ISSN No:-2456-2165

Clinical Profile of Ptosis in a Tertiary Care Centre

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Abstract:-

- ***Aims:***

To find out common types and causes of ptosis in a given population, its ocular associations and to see if there are any preventable causes for ptosis.

- ***Methods and Material:***

A retrospective analysis of 99 patients with ptosis seen in ophthalmology OPD of a tertiary care centre. A total of 85 patients were finally included in the study excluding pseudoptosis and those without final diagnosis. Detailed history, clinical ocular examination had been conducted as per predefined proforma.

- ***Results:***

The commonest age of presentation was in the first two decades of life while most common age of onset of ptosis was before the age of 1 year. Males (63.52%) were more affected than females (36.47%) (1.7:1 ratio).Unilateral ptosis(64.70%)(Left>Right Sided)was more common than bilateral(35.29%) ptosis. Maximum patients had congenital ptosis (62.35%) as compared to acquired ptosis(37.64%). Myogenic ptosis(congenital and acquired)(54.11%)was most common; followed by neurogenic(19.9%);traumatic(15.29%) and aponeurotic(10.58%).The causes of neurogenic ptosis were third nerve palsy – congenital as well acquired like synkinetic ptosis, myasthenia gravis and congenital fibrosis of extra-ocular muscles (CFEOMs)(Type1).

- ***Conclusions:***

Though congenital ptosis was the most common type of ptosis seen in clinical practice, one should keep in mind that the acquired cases especially the neurogenic and traumatic cases are the ones which are going to require more intensive evaluation and investigations.There is no single step approach to a patient with ptosis; each type has its unique properties and hence a unique approach.

Keywords:- *Causes, Ptosis, Types.*
Key Messages:- *Though congenital cause is most common, acquired causes of ptosis must be be ruled out.*

I. INTRODUCTION

Blepharoptosis or ptosis is drooping or inferior displacement of the upper eyelid.[1]It is a common cause of reversible peripheral vision loss in adults, but may result in the development of amblyopia in children.[1][2] The prevalence of ptosis has been found to be ranging from 2.23% (95% CI: 1.73 – 2.74)[3] to 13.5% (95% CI: 12.1 – 14.9)[4] in adults whereas the prevalence of childhood ptosis is estimated at 1:10,000.[5] In a study in the rural population in Maharashtra, India the prevalence of ptosis amongst school children was found to be 0.58%.[6]

Ptosis can be congenital or acquired. It has been classified by Frueh according to causative mechanism as neurogenic, myogenic , aponeurotic and mechanical[7]. Beard classified ptosis in a more descriptive manner as due ptosis due to levator maldevelopment and other myogenic causes, aponeurotic, neurogenic, mechanical and apparent ptosis[8]. Congenital ptosis of the simple type (myogenic variety) is usually the most common type of ptosis seen[9].

Despite being mostly a non-progressive disease, congenital ptosis can cause cosmetic, functional and psychosocial problems in children. It has been reported that the prevalence of amblyopia, strabismus and refractive errors among patients with congenital ptosis were much higher than those among the general population.[2],[10] Early diagnosis and treatment of congenital ptosis will contribute to prevention and management of these ocular abnormalities.

Not only does ptosis have a significant effect on vision, acquired ptosis can also be the presenting manifestation of many more serious neurological diseases. Keeping this in mind, this study was designed to find out common types and causes of ptosis in the given population.

II. SUBJECTS AND METHODS

A retrospective analysis of 99 patients with ptosis seen in ophthalmology OPD of a tertiary care centre from January 2010 till October 2019 was carried out. Those without a final diagnosis or with pseudoptosis were excluded. A total of 85 patients were finally included in the study. Detailed history, clinical ocular examination had been conducted as per predefined proforma.

ISSN No:-2456-2165

Unilateral ptosis was defined as either a measured palpebral fissure asymmetry of ≥ 1mm between the two upper eyelids or a marginal reflex distance (MRD) of < 2.5 mm. Bilateral ptosis was defined as a MRD of < 2.5 mm in both eyes.[5]

Ptosis was classified as myogenic (congenital and acquired), aponeurotic, neurogenic (congenital and acquired), mechanical and traumatic.[1]Patients were considered to have a congenital form of ptosis if they presented to a physician within the first few months of life unless an acquired etiology was specifically noted in the chart. Late-presenting cases were deemed congenital if symptoms were observed within the first few months of life and verified by a photograph or parental history. If above history was absent then patient was considered to have acquired ptosis.[5]Ptosis due to congenital fibrosis of extra-ocular muscles (CFEOMs) has been included as neurogenic in origin considering that this entity is now included under the category of Congenital Cranial Dysinnervation Disorders.[11]

Amount of ptosis was calculated by deducting the Margin reflex distance 1 (MRD1) of the affected eye from that of the normal eye in unilateral ptosis and from an assumed normal MRD1 of 4.5 mm in case of bilateral ptosis. Mild ptosis was defined as a difference of 2mm, moderate as a difference of 2- 4mm and severe as a difference of more than 4 mm.

