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Ophthalmology

A CLINICO-PATHOLOGICAL STUDY OF PTOSIS

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ABSTRACT

Background: Blepharoptosis, or ptosis of the eyelid, refers to drooping of the upper eyelid that usually results from a congenital or acquired abnormality of the muscles that elevate the eyelid. Ptosis of upper eyelid is a common condition encountered by every ophthalmologist especially an oculoplastic surgeon and accounts for more than 50% of all major oculoplastic surgeries[8]. The etiology of ptosis is varied and multifactorial[12]. The treatment modalities for ptosis varies depending on the etiology, amount of ptosis, levator function and various other factors. This study is made to assess the various causes, pathogenesis and the various modalities of management.

Methods: This prospective study was conducted among patients presenting with ptosis to the department of ophthalmology, of a tertiary eye center in southern India, diagnosed on the basis of history, clinical symptoms and signs, and by pathological study. 66 eyes of 56 patients were studied during period of two years beginning from oct- 2017 to oct -2019.

Results: In our study congenital ptosis was the commonest type - 46 cases (82%) and acquired variety of ptosis comprised 10 cases (18%). Congenital ptosis commonly presented in the first two decades of life (65%) both in males and females. The mean age of incidence was 15 yrs. Males (56.5%) were slightly more commonly affected than females(43.4%) Acquired ptosis presented mostly after 4th decade and the incidence increased with age. There was no sex predilection among the acquired type. Among congenital ptosis, simple (i.e) dystrophic type was the most common (78.2%) compared to complicated type (21.7%). Among the cases with acquired ptosis, 6 were myogenic/ aponeurotic (60%), 2 were mechanical due to plexiform neurofibromatosis (20%), 1 each was traumatic and neurogenic (longstanding 3rd nerve palsy (10%). All the cases of congenital simple ptosis showed dystrophic changes like fibrocollagenous tissue replacing normal skeletal muscle, decrease in the diameter of fibres, fibrous and fatty infiltrate. Aponeurotic ptosis, LPS showed atrophy of muscle with mild fatty infiltration without any fibrous component.

Conclusions: Commonest type of ptosis is congenital ptosis, with simple congenital ptosis (dystrophic type) predominantly seen and commonly presenting in the 1st and 2nd decades of life. More commonly observed in males than in females..Unilateral ptosis was more common in this study with left eye involved predominantly. There was a correlation between LPS function and amount of fibrofatty infiltrate,in simple congenital ptosis. Eyes with poor LPS function show severe fibrosis. LPS resection gave very good results in mild to moderate ptosis and brow suspension was prefered in severe ptosis with poor levator action. Among the acquired ptosis, aponeurotic type is the most common with fatty infiltrate is the commonest finding, which also explains the lack of lid lag in aponeurotic type of ptosis. surgical management by LPS plication gave good results.

KEYWORDS : Frontalis sling surgery, levator muscle, Muller's muscle, ptosis

BACKGROUND

ANATOMY

The muscles concerned with elevation of upper eyelid are levator palpebrae superioris (LPS), Muller's muscle, and frontalis.[1] LPS is the major muscle involved which is innervated by the CN III(oculomotor nerve). The levator originates from the lesser wing of the sphenoid bone. As it traverses the orbit, it broadens and becomes a fibrous aponeurosis that inserts on the anterior aspect of the tarsal plate. The upper eyelid skin crease is formed by attachments of the aponeurosis to the orbicularis muscle and skin.[2] Muller's muscle is a smooth muscle that arises from the undersurface of the levator and inserts into the superior tarsus. The Muller's muscle contributes 1–2 mm of eyelid elevation. Muller's is under sympathetic control and when gets fatigued or dysfunctional, leads to mild ptosis[9]. The frontalis muscle lifts the brows, and it is innervated by CN VII (facial nerve).

CLASSIFICATION OF PTOSIS

Blepharoptosis or ptosis can be classified as: based on age of onset

1. Congenital : at birth or <1yr of age
2. Acquired : after 1yr of age

ETIO-PATHOLOGIC TYPES OF PTOSIS

CONGENITAL PTOSIS:

The majority of congenital ptosis is due to myogenic dysgenesis of the levator muscle. Congenital ptosis may occur through autosomal dominant inheritance.

1. ISOLATED CONGENITAL PTOSIS

A. SIMPLE CONGENITAL PTOSIS:

due to dystrophy of the levator muscle is the most common type of congenital ptosis. The patients usually manifest a lid lag on down-gaze and higher than normal or poorly formed lid crease. Rarely, lagophthalmos may be seen.

