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Inflammation as an Etiology of Acquired Ptosis in Children

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ABSTRACT

Ptosis or blepharoptosis is the downward displacement of the upper eyelid caused by a dysfunction of the eyelid elevator. There are many etiologies which can cause ptosis including congenital ptosis, blepharophimosis, aponeurotic dehiscence, acquired third nerve palsy, traumatic structural ptosis, and inflammation. To report a case of inflammation acquired ptosis in children. A Twelve year-old boy came to hospital with drooping of his right eyelid since 10 days before. There was no other complain on both eye. From the examination, visual acuity was 20/20 in both eyes. Ptosis examination for right eye/left eye, palpebral aperture 0/6mm, MRD1 0/1mm, levator function 0/14 mm. On the right eye there was restrictive movement to superotemporal gaze. For the anterior and posterior segment were normal for both eyes. In the head CT scan, there was no abnormality. In the laboratory test, there was increase in eritrosit sediment rate. We consulted the patient to the pediatric neurology department. The ptosis resolved after steroid administration. Inflammation can be the cause acquired ptosis in children and get respond very well with steroid administration.

Keyword: Inflammation, etiology, acquired, ptosis, children

INTRODUCTION

Ptosis or blepharoptosis is the downward displacement of the upper eyelid caused by dysfunction of the eyelid elevator. There are many etiologies which can cause ptosis in children including congenital ptosis, blepharophimosis, aponeurotic dehiscence, acquired third nerve palsy, traumatic structural ptosis, and inflammation. History taking is vital for determining the cause of ptosis. Morbidity of ptosis is associated with blockage of visual axis in severe cases or induced astigmatism and amblyopia in children.

CASE PRESENTATION

A 12-year-old boy was referred to the eye clinic with a 10-day history of acute onset painless eyelid drooping on both eyes. Persistent ptosis throughout the day. He did not report any double vision or visual obscuration. He also had headache since 1 month before. He had normal milestone, with no preceeding viral illness, fever, malaise or fatigue. No history of head trauma.

Table 1. Ptosis Examination

	Right Eye	Left Eye
Palpebral aperture	0 mm	6 mm
Superior margin-reflex distance	0 mm	1 mm
Levator function	0 mm	14 mm
Lid crease	5 mm	5 mm
Lagophthalmus	-	-
Bell's Phenomenon	Good	Good

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Apart from its position the lid was normal in appearance with no mechanical cause for the ptosis such as swelling, palpable mass or subtarsal abnormality. Visual acuity on both eyes were 20/20. Pupillary reflects were normal. The eye was straight on primary position with restrictive movement to superotemporal gaze on right eye. There was no proptosis with normal anterior and posterior segment of the eyes. No other neurological deficit was found. The patient underwent laboratory tests, with normal result. Head CT-scan with contrast was requested to rule out any abnormality in the brain and showed a normal cerebral appearance, no retro-orbital mass and aneurysm. All of the extraocular muscles were normal, with no optic nerve compression, no dilation of the superior ophthalmic veins and symmetrical cavernous sinuses.

Based on the ptosis condition and abnormality of the eye movement, we suspected the patient had superior division third nerve palsy. Patient was referred to pediatric neurology department to rule out systemic diseases related to the etiology of ptosis.

The patient was started on 24 mg of oral methylprednisolone at advised by the pediatrician and reducing to 20 mg after a week. One week after steroid administration, the ptosis was completely resolved. The steroid were slowly reduced until 8 weeks.

Based on ophthalmology and systemic examination with normal result and it well response with steroid administration we suggest inflammation as the cause of the ptosis and eye movement abnormality.

Figure 1. Initial Presentation of the Patient with Right Eye Ptosis

Figure 2. One week after after steroid administration

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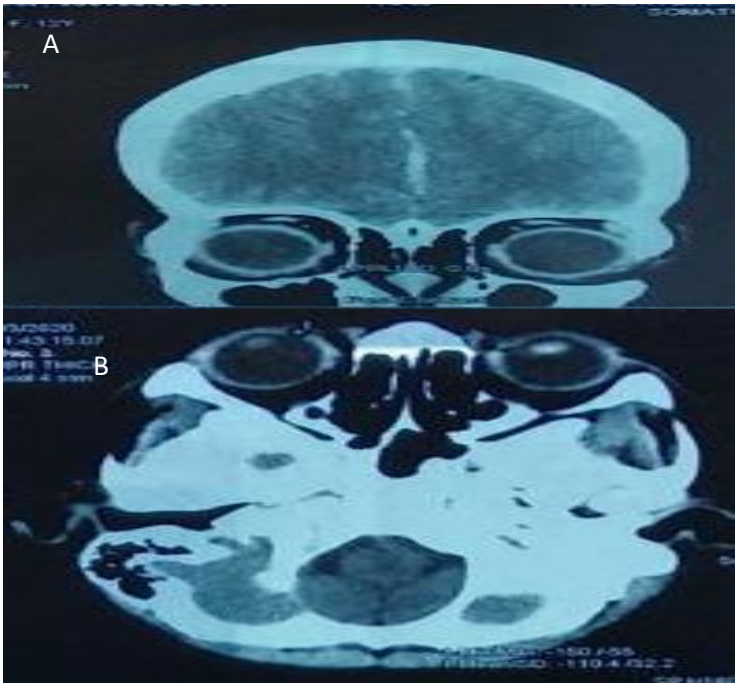

Figure 3. Head CT scan A. coronal view. B. Axial view

DISCUSSION

There were several differential diagnosis for ptosis which could be considered such as preseptal cellulitis, insect bite, idiopathic orbital inflammatory disease/ myositis, trauma, ophthalmoplegic migraine, oculosympathetic paresis, third nerve paresis, and myasthenia gravis, if the ptosis happen acute, idiopathic, and transient (Griepentrog, 2011).

