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Slow But Malignant: Adenoid Cystic Carcinoma of Lacrimal Gland- A Case Report

KEYWORDS: Adenoid cystic carcinoma, lateral orbitotomy, proptosis,

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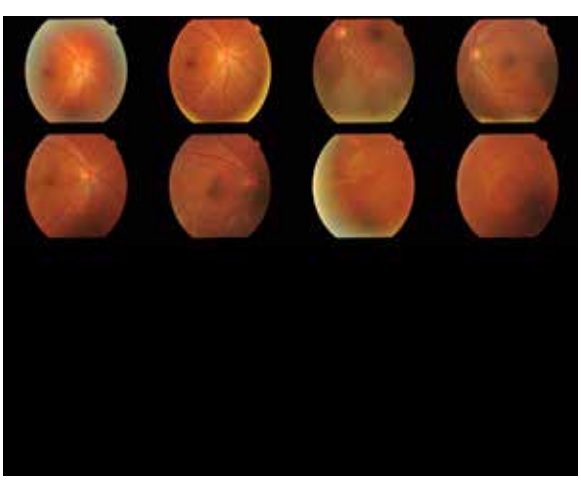
ABSTRACT *Introduction- Adenoid cystic carcinoma is a epithelial malignancy of lacrimal gland which presents with painful non-axial proptosis and associated with bone destruction. It occurs mostly in 4-6th decade of life with female preponderance.Purpose: To analyze the clinical profile of a painless non-axial proptosis & document a clinico-pathological picture.Material and methods: We present a case of 3rd decade young adult female who presented with gradually increasing painless abaxial proptosis of RE since two and half years. Her MRI- findings revealed soft tissue mass in right orbital fossa with no perineural invasion suggesting benign soft tissue mass. She underwent Lateral orbitotomy and tumor debulking procedure followed by post-operative radiotherapy. Histopathological analysis revealed basaloid cells with cribriform pattern which is hallmark of ACC.Conclusion- Though the presentation and imaging studies favour benign pathology, this case shows the importance of histopathological analysis which helped us in confirming the diagnosis of ACC.*

INTRODUCTION-

Adenoid cystic carcinoma (ACC) of the lacrimal gland is one of the malignant neoplasm found in orbit. It is also described as 'Cylindroma' by Billroth in 1859. It represents about 1.6% of all orbital tumors and 25-30% of all epithelial lacrimal gland tumors.[1][2] The peak incidence is in 4 th to 6th decade (average age of 50 years at diagnosis) with slightly higher preponderance noted in females. Often the initial presenting sign is abaxial (nonaxial) proptosis with inferior and nasal deviation of globe.ACC may cause pain because of perineural invasion [3] and bony infiltration by tumor. It is characterized by rapid course with a history of less than 1 year and early onset of pain which helps us to differentiate this malignant tumour from benign mixed tumor, which presents with progressive proptosis for more than a year and is painless.We document a case of young adult female with Adenoid cystic carcinoma who represented with painless unilateral non-axial proptosis.



CASE REPORT-

A 32 year old female presented in the Ophthalmology Department in M.G.M Medical college & MYH hospital Indore, India with slowly progressive bulging of right eye (Fig.1) since two and half year and painless progressive loss of vision in right eye since one year.There is no history of pain, trauma, and headache. The patient has no complaints of nasal discharge or nasal obstruction which suggested no extension through nasolacrimal duct to nose. There was no significant past, family and personal history. Clinically she was well oriented to time, place and person. Ophthalmic evaluation of the patient revealed visual acuity of 6/18 with pinhole 6/12 and abaxial proptosis (on Luedde's exopthalmometry 23mm axial and 5mm vertical displacement), ocular movements restricted in all cardinal gazes and pupil showed RAPD in the right eye. The fundus examination of right eye was normal (Fig 2).

