Ptosis Due to Systemic Diseases

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Acquired ptosis is a commonly seen condition. Although a vast majority of the ptosis is senile in origin, a certain percentage of the cases are associated with systemic diseases. There are no local or international data on prevalence of acquired ptosis due to systemic disease. For ptosis cases with a systemic association, early and correct diagnosis is of paramount importance. One of the good examples is unilateral ptosis due to surgical third nerve palsy, which is in turn due to an intracranial aneurysm.

Although the aetiologies of acquired ptosis are manifold, they can be classified into 4 main groups:

- Neurogenic (e.g. myasthenia gravis, Horner’s syndrome, third nerve palsy);
- Aponeurotic (also called senile ptosis, in which the levator palpebrae aponeurosis disinserts from the upper lid tarsal plate. Traumatic ptosis, though with a different aetiology, shares the same pathological change)
- Mechanical (due to increase of upper lid weight/rigidity, such as scar, oedema, redundant skin, tumour etc)
- Myogenic (as in myotonic dystrophy, chronic progressive external ophthalmoplegia etc)

Based on the above classification, neurogenic and myogenic ptosis are both associated with certain underlying systemic diseases. As the list of systemic diseases with ptosis as one of the signs will be very long, only several of the commonly seen entities will be discussed.

Myasthenia gravis (MG)

MG is a common neurogenic ptosis. As the site of pathology is at the neuromuscular junction, it can also be classified as neuromuscular or even as myogenic ptosis. Studies have shown that up to 70% of MG patients present with ptosis, and 90% of MG patients will eventually have ptosis.¹ The ptosis in MG can be either unilateral or bilateral. Severity of the ptosis depends heavily on time of the day, both due to fatigue and drug effect. Apart from the classical Tensilon challenge, blood test and electromyography, we now also know that ice applied to eyelids is also a sensitive way to differentiate myasthenic ptosis from others, probably as coldness can temporarily inhibit the activity of cholinesterase.²

Myasthenic ptosis can be treated either by drug, by surgery, or both. As diurnal variation of the ptosis is a typical feature in MG, the patient, if chooses to receive surgery, must be warned that the surgical outcome will still vary at different hours of the day, and revision of the surgery is not uncommon. (Figures 1A and 1B)

Third nerve palsy

Oculomotor palsy can cause squint, ptosis, or both. The ptosis can also be unilateral or bilateral, as the subnucleus to the levator muscle is a shared, midline structure in the brainstem. If the palsy is due to extrinsic compression by tumour or aneurysm, the ipsilateral pupillary response will also be affected and pupil size usually becomes larger. If a patient presents with sudden onset of unilateral ptosis with the eye on the same side in a down and out gaze, and the ipsilateral pupil is either enlarged or sluggish in reaction to light, surgical third nerve palsy must be put on the top differential diagnosis; and CT/MRI should be arranged as early as possible. Medical third nerve palsy, on the other hand, spares the pupil. Typically, the signs of medical third nerve palsy will slowly improve in weeks to months, so surgery should only be considered when the spontaneous improvement of ptosis and ocular motility are unsatisfactory after six months. As the levator function in a patient with third nerve palsy is typically poor or even absent, frontalis sling procedure is usually required to correct the residual ptosis. (Figure 2)
Horner’s Syndrome

Horner’s syndrome has the classical triad of ptosis, miosis and anhidrosis. The ptosis is due to the result of interrupted innervation to the sympathetic, autonomic Muller’s muscle rather than the somatic levator palpebrae superioris muscle. While Horner’s syndrome affects lid position and pupil size as a surgical third nerve palsy does, the ptosis in Horner’s syndrome is typically mild (less than 2mm); and the pupil is smaller on the affected side. The small pupil in Horner’s syndrome is also due to the interrupted sympathetic innervation. There are several locations where the sympathetic innervation can be interrupted; while 4% cocaine instilled to the eyes can confirm the diagnosis of Horner’s syndrome, Hydroxyamphetamine eye drops can differentiate the location of the lesion.

As the ptosis in Horner’s syndrome is typically less than 2mm, an internal approach (i.e. incision via the conjunctivum instead of eyelid skin) is particularly useful in the surgical correction; as the internal approach, although limited in the extent of correction, has no skin scar after surgery. (Figures 3A and 3B)

Chronic Progressive External Ophthalmoplegia (CPEO)

CPEO gets its name because of its early onset, progressive in severity, affects lid position and only external eye movement (it doesn’t affect the movement of the pupil, an intraocular structure, hence the name external ophthalmoplegia). (Figure 4)

CPEO is a mitochondrial cytopathy, being the commonest cause (43%) of myogenic ptosis. Patient with this disease develops ptosis during young adulthood; then squint; and then bilateral premature cataract at a later stage. Typically, patient with CPEO has very poor Bell’s phenomenon (the protective up-rolling of eyeball when eyelids are closed); so they may have exposure keratopathy after ptosis correction. Because of this, we tend to slightly under correct the ptosis, so as to reduce the risk of corneal exposure after surgery.

Conclusion

As mentioned above, while senile degenerative ptosis is still by far the most common cause of ptosis, we need to be vigilant in picking up those with a systemic cause behind. In neurogenic and myogenic ptosis, the drooping of eyelid can be the first sign of a serious yet treatable systemic disease.

References