DESCRIPTIVE STUDY ON LACRIMAL GLAND LESIONS IN A TERTIARY EYE CARE CENTRE

DISSERTATION SUBMITTED FOR

M.S Degree (Branch III) Ophthalmology

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CERTIFICATE

Certified that this dissertation entitled “DESCRIPTIVE STUDY ON LACRIMAL GLAND LESIONS IN A TERTIARY EYE CARE CENTRE” submitted for Master of Surgery (Branch III) Ophthalmology to the Tamil Nadu Dr.M.G.R. Medical University, Chennai is the bonafide work done by Dr. R.VIJAY, under our supervision and guidance in the Department of Orbit and Oculoplasty at Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai from May 2012 to April 2015.

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I, Dr. R.VIJAY solemnly declare that the dissertation titled “DESCRIPTIVE STUDY ON LACRIMAL GLAND LESIONS IN A TERTIARY EYE CARE CENTRE” has been prepared by me. I also declare that this bonafide work or a part of this work was not submitted by me or any other for any award, degree, diploma to any other university board either in India or abroad.

This dissertation is submitted to the Tamil Nadu Dr.M.G.R Medical University, Chennai in partial fulfillment of the rules and regulation for the award of M.S. Ophthalmology (Branch III) to be held in April 2015.

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INTRODUCTION

There has been an increasing interest among ophthalmologists to study the management algorithms of lacrimal gland lesions.

Unlike other ophthalmic conditions, lacrimal gland lesions have difficulties in clinical examination, diagnosis and surgical approach, as final diagnosis cannot be accurately predicted. Occurrence of these lesions can have relationship with age, gender, demographic and other socioeconomic factors. Large number of various lesions in lacrimal gland have lead to varieties in the series published by individual authors and different institutions.

The present study has been aimed to study the demographic profile, clinical features, treatment options and outcome of lacrimal gland lesions that have been reported to Aravind Eye hospital, Madurai from the year 2012 – 2014.
INCIDENCE

Mayo case series (1948 – 1997)

Histological study of 83 cases of lacrimal gland lesions. It includes 29 cases (35 percentage) of Adenoid cystic carcinoma, 26 cases (31 percentage) of benign mixed tumour, 16 cases (19 percentage) of malignant mixed tumour, 7 cases (8 percentage) of adenocarcinoma, 2 cases (2.5 percentage) of mucoepidermoid carcinoma, 2 cases (2.5 percentage) of squamous cell carcinoma and 1 case (1 percentage) of apocrine carcinoma.

Reese’s study: Clinicopathological study of 112 cases of lacrimal gland lesions. It included 50% Epithelial and 50% Non Epithelial lesions. Out of Epithelial lesions 50% were pleomorphic adenoma, 25% were adenoid cystic carcinoma and 25% were remaining other types of carcinoma. Among Non epithelial lesions 50% were lymphoid lesions and 50% were pseudotumours.

Pseudotumours and lymphoproliferative disorders are being more common. It is important to differentiate between these groups, as some of these conditions are life threatening.
Lacrimal gland lesions are experienced in various age groups. Primary tumours with rapid progression is mostly seen in children and adolescent. Lymphomas are predominantly seen in adult life. Primary and secondary tumours has equal occurrence in middle age. Secondaries are more common in old age.
ANATOMY OF LACRIMAL GLAND

Lacrimal gland is located in the lacrimal fossa, which is formed by the orbital plate of frontal bone in the anterolateral part of orbital roof. Levator aponeurosis divides the lacrimal gland into superior (orbital) and inferior (palpebral portions). Both the portions are continuous with each other posteriorly.

ORBITAL PART:

Larger than palpebral part. Two surfaces are present in this part (Superior and inferior). There are two borders (anterior and posterior). There are two aspects (Medial and Lateral).

Superior surface

It is convex in shape and related to the periorbita, which lines part of frontal bone, forming lacrimal gland fossa. Fine trabeculae attaches periorbita and superior surface of orbital part.
Inferior Surface

It is concave in shape. It is related with levator palpebrae superioris muscle and lateral horn of levator aponeurosis.

Anterior Border

It is sharp and lies parallel to orbital margin. It is related with septum orbitale.

Posterior Border

It is round in shape. It is continuous with palpebral lobe of lacrimal gland.

Lateral aspect

It lies on lateral rectus muscle.

Medial aspect

It is related with levator palpebrae superioris muscle
**PALPEBRAL PART:**

Size of orbital part is 3 times more than palpebral part. It lies over the orbital part of lacrimal ducts.

On everting upper lid, this part of the gland can be seen through conjunctiva. Palpebral part of gland is continuous with orbital part posteriorly. From the main lacrimal gland, 10-12 ducts pass down and open in lateral aspect of superior fornix. Some ducts open in lateral aspect of inferior fornix.

Excision of palpebral lobe of lacrimal gland is similar to excision of entire gland which aborts the whole secretory function of gland, since most of the ducts pass through palpebral lobe of gland only.

**HISTOPATHOLOGY:**

Lacrimal gland is a serous acinous gland, which is tubuloalveolar and the structure of lacrimal gland is similar to salivary gland.

It divided into three major parts – glandular tissue, Stroma, and Septa. Outer limit of gland is lined by capsule.
Glandular tissue is contributed by acini and ducts which are arranged in lobes. Fibrovascular septa separates this lobules from each other.

Pyrimidal cells lining the acini, are surrounded by layer of flattened myoepitheliel cells. These cells have nucleus which is round in shape and situated towards base. These cells are serous in type and have secretory granules which are eosinophilic in nature. These cells are responsible for tear secretion which is extracted by myofibril contraction.

Secretion of acinar units is first drained by intralobular part of the gland, followed by extralobular part and finally by lacrimal ducts.

Lacrimal gland stroma contributed by mesodermal tissue is formed by lymphoid tissue, blood vessels and elastic tissue.

**Blood supply**

Lacrimal Artery, a branch of ophthalmic Artery, supplies lacrimal gland mainly. Transverse facial Artery also supplies the gland additionally.
**Venous Drainage**

Lacrimal gland is drained by lacrimal veins which is further drained by ophthalmic veins.

**Lymphatic Drainage:**

Lacrimal gland is drained into preauricular lymph nodes.

**NERVE SUPPLY:**

- Sensory nerve supply from lacrimal nerve, a branch of ophthalmic division of trigeminal nerve.
- Sympathetic nerve supply from carotid plexus of the cervical sympathetic.
- Secretomotor fibres derived from superior salivary nucleus.

**Accessory Lacrimal Glands:**

1. Glands of wolfring
2. Glands of Krause
3. Infraorbital glands
Glands of Krause:

These glands lie in fornices (sub conjunctiva). In upper fornix, about 40-42 glands are present. In lower fornix about 6-8 glands are present.

Glands of Wolfring

These glands are located in superior tarsus (upper border) and also in inferior tarsus (lower border).

CLASSIFICATION OF LACRIMAL GLAND LESIONS:

1. Neoplasm:

   Benign: Pleomorphic adenoma
   Warthin’s tumour
   Oncocytoma
   Myoepithelioma

   Malignant: Adenoid cystic carcinoma
              Carcinoma in Ex pleomorphic adenoma
              Mucoepidermoid carcinoma
              Adenocarcinoma
              Undifferentiated carcinoma
2. Inflammatory:

Acute dacryoadenitis : Mumps, Influenza, Diphtheria, Infectious mononucleosis.

Chronic dacryoadenitis : Tuberculosis, Sarcoidosis, Lympho proliferative disorders, Sjogrens syndrome, Wegeners granulomatosis

3. Structural : Dacryops

CLINICAL PRESENTATION :

Lacrimal gland lesions clinically presented as 4 categories.

1. Benign tumours

2. Malignant tumours

3. Lymphoproliferative disorders

4. Inflammatory lesions
BENIGN TUMOURS:

Presented as chronic, painless, slowly progressive, non axial proptosis. Lacrimal fossa remodelling is seen radiologically.

PLEOMORPHIC ADENOMA:

It is the most common benign epithelial tumour. Typically presents in the second and fifth decade. This tumours are distributed equally between males and females. Clinically as chronic slowly progressive swelling in the upper outer quadrant causing non axial proptosis.

Histology reveals evidence of both epithelial and mesenchymal differentiation. Proliferation of benign epithelial cells are arranged in double layer to form lumen. Stromal differentiation can be seen in the formation of bone and cartilage.