PATHOLOGY: In congenital ptosis there is evidence of true muscular dystrophy of the levator muscle as demonstrated by: loss of cross striations, random decrease in muscle fibre diameter, sarcolemmal retraction, proliferation of the sarcolemmal nuclei, fibrous and fatty tissue displacement of the striated fibres. The muscle fibres become atrophic, pale and flabby associated with hypertrophy and pseudohypertrophy in the same area[11].

Hyaline degeneration and vacuolisation was also seen. The intrafusal muscle fibres of muscle spindles are preserved after large portions of the muscle disappeared with an increase in the endomysial collagen (fibrous tissue in the muscular septa) and fat deposition within the muscle fibres[11].

B. SYNKINETIC PTOSIS :

Rarely, congenital ptosis can occur due to an aberrant innervation of the levator muscle by the mandibular branch of the trigeminal nerve, resulting in Marcus Gunn Jaw-Winking Syndrome. In this syndrome, there is a brisk upper eyelid retraction when the ipsilateral pterygoid muscle contracts during mastication, jaw thrusting to the contralateral side, jaw protrusion, chewing, smiling, or sucking.

PATHOLOGY: Histological sections from both sides of patients affected by MGP showed loss of muscle fibres in the affected levator muscle relative to the clinically normal side with central accumulation of mitochondria within individual muscle fibres.

Morphometry of the muscle fibres suggests that there is both atrophy and compensator hypertrophy of the remaining muscle fibres in the affected levator muscle.[3]

Muscle fibre type grouping suggests that the underlying process is a neurogenic atrophy. For example, in an area in which type 2 muscle fibres lose their nerve supply and are reinnervated by an adjacent

sprouting nerve that has a type1 pattern of electrical stimulation, then the previous type 2 muscle fibres undergo physiological and biochemical adaptation to become type 1 fibres as recognized by their histochemical staining reaction. This may result in focal areas assuming a monomorphic (rather than the normal speckled) type of fibre distribution.

C. APONEUROTIC PTOSIS:
Congenital aponeurotic defects result from a failure of the aponeurosis to insert on the anterior surface of the tarsus or from birth trauma following forceps delivery. The skin crease may remain normal or high depending on where the aponeurosis is affected. The levator function is usually good and there is no lid lag on down-gaze.

2. COMPLICATED CONGENITAL PTOSIS
A. ASSOCIATED WITH SUPERIOR RECTUS WEAKNESS :
Due to close embryological development of the levator and superior rectus muscles.

B. BLEPHAROPHIMOSIS SYNDROME:
is another rare autosomal dominant inherited condition. It presents with the characteristic features of epicanthus inversus, telecanthus, and ptosis. Non-ocular associations include ovarian failure, arched palate and cardiac defects.

PATHOLOGY: The most striking feature is thin, long anterior part of the LPS.

This consists of a disorganized, thin, long aponeurosis. However, in the posterior part of the LPS, organized thick structure suggestive of a muscle belly is seen. Histopathologic examination revealed posteriorly well-formed striated muscle fibers in patients with BPES. These striated muscle fibers are comparable to those of the normal tissue but are more intermixed with collagenous tissue and little fatty degeneration.

C. NEUROGENIC PTOSIS-HORNER'S SYNDROME:
Occurs in infancy, presents with ptosis, miosis, anhydrosis, and progressive heterochromia ipsilateral to the affected side. The lesion may occur anywhere along the oculosympathetic pathway.

D. CONGENITAL THIRD CRANIAL NERVE PALSY :
may be partial or complete. It may present with ptosis together with inability to depress, elevate, or adduct the eye. The pupil may be dilated.

3. OTHER CAUSES OF CONGENITAL PTOSIS
A. BIRTH TRAUMA
B. DUANE SYNDROME:
In this condition, the sixth cranial nerve fails to innervate the lateral rectus muscle. Instead, the muscle acquires an innervation from the third cranial nerve. Although the synkinesis produced does not involve lid innervations, enophthalmos with pseudoptosis may result.

C. PERIORBITAL TUMOR or other deep orbital tumors near the superior orbital fissure may produce proptosis with ptosis.

D. CONGENITAL FIBROSIS OF THE EXTRAOCULAR MUSCLES (CFEOM):
Is a non-progressive, autosomal dominant ocular disorder, resulting in fibrosis of the extraocular muscles. This disorder is characterized by bilateral ptosis and external ophthalmoplegia, with a compensatory backward tilt of the head. The pathophysiology of this disorder could be due to a primary neurogenic or myopathic etiology.