In this patient, isolated bilateral ptosis was not typical of neurogenic cause. Usually in children, third nerve palsy can be congenital or it can be acquired because of some condition such as trauma, inflammation, or viral infection. In adult, intracranial aneurysm, microvascular infarction, inflammation, trauma, tumor, or infection are the most common causes of third nerve palsy (Griepentrog, 2011; Marenco et al., 2017).

Myogenic causes of ptosis include myasthenia gravis. The finding such as absence of variability, fatiguability and Cogan's twitch sign, as well as the isolated acute onset make this less likely. This patient did not exhibit symptoms of systemic muscle fatigue if they have ocular myesthenia but variable diplopia is often present. Ptosis may be asymmetric but is usually bilateral. The ice pack test also negative (Young & Leavitt, 2016; Mittal et al., 2011).

Another important differential diagnosis is Orbital infection, and usually patients sign and symptom more diffuse and commonly treated with intravenous antibiotics and closely monitored before the diagnosis of idiopathic orbital inflammation is made. However, in this patient, we found absent orbital signs, no erythema, pain or swelling and no leukocytosis, so we did not think about treatment indication with intravenous antibiotics. From the CT scan findings and the progression of his symptoms showed nothing in the 10 days from onset to starting steroids, the diagnosis of infection was very unlikely and did not need further investigation (Arat et al., 2013; Önder et al., 2016; Janicek, 2015).

Idiopathic Orbital Inflammation Syndrome (IOIS), known as orbital pseudotumor is another important differential diagnosis. IOIS is defined as a benign intraorbital process confined to the orbit and also extra orbital involvement. Clinically, orbital pseudotumor has been categorized as myositis, dacryoadenitis, anterior, apical and diffuse process.

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Pathomechanism in IOI can be caused by imbalance of inflammatory cytokines, such as high level of interleukin (IL) -2, IL-8, IL-10, IL-12, interferon (IFN)- γ, and tumor necrosis factor (TNF)- α.

There are several difference symptoms between adult and children. In adult, usually presenting with exophtalmus but in children presenting more in ptosis. Optic disc edema and uveitis can be found in the ophthtalmology examination. Constitutional signs and symptoms of IOIS are headache, fever, anorexia, emesis, lethargy, abdominal pain, and eosinophilia. There was no sign of uveitis and optic disc edema in ophthalmology examination for this patient. In the laboratory finding, to make the right diagnose, we need to do head CT scan. The result, we can find enlargement of the muscle belly of one (or more) extraocular muscles with the involvement of their tendinous insertions. Additional inflammation can be seen in surrounding tissues such as orbital fat, optic nerve sheath, uvea, sclera, or even diffuse orbital involvement, including the lacrimal gland. Another appearance such as an infiltrative mass and extends outside of the orbit via superior or inferior orbital fissures. The extension into the cavernous sinus, meninges, and dura can occur. In this patient, from the CT scan we didn't find any enlargement of the muscle belly of extraocular muscle and surrounding tissues (Naphade et al., 2012; Yan et al., 2006).

Although in CT scan we did not find any abnormality, in this patient might be diffuse type.

The main cause of ptosis condition in this patient suspected to be IOI. Another examination should be done such as C reactive Protein, Electomyography (EMG), MRI. There were several therapy such as corticosteroid, Non–Steroid Anti Inflamation Drug, anti metabolites, external beam radiotherapy. The first line management of IOI is corticosteroid. The mechanism of steroid in this condition for the anti-inflammatory and immunosuppressive effects. Mechanism action of corticosteroid in effect of anti-inflammatory by inhibiting phospholipase A2 and cyclooxygenase pathway. For effects of immunosuppressive are because of inhibition of IL and IFN synthesis, inhibition of major histocompatibility antigen expression, and cytotoxic effect on T lymphocytes. Many patient show improvement after this therapy. The starting dose of 1mg/kg/day, and then slow tapering over 6-8 weeks. There are some systemic side effect which need attention while using long term corticosteroid such as cushing syndrome, hyperglycemia, hypertension, adrenal suppression, weight gain, avascular bone necrosis, and growth retardation. In this patient we treated with steroid, and the condition of the ptosis resolved.

CONCLUSION

Inflammation can be the cause of acquired ptosis in children and very responsive with steroid administration. Therefore acquired ptosis in children need a comprehensive investigation from the history taking, physical examination, and other diagnostic modality, such as imaging, laboratory. Furthermore we need collaboration with related department to determine the cause so we can give comprehensive therapy.

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