Immunohistochemistry reveals epithelial element and chondroid elements staining separately.
CT scan reveals a well circumscribed, pseudoencapsulated lesion with expansion and remodeling in the lacrimal fossa without bony erosion. This tumour can undergo malignant transformation.

Treatment of choice is Excision biopsy through Lateral orbitotomy.

**WARTHIN’S TUMOUR: (CYSTADENOLYMPHOMA)**

It is a epithelial tumour of lacrimal gland which is very rare. Histology reveals epithelial columnar cells arranged in solid nests or lining the cystic spaces. The deep layer of epithelium is cuboidal and superficial layer is columnar, and cytoplasm has finer granules. It resembles oncocytic cells.

**ONCOCYTOMA:**

Rare tumour occurring secondary to metaplasia of ductal cells. Histology reveals large eosinophilic cells rich in mitochondria.

This cells are polygonal and cytoplasm has fine granules which are eosinophilic.
**MYOEPITHELIOMA:**

Rare tumour with biological behavior similar to that of a pleomorphic adenoma. It consists of 5 subtypes as spindle, plasmacytoid, epithelial, clear and mixed.

It is contributed mainly by myoepithelial cells. Spindle cells and plasmacytoid types are some variants of this tumor.

**MALIGNANT TUMOURS:**

Subacute presentation of short duration (4-6 months). Radiologically presented as infiltration of adjacent structures, calcification and irregular bony erosion.

**ADENOID CYSTIC CARCINOMA: (CYLINDROMA)**

Most common malignant epithelial tumour of lacrimal gland comprising 50 percent of malignant tumours of lacrimal gland and 25 percent of all lacrimal gland tumours. Presented with bimodal distribution which peaks in second and fourth decades. Clinically presents as proptosis of shorter duration with pain and paraesthesia.
Histology revealed ductal cells forming spaces into which basement membrane like material is deposited. This confirm a cribriform or Swiss-cheese appearance to tissues.

Five histological patterns are seen in this lesions as follows.

1. Cribriform type (most common subtype)
2. Sclerosing type
3. Basaloid type (worst prognosis)
4. Comedocarcinoma
5. Ductal / tubular type

Imaging reveals irregular mass with bony erosion (70%), and occasional calcification (20%). Features on CT imaging are larger sized tumours, with calcification and bony erosions and with extension along lateral orbital wall. MRI identifies perineural infiltration and cavernous sinus involvement. Immunohistochemistry reveals cribriform and tubular areas which stains positive for Alcian blue, S100 and CEA.
PRIMARY ADENOCARCINOMA:

Rare tumour with clinical features similar to Adenoid cystic carcinoma. Histology reveals pleomorphic and mitotically active cells arranged in sheets and cords.

Pathologically they are classified into Polymorphic, low grade adenocarcinoma, Ductal adenocarcinoma, Acinic cell adenocarcinoma and Basal cell adenocarcinoma.

CARCINOMA IN Ex. PLEOMORPHIC ADENOMA

May arise denova, as consequence of malignant transformation following incomplete excision of benign adenoma or as malignant transformation of a presumed benign adenoma. Presented as well circumscribed pseudo capsulated lesion.

Clinically lacrimal gland mass is indolent. Rapid growth indicates malignant transformation or with recurrence of previously excised lacrimal mass.
Treated with mass excision followed by radiotherapy. Systemic evolution for metastasis especially to lung is noted.

Indication of orbitectomy is orbital infiltration with bony involvement.

**MUCOEPIDERMOID CARCINOMA:**

Common neoplasm of major salivary glands which rarely affects the lacrimal gland. Histology reveals epidermoid and mucus secreting cells arranged in a pattern of cords and islands.

Clinically presented with slow or rapid growing mass in lacrimal fossa. Bony invasion can also occur rarely.

**ACINAR CELL CARCINOMA:**

Occasionally involves the lacrimal gland. Grossly represented as neoplasm of multipotential ductal cells. Histology reveals solid acinic growth pattern with microcystic spaces containing mucoid material.
SQUAMOUS CELL CARCINOMA:

It is a very rare tumour. Histology reveals pure proliferation of keratinized moderate to well differentiated squamous cells.

Orbital exenteration is recommended.

MALIGNANT MYOEPITHELIAL CARCINOMA:

Very rare low grade neoplasm with shorter duration of presentation. CT scan imaging shows well circumscribed mass with foci of calcification. Histology reveals encapsulated mass composed predominantly of spindle cells and large clear myoepithelial differentiated cells. Immunohistochemistry reveals focal positivity for smooth muscle actin, vimentin and glial fibrillary acidic protein. Clear cell myoepithelial carcinoma is managed by Orbital Exenteration.

CARCINOSARCOMA:

Very rare tumour that may arise from a pleomorphic adenoma.

SECONDARY TUMOURS:

Sinus tumours rarely invade the lacrimal gland. However basal cell carcinoma from lateral canthus and conjunctiva can invade lacrimal gland.
Sebaceous cell carcinoma from the lid can also presents with mass in the lacrimal fossa.

**LYMPHOPROLIFERATIVE DISORDERS:**

50% of orbital lymphomas arise in the lacrimal fossa. Lymphoproliferative disorders represent a group with dense cellular infiltrate that is composed predominantly of monoclonal B cells and of Non Hodgkins lymphoma type.

T cell lymphoma is rare in orbit. It includes reactive and lymphomatous lesions of Non Hodgkins lymphoma.

Ocular adnexal lymphoma constitute 6 -8 % of all lymphoma.

Occurs in 6th – 7th decade of life with a slight female predominance.

Usually occurs in the anterior orbit as fleshy pink subconjunctival tumefaction (salmon patch), tends to mould the shape of the globe.

On palpation they are nodular and rubbery in consistency with well defined margins. 35% of the patients progress to systemic lymphoma.
Histology reveals lymphoid infiltrates consisting of monomorphous, atypical population of cells. Subdivided according to R.E.A.L (Revised European American classification of Lymphoma) which includes

- Extranodal marginal lymphoma
- Diffuse large B-cell lymphoma
- Follicular centre lymphoma
- Mantle cell lymphoma
- Lympho plasmacytic lymphoma

Immunohistochemical analysis with various cytogenic markers help in precise classification and predicting prognosis.

CT or MRI reveals fairly well defined homogenous mass isodense to extramuscles. Usually involves anterior, superior or lateral orbit. They almost have an extraconal component. They tend to mould around pre-existing structures without eroding bone or expanding the orbit. Lacrimal gland lesions are well defined commonly with a lobulated or nodular edge.
INFLAMMATION:

Dacryoadenitis:

Typically presented as acute and subacute dacryoadenitis with pain and tenderness in lacrimal gland. There may be minimal evidence of proptosis with downward and inward displacement of globe. Common differential diagnosis are sarcoidosis, sjogren’s syndrome, wegener’s granulomatosis and idiopathic sclerosing inflammation.

On imaging, lacrimal gland is enlarged, with contrast enhancement and irregular margins. Ultrasound features of mass are internal reflectivity echoes and adjacent muscle thickening.

Histology revealed polymorphous cellular infiltration with edema and vascular dilatation and with no evidence of lacrimal gland destruction.

If there is destruction of lacrimal gland, possibility of organ specific immune disorder should be considered.
**Idiopathic Sclerosing Inflammation:**

Cicatrical infiltration with mild inflammation and mass effect present. Corticosteroids, Cyclophosphomide and azothioprine are also tried in this cases.

**Sarcoidosis:**

Seven percent of lacrimal gland mass was associated with sarcoidosis. On imaging, circumscribed mass with infiltrative margins seen. It is usually associated with other ocular findings. It is treated with oral corticosteroids and other immunosuppressants.

**Sjogrens Syndrome:**

Lacrimal gland involvement in sjogrens syndrome is presented with lymphocytic infiltration with mild dacryoadenitis and decreased secretion of tears.

This damage due to T cell infiltration and auto antibody involvement.

Pathologically glands with periductal lymphocytic infiltration, acinar atrophy and fibrosis are seen.
**DACRYOPS:**

Secondary cystic lesion which arise from the palpebral lobe of lacrimal gland. It is transparent and do not need biopsy for diagnosis.

These swellings are mobile, tense in nature and have fluctuation. On everting upper lid these cysts are transilluminant.