E. KEARNS-SAYRE SYNDROME:
is a mitochondrial deletion disorder that is characterized by progressive external ophthalmoplegia, heart block, retinitis pigmentosa, and central nervous system manifestations. This condition begins in childhood but is rarely present at birth. It generally becomes symptomatic in the first or second decade of life.

F. MYOTONIC DYSTROPHY:
is an autosomal dominant disorder characterized clinically by myotonia and progressive muscle weakness usually beginning in the muscles of the hands, feet, neck, or face and then progressing to other muscle groups, such as the heart.. Patients may present with polychromatic cataracts, gonadal atrophy, or premature thinning and/or loss of hair.Bilateral ptosis may be seen.

G. MYASTHENIA GRAVIS causes unilateral or bilateral fluctuating ptosis. This may be seen in children associated with extraoccular muscle palsies.

H. PSEUDOPTOSIS*:* -
Decreased orbital mass e.g., unilateral smaller eye, fat atrophy, blowout fracture may produce the appearance of ptosis secondary to the decreased volume of orbital contents.

ISOLATED ACQUIRED PTOSIS
A. APONEUROTIC PTOSIS: Is the most common cause of acquired ptosis.
- Involutional changes (degeneration of the levator muscle or thinning of aponeurosis) dehiscence, or disinsertion of the levator aponeurosis are common in old age.
- In younger patients, repeated manipulation of upper eyelid during contact lens wear may cause disinsertion of levator aponeurosis.
- Aponeurotic ptosis occurs frequently after ocular surgery (cataract surgery and glaucoma filtration surgery) and also following trauma. A variety of mechanisms may be responsible, such as direct injury to the aponeurosis, stretching or damage from postoperative swelling, use of rigid eyelid speculum, myotoxic effects of local anesthetics, or damage to the levator muscle from bridle sutures[12].
- The ptosis could be unilateral or bilateral and vary in severity.
- Either raised, multiple or absent upper lid crease due to slackened attachments of the aponeurosis to the tarsal plate and orbicularis oculi may be seen.There is also thinning of the upper eyelid and a deep superior eyelid sulcus.
- The levator function is essentially normal. Ptosis typically worsens in down-gaze and obstructs vision, especially when reading.

NON-ISOLATED ACQUIRED PTOSIS
B . NEUROGENIC PTOSIS: Can be congenital or acquired in origin.
- *Acquired Horner's syndrome* can be secondary to trauma, neoplastic insult, or vascular disease of the sympathetic pathway. All stigmata of congenital Horner's syndrome, excluding iris and areola hypopigmentation are present.
- *Raeder paratrigeminal syndrome* occurs in middle-aged men with daily ipsilateral headaches and the stigmata of acquired Horner's syndrome.
- *Dysfunction of the third cranial nerve* can result from a myriad of acquired insults. Trauma, multiple sclerosis, vasculopathy and infection are all potential etiologies.
- *Synkinetic neurogenic ptosis* is the product of innervational anomalies. Marcus Gunn jaw winking and post-traumatic ptosis are two examples of this.

C. MYOPATHIC PTOSIS: results from any pathology that affects the levator muscle.
- This is usually seen in mitochondrial myopathies, such as:
- *Chronic progressive external opthalmoplegia* presents with bilateral slowly progressive symmetric ptosis, orbicularis oculi weakness that prevents complete closure of the eyes and impaired eye movements.
- *Oculopharyngeal Muscular Dystrophy* is a rare, autosomal dominant myopathy that is characterized by late-onset ptosis, progressive tongue atrophy, dysphasia, dysarthria, and proximal lower extremity weakness.
- *Myotonic Dystrophy* is another autosomal dominant disorder with clinical findings of ptosis and muscle dystrophy of the face, jaw and neck. Other associated abnormalities include ophthalmoparesis, cataract, frontal balding, cardiac conduction defects and variable intellectual impairment.
- The levator muscle may also be damaged by a variety of inflammatory (sarcoidosis), infiltrative (lymphoid) or ischemic processes affecting the orbit.

D. NEUROMUSCULAR PTOSIS: Myasthenia gravis
- This is an autoimmune disorder whereby antibodies block, alter, or destroy the postganglionic acetylcholine receptors in the neuromuscular junction of skeletal muscles, thereby preventing muscle contraction.
- The ptosis may be unilateral or bilateral and frequently asymmetric. It may be isolated or associated with varying degrees of ophthalmoplegia, resulting in coexisting diplopia.

