LACRIMAL FOSSA MASS - MANAGEMENT DILEMMA

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INTRODUCTION

Lacrimal fossa mass poses an interesting diagnostic challenge for the ophthalmologist. It usually presents with lid edema, ptosis and globe displacement. Clinician usually expects inflammatory, lymphoproliferative and neoplastic lesions. Patient's age and durations of symptoms are important considerations. Lymphoproliferative/malignant lacrimal lesions are common in fifth and sixth decades. Pain occurs early in inflammations and late in malignancies[1]. Orbital echography and CT (computerized tomography) are necessary to know the details. On CT, dacryoadenitis tends to show the gland to be enlarged in an oblong configuration. Neoplastic processes are often more nodular and rounded. Definite diagnosis is given by histopathology in majority of the cases but sometimes it remains doubtful. Role of infectious agents[2] and genetic abnormalities[3] in the aetiology of ocular adnexal neoplasia are being investigated.

CASE REPORT

A 65 year old male presented with severe pain in left orbital area of 3 months duration. There was no history of trauma, nasal problem, malignancy or hospitalization. He had abnormal head posture (Fig.1, left) with chin elevation and head tilt to right side. Inferior dystopia and ptosis became obvious when chin and head were straightened (Fig.1, Middle). There was proptosis (Fig. 2), lid swelling, tenderness and moderate restriction of elevation of the left eye. Right eye was normal. There was no lymphadenopathy.

Blood CP, total and differential leukocyte count, Liver function tests, X ray chest were within normal limits. Ultrasound B scan of left eye revealed an ovoid mass 17*17 mm below the upper eyelid. Ultrasound abdomen did not reveal any abnormality. CT scan (Fig. 3) revealed a concavo convex extraconal soft tissue density mass, almost homogenous, in the left orbit supero laterally. No localized bone erosion was seen.

Patient was given Augmentin 1.2 G 12 hourly and Prednisolone 60 mg orally daily for 4 weeks. It resulted in moderate improvement of signs and symptoms (Fig 1 Right). Lateral orbitotomy (Fig. 4) was performed and the dissection was extended superiorly to excise the whole mass.

First histopathological examination revealed sheets and starnds of atypical lymphoid cells having hyperchromatic, irregular nuclei and scant cytoplasm with scarce mitoses suggesting "atypical lymphoid infiltrate of lacrimal gland". Review of slides at tertiary referral center gave diagnosis of poorly diffrentiated adenocarcinoma of lacrimal gland. On immunohistochemistry it was positive for CKAE1/AE3. Bone scan, liver-spleen scan did not reveal any metastasis. Patient was given radiotherapy and chemotherapy. Six months follow up has not revealed any recurrence.

DISCUSSION

This case posed an interesting clinical puzzle. Severe pain, proptosis, periorbital edema leading to ptosis, reduced extraocular motility and favourable response to antibiotics and steroids were suggestive of inflammatory process. However persistence of large mass after medical therapy demanded surgical exploration and histopathological examination. Histopathological report added to the confusion as initial diagnosis was 'atypical lymphoid infiltrate of lacrimal gland'. Oblong mass conforming to adjascent structures is usually due to lymphoproliferative disorders. It is known that most are non Hodgkin's lymphomas (NHL)[4] but multiple types and rarity of many types have made the classification and treatment of orbital lymphoma difficult[5]. Intraorbital NHL can resemble pseudotumour or other tumours; differentiation is often difficult on radiological criteria. This patient had no systemic

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involvement though systemic disease has been reported in 40% of atypical hyperplasias with 19% mortality rates at 5 years[6]. Later histopathological diagnosis was 'poorly differentiated adenocarcinoma of lacrimal gland'. Immunohistochemistry was suggestive of epithelial neoplasia. Clinical points in favour of this diagnosis were his age (rare for primary or pleomorphic adenocarcinomas to occur before 30 years of age)[7,8], short history and pain. Pain has been found to be less often and later with adenocarcinoma (12% of malignant tumors of the lacrimal gland)[7,9] than with adenoid cystic carcinoma. Points against carcinoma were absence of calcification, bone erosion and no recurrence after six months of follow up. Calcification has been found in most of the cases[7], erosion of bone in all[7] and recurrences very common[7,8,10] in primary adenocarcinoma.