They enlarge very slowly. As cysts are discharging intermittently variability in size is commonly present. All cases presented with unilateral or bilateral involvement.

Histology reveals double layer of epithelium lining the cyst.
CLINICAL APPROACH

Important factors in clinical evaluation are

1. Clinical examination of lacrimal gland mass
2. Evaluation of proptosis
3. Histopathological Examination
4. Imaging

CLINICAL EXAMINATION OF LACRIMAL GLAND MASS:

1. INSPECTION:

   Change in skin colour and texture is seen. Lid contour may present as S shaped in lacrimal gland involvement which is surrounded by edema due to pressure signs and lymphatic drainage interference. Width of palpebral fissure measured and compared with other eye.

2. PALPATION:

   On palpation warmth, consistency, and tenderness is noted. Resistance to retropulsion is also seen. Mobility is restricted in secondary tumours rather than in malignant tumours.
EVALUATION OF PROPTOSIS:

With Hertle’s Exophthalmometer forward distance of frontal surface of the eye in relation to lateral rim of the orbit is measured and compared with other eye in unilateral proptosis. Normally it measures 16 – 17 mm in adults and slightly less in children. Usually globe deviation is opposite to the position of growing tumour. Lateral displacement is determined by measuring distance between the pupils of the eye and midline of the bridge of the nose. Anterior tumours mostly produce displacement and posterior tumours produce proptosis.

Fundus examination with direct ophthalmoscopy may reveal fine continuous radial lines of choroidal folds due to pressure effect.

HISTOPATHOLOGICAL EXAMINATION:

Aim of biopsy is to remove portion of orbital mass and better inspection of histopathological study to establish the diagnosis. Excision biopsy requires almost total removal of the tumour, primarily a method for benign tumour and secondarily as a diagnostic procedure. Incision biopsy is done for lesions not amenable to complete excision. FNAC and needle biopsy for lymphomas is rarely done.
INVESTIGATIONS

ULTRASOUND:

Two echoes are used most commonly. A scan to detect particular tissue component. B scan is used to localize the tumour.

CT SCAN:

Tomography is a type of body section Roentgenography permitting study of tissue shadows in one plane of focus of X ray machine and depend on difference in tissue electron density.

Principle of CT:

X- ray tube of machine emits a thin, collimated beam of X –rays. During passing through tissues, this beams are attenuated and detected by array of special detectors.

X-ray photons within detectors generate electric signals. Standard radiation dose is 3-5 rads and for high resolution CT is 10 rads.

Spatial resolution of a CT scan depends on slice thickness. Thinner slice effects higher resolution.
2mm cuts are optimal for eye and orbit. 1mm cuts are useful in orbital apex and evaluation of orbital mass.

Intraocular tumor with proptosis are common indication for CT Scan.

**Hounsfield Units:**

Represent a scale of radiation attenuation values of tissues. The number assigned is called Hounsfield number.

This number can range from -1000 to +1000 HU and above. Higher the number, greater the attenuation of X-rays and higher the tissue density.

**Contrast enhancement:**

Contrast enhancing lesion is bright or more intense after contrast medium infusion. An increase is Hounsfield value is more reliable indicator of contrast enhancement.

**Views**

Commonly used are axial, coronal and sagittal sections. Lateral view is important in lacrimal gland tumours to know the density, homogeneity of
lesion, lobular involvement within the gland, extension beyond the gland and bony changes like moulding of lacrimal fossa, erosion, invasion and destruction of adjacent bone.

Benign growth present as pseudoencapsulated moulding of lacrimal gland fossa with remodeling.

Bony destruction is seen in Adenocarcinoma, Squamous cell carcinoma and in secondary neoplasm. Small flecks of calcification can also be seen in CT scan.

**MRI:**

**Principle**

Depends on rearrangement of Hydrogen Nuclei, where a tissue is exposed to a strong electromagnetic pulse.

When the pulse subsides nuclei returns to normal position, re- radiating some of energy they have absorbed.
Sensitive receivers pickup this electromagnetic echo. These signals are analyzed, computed and displayed as a cross sectional image.

**Contraindications of MRI**

1. Presence of metal (pacemaker, aneurysmal clips)
2. Cardiac bypass surgery patients.
3. Claustrophobic patients.

**Features to be seen in MRI:**

1. Size, Shape and Site of tumor.
2. Circumscription of tumor.
3. Margin of tumor – smooth (benign) or irregular (malignant)
4. Effect on surrounding structures – fossa formation (benign) or Hyperostosis
5. Consistency – Homogenous (benign) or Heterogenous (malignant)

Non invasive, costly but not associated with radiation. Images are obtained in varieties of planes without repositioning patients.
Structure of the globe is better delineated in MRI than CT and more useful to view the intracanalicular portion of the optic nerve.

MANAGEMENT

CHEMOTHERAPY:

Investigative tools like CT scan and Ultrasound distinguish lymphomas which are radiosensitive and pseudotumours which are steroid responsive. However tissue examination is needed for confirmation. For pseudotumours 60 – 80 mg of oral prednisolone per day is the treatment of choice. Intraarterial cytoreductive chemotherapy may improve survival in adenocarcinoma. Surgical excision followed by chemotherapy is used for patients with systemic diseases.

RADIOThERAPY:

Radiotherapy (30-40Gy) followed by surgical excision is given for localized lymphoma. Radiotherapy of 6400-6800 Gy followed by surgical excision is given in Adenoid cystic carcinoma.
SURGICAL APPROACH FOR INCISIONAL BIOPSY:

Skin incision is made at the beginning of lateral canthal tendon and extended posteriorly 20 – 25 mm. Upper half of the lateral canthal tendon is detached from the bone. The upper lid and attached soft tissue is reflected upwards so that gland is identified for biopsy.

SURGICAL APPROACH FOR EXCISION BIOPSY: (LATERAL ORBITOTOMY)

Extended upper lid crease incision is made on the skin with scalpel blade and dissected through orbicularis muscle and deep fascia upto periosteum of the orbital rim. Periosteum along the lateral orbital rim is cut and elevated. Greater wing of sphenoid is removed to provide adequate exposure of the deep orbit. The periorbita is opened and the lesion is located, excised and sent for biopsy.

Complete excision without violating the pseudocapsule is the treatment of choice in benign lesions especially pleomorphic adenoma.
SUMMARY OF TREATMENT ALGORITHM FOR LACRIMAL GLAND LESIONS:

Tumour mass with predominant inflammatory features and without bony changes, steroids is the treatment of choice.

Tumour mass without inflammatory features, but with bony changes treated with biopsy, followed by specific therapy.

Tumour mass with benign features and no bony changes with compression or expansion of the lacrimal fossa is treated with complete excision and biopsy.

Tumour mass with malignant features and with bony changes including erosion and destruction of the bone is treated with excisional biopsy followed by radiotherapy with or without chemotherapy.
REVIEW OF LITERATURE

1. In a retrospective analysis of lacrimal gland lesions by J.E. WRIGHT (1992) with 50 cases observed that cranioorbital resection is done for 11 patients with malignant tumors. Combined tumor resection with radiotherapy improved patient survival in adenoid cystic carcinoma.

2. JUNG YOUNG reported a rare clinical case of 50 year female with pleomorphic adenocarcinoma who presented with spinal and intracranial metastasis.

3. In a clinical study of patients with Adenoid cystic carcinoma involving lacrimal gland (ASHTON series), bony invasion is very common in adenoid cystic carcinoma. Biological behavior of more aggressiveness was seen in basaloid variety.

4. SHIELD’s reported that 6% of lacrimal gland lesions were dacryops (8 of 142 patients). Mean age at time of presentation of dacryops was 38 years.
5. In case series conducted by WRIGHT, 50% of patients with adenoid cystic carcinoma presented with recurrences within two years. Mean age of presentation of adenoid cystic carcinoma is 41 years.

6. ICE and colleagues studied survival of adenoid cystic carcinoma in 26 patients and observed that only 50% of adenoid cystic carcinoma patients survived after 2.5 years. Intracranial extension is the major cause of death in adenoid cystic carcinoma.

7. In a largest series of tumors in lacrimal gland by EVIATOR, found that Orbital Exenteration and radiotherapy is beneficial for Mucoepidermoid carcinoma (High grade). Excision with or without radiotherapy is useful in lesions with low grade potential.