- The hallmark feature of myasthenia gravis is the fluctuating weakness and fatigability of the muscles, typically worsening with activity.
- Exaggerated ptosis with sustained upward gaze for at least 30 seconds, positive Cogan's upper eyelid-twitch sign and reversal of ptosis with rest and local application of ice pack may be appreciated.

E. MECHANICAL PTOSIS: occurs as a result of excessive weight, usually from a neoplasm on the upper eyelid, making it too heavy for the levator muscle to perform its function.

- The most common causes are benign or malignant neoplasm of the eyelid, such as a hemangioma, chalazion, neurofibroma and dermoid cyst with greater ptosis occurring in the area of the mass.
- Cicatricial changes in the tarsal conjunctiva and superior fornix following trachoma may result in a restrictive type of ptosis.
- Blepharochalasis is a rare condition of unknown etiology that affects young people and is usually hereditary. It manifests with repeated transient attacks of eyelid edema and erythema that starts around puberty and leads to ptosis during the attacks due to odema and later due to fibrosis that sets in.
- Ptosis can also be seen following enucleation because the absence of support to the levator by the globe permits the lid to droop
- Entrapment of the levator in orbital fracture or encroachment by an orbital foreign body may also mechanically interfere with the function of the levator leading to ptosis.

F. PSEUDOPTOSIS: is a form of ptosis that occurs due to abnormalities other than those found in the eyelid elevators. Pseudoptosis may be found on the side of the eye that is abnormal in size, shape, or position; for example, anophthalmos, microphthalmos and phthisis bulbi.

- Pseudoptosis may be seen in the contralateral normal eye of persons with unilateral lid retraction and proptosis from thyroid ophthalmopathy, as well as those with fixating hypertropic eye.

H. TRAUMATIC PTOSIS: Traumatic blepharoptosis may develop following an eyelid injury with ensuing damage to the lid elevator muscles, the levator aponeurosis or disruption of the neural input. Hence, eyelid trauma may lead to a myogenic, aponeurotic or a neurogenic ptosis.

G. BROW PTOSIS: Brow ptosis is a condition in which the eyebrow droops or sags. Laxity in the forehead muscles allows the eyebrows to fall. With this, the skin below the eyebrow also falls into the upper eyelid space, making the upper eyelid fold heavy. More common in above 50 years and those with dermatochalasis(a condition with redundant skin and muscle of the eyelid).

MATERIALS AND METHODS

This prospective study was conducted among patients presenting with ptosis to the department of ophthalmology, of a tertiary eye center in southern India and diagnosed on the basis of history, clinical symptoms and signs, and by pathological study. 66 eyes of 56 patients were studied during period of two years beginning from oct- 2017 to oct -2019.

Inclusion Criteria:

patients with ptosis, congenital or aquired which were managed Surgically and LPS muscle specimen subjected to histological examination.

Exclusion Criteria:

cases of ptosis non surgically managed like ocular Myasthenia, Honer's Syndrome ,Acute third nerve palsy, CPEO, Mitochondrial Myopathies.

PROCEDURE:

all the patients first underwent thorough clinical examination including history, general examination, ocular examination including measurement of amount and degree of ptosis, levator function, slit lamp examination, extraocular movements,and special tests performed where required to confirm diagnosis.

The surgical procedure was performed in each case depending on amount of ptosis and LPS action, and the excised LPS muscle, usualy varying in length from 10 mm to 20mm was excised and placed in bottle containing 10% formalin and taken to pathology lab, where cut sections were prepared and stained by haematoxylin and eosin.

EXAMINATION

The examination of a patient with ptosis should be proper and should aim at confirmation of the diagnosis and decision of treatment.

SIGNS

Examination starts from the moment a patient enters the op room head posture and face turn if any should be noted. Chin-up position is the most commonly encountered posture