Contrasting clinical features and uncommon occurrence of lacrimal fossa masses, make such cases a clinical challenge.

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Fig 1.

Left. Compensatory chin elevation and head tilt to right side

Middle. Inferior dystopia and ptosis becoming prominent with chin/head straightened

Right. Improvement in dystopia after 4 weeks course of antibiotics and steroids.

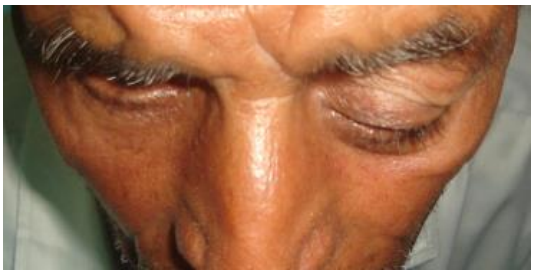

Fig 2.Left proptosis

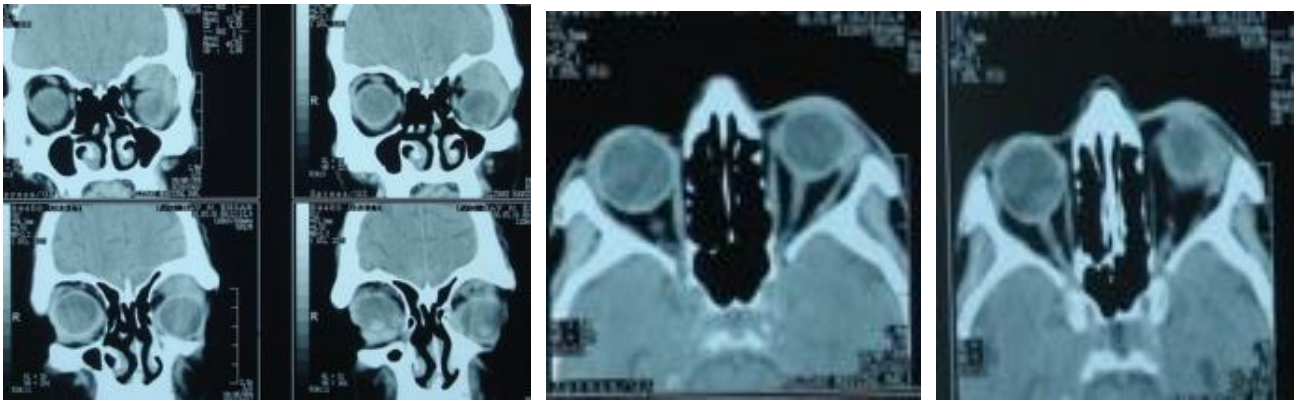

Fig-3: CT scan Left: Coronal view, Right: Axial view. Concavo convex soft tissue density mass, almost homogenous was noted in the left orbit supero laterally.

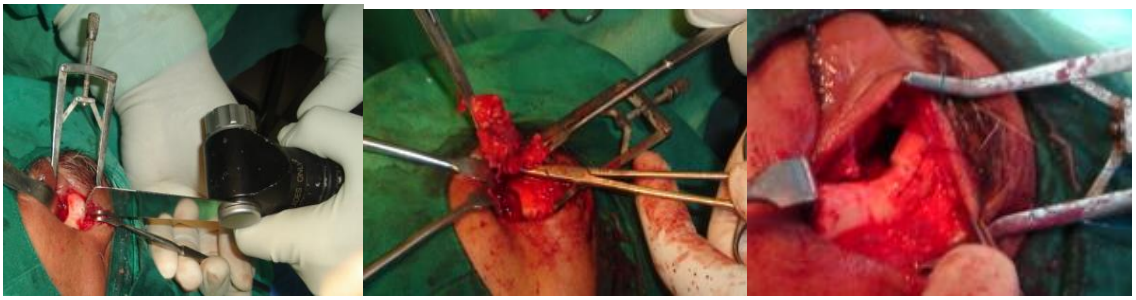

Fig-4: Left: Lateral Orbitotomy, Middle: Excision of the mass, Right: Bone sutured into place