A RARE CASE OF RIGHT SIDED CONGENITAL PTOSIS DUE TO FOCAL DEHISCENCE OF THE CRIBRIFORM PLATE

Dr. Anoop Singh Gurjar*¹, Dr. Manish Parakh², Dr. Manisha Gurjar³, Dr. Sushma Kushal Kataria⁴, Dr. Poonam Parakh⁵,

¹Assistant Professor, Dept. of Anatomy, Dr. S.N. Medical College, Jodhpur, Rajasthan
²Professor, Dept. of Pediatric Medicine, Dr. S.N. Medical College, Jodhpur, Rajasthan
³Assistant Professor, Dept. of Biochemistry, Dr. S.N. Medical College, Jodhpur, Rajasthan
⁴Professor, Dept. of Anatomy, Dr. S.N. Medical College, Jodhpur, Rajasthan
⁵Assistant Professor, Dept. of Obs. and Gyn., Dr. S.N. Medical College, Jodhpur, Rajasthan

*Correspondence for Author
Dr. Anoop Singh Gurjar
Assistant Professor, Chief Warden Bungalow, Near Medical Boys Hostel, M.D.M. Hospital Campus, Jodhpur, Rajasthan, India.

ABSTRACT
Ptosis results from a congenital or acquired weakness of the levator palpebrae superioris (LPS) and the Muller’s muscle responsible for raising the eyelid, damage to the nerves which control those muscles or laxity of the skin of the upper eyelids. Ptosis may be isolated or may signal the presence of a more serious underlying neurological disorder. Here we report a case of congenital ptosis occurring due to focal dehiscence and cephalocele of the cribriform plate. The case is being reported due to rarity of this etiology and we hope that this will add to the existing knowledge and list of causes of congenital ptosis.

KEYWORDS: Ptosis, Congenital, Dehiscence, Cribriform, Cephalocele.

INTRODUCTION
Ptosis also known as blepharoptosis refers to a clinical condition in which there is drooping or falling of the upper or lower eyelid. In severe cases the drooping of eyelid can cover part or entire pupil and may interfere with the vision.[1, 2] In cases of uncorrected congenital ptosis, the pediatric patient may develop amblyopia and suffer lifelong visual impairments. Ptosis usually results from under action of the eyelid protractors relative to the eyelid retractors,
causing the eyelid to be lower than its normal anatomic position. It can occur in isolation or may be an early presenting symptom of a serious underlying disease.

The closure of the eyelids is facilitated by the protractors of the eyelids i.e. circumferential orbicularis oculi muscle, which is innervated by the facial (seventh cranial) nerve. The elevators of the upper eyelid are the LPS and the Muller’s muscle. The LPS is the main upper eyelid elevator and is innervated by the oculomotor (third cranial) nerve. The 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 is innervated by the sympathetic nervous system. The muscle is responsible for the over-elevation of the eyelid when a patient becomes excited or fearful and leads to mild ptosis with fatigue or inattention.

Ptosis can be broadly classified into congenital and acquired depending on the age of presentation.[3] Ptosis is considered congenital if present at birth or diagnosed within first year of life and acquired if it is diagnosed after 1 year of age. It is usually an isolated presentation but in certain cases it may signal the presence of a more serious underlying neurological disorder. In a study from Nigeria most common cause of ptosis was congenital (56%) and in most of the cases it was unilateral.[4] Ptosis can also be classified into aponeurotic, neurogenic, myopathic, neuromuscular, neurotoxic, mechanical, traumatic, and pseudoptosis. This classification provides an insight into the mechanical basis of ptosis.

Mostly congenital ptosis is isolated and includes Simple congenital ptosis due to myogenic dysgenesis of the levator muscle. In these cases, rather than having normal muscle fibers, fibrous and adipose tissue are present in the muscle belly instead. Thus there is a reduction or absence of functional muscle, impairing the ability of the levator to contract and elevate the eyelids.[5]

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 or Synkinetic ptosis. 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. This phenomenon is discovered early as the infant is bottle or breastfed.[6]
Congenital aponeurotic ptosis may also 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.

Non-isolated congenital ptosis includes Blepharophimosis Syndrome, Congenital Third Cranial Nerve Palsy (may be partial or complete) and Neurogenic Ptsis-Horner’s Syndrome. Due to close embryological development of the levator and superior rectus muscles, congenital ptosis may also be associated with superior rectus weakness. Other cause of ptosis includes Duane syndrome, periorbital tumor or other deep orbital tumors, congenital fibrosis of the extraocular muscle, Kearns-Sayre syndrome, Myasthenia gravis and pseudoptosis. [7]

In the context of the aforementioned discussion we present a case of congenital ptosis occurring as a result of focal dehiscence of the cribriform plate with small cephalocele. The case is being reported due to rarity of this etiology and we hope that this will add to the existing knowledge and list of causes of congenital ptosis.