Pseudo ptosis was defined as that ptosis when seen in the following situations - contralateral eyelid retraction or proptosis, hypotropia, hypoglobus, microphthalmos, enophthalmos, pthisis bulbi, aberrant re-innervation of facial nerve.[1]

The study protocol was approved by the Institutional Ethical Committee and adhered to the tenets of the Declaration of Helsinki.

III. RESULTS

85 patients were included in this study. The commonest age of presentation to the ophthalmologist was in the first two decades of life (23 patients / 27.05% before 10 years of age; 18 patients / 21.17% between 11 – 20 years of age), while 53 (62.35%) patients or their caretakers gave a history of noticing that the onset of the ptosis before the age of one year.(Table 1)

Table No 1 – Distribution of ptosis patients as per age of onset and age at diagnosis

Age of patient (Years)	No. of patients (%) At Onset	No. of patients (%) At Diagnosis
< 1 year	53(62.35)	2(2.35)
1-10	5(5.88)	23(27.05)
11-20	5(5.88)	18(21.17)
21-30	6(7.05)	18(21.17)
31-40	1(1.17)	7(8.23)
41-50	2(2.35)	1(1.17)
51-60	7(8.23)	8(9.41)
61-70	6(7.05)	8(9.41)
Total	85	85

Males (54/63.52%) were more affected than females (31/36.47%) in the ratio of 1.7:1.
55 patients (64.70%) had unilateral ptosis while 30 patients (35.29%) had bilateral ptosis. Of the 55 patients with unilateral ptosis, 33 (60%) had left sided ptosis and 22 (40%) had right sided ptosis.
Maximum patients had congenital ptosis (62.35%) as compared to acquired ptosis (37.64%). (Table 2)

Table No 2 – Distribution of patients as per type of ptosis

Type of ptosis	No. of patients (%)
Congenital	53(62.35)
Acquired	32(37.64)
Total	85

Myogenic ptosis, congenital (51.76%) as well as acquired (2.35%) were the maximum in number in 46 (54.11%)patients ; neurogenic in 17(19.99%) patients; aponeurotic in 9 (10.58%) patients, and traumatic in 13 (15.29%) patients. (Table 3)

Table No 3 - Distribution of patients as per cause of ptosis

Cause of Ptosis	Sub type	Total no (%)	Total No of Patients (%)
Myogenic	Congenital	44 (51.76)	46 (54.11)
	Acquired	2 (2.35%)	
Aponeurotic	Congenital	0	9 (10.58)
	Acquired	9 (10.8%)	
Neurogenic	Congenital	9 (10.58%)	17 (19.99)
	Acquired	8 (9.4%)	
Mechanical	-	0	
Traumatic	-	13 (15.29%)	13(15.29)
Total		85	85 (100)

The causes of neurogenic ptosis were third nerve palsy – congenital (2 /85; 2.35%)as well acquired (4 /85; 4.70%); synkinetic ptosis with Jaw Winking phenomenon (2 /85; 2.35%), myasthenia gravis (4 /85; 4.70%) and five (5.88%) cases of congenital fibrosis of extra-ocular muscles (CFEOMs)(Type1).

The average age for the presentation of myogenic ptosis was 19.81 years (range: 8 months – 70 years and the M:F ratio was 1.87:1; that of neurogenic ptosis was 25.29 years (range from 2 – 60 years) and M:F ratio was 1.42:1; of aponeurotic ptosis was 60.33 years (range 30 – 70 years) and M:F ratio was 2:1 while that of traumatic ptosis was 37.07 years (range: 10 – 65 years) and the male: female ratio was 1.6:1.