8. Study of 262 consecutive orbital tumors by PALANISAMY SUNDARRAJ revealed that incidence of pleomorphic adenocarcinoma is almost equal to adenoid cystic carcinoma.
9. A Comprehensive review of lacrimal gland lesions (26 patients) by HIND. M. ALKATAN found that incidence of malignant tumors is slightly higher than benign tumors.

10. EDWIN CHAN (HONG KONG STUDY) from 23 patients identified that lymphoproliferative disorders are more common above 60 years.

11. AUSTRALIAN COHORT of 263 patients with lacrimal gland biopsies revealed that non specific etiology is more common in two thirds patients with dacryoadenitis.

12. WASEE TULVATANA (THAILAND) studied 68 patients with lacrimal gland lesions and recognized, that lymphoma is the most common malignant lacrimal gland tumor.

13. RODRIGO SANTUS -10 year follow up of 180 cases with lacrimal gland lesions revealed that malignant tumor is more common than benign tumour in lacrimal gland.
14. Retrospective analysis of lacrimal gland tumors for a period of 15 years (63 cases) by M. KOHLI observed that epithelial tumors are uncommon below 10 years.

15. Dr. USHA KIM (ARAVIND EYE HOSPITAL) case series of 78 patients between 2005 and 2008. Two patients had Rosai-Dorman disease and one had neurofibroma in benign category. Among 29 patients with pleomorphic adenoma one patient had recurrence. Among 12 patients with adenoid cystic carcinoma two patients underwent Exenteration for recurrence. Among 2 patients with Mucoepidermoid carcinoma one patient underwent excision and another underwent Exenteration followed by radiotherapy.
AIMS AND OBJECTIVE

Aim:

To study the demographic profile, clinical features, treatment options and outcome of Lacrimal gland Lesions.

Objective:

1. To determine demographic profile of patients presenting with Lacrimal gland lesions including age and gender distribution.
2. To describe various clinical presentations of Lacrimal gland lesions.
3. To study the management algorithms, and histological features of different lacrimal gland lesions.
4. To determine the treatment outcomes in patients with various Lacrimal gland lesions.

Methodology

1. Cases presenting with Lacrimal gland lesions from the period of June 2012 to July 2014 are included for this study.
2. Retrospective, descriptive, case series conducted from June 2012 to July 2014.
History

1. Chief complaints:
   - Protrusion of eyeball (Unilateral/Bilateral)
   - Associated with lid swelling and drooping of Lids
   - May be associated with pain

2. History of Present illness
   - Onset
   - Course
   - Duration
   - Progression of symptoms

3. History of co-existing Systemic illness.

Past H/o

History of previous treatment/surgery

History of similar episodes in past

General examination

- Facial contour
- Asymmetry
- Pallor
- Cyanosis
- Clubbing
- Jaundice
- Lymphadenopathy

**Ocular Examination**

- Visual Acuity
- Head Posture
- Ocular Alignment

1. **Inspection**
   
   - Eyebrows
   - Eyelids
   
   **Mass**
   
   - Unilateral/ Bilateral
   - Pulsatile/ Not pulsatile
   - Variation with posture
   - Variation with valsalva
2. Palpation

- Warmth
- Tenderness
- Reducibility
- Pulsation
- Thrill
- Compressibility
- Finger insinuation
- Resistance to Retropulsion
- Variation with posture

**Palpation of Mass**

- Size
- Shape
- Number
- Margin
- Position
- Relation to Eyeball
3. **Auscultation**

- Bruit

**S/L Evaluation**

- Conjunctiva - Congestion / edema
  
  chemosis / dilated episcleral vessels.

- Cornea

- Pupil – Size/Shape/RAPD

**Systemic**

- Para Nasal sinus.

- Thyroid / CNS/RS/CVS

- Lymph Node enlargement

**Radiology**

- X –Ray

- USG – B Scan

- CT

- MRI
Histopathologic Examination

- Macroscopic
- Microscopic
- Immunoreactivity
CASE REPORTS

1. Name: Priyanka Mondal   Age: 17 years   Sex: Female

Came to orbit clinic for Right eye Protrusion for 2 years

Swelling with insidious onset, not associated with pain

Not warmth, Firm in consistency

Non tender mass in superotemporal quadrant.

Finger insinuation not possible

Visual acuity: Right Eye – 6/6 and Left Eye – 6/6.

Fundus: Normal
CT Orbit:

Mass in Superolateral angle of orbit involving lacrimal gland tethering superior rectus –levator complex suggesting pleomorphic Adenoma. There was no perineural spread, no calcification and no necrosis.

Management

Superolateral orbitotomy and mass excision and sent to histopathological examination.
POST OPERATIVE  (Lateral Approach)

HPE Report:

Evidence of both epithelial and mesenchymal differentiation.

Proliferation of benign epithelial cells are arranged in double layer to form lumens. Stromal differentiation can be seen in the formation of bone and cartilage suggestive of Pleomorphic Adenoma

Follow up:

In two year follow up there was no recurrence seen in this patient.
2. Name : Parvathi  Age: 45 years  Sex: Female

Came to orbit clinic for drooping of left upper lid and swelling in supero lateral aspect of orbit for 2 months.

Non tender mass with firm to hard in consistency

EOM: Restricted in Abduction

Visual Acuity in Both eyes - 6/6

Fundus: Normal

CT Scan:

Cystic lesion is supero lateral aspect of left orbit along the lateral canthal region indenting globe representing dermoid cyst. There is no evidence of calcification and malignant changes.

Management:

Excision through lateral orbital approach under General anesthesia and cyst separated and excised in toto along with prolapsed lacrimal gland.

HPE: Dacryops (Lacrimal Ductal cyst)

Followup : No recurrence
3. Name: Rani  Age: 41 years  Sex: Female

Came to orbit clinic with complaints of left protrusion of eyeball – 20 days

O/E: S- Shaped ptosis present not obscuring visual axis.

   Firm mass in lacrimal gland, not freely mobile, no retropulsion

EOM: Full

Visual Acuity: Right Eye – 6/6

Left Eye – 6/6 Partial

Anterior Segment: Normal

Pupil: Reacting
CT Orbit: Lacrimal gland lesion with cystic spaces and associated with bony destruction of orbit suggestive of Adenoid cystic carcinoma.

Management: Left superolateral orbitotomy and excisional biopsy.

Post operative (Lateral Approach)

HPE: Adenoid cystic carcinoma (basaloid variant)

Followed by: Postoperative Radiotherapy and chemotherapy.
4. **Name**: Haneef S.Iyad  **Age**: 14 years  **Sex**: Male

Came to orbit clinic with drooping of Left upper lid - 3 months which was painless/ progressive.

Swelling is firm in consistency, not fixed to underlying structures.

Swelling - not warmth, not tender,

No associated systemic illness

No previous episodes.

**Visual acuity**: Right Eye - 6/6

Left Eye - 6/6.

**Management**: Left eye cyst explored and dissected removed in toto and sent for histopathological examination.

**HPE**: Dacryops with squamous metaplasia.
5. Name: Janani Age: 12 years Sex: Female

Came to orbit clinic for Left eye swelling in superotemporal aspect of orbit for 2 months.

Swelling is firm in consistency,

Not warmth, not tender, Not mobile.

Not fixed to underlying structures

Visual acuity : Both eyes - 6/6

Anterior Segment : Normal

Pupil : Reacting

Extra ocular movements : Normal

Management:

Left – Dacryops excision done.

HPE Report:

Lacrimal ductal cyst (dacryops)
6. **Name:** Ali Rizween  
   **Age:** 26 years  
   **Sex:** Male

Came to Eye O.P. for Right Eye protrusion- 6 months

Insidious onset, gradual and progressive

**O/E:** Right Eye – Eccentric proptosis with firm, tender mass in superolateral quadrant.

**Visual Acuity:** Right Eye -6/18 and left eye-6/6

**Anterior Segment:** Normal

**Fundus:** BE- Normal

**CT Orbit:**

- lacrimal gland lesion without bony remodeling and without calcification seen

**Management:**

Superolateral orbitotomy with excision biopsy without violating capsule.

**HPE:**

Pleomorphic Adenoma with squamous metaplasia and Keratin production.
7. Name: Sasikumar  Age: 27 years  Sex: Male

Came to orbit clinic for swelling in the superotemporal aspect of left eye for 6 months

**O/E:** Left Eye Inferior dystopia with resistance to retropulsion.

Orbital margins intact.