**CASE REPORT**

A seven year old Indian male child presented with hyperactivity and drooping of right eyelid appreciated during early infancy and currently hampering his vision from the right eye. He had no history of head trauma, absence of sweating on face, squint, diplopia or diurnal variation in the drooping of eyelid. He was born at term to non-consanguineous healthy parents, third gravida mother by vaginal delivery with insignificant perinatal, natal and family history. He had an age appropriate neurodevelopment and attending kindergarten. On examination he had anemia, was fidgety and hyperactive, well oriented, pupils were 5 mm on both the sides with brisk direct light reaction and had no relative afferent papillary defect, had right sided ptosis and mostly kept his neck in right lateral flexion, eye movements were full in all cardinal gaze directions, had no esotropia or exotropia, had no facial asymmetry, had normal gag, bilaterally symmetrical palatal arches with no pooling of secretions and no atrophy or fasciculation of tongue. Had normal motor and sensory system examination with no signs of meningeal irritation, cerebellar or autonomic dysfunction.

Routine laboratory investigations including complete blood counts, Random Blood Glucose, Blood Urea, Serum Creatinine, AST, ALT, Blood T3, T4 and TSH were within normal limits. On MR imaging the coronal images through anterior orbit revealed apparent focal dehiscence
of the right cribriform plate and minor abnormal inferior displacement of right gyrus rectus compared to the left side raising suspicion of small cephalocele. The brain parenchyma including both cerebral hemisphere, cerebellum and brain stem was normal.

Figure No.: 1: Seven year old patient with right sided congenital ptosis.

Figure No.: 2: MR imaging through anterior orbit (coronal section) showing a dehiscence of the cribriform plate on the right side

DISCUSSION
The orbits are conical or four-sided pyramidal cavities which open into the midline of the face and point back into the head. Each consists of a base, an apex and four walls. The apex lies near the medial end of superior orbital fissure and contains the optic canal (containing the optic nerve and ophthalmic artery) which communicates with middle cranial fossa. The roof
(superior wall) is formed primarily by the orbital plate of frontal bone and also the lesser wing of sphenoid near the apex of the orbit.

The LPS which is a skeletal muscle originates on the lesser wing of the sphenoid bone, just above the optic foramen. It broadens and becomes the levator aponeurosis. This portion inserts on the skin of the upper eyelid, as well as the superior tarsal plate. The superior tarsal muscle, a smooth muscle, is attached to the LPS, and inserts on the superior tarsal plate as well. Levator palpebrae superioris is innervated by the superior division of the oculomotor nerve (Cranial Nerve III). The superior tarsal muscle is sympathetically innervated and is occasionally considered to be a part of the LPS.

The medial wall of orbit is formed primarily by the orbital plate of ethmoid, as well as contributions from the frontal process of maxilla, the lacrimal bone and a small part of the body of the sphenoid. It is the thinnest wall of the orbit, evidenced by pneumatized ethmoidal cells. The cribriform plate of the ethmoid bone (horizontal lamina) is received into the ethmoidal notch of the frontal bone and roofs in the nasal cavities. Pathology in the lesser wing of sphenoid bone, orbital plate of ethmoid bone including cribrifom plate, the belly and aponeurosis of LPS, the superior tarsal muscle may result into dysfunction of the Levator palpebrae muscle.

In the current patient there was a dehiscence of more than 3.5 mm in the cribriform plate on the right side and this dehiscence was reaching the right orbit up to the medial border and superior surface of LPS, in the roof and medial wall of orbit. This dehiscent portion resulted into poor support to the LPS resulting into its misalignment and malfunction, thereby causing ptosis on the affected side.

Dehiscences of the ethmoidal cells are not rare and have a particular clinical significance. Tonndorf investigated 100 human skulls including fetal skulls and showed that the cribriform foramina opened into a frontal recess, that is lateral to the middle nasal concha, in 10 cases. A review of the literature did not reveal any other study investigating the dehiscences in the cribriform plate of ethmoid bone and its association with ptosis. Dehiscences of the walls of the anterior ethmoidal canal 6.9 mm long were found in 93% of cases in one series and in another series 2 mm long dehiscences were seen in 39% of cases (Lang 1989).
Apart from the possibility that a congenital dehiscence of the cribriform plate with resultant cephalocele may result into congenital ptosis, a dehiscence in the orbital plate of the ethmoid bone in old age due to excessive pneumatisation may also result into ptosis.

CONCLUSION

Congenital ptosis may be rarely caused by a malfunction of the LPS muscle due to focal dehiscence of the cribriform plate and resultant cephalocele. This etiology is yet underreported and therefore requires more case reports and insight into the mechanistic aspect of this etiology to explore accurate surgical management.

REFERENCES