- Frontalis overaction: patient may compensate the ptosis by lifting eyebrows.
- External examination should include the palpation of eyelids and orbital rim
- Features suggestive of BPES: Telecanthus, epicanthus inversus, hypoplasia of the superior orbital rims, horizontal shortening of the eyelids, ear deformities, hypertelorism, and hypoplasia of the nasal bridge[4]
- Cigarette paper appearance of lid skin due to recurrent edema occurs in blepharochalasis
- Proptosis or enophthalmos should be ruled out which may contribute to pseudoptosis. Exophthalmometry with Hertel's exophthalmometer is relevant in this regard
- Strabismus if present should be evaluated. Cover/uncover test should be done in all cases of ptosis. It should be noted that hypertropia can mimic ptosis
- The presence of lagophthalmos should be assessed-ptosis surgery can cause worsening of the same
- Bell's phenomenon: Grading should be done. Good bell's is always needed for the health of cornea after surgery[5]
- Lid position in downgaze: Lid lag in downgaze (higher position of upper eyelid in downgaze) in the absence of trauma/surgery is suggestive of dysgenesis of levator muscle (commonly in congenital ptosis) since the dysgenetic muscle is not able to relax properly[6]
- Synkinesis: The variation of the amount of ptosis with jaw movements is seen in Marcus Gunn jaw-winking ptosis, and variation in ptosis with ocular movements is noted in aberrant regeneration of the oculomotor nerve or the facial nerve, and some types of Duane's retraction syndrome
- Extraocular movements affected in CPEO, myasthenia as well as in the third nerve palsy
- Best corrected visual acuity and cycloplegic refraction should be done especially in children to asses amblyopia and visual problems
- Anterior segment examination with special emphasis on pupils: pupils affected in Horner's, third nerve palsy, etc.
- Posterior segment examination: abnormal retinal pigmentation seen in Kearn-Sayre syndrome
- Cogan's lid twitch: Elicited by having the patient look in downgaze, followed by upgaze. As the affected eye saccades up, the upper lid overshoots. Seen in myasthenia[6]
- Herring's law-Hering's law of equal innervation states that the reciprocal eye muscle of each eye is innervated equally. As such, manual elevation of the more ptotic eyelid decreases the muscle strength required to keep the lid elevated, and so the contralateral LPS relaxes and causes ptosis in the other eye.

MEASUREMENTS

- **Margin-reflex distance 1 (MRD1):** The distance between the central corneal light reflex and upper eyelid margin with eyes in primary position. Normal MRD 1 is 4–5 mm
- **Margin-reflex distance 2 (MRD2):** The distance between the central corneal light reflex and lower eyelid margin with eyes in primary position
- Palpebral fissure height (PFH): It is the distance between the upper and lower eyelid margins at the axis of the pupil.(MRD1 + MRD2 = vertical PFH)[5],[6]
- **Levator function:** Berke's method estimated by measuring the upper eyelid excursion, from downgaze to upgaze with frontalis muscle function negated and with the head positioned in the frontal or Frankfort plane. The amount of lid elevation is recorded in millimeters (mm) of levator function.
- The classification of levator function:
- Poor: 0–4 mm lid elevation
- Fair: 5–11 mm lid elevation
- Good: 12–14 mm lid elevation
- Normal: >15 mm lid elevation.
- **Iliff test:** It is used to assess levator function in infants. Upper eyelid of the child is everted as the child looks down. If the levator action is good, lid reverts on its own.
- **Margin Crease Distance (MCD):** Upper eyelid crease position is the distance from the upper eyelid crease to the eyelid margin. It is

normally 7–8 mm in males and 9–10 mm in females. High skin crease suggests aponeurotic defect. The depth of skin crease is a guide to determine the levator function in young children

- **Margin limbal distance (MLD):** Putterman's method-measurement of the distance between the middle of upper eyelid margin to the 6 o'clock limbus in extreme upgaze. Normal is about 9 mm

TESTS

- **Fatigue test:** MRD1 should be measured first. Then the patient should be asked to look up for 2 min after which the MRD 1 is to be measured again. Worsening of ptosis is seen in myopathies, myasthenia as well as senile aponeurotic ptosis
- **Ice test:** Glove containing ice pack is applied on the closed ptotic eye for 2 min. If the lid elevates by 2 mm or more, it is suggestive of myasthenia
- **Tensilon test:** In cases of suspected myasthenia, 2 mg of edrophonium is injected slowly in 15–30 s. The needle is left *in situ*, and the remaining 8 mg is injected slowly if no adverse reaction is observed within 1 min. If myasthenia is the cause, ptosis improves after the injection[1]
- **Phenylephrine test:** Sympathomimetic agents, such as phenylephrine or apraclonidine, can be instilled under the eyelid to test the function of Muller's muscle
- **Schirmer's test:** To evaluate tear function
- Tear breakup time in individuals suspected of having dry eyes due to the potential risk of incomplete eyelid closure and exposure keratopathy following surgical correction
- **Corneal sensitivity** should be tested in all cases.