IV. DISCUSSION

Our study of 85 patients was done to find out which are the commonest types of ptosis and their causes seen in our population. Presence of ptosis is usually accompanied by some ocular or systemic association; if nothing serious at least an anomalous head posture in the form of a chin elevation or an elevation of the brow as a compensatory mechanism. Ptosis since early childhood can be an indicator of the presence of amblyopia or co – existing strabismus. Similarly a recent sudden onset ptosis can be a presenting sign of a neurological disorder like a third nerve palsy due to an intra-cranial lesion. A gradually progressive ptosis in a teenager could indicate the onset of chronic progressive external ophthalmoplegia and its attendant systemic complications.

Majority of the patients who presented to us were within the second decade of life. Almost similar findings were seen in a study conducted by Baiyeroju et al[12], in which 52% of patients were less than 16 years of age[13] and by Sarika et al[13] , in whose study, most of the ptosis patients (206/490; 42%) were in the age range of 1-19 years.

Our study showed a tendency towards involving males as compared to females. Lee YounGonet al in a Korean study with a large study population and spread over 24 years also showed a similar tendency.[14]Sarika et al[13] also had a slightly higher male preponderance, whereas Baiyeroju et al[12] showed an equal 1:1 male to female ratio. Another large scale Korean study on childhood ptosis showed that males (52.8 % ± 0.6) were more affected by ptosis as compared to females.[15]

Unilateral ptosis was seen predominantly in our series. It was seen in 64.70% patients and this was comparable to the other series[5],[9],[13]. 60% of the patients with unilateral ptosis had affection of the left side. Left eye predominance was noted for the first time by Griepentrog et al who noted a $p<0.001$ in their study[5]. Lee Youn Gon et al have also noted the same observation[14]. This left sided dominance has been postulated to be due to underlying failure of proper motor innervations[5]; similar to the congenital miswiring of the medial and lateral rectus muscles associated with an absent or hypoplastic abducens nerve seen in Duane's retraction syndrome another congenital disorder with a propensity to affect the left side[16].

Congenital ptosis (62.35%) was much more common than acquired ptosis (37.64%) in our study. Lee younGonet al[14] in their study of 2,328 patients had 78% patients with congenital ptosis and only 22% with acquired ptosis. Others have also shown similar findings.[12],[13],[15]

Our study classified ptosis according to the mechanisms as myogenic, aponeurotic , neurogenic, mechanical and traumatic[1],wherever applicable they were further subdivided as congenital or acquired. In our series of 85 patients, only the myogenic and neurogenic types were further subdivided in to congenital and acquired. Most of our patients (51.76%) were of the myogenic variety, that too of the congenital type. This is also referred to as simple congenital ptosis[12] or isolated simple congenital ptosis.[11]Simple congenital ptosis has been found to be the most common type of congenital ptosis; Griepentrog et al - 81 / 96 (75%)[3]; Lee YounGonet al[12] -1,715 / 1,815 (73.7%); Baijeroyu et al – 14 / 25(56%)[13].Only two patients of the myogenic variety had chronic progressive ophthalmoplegia (CPEO). The mean prevalence of CPEO has been estimated to be about 1 in 30,000 in the general population[17] and bilateral and often asymmetric ptosis can be its presenting complaint[18]

We saw a considerable number of patients with neurogenic ptosis at 19.99%. Special attention is required in these patients because of associated debilitating neurological problems. There were an almost equal number of congenital and acquired cases in our study. Our study had a slightly higher number of neurogenic cases compared to other studies; comparable only to Sarika et al (22.7%)[13]. The prevalence in other studies ranged from Lee Yong Gun et al[14]- 1.9%, Lim et al[19] – 5.6%, Lee V et al [9] - 10.88% to and Thapa et al[20] - 15.2%. The causes of neurogenic ptosis were similar to other studies.[13],[18],[19] and included third nerve palsy – congenital as well acquired; synkinetic ptosis with Jaw Winking phenomenon, myasthenia gravis and five cases of congenital fibrosis of extra-ocular muscles (CFEOMs)(Type1).

The prevalence of CFEOMs has been found to 1/230,000 cases[21]; but the number of cases of CFEOMs were higher than expected as we had a family present to us consisting of the proband, his mother, brother, maternal uncle and his daughter in our case series with an autosomal dominant inheritance. CFEOMs is now classified as to be a part of the congenital cranial disinnervation disorders and hence ptosis due to this entity is classified as neurogenic in origin[11]. It is distinguished from congenital third nerve palsy from the comparatively more profound limitation of ocular movements which are seen in CFEOMs especially of upgaze. Perverse convergence is also noted in patients with CFEOMs on attempting up gaze.