Mass firm in consistency.

Finger insinuation not possible.

**EOM:** Normal

**Anterior segment:** Normal

**Visual Acuity:** Right Eye - 6/6 and Left Eye - 6/6
CT scan:

Heterogenously enhancing soft tissue lesion displacing globe downwards and cause bony indentation due to mass effect.

Management: Superior orbitotomy and mass excision biopsy

Post operative (Lateral Approach)

HPE: Lacrimal gland – Pleomorphic Adenoma
8. Name: Mahesh      Age: 39           sex: Male

Came to orbit clinic with complaints of Bilateral Gradual progressive swelling of superolateral orbit for 2 years.

**Finger insinuation**: Not Possible

**EOM**: Full

**Visual Acuity**: in Right Eye – 6/6 and in Left Eye – 6/6

**Pupil**: 2mm, Reacting

**Fundus**: Normal

**CT report**: B/L lacrimal gland enlargement – Suggestive of lymphoproliferative disorder.

**Management**: Patient started on systemic steroids. After stopping steroids, mass recurred.

**Management**: Left Eye - Incisional Biopsy of Lacrimal gland mass.

**HPE Report**: Non Hodgkin’s lymphoma

Patients was advised chemotherapy and regular follow up
9. **Name:** Maragatham       **Age:** 37 yr       **Sex:** Female

Came to orbit clinic with complaints of left Eye Protrusion – 3 years.

**O/E:** Upper lid and Lower lid congestion with inferior dystopia.

Eccentric proptosis and Restricted Retropulsion was present.

**Extra ocular Movement:** Full

**Pupil:** 3mm, reacting to light.

**Visual acuity:** Right Eye -6/6, Left Eye – 6/6- Partial

**CT ORBIT:** Well defined, extra conal, soft tissue density with attenuated mass lesion in superolateral compartment of left orbit- Lacrimal gland mass.

**Management:** Lateral orbitotomy and mass excision.

**HPE Report:** Pleomorphic Adenoma

**Follow up CT:** No residual / recurrent lesions.
10. Name: Abdul Rasheed  Age: 7 years  Sex: Male

Came to for orbit clinic with complaints of

Swelling in Right Eye – Upper lid for past -3 months

O/E: Right Eye – Swelling in the supero lateral aspect of Right Eye Orbit

BE:

Visual Acuity: 6/6
CT Orbit:

Hyper dense lesion and enlargement of Right Eye - lacrimal gland blends with lateral rectus.

Management:

Superolateral mass excision under incisional biopsy
HPE:

Structure of fibrofatty tissue with dense hyalinised collagen and vascular proliferation. Perivascular lymphocytic infiltration with interspersed eosinophils are seen suggestive of Idiopathic sclerosing orbital inflammation.

Immunohistochemistry:

LCA: Diffuse positive reactivity

Post Operative Period:

Started on tab. prednisolone in tapering doses.
11. Name: Sundarajan  Age: 43 years  Sex: Male

Came to orbit clinic for pain with complaints of Right Eye – protrusion for 2 months.

Not warmth, firm in consistency

Swelling not associated with retropulsion

Not fixed to underlying structures

Finger insinuation not possible.

O/E: Right Eye – Eccentric proptosis with firm tender mass in superotemporal aspect.

Visual Acuity: BE – 6/6

CT Scan: Heterogenous mass in superolateral aspect of Right orbit without Bony erosion tethering lateral rectus.

Management: Right Eye – Lateral orbitotomy excision Biopsy followed by chemotherapy and Radiotherapy.

Follow up: In one year, recurrence of Adenoid cystic carcinoma and advised chemotherapy with Adriamycin and cisplatin.
12. Name: Alphonsa  Age: 45 years  Sex: Female

Came to orbit clinic with complaints of swelling in Right superotemporal aspect for 2 years

Swelling is Painless and Progressive

Rubbery consistency

Finger insinuation not possible

O/E:

Right Eye ‘S’ shaped ptosis

Firm mass palpable

CT Orbit:

Mass replacing entire lacrimal gland and blends with globe and inferior rectus muscle.

Management:

Right Lateral orbitotomy and incisional Biopsy.

HPE:

Non Hodgkins lymphoma – B Cell type and started chemotherapy.
13. Name: Fathima  Age: 54 years  Sex: Female

Came to orbit clinic for Right Eye – Protrusion for one month.

O/E: Right Eye - Proptosis

Firm mass palpable in Right Lacrimal gland area

Finger insinuation not possible

Abduction restricted.

On palpation - mass not warmth,

rubbery in consistency

not pulsatile, no bruit present

orbital margins intact.

No change in size of mass in various postures

Systemic examination: Generalised lymphadenopathy

Hepatosplenomegaly

Anterior segment examination: Normal

Pupil: reacting to light.
**Visual Acuity:**  
BE - 6/24

Complete hemogram - Normal

Chest X Ray - Normal

Ultrasound abdomen - Fatty liver

BMA - Leucopoiesis with small to medium sized atypical cleaved nucleated lymphoid cells upto 50 percent. - lymphomatous infiltration of bonemarrow with leukemic manifestation.
CT Orbit:

Lacrimal gland mass involving Right globe and lateral rectus muscle with Bony erosion.

DIFFERENTIAL DIAGNOSIS:

1. pleomorphic adenoma
2. Lymphoproliferative disorders

Management:

Lateral orbitotomy and excision biopsy
**HPE Report:**

B Cell type – Follicular lymphoma/ blastic differentiation. Lymphoid cells arranged in follicles.

Followed by:

**Chemotherapy** (chlorambucil and prednisolone) – advised for follow up.
Name : Thangamma David  Age:65 years  Sex: Female

Came to orbit clinic with complaints of

Swelling in Right  Superolateral aspect of orbit for  3 months

Not warmth, not mobile

Finger  insinuation  not possible.

O/E: Right Eye  -Mechanical ptosis

    Swelling in superolateral quadrant

    Salmon patch like appearance in tarsal conjunctiva.

Visual Acuity:  Right Eye  -  6/18

    Left Eye  -  6/12

CT Orbit:  Soft tissue density mass in the superolateral aspect of right orbit.

Management:  Mass excision biopsy.

HPE Report:

    Necrobiotic Granulomatous inflammation (nonspecific). Patient was started on steroids.
15. **Name:** Hamza  **Age:** 24 years  **Sex:** Male

Came to orbit clinic with complaints of

Left Protrusion of Eyeball for 10 days

No history of trauma

No history of previous episodes

No history of fever with seizures

**O/E:** Eccentric proptosis seen

mass felt in supero lateral region.

Globular in shape

Smooth surface

Mass firm in consistency

Mass not warmth, not tender

Not fixed with underlying structures

Finger insinuation not possible.
Visual Acuity:  Right Eye - 6/6 and  Left Eye - 6/9

Investigation:

Complete hemogram - Normal
Chest x ray - Normal

CT Orbit:

Extraconal soft tissue density lesion in left supero lateral aspect of orbit without Bony erosion. Suggestive of pleomorphic adenoma.

Management:

Orbitotomy and excision biopsy.

HPE:  Adenoid cystic carcinoma

Post operative Management:

Chemoradiotherapy and follow up
16. **Name:** Chinnaiah  **Age:** 41 years  **Sex:** Male

Came to orbit clinic with complaints of painless, gradual, swelling in Left Orbit for 20 days

Associated with Inferior dystopia and eccentric proptosis. Swelling is firm in consistency. Not freely mobile, not associated with retropulsion.

**Visual Acuity:**
- Right Eye - 6/6 Partial and
- Left Eye - 6/6

**Anterior Segment:** Normal

**EOM:** Full

**Pupil:** Reacting

**Fundus:** Normal

**CT Report:** Left lacrimal gland Neoplasia with bony erosion with mild infiltration of Lateral rectus muscle

**Management:** Lateral orbitotomy with mass excision Biopsy.

**HPE report:** Hemangiopericytoma (intermediate grade) solitary fibrous tumor complex. Diffuse CD34 and vimentin positive.

**Follow up:** No recurrence.
17. Name: Gnanaoli  Age: 33 years  sex: Male

Came to orbit clinic with complaints of Bilateral mass in superolateral quadrant.