MANAGEMENT OF PTOSIS

NON SURGICAL MANAGEMENT Mild Congenital Ptosis: Routine monitoring every 3-12 months for signs of amblyopia, strabismus, and abnormal head postures; surgery may be indicated if these signs are present.

Ptosis In Ocular Or Generalized Myasthenia Gravis: Responsive to medical therapy (cholinesterase inhibitors, corticosteroids, azathioprine and diaminopyridine)

Chronic Progressive External Opthalmoplegia: Managed with crutch spectacles

SURGICAL MANAGEMENT

Basically two types of surgeries are done to correct ptosis:

I. shortening/tightening of levator-muller-tarsal complex:
   1. Fasanella-servat : muller-tarsus complex is resected
   2. Levator resection
   3. Reinsertion of levator aponeurosis
   4. Resection of muller's muscle and conjunctiva

II. Pulling the lid up to the frontalis at brow:
   1.Frontalis sling procedure

Congenital Ptosis with Strabismus: Surgical correction of strabismus prior to ptosis surgery. Ptosis correction usually performed before amblyopia sets in.

Mild Ptosis, Levator Function > 10 mm: Müller's muscle-conjunctival resection[9] or Fasanella-Servat procedure[7,8] (The upper border of the tarsus is excised with the lower part of muller's and overlying conjuctiva)

Moderate Ptosis, Levator Function 5 – 10 mm: Levator palpebrae superioris resection

There is no exact amount but the approximate amounts are:

a) 3-4mm of resection corrects 1mm of ptosis

b) Atleast 10mm of resection is required to correct congenital ptosis

c) In old people where ptosis varies diurnally not more than 8mm should be resected

d) According to LPS function :

LPS 8-10mm : 10-18mm resection
6-7mm : 18-20mm
4-5mm : 22-26mm

e) If superior rectus is weak, an additional of 4mm is to be resected.

Severe Ptosis, Levator Function < 5 mm: Brow-frontalis suspension[9]

PRINCIPLE: This surgery connects the eyelid to the brow with a sling material and utilizes the power of the frontalis muscle to elevate the poorly functioning eyelid.

- Most of the techniques are based on multiple cutaneous stab incisions at the level of the tarsus and the eyebrow, through which the sling material is passed in a sub-orbicularis plane.
- There are various suture designs: a single triangle, double triangle, single rhomboid (Friedenwald-Guyton procedure), double rhomboid (Iliff procedure), double trapezoid (Wright procedure), single pentagon (Fox procedure), and double pentagon configurations (Crawford procedure).
- Although fascia lata is most commonly used due to its long-lasting effect and low rate of complications, it has several limitations such as difficulty of harvesting, insufficient amounts in small children, and postop donor-site complications.
- Other sling materials are polypropylene suture, nylon suture, silicone, mersilene mesh, gore-tex but these materials have a higher risk of extrusion, infection, granuloma, formation and breakage after trauma.

INDICATIONS:

- ptosis with poor levator muscle function. i.elevator function of 4 mm or less.
- chronic progressive external ophthalmoplegia (CPEO)
- muscular dystrophy
- third nerve palsy
- myasthenia gravis
- aponeurotic ptosis in elderly patient, associated with a poor Bell's phenomenon

RESULTS

Table 1: Age & Sex Distribution

AGE	Congenital PTOSIS			Acquired PTOSIS		
	Male	Female	Total	Male	Female	Total
0-19y	18	12	30	-	-	-
20-39y	8	8	16	-	-	-
40-59y	-	-	-	2	2	4
>60y	-	-	-	3	3	6
TOTAL	26	20	46	5	5	10

- Congenital ptosis was common among first 2 decades in males and females
- Males (56.5%) were slightly more commonly affected than females (43.4%)
- among congenital ptosis.
- Aquired ptosis was observed after 4th decade and incidence increased with age.

Table 2 : Involvement Of Eye

	RE	LE	BE
CONGENITAL	14	27	5
ACQUIRED	3	6	1
TOTAL	17	33	6

- Unilateral ptosis(89.2%) was more common tham bilateral ptosis (10.7%), in both congenital and aquired type.
- Among unilateral ptosis, LE is more commonly involved (60%).

