Challenging Case

A 16-year-old Girl with Diplopia and Unilateral Upper Lid Ptosis

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CASE PRESENTATION

A 16-year-old girl presented with drooping of the left upper eyelid and diplopia in primary position, which was worse in right gaze. Both eyes had visual acuity of 20/20, intraocular pressure of 16 mmHg, and normal slit lamp and fundus examinations. The response to color vision testing using Ishihara plates was 12/12 in both eyes. The video shows the ocular motility findings, and Figure 1 depicts the results of the visual field examination [Video 1]. Results of magnetic resonance imaging of the brain are shown in Figures 2 and 3. The parents did not accept to perform any further work-up.

Herein, we address two questions regarding the above-mentioned case to three experts in the related field.

Question 1: What are the possible causes of these findings?

Question 2: What is your treatment plan for this patient?

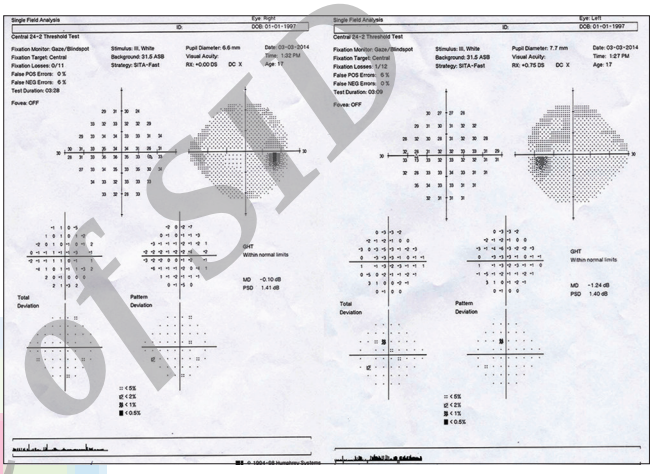

Figure 1. Visual fields of the right and left eyes.

Mark L. Moster, MD

First I would like to discuss possible causes of the condition based on the examination, prior to obtaining the MRI scan. This girl has findings most consistent with partial left CNIII palsy. There is ptosis, together with adduction, elevation and depression deficit. It is hard to tell if the pupil is involved on the video. It appears that the optic nerve and the 6th nerve are intact, and we lack data on the 4th and 5th cranial nerves.

We are also unaware whether the onset of the condition was sudden or slowly progressive. If the palsy was sudden onset, an aneurysm at the junction of the internal carotid and posterior communicating artery would be at the top of the list of the differential diagnoses except in very young children. Another cause in a young person is ophthalmoplegic migraine. In an older person with vascular risk factors, an ischemic peripheral third nerve palsy is most common. If a slowly progressive CNIII palsy is present, a structural lesion with compression of the nerve is more likely.

Once we have the MRI in this case, we see a lesion in the orbital apex extending to the anterior cavernous sinus. Based on the appearance on the current images, differential diagnoses would include mainly a cystic schwannoma or a venolymphatic malformation. The bright T2 and dumbbell shape is consistent with a schwannoma.

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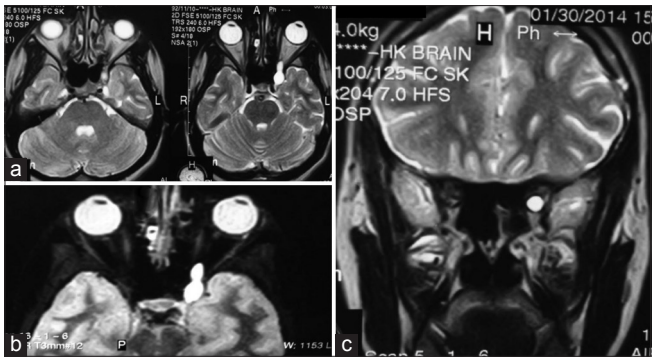

Figure 2. T2 weighted MRI of the orbit; (a and b) axial views, (c) coronal view.

I would consult oculoplastics/orbital specialists and neurosurgeons. A diagnosis would be made by biopsy, but one must be careful about bleeding if the lesion appears to be vascular. Resection of the lesion might be tried but could be difficult because of the potential for visual loss or other cranial neuropathies from operating at the orbital apex or anterior cavernous sinus. If surgical removal is not feasible and deficits are progressive or if the lesion grows significantly, I would consider highly focused radiotherapy for either schwannoma or venolymphatic malformation. If the lesion turns out to be a venolymphatic malformation, partial cauterization to shrink the lesion might also be considered.

If the deficit is longstanding and the lesion remains stable on sequential MRIs, then the option of observation without intervention may also be considered, since optic nerve function is normal.

Masoud Aghsaeifard, MD

The clinical features of the patient are compatible with partial third nerve palsy with aberrant innervation. Aberrant innervation is seen with parasellar mass lesions, trauma, or congenital lesions. Based on the history, the cause of the condition might be a parasellar lesion. The MRI depicts an orbital apex or superior orbital fissure lesion with a signal similar to CSF, i.e., hypointenseon T1 and hyperintenseon T2 without significant enhancement which is compatible with a cystic lesion. Differential diagnoses include the following: Arachnoid cyst, orbital encephalocele, perioptic cystic lesion, infectious cyst (hydatid cyst) and orbital meningocele.

Padmaja Sudhakar, MD

This is a 16-year-old girl of likely Indian origin whose clinical examination seems consistent with partial left third nerve palsy. Involvement of the pupil has not been described in the synopsis of the case or demonstrated in the video. Brain MRI depicts a cystic lesion at the left orbital apex. She does not appear to have involvement of 4th nerve, 6th nerve, or the optic nerve.

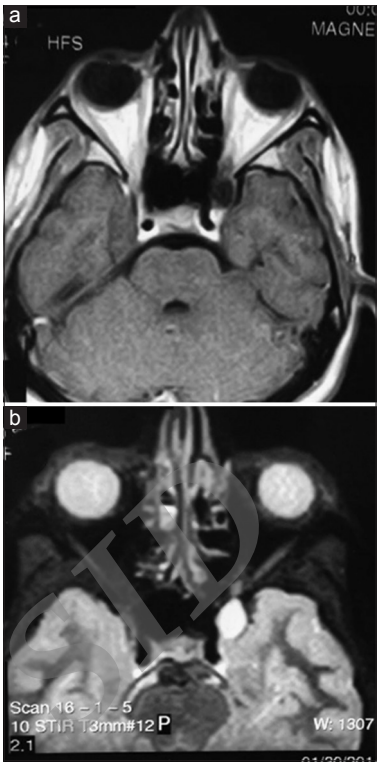

Figure 3. Axial MRI of the orbit; (a) post contrast T1 weighted image, (b) short T1 inversion recovery (STIR) image.

The orbital apex refers to the posterior aspect of the orbit adjacent to the superior orbital fissure and optic canal, and freely communicates with the inferior orbital fissure and pterygopalatine fossa. The orbital apex is an important landmark where several neurovascular structures enter the orbit from the cranium. These include structures that exit within and external to the annulus of Zinn. An orbital apex syndrome may present with ophthalmoplegia due to involvement of the oculomotor (3rd) nerve, trochlear (4th) nerve, or abducens (6th) nerve, or can cause optic nerve dysfunction. Involvement of the ophthalmic branch of the trigeminal nerve may also occur.

There are a variety of lesions that can affect the orbital apex and manifest with the orbital apex syndrome. These include neoplastic lesions, vascular lesions, inflammatory lesions, infectious processes and traumatic lesions. In this particular case, the lesion seems to involve the orbital apex and cavernous sinus. It is a smooth, elliptