of aggregate sized 7x5x1.5cm and the histopathological slide revealed multiple myoepithelial cells with a tubular gland like structures in a myxoid background giving a diagnosis of Pleomorphic Adenoma of lacrimal gland. [Fig 4]

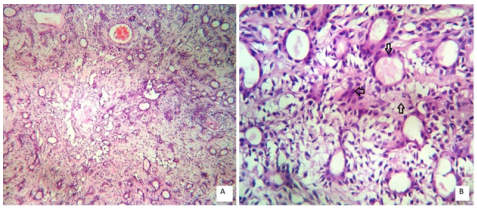

Fig.4 Histopathological picture

A- Low magnification;

B- High magnification Down arrow-tubular gland like structure, Up arrow- mixoid background

Side arrow- myoepithelial cells

In the immediate Post-op and 1 month post-op period, proptosis reduced significantly but ptosis was present. On 3 month post-op follow-up, ptosis reduced significantly [Fig 1(B)] but defect in the superior rectus function was noticed. Unaided vision remained unchanged with 6/18(OD) & 6/12(OS). Best Corrected visual acuity is 6/6 (OU).

DISCUSSION

Pleomorphic adenoma of lacrimal gland is usually extraconal mass but in this case it was found to be intraconal extension.

The excision of pleomorphic adenoma of lacrimal gland is recommended to be en masse but in this case it was not possible to remove en masse and had to be excised in piecemeal which have a significant rate of recurrence or malignant transformation and thus this patient have to be followed up regularly.[3]

In this case, since the major mass was intraconal, there was a high chance of optic nerve compression leading to optic atrophy followed by decrease in vision. But in our case the timing of surgery was early enough to prevent the same.

In this case since the mass was extensive, there was significant manipulation of the tissues which lead to haemorrhage of levator muscle resulting in ptosis lasting for few months. [5]

As from the MRI we can see that the intraconal mass is stretching the superior rectus chronically due to its shape and position, it may take 1-3 months to regain the correct tension. This may have resulted in defect of superior rectus function on 3 month follow-up.[6]

However in long term follow-up, if the rectus function is not regained, intervention maybe required.[4]

Pleomorphic Adenoma cases have also been reported by GE Rose et al [7] ; S Tonya Stefuo, Cathy DiBernards et al [8]; S Fenton, SieGo [9].

CONCLUSION

Pleomorphic Adenoma of lacrimal Gland must be considered in the presence of long term painless proptosis with dystopia. Patients suspected to have pleomorphic adenoma, prior biopsy should not be performed in order to prevent tumour seeding into adjacent orbital tissue. Pleomorphic adenoma have a good prognosis provided excision is complete and without disruption of the capsule. It is crucial to have a long term follow-up to monitor recurrence or malignant change.

Declaration of patient consent

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

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Nil

Conflict of interest

There are no conflict of interest.

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