Table 3: Types Of Congenital PTOSIS

Type Of PTOSIS	Unilateral	Bilateral	Total
SIMPLE CONGENITAL	32	4	36
COMPLICATED CONGENITAL	9	1	10

- Among congenital ptosis, simple (i.e) dystrophic type was the most common (78.2%) compared to complicated type (21.7%).
- Complicated congenital ptosis comprised of synkinetic ptosis with marcusgunn phenomenon 4 cases, ptosis with double elevator palsy 2 cases, Congenital neurogenic ptosis 1case, Congenital ptosis with superior rectus weakness 2 cases,

Table 4: Types Of Acquired PTOSIS

Type Of PTOSIS	Unilateral	Bilateral	Total
MYOGENIC	5	1	6
NEUROGENIC	1	-	1
MECHANICAL	2	-	2
TRAUMATIC	1	-	1
TOTAL	9	1	10

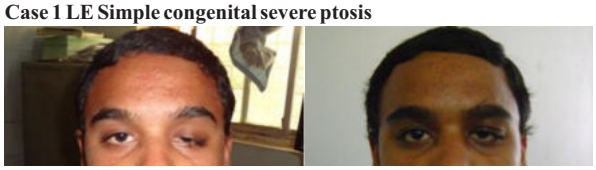
- Among aquired ptosis, most common was myogenic/ aponeurotic type (60%), mechanical ptosis (20%) due to infiltration of neurofibromatisis and inflammatory infiltrate in blepharochalasis, neurogenic (10%), traumatic (10%).

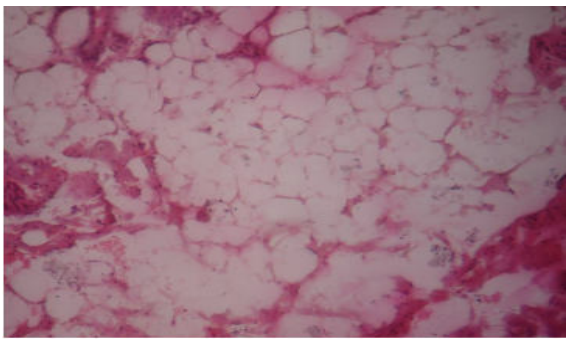
Table 7: Histopathology Of LPS

SEVERITY OF PTOSIS	CONGENITAL		AQUIRED	
	FINDINGS	NO OF CASES	FINDINGS	NO OF CASES
MILD	Dystrophic changes with moderate amount of fibrous and fatty component	10	-	-
MODERATE	Dystrophy with areas of fibrocollagenous tissue with moderate amount of fatty infiltration	19	Atrophic muscle fibres with mild fatty infitration	4
SEVERE	Atrophic muscle, with most of muscle replaced by adipose tissue	17	Atrophic smooth muscle fibres with few skeletal muscle fibres at periphery and mild fatty infiltration	4
			Bundles of neoplastic nerve fibers with wavy nucleus intermixed with collagen in a myxomatous matrix and fibroadiopose tissue. Suggestive of neurofibroma.	2

- Congenital simple ptosis, the histology of LPS was consistent with dystrophic features of fibrocollagenous tissue replacing normal skeletal muscle along with fibrofatty infiltration that increased as the severity of ptosis increased.
- The presence of fibrous tissue accounts for lid lag in congenital simple ptosis
- Aponeurotic ptosis, LPS showed atrophy of muscle with mild fatty infiltration without any fibrous component[12] corelating with lack of lid lag in aponeurotic ptosis.
- Among 2 cases of plexiform neurofibromatosis, LPS muscle showed infiltration with neoplastic nerve fibers with wavy nucleus intermixed with collagenous matrix

Case 1 LE Simple congenital severe ptosis

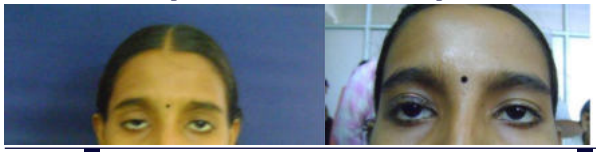


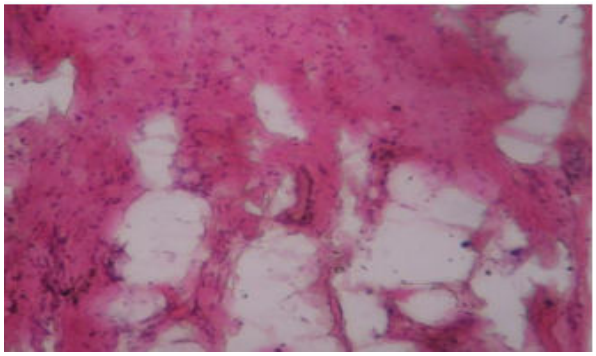


HPE- LPS shows fibrosis and fatty infiltration

Case 2 RE Simple Congenital Moderate Ptosis

Pre op **Post op**

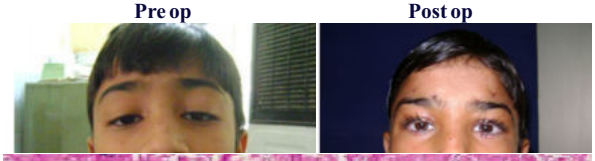


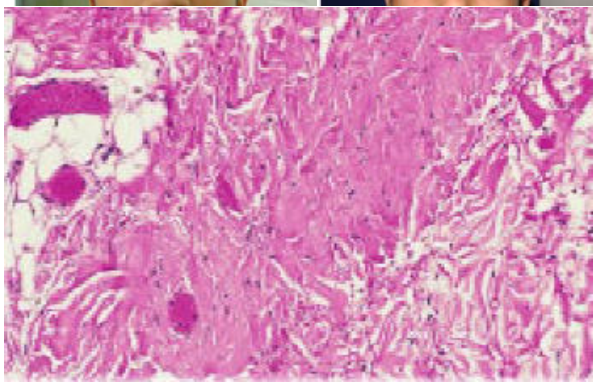


HPE- LPS Shows Fibrocollagenous Infiltrate

Case 3 Bilateral Simple Congenital Severe Ptosis

Pre op **Post op**





HPE -LPS Showing Fibrocollagenous Infiltration And Fatty Infiltration

DISCUSSION

EPIDEMIOLOGY OF PTOSIS

In our study congenital ptosis was the commonest type - 46 cases (82%) and acquired variety of ptosis comprised 10 cases (18%). Congenital ptosis commonly presented in the first two decades of life (65%) both in males and females.The mean age of incidence was 15 yrs. Males (56.5%) were slightly more commonly affected than females(43.4%)Acquired ptosis presented mostly after 4thdecade and the incidence increased with age. There was no sex predilection among the acquired type.

Unilateral ptosis was more common in both congenital (89.1%) and acquired (90%) types. Among unilateral cases LE was predominantly involved(27 cases (66%) in congenital and 6 cases (60%) in acquired). RE involvement included 14 cases (34%) in congenital and 3 cases (30%) in acquired type. Bilateral involvement was 10% in both types.

ETIOLOGY OF PTOSIS

Among congenital ptosis, simple (i.e) dystrophic type was the most common (78.2%) compared to complicated type (21.7%). Complicated congenital ptosis comprised of synkinetic ptosis with marcusgunn phenomenon (4 cases) , Congenital ptosis with double elevator palsy (2 cases), congenital ptosis with superior rectus weakness(1 case).

Among the cases with acquired ptosis, 6 were myogenic/ aponeurotic (60%), 2 were mechanical due to plexiform neurofibromatosis (20%), 1 each was traumatic and neurogenic(longstanding 3rd nerve palsy (10%).

HISTOTOPATHOLOGY OF PTOSIS

All the cases of congenital simple ptosis showed dystrophic changes like fibrocollagenous tissue replacing normal skeletal muscle, decrease in the diameter of fibres, fibrous and fatty infiltrate. Fatty infiltrate increases as the severity of ptosis increases. The presence of

fibrous tissue accounts for lid lag in congenital simple ptosis.

Aponeurotic ptosis, LPS showed atrophy of muscle with mild fatty infiltration without any fibrous component corelating with lack of lid lag in aponeurotic ptosis.

CONCLUSION

Commonest type of ptosis we came across in this study is congenital ptosis.

Ptosis was more commonly observed in males than in females.Patients with ptosis more commonly presented among the 1st and 2nd decades of life.Unilateral ptosis was more common in this study with left eye involved predominantly.

Among the congenital ptosis,simple congenital ptosis(dystrophic type)is more common.

There was acorrelation between LPS function and amount of fibrofatty infiltrate, in simple congenital ptosis.Eyes with poor LPS function(85% of lids examined)show severe fibrosis.Predominance of fibrous tissue infiltrate might be responsible for lid lag in simple congenital ptosis. LPS resection gave very good results in mild to moderate ptosis and brow suspension was prefered in severe ptosis with poor levator action.

Among the acquired ptosis,aponeurotic type is the most common one we came across.

Fatty infiltrate is the commonest finding in aponeurotic dehiscence which also explains the lack of lid lag in aponeurotic type of ptosis. Managed surgically by LPS plication which gave good results.

Further investigations such as immunohistochemistry is required in order to know the exact correlation between fibrosis and LPS function.

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