No H/o of trauma ,

No H/o of previous episodes in past

O/E:

Soft tender mass in superolateral quadrant of Both Eyes.

Not warmth, not tender

Finger insinuation not possible

Orbital margins intact

General Examination

B/L Parotid gland involvement

B/L submandibular gland involvement with

multiple enlarged lymph nodes and hepatosplenomegaly.

Visual Acuity: Right eye - 6/9
Left eye - 6/9

Both eyes

- S shaped ptosis
- palpebral lobe of lacrimal gland enlargement
- subconjunctival hemorrhage
- orbital margins intact. EOM - Normal
**CT Orbit (axial view)**

Bilateral Lacrimal gland enlargement.

**CT Abdomen:**

Diffuse bone marrow changes replacing axial and appendicular skeleton with multiple lymphadenopathy and Hepatosplenomegaly.

**Complete hemogram:**

- **TC**: 1,20,000
- **DC**: Atypical cells 80 percent.
- **ESR**: 30 mm/hr
**Peripheral Smear** - Presence of large atypical cells with irregular nuclei and prominent nucleoli.

**Bone marrow aspiration** - Lymphomatous infiltration of bone marrow with leukemic manifestations.

**CT Brain** – No evidence of subarachnoid hemorrhage and intraventricular hemorrhage.

**MRI Spine / Abdomen**

Diffuse marrow changes replacing axial and appendicular skeleton

Multiple enlarged lymph nodes and hepatosplenomegaly.

**Peripheral Smear:**

Presence of large atypical cells with irregular nuclei and prominent nucleoli.

**HPE Report:**

Monomorphous atypical lymphoid cells with prominent nucleoli Suggestive of B cell type Non Hodgkin’s lymphoma.
Post Operatively:

Treated with chemotherapy- chlorambucil and prednisolone.

Follow Up : No recurrence was seen
18. Name: Pandi  Age: 50 years  Sex: Male

Came to for orbit clinic with Right Eye defective vision for 6 months

O/E:

Right sided proptosis with inferior dystopia

Mass in superotemporal quadrant of orbit with firm consistency

Mass not fixed to underlying structures

Not associated with retropulsion

Size of mass not varies with posture.

Anterior segment: Normal

EOM: Restricted in abduction and elevation in abduction.

Visual Acuity: Right Eye -6/18  Left Eye - 6/6

Fundus: Choroidal folds present

CT Orbit: Right side lacrimal gland neoplasm with mild bony erosion.

Management: Right – Lateral Orbitotomy with mass excision biopsy

HPE: Pleomorphic Adenoma.
19. **Name:** Maria Nevis  **Age:** 43 years  **Sex:** Female

Came to orbit clinic with Left swelling in Superolateral orbit – 5 years

Sudden onset with rapid growth

**O/E:** Mass in superolateral quadrant

Warmth, firm in consistency

Finger insinuation not possible

No pulsatile, no change in size after varying posture.

No similar treatment episodes in past.

**Extra ocular movements:** Normal

**Visual Acuity:** Both Eyes – 6/6.

**IOP** Both Eye – 15 mm Hg

**Management:**

Left lacrimal gland incisional biopsy done.

**HPE report:**

Pseudotumor.
20. Name: Ramamma  Age: 60 years  Sex: Female

Came to orbit clinic with complaints of

   Right Eye  protrusion of Eyeball –for 5 months

   Swelling sudden onset rapid growth

O/E: Right side – Lacrimal gland swelling chemosis and congestion at rectus insertion site

Visual acuity: BE – 6/18.

CT Scan: Heterogenous lesion in superolateral compartment of Right lacrimal gland with laterial rectus and optic nerve involvement.

Thyroid: Cystic lesion with calcification with multiple lymph nodes.

Management: Superolateral orbitotomy with mass excision Biopsy.

HPE report: Adenocarcinoma of Lacrimal gland (Basoloid type).
21. **Name:** Kannan  
**Age:** 40 years  
**Sex:** Female

Came to Orbit clinic for Right protrusion of eyeball for 6 months.

**O/E:** Eccentric Proptosis with firm swelling in superotemporal quadrant.

- Not warmth, firm in consistency
- Not fixed to underlying structures
- Finger insinuation not possible
- Not pulsatile, No bruit felt.

**Visual Acuity:** Both Eye - 6/6

**Extra ocular movements** - Normal

**CT Orbit:** Right Eye – Lacrimal gland mass with bone remodeling with no perineural invasion, with no calcification.

**Management:**

- Right Eye – lateral orbitotomy with mass excision Biopsy.

**HPE Report:**

- Pleomorphic Adenoma.
22. **Name**: Johnson  
**Age**: 18 years  
**Sex**: Male

Came to orbit clinic with complaints of swelling in left lacrimal gland area – One month sudden onset, rapid growth

**O/E:** Firm mass felt in lacrimal gland area

Resistance to retropulsion.

Not warmth, Not pulsatile

**CT Orbit:** Well defined lobulated, soft tissue density lesion in Left lacrimal gland with Bony erosion infiltrating lateral rectus and Infratemporal fossa and frontal lobe.

**Management:** Lacrimal gland mass incision biopsy.

**HPE Report:**

Lacrimal gland acini with infiltration of round to oval cells having contorted and indented nuclei with pale eosinophilic cytoplasm. Focal collections of eosinophils with prominent nucleoli and necrosis seen. Special stain for AFB- Negative- suggestive of Histiocytosis more in favour of Langerhan cell histiocytosis.

**Follow Up**: No recurrence was seen
23. **Name: Ashokan**  **Age: 58 years**  **Sex: Male**

Came to orbit clinic for Swelling Right lacrimal gland area – 3 months.

Insidious onset, painful

Progressive in nature

Not associated with fever

Not associated with other systemic illness

**PAST H/o**

No similar episodes in past

No H/o of trauma

**O/E:**

Firm mass in superotemporal aspect of lacrimal gland

Firm in consistency

Smooth surface

Warmth. Mild tenderness present.

Not fixed to underlying structures
Visual Acuity

Right Eye - 6/6
Left eye - 6/6

EOM- Normal

Investigations:

Complete hemogram- Normal
ESR- 45 mm/hr

CT Brain:

Hyper intense signal along superolateral aspect of Right lacrimal gland.
There was no evidence of calcification and perineural invasion. No evidence of malignant changes.

Management:

Incisional biopsy for lacrimal gland mass.

HPE:

Right Dacryo adenitis (Non specific)

Patient was Treated with steroids and advised follow up.
24. Name : Vijayan  Age : 64 years  Sex : Female

Came to orbit clinic with complaints of

Mass in Superolateral quadrant

Associated with pain, swelling – one month.

Not associated with lid swelling

Not associated with drooping of eye lids

O/E:  Mass is firm in consistency

Finger insinuation not possible

Not warmth, not tender

Irregular surface.

No H/o of DM and Hypertension

No H/o of trauma

No H/o of contact with tuberculosis patients

No H/o mouth ulcers, fever, joint pain

No H/o of genital ulcers, urethral discharge
Previous recurrent history of dacryoadenitis which was treated with steroids,

**Visual Acuity:**

- Both Eye - 6/6

**Anterior segment examination:**

- Facial contour – normal
- Eye lids – normal
- Eccentric proptosis
- RE- Temporal congestion

No evidence of scleritis or sclera abscess.

**MRI (Brain and Orbit):**

Swelling in superolateral aspect of lacrimal sac and gland.

- Mantoux : Reactive (15mm)
- ESR : Elevated (>150mm/Hg)
- X-ray chest : Hilar lymphadenopathy
- Nested PCR : Myco bacterium Tuberculosis positive.
- Serum ANA : Negative
- IgM for TB : Negative
IgG for Toxoplasmosis : Negative
IgM for Toxoplasmosis : Negative
Mantoux : 15mm. Reactive
Urine Bence Jones protein : Negative
Urine routine : 2 to 3 pus cells
ESR : More than 150 mm/hr

CT Orbit:

Enlargement of right lacrimal gland, with edema, no evidence of calcification, bony changes.

Management:

Right lacrimal gland – mass Incisional biopsy through skin crease incision, gland identified, biopsy taken and material sent for culture and sensitivity, biopsy

HPE Report:

Structure of lacrimal gland acini in lobules with destruction by infiltration of lymphocytes and plasma cells suggestive of TB Dacryoadenitis Patient was started Anti tuberculous Therapy. He responded well with no recurrence.
25. Name: Jolsna Benny  Age: 36 years  Sex: Female

Came to orbit clinic for swelling in Right superotemporal orbit - one year.