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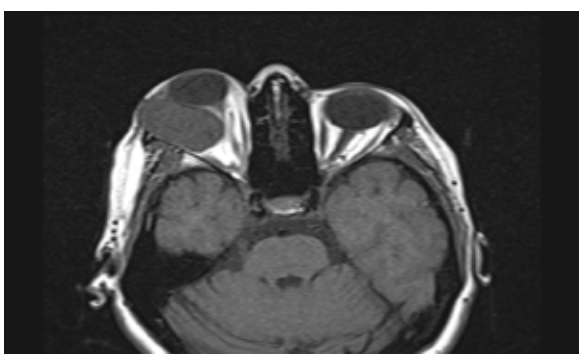
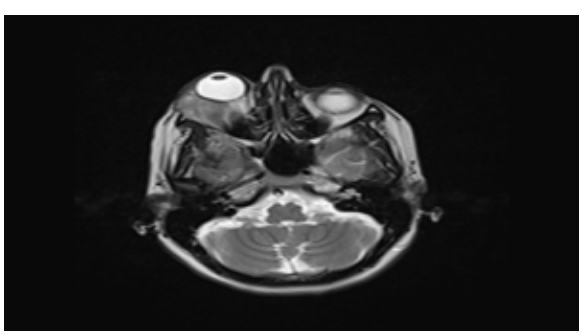
INVESTIGATION

Routine blood, thyroid profile and urine investigations were in within normal limits

MRI Brain and Orbit revealed 4×3×3 cms sized smoothly marginated soft tissue lesion in the supero-lateral right orbital fossa, extraconal in origin and causing extrinsic indentation on the right globe which appears displaced inferiorly and infero-medially (Fig 3).The lesion appears isointense on T1 and heterogeneously hyperintense on T2 (Fig 4). Normal lacrimal gland is not visualized. No intrinsic signal abnormality seen in optic nerve.The image morphology was more in favour of lacrimal gland origin most likely benign soft tissue mass.



T-1 SAGITAL AXIAL (Fig. 3)



T-2 SAGITAL AXIAL (Fig. 4)

TREATMENT AND FOLLOW UP

Patient was referred to neurosurgeon and surgery was done under general anesthesia with informed consent. She underwent tumor debulking procedure through lateral orbitotomy approach. Mass was sent for histological examination. Histological examination revealed Tumour cells consisting of basaloid epithelial cells arranged in cribriform pattern is characteristic of Adenoid cystic carcinoma.(Figure-5)

HISTOPATHOLOGICAL PICTURE-

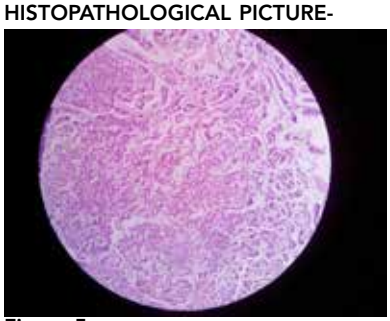


Figure-5

Discussion-

Adenoid cystic carcinoma of lacrimal gland is malignant neoplasm & has slightly more preponderance in females, grows within an year and is known for recurring even after therapy. It is second most frequently occurring lacrimal gland tumor after pleomorphic adenoma. Despite their rarity, ACC should be kept in the differentials of abaxial proptosis when patient presents with proptosis over a period of 1 year, accompanied by pain, diplopia and restricted ocular movements. Orbital pain as a result of perineural spread is commonly seen in 10-40%cases. Orbital imaging, MRI Scan is important to establish the tumor extension , to plan treatment and to allow radiologic and clinical follow up.The histopathologic appearance of ACC is characteristic and consists of sheets of epithelial cells arranged in either solid or cribriform patterns that mimic a glandular structure. Five histopathologic subtypes have been identified: Cribriform, sclerosing, basaloid, comedocarcinoma, and tubular (ductal). Lower tumor grades are associated with a predominantly cribriform pattern,and these patients have longer survival rates.[4].The treatment of ACC is controversial. Some practitioners advocate a globe-sparing approach[5] with local excision of the orbital mass followed by supplemental external beam radiation therapy or brachytherapy.[6]Local recurrence is common, occurring in nearly half of patients within two years,with soft tissues or orbital bone as the most frequent sites. The tumor often infiltrates and spreads through bone[7][8]. Planned combinations of surgery with preoperative or more commonly postoperative radiotherapy have shown improvement in both local control and survival.

Conclusion-

Despite its painful nature & older age of presentation, we report a case of Painless Adenoid cystic Carcinoma in young adult female who was managed promptly. Such cases should be kept in the differentials of abaxial proptosis when patient presents with proptosis over a period of 1 year duration. Though the presentation and imaging studies favours benign pathology, this case shows the importance of histopathological examination which helped us in confirming the diagnosis of ACC.

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