Myasthenia gravis is clinically a very important cause of neurogenic ptosis to be kept in the differential diagnosis. Patients present with variable asymmetric ptosis and non -

specific diplopia which is due to external ophthalmoplegia not conforming with any typical ocular nerve palsy. Though generalized myasthenia gravis (MG) is usually diagnosed in females, pure ocular myasthenia gravis(OMG)is seen in males above the age of 40 years[22]. Diagnosis of myasthenia gravis is aided by a history of improvement in ptosis with rest, ice pack test and eliciting signs like the peek sign and Cogan's lid twitch sign[22]. In our series, of the four patients three were females and one was a young boy of ten years. All patients had come with ocular myasthenia gravis. Female preponderance was also noted by Chithra et al in a study of 40 patients of Myasthenia gravis in South India[23]. Of their 40 patients with myasthenia gravis, 16 had OMG.

We had two children with congenital third nerve palsy. It was seen in 39% of the cases of third nerve palsy in a tertiary care centre[23]. The same authors noted that children with congenital etiology (without associated CNS disease) improved on amblyopia treatment. Both our patients had mild amblyopia and they responded to patching. All the cases of acquired third nerve palsy in our study were pupil sparing – due to presumed microvascular disease. The common causes of acquired third nerve palsy are presumed microvascular (42%), trauma (12%), compression from neoplasm (11%), post-neurosurgery (10%), compression from aneurysm (6%), other cause (5%), stroke (4%), undetermined (4%), pituitary apoplexy (2%), Tolosa-Hunt syndrome (2%), and giant cell arteritis (1%)[24].

Though it is important to pay attention to the involvement of the pupil, anisocoria alone cannot distinguish the cause of the third nerve palsy. Fang et al noted that 16% of their patients with presumed microvascular disease as etiology of the third nerve palsy had pupil involvement[24]. Pupil involving third nerve palsy is usually due to compression of the pupillary fibers lying on the top portion of the nerve supplying due to a space occupying lesion in the brain like an aneurysm of the posterior communicating artery. Pupil sparing third nerve palsy is due to the atherosclerotic changes in the vasonervorum – the small blood vessels supplying the nerve itself.

Aponeurotic ptosis was seen in 9 (10.58%) patients in our series. Majority of the studies had a similar prevalence of aponeurotic ptosis (Lee Yong Gun et al[14]– 11.5% and Thapa et al[20]– 10.7%.) or even lesser (Lee V et al[9]). The exception was a study by Lim et al[19] who had a very high prevalence of aponeurotic ptosis at 60.2%. They have explained this on the basis of the higher average age in their study of the presenting patients with aponeurotic ptosis at 62 years[19]. Balasubrahanian et al also had a slightly higher prevalence of aponeurotic ptosis in their study at 23.85%.[25]

Traumatic ptosis was seen in 15.29% cases in our series. In our study it is the third most common cause of ptosis after myogenic (predominantly of the congenital variety) and neurogenic. It was the second most common cause in a study by Lim et al[19] and most common in a study by Thapa et al[19]. The average age of presentation of was 37.07 years(range: 10 – 65 years) and the male: female ratio was 1.6:1. Ocular manifestations of trauma are predominantly seen in younger males, be it traumatic ptosis, traumatic hyphaema[23]or traumatic optic neuropathy[24]. The demographic profile of our patients fit the norm[25].

Almost all our patients had ptosis due to injury to the lid – levator muscle or aponeurosis, while one patient had traumatic ptosis due to injury to third nerve during surgery for pituitary macroadenoma. Traumatic ptosis could be due to variety of mechanisms – myogenic, aponeurotic, neurogenic, mechanical and mixed mechanism25. Jacobs et al[25]have encouraged classification of traumatic ptosis along the above sub-classification as they have noted that there are meaningful differences in presentation depending on the underlying mechanism. They have further made recommendations for management and prognostication as per the subtype[25].Transient isolated traumatic neurogenic ptosis after mild head trauma has also been reported[26]. The fallacy of our study was that we have not subclassified all the cases of traumatic ptosis.

We had no case of mechanical ptosis in our study.

V. CONCLUSION

Our study showed that though congenital ptosis was the most common type of ptosis seen in clinical practice, one should keep in mind that the acquired cases especially the neurogenic cases are the ones which are going to require more intensive evaluation and investigations. Traumatic ptosis has to be evaluated in detail so that we can provide the best possible cosmetic and functional prognosis to the patient. There is no single step approach to a patient with ptosis; each type has its unique properties and hence a unique approach.

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