O/E: S- Shaped ptosis. Mass is firm to hard in consistency.

Anterior segment: Normal

EOM: Restricted in abduction and elevation in Abduction.

Visual Acuity: Right Eye -6/6  Left Eye - 6/6.

CT Orbit: Right – Lacrimal gland Neoplasia with mild Bony erosion.

Management: Right Eye – Lateral orbitotomy with mass excision Biopsy.

HPE: Pleomorphic Adenoma (with mixed epithelial cells and mesenchymal cells)

Follow up: No recurrence.
26. Name: Geetha  Age: 33 years  Sex: Female

Came to orbit clinic with complaints of swelling in superotemporal aspect of Left orbit for 3 months.

Swelling was insidious in onset.

Not associated with pain

Not warmth, not tender

Finger insinuation not possible.

O/E: Swelling – cystic in consistency

Transillumination was positive

No bruit felt on auscultation

Was not associated with pulsation

Visual Acuity: BE – 6/6.

Management:

Left Excision of mass in toto done and sent for histopathology.

HPE: Inflamed ductal cyst (Dacryops)
27. Name: Muthu Vadivoo    Age: 64 years    Sex: Female

Came to orbit clinic for protrusion of Left Eyeball – 2 months

O/E: Left Eye – eccentric Proptosis

Resistance to retropulsion

Firm in consistency

Finger insinuation not possible superiorly and laterally.

Visual Acuity: Right Eye -6/6 – partial and Left Eye -6/36.

Investigations:

Serum RA factor - Negative, ANA screening - Negative

CT orbit: Soft tissue density mass in superolateral aspect of left orbit
Management: Lateral orbitotomy with incisional biopsy done.

HPE Report:

Necrobiotic granulomatous inflammation suggestive of sarcoidosis.

Post operatively treated with Steroids.
ANALYSIS

In this series, 27 cases of lacrimal gland lesions reported in our Orbit clinic, Aravind Eye hospital from June 2012 to July 2014.

Table : 1

| S.No | Lacrimal Gland Lesions                  | No. of Cases | Sex       |
|------|----------------------------------------|--------------|-----------|
|      |                                        |              | Male  | Female |
| 1    | Benign                                 |              | 8     | 4      | 4      |
|      | Pleomorphic Adenoma                    |              |        |        |        |
| 2    | Malignant                              |              | 3     | 2      | 1      |
|      | Adenoid Cystic Carcinoma               |              |        |        |        |
|      | Hemangio Pericytoma                    |              | 1     | 1      | -      |
|      | Adenocarcinoma                         |              | 1     | -      | 1      |
| 3    | Inflammatory                           |              | 3     | 2      | 1      |
|      | Pseudo Tumor                           |              |        |        |        |
|      | Sarcoidosis                            |              | 1     | -      | 1      |
|      | Nonspecific Adenitis                   |              | 1     | -      | 1      |
| 4    | Infective                              |              | 1     | 1      | -      |
|      | TB Adenitis                            |              |        |        |        |
| 5    | Lympho proliferative Disorders         |              | 2     | 1      | 1      |
|      | Non Hodgkin’s lymphoma                 |              |        |        |        |
|      | Atypical lymphoid hyperplasia          |              | 1     | 1      | -      |
|      | Langerhan cell Histiocytosis           |              | 1     | -      | 1      |
| 6    | Structural                              |              | 4     | 1      | 3      |
|      | Dacryops                               |              |        |        |        |
|      | Total                                  |              | 27    | 13     | 14     |
Table: 2

Sex Ratio

|       | Male | Female |
|-------|------|--------|
| Male  | 13   |        |
| Female|      | 14     |

![Male and Female Composition](image.png)
| S.No | Type of Tumor                          | Total No. of Case | 0-10 yr | 10-20 yr | 20-30 yr | 30-40 yr | 40-50 yr | 50-60 yr | 60-70 yr |
|------|---------------------------------------|-------------------|---------|----------|----------|----------|----------|----------|----------|
| 1    | Pleomorphic Adenoma                   | 8                 | -       | 1        | 3        | 2        | 1        | 1        | -        |
| 2    | Dacryops                              | 4                 | -       | 2        | -        | 1        | 1        | -        | -        |
| 3    | Pseudo Tumor                          | 3                 | 1       | -        | -        | -        | 1        | 1        | -        |
| 4    | Non Hodgkins lymphoma                 | 2                 | -       | -        | -        | 1        | 1        | -        | -        |
| 5    | Adenoid cystic Carcinoma              | 3                 | -       | -        | -        | -        | 2        | -        | 1        |
| 6    | Sarcoidosis                           | 1                 | -       | -        | -        | -        | -        | -        | 1        |
| 7    | Atypical lymphoid Hyperplasia         | 1                 | -       | -        | -        | 1        | -        | -        | -        |
| 8    | Langerhan cell Histiocytosis          | 1                 | -       | 1        | -        | -        | -        | -        | -        |
| 9    | TB Adenitis                           | 1                 | -       | -        | -        | -        | -        | -        | 1        |
| 10   | Adenocarcinoma                        | 1                 | -       | -        | -        | -        | -        | -        | 1        |
| 11   | Hemangiopericytoma                    | 1                 | -       | -        | -        | -        | 1        | -        | -        |
| 12   | Nonspecific Adenitis                  | 1                 | -       | -        | -        | -        | -        | -        | 1        |
Table: 4 Demographic profile

| State            | Tamil Nadu | Kerala | Andhra pradesh | West Bengal | Maldives |
|------------------|------------|--------|----------------|-------------|----------|
| No of cases      | 18         | 5      | 2              | 1           | 1        |
Table:5

| Procedure                  | Count  |
|----------------------------|--------|
| Lateral Orbitotomy        |        |
| Excision Biopsy            | 17     |
| Incisional Biopsy          | 5      |
| Chemo Radiotherapy (Post Excision) | 3  |
| Chemotherapy (Post Excision)| 2      |
RESULTS

1. A total of 27 patients presented to our orbit department, Aravind Eye Hospital with Lacrimal gland lesions from June 2012 to July 2014 of whom 13 patients were males (48%) and 14 (52%) were females. The age of presentation ranged from 7-64 years.

2. The patients were clinically classified into six categories. – Benign, Malignant, Inflammatory, Infective, lymphoproliferative and structural lesions.

3. Benign and malignant tumours are completely rare below 10 years.

4. There is significant increased incidence of structural lesions. (Dacryops) presented in this study. Among structural lesions 4 patients (15%) presented with Dacryops. There were 3 female and 1 male patient. Patient with dacryops underwent lateral orbitotomy and mass excision in toto. All the four patients responded well with no recurrence.

5. Rare tumors of Hemangiopericytoma, Adenocarcinoma, Langerhan cell Histiocytosis were reported in this case series.

6. In Dacryoadenitis (6 patients) (22%) 3 patients were male and 3 were female. A case of Tuberculous adenitis involving lacrimal gland was
identified which was confirmed with nested PCR. He was treated with excision biopsy followed by Anti Tuberculous drugs with no recurrence.

7. In this category of Dacryoadenitis, two patients presented with Necrobiotic granulomatous inflammation for which both of them underwent excision biopsy and one was diagnosed as sarcoidosis and another was diagnosed as Nonspecific adenitis. Both of them treated with steroids with no recurrence.

8. Remaining 3 patients with dacryoadenitis were diagnosed as pseudo tumor. They responded well with steroids.

9. Among 8 benign tumors (29%) reported in this series, all patients presented with pleomorphic adenoma. 4 patients were male and 4 were female with equal sex preponderance. The age of presentation ranged from 10 – 60 years. Duration of symptoms ranges from 4 months to 6 years.

10. The commonest symptom in these patients with pleomorphic Adenoma is painless, insidious onset, orbital mass. The treatment included complete excision without violating the capsule. There was no recurrence during follow up.
11. Among 4 patients (14%) presented with lymphoproliferative disorders, two patients presented with bilateral lacrimal gland enlargement. One patient was diagnosed as Langerhan cell Histiocytosis for which, he underwent incisional biopsy followed by steroids with no recurrence.

12. Two patients were diagnosed as Non Hodgkins Lymphoma (B cell type). They underwent excision Biopsy followed by chemotherapy alone. These 2 patients responded well with treatment with no recurrence. Remaining one patient with Atypical lymphoid Hyperplasia, was treated with steroids.

13. Five patients (18%) in this study were diagnosed as malignant lesions, 3 had Adenoid cystic carcinoma, one with Adenocarcinoma and one with Hemangiopericytoma.

14. Among 3 patients (11%) with Adenoid cystic carcinoma 2 were male and one was female. These patients treated with lateral orbitotomy and excision biopsy followed by Chemo Radiotherapy and regular follow up.

15. Hemangiopericytoma (intermediate grade) was identified in one male patient. Adenocarcinoma (basaloid types) was reported in one female patient. Both of them underwent mass excision biopsy through lateral orbitotomy, with no recurrence during followup.
DISCUSSION

A total of 27 patients presented to our orbit department, from Aravind Eye Hospital with Lacrimal gland lesions from June 2012 to July 2014. This study is comparable to Henderson Series (1994), Rootman Series (1999), Reese (1956), Ashton (1975), Stewart (1979), Shields (1989), British Columbia orbit clinic series (1976-1999) Dr.Usha kim, Aravind Eye Hospital series (2005-2008) et al.

This study has its own unique features as Benign and malignant tumours are completely rare below 10 years and increased incidence of structural lesions (Dacryops). For Dacryops complete excision biopsy in toto was done.

This study also features rare tumors of Hemangiopericytoma, Adenocarcinoma, Langerhan cell Histiocytosis in this case series. Hemangiopericytoma is not included in other comparable case series like British Columbia orbital clinic series.

In all nonspecific dacrtyoadenitis cases, whom not responding to medical treatment, infective etiology especially tuberculosis should be ruled out. Though tuberculosis is common in our country only one case reported with dacrtyoadenitis due to tuberculosis which was confirmed with nested PCR.
Lacrimal gland lesions has wide range of clinical presentation and Histopathological diagnosis. Lacrimal gland lesions can be divided into two groups. First group with lesions conforming to lacrimal gland (Benign lesions) which presents with painless lacrimal gland mass with insidious onset. Pleomorphic adenoma is the common benign tumor in this study. All patients need excision biopsy without violating capsule and followup is needed.

The second group is with lesions not conforming to clinical picture of lacrimal gland. This group contains variety of lesions like structural lesions (Dacryops), lymphoma, malignant tumors, dacryoadenitis.

Adenoid cystic carcinoma is the common malignant tumor arising from lacrimal gland. This tumor has high recurrence rate of of 40 to 100 percent in various studies. In our study we found recurrence of 75 percent after a follow up for 2 years. 6 months and 1 year followup is needed in these cases to detect metastasis especially to lung.

Lymphoproliferative disorders involving lacrimal gland presented with equal gender preponderance and bimodal presentation which is more in third to fifth decade.
Cases with Bilateral lacrimal gland involvement and with suspicious malignancy immediate complete metastatic workup with immunohistochemistry confirmation are needed to prevent recurrence and to improve life expectancy of patients.

All patients with lacrimal gland lesions need lateral orbitotomy and excision biopsy.

In some cases with suspected infective and nonspecific inflammatory etiology and cases with orbital invasion (where complete excision is not possible) are subjected to incisional biopsy.

Early and accurate diagnosis with proper followup and counselling is needed in all lacrimal gland lesions, so that satisfactory outcome can be expected.
CONCLUSION

1. A total of 27 patients presented to our orbit department, Aravind Eye Hospital with Lacrimal gland lesions from June 2012 to July 2014.

2. Benign and malignant tumours are completely rare below 10 years.

3. There is significant increased incidence of structural lesions (Dacryops) presented in this study.

4. Rare tumors of Hemangiopericytoma, Adenocarcinoma, Langerhan cell Histiocytosis were reported in this case series.

5. Acute dacryoadenitis can occur at any age. Complete ophthalmic examination with appropriate investigations is needed to identify underlying etiology so that treatment plan can be tailored to each patient individualy.

6. Benign tumours commonly seen in age group of 20 - 40 years and malignant tumours are common in 60 – 70 years.

7. Most common benign tumour is pleomorphic adenoma. Since they underwent complete excision without violating capsule, there was no recurrence seen in this case series.
8. Adenoid cystic carcinoma is the common lacrimal gland malignant tumour with recurrence rate of 75% in 2 years follow up in this series.

9. Incisional biopsy has also important role in this study in some cases where complete excision is not possible.

10. Post surgical Chemoradiotherapy is beneficial in Adenoid cystic carcinoma.

11. Post Surgical Chemotherapy is beneficial in Non Hodgkin’s lymphoma.
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CLINICAL PROFORMA

Name : 
Age : 
Sex : 
M.R.No. : 
Diagnosis : 
Complaints : 

Clinical Evaluation

BCVA

Pre op Evaluation

Visual Acuity

General Examination

- Facial contour
- Asymmetry
- Pallor
- Cyanosis
- Clubbing
- Jaundice
Ocular Examination

- Visual Acuity
- Head Posture
- Extraocular movements
  
  Y-1   N-2

4. Inspection

- Eyebrows
- Eyelids

Mass

- Eccentric proptosis
  
  P-1   A-2

- Unilateral/ Bilateral
  
  1   /   2

- Inferior dystopia
  
  P-1   A-2

- Pulsatile/ Not pulsatile
- Variation with posture
- Variation with valsalva
5. **Palpation**

- Warmth
- Tenderness
- Reducibility
- Pulsation
- Thrill
- Compressibility
- Finger insinuation
- Resistance to Retropulsion
- Variation with posture
- Transillumination

P-1  A-2

**Palpation of Mass**

- Size
- Shape
- Number
- Margin
- Position
Relation to Eyeball

Consistency

Firm -1  Cystic-2  Rubbery -3

6. Auscultation

Bruit

S/L Evaluation

Conjunctiva - Congestion / edema

chemosis / dilated episcleral vessels.

Cornea

Pupil – Size/Shape/RAPD

Systemic Involvement

Y-1  N-2

Para Nasal sinus.

Thyroid / CNS/RS/CVS

Lymph Node enlargement

Radiology

X-Ray
- USG – B Scan
- CT
- MRI

**Biopsy**

- Excisional / Incisional
  
  1 / 2

**Histopathologic Examination**

- Macroscopic
- Microscopic
- Immunoreactivity
  
  P-1       A-2

**Nature of the lesion**

Benign -1
Malignant -2
Inflammatory -3
Infective -4
Structural -5
Lymphoproliferative -6
Management

- Orals steroids
  Y-1      N-2
- Postop chemoradiotherapy -1
- Postop chemotherapy -2
- Postop radiotherapy-3
- Nil -4

Follow up

Recurrence

Y-1      N-2
ABBREVIATION

CT  -  Computed Tomography
MRI -  Magnetic Resonance Imaging
RAPD -  Relative afferent pupillary defect
BMA -  Bone marrow aspiration
HPE -  Histopathological examination
TC  -  Total Count
DC  -  Differential Count
INTRODUCTION

There has been an increasing interest among ophthalmologists to study the management algorithms of lacrimal gland lesions.

Unlike other ophthalmic conditions, lacrimal gland lesions have difficulties in clinical examination, diagnosis, and surgical approach; as final diagnosis cannot be accurately predicted. Knowledge of these lesions can have a relationship with age, gender demographics, and other socioeconomic factors.

A large number of various lacrimal gland lesions occur in the series performed by the authors in different institutions.

The present study has been aimed to study the demographic profile, clinical features, treatment options, and outcome of lacrimal gland lesions that have...
INTRODUCTION

There has been an increasing interest among ophthalmologists to study the neoplastic degeneration of lacrimal gland tumors. Unlike other exophthalmic conditions, lacrimal gland tumors have difficulties in clinical examination. However, a singular approach to the differential diagnosis cannot be accurately performed. Various theories of these tumors have been developed with age, gender, demographic and other environmental factors. A large number of cases have been rarely documented. In contrast, various authors have tried to establish the various factors in the work published by individual authors and different institutions. The present work has been aimed to study the demographic profile, clinical differences, treatment options and outcomes of lacrimal gland tumors that have been reported in the Author's Eye Hospital, Mumbai, from the last 2015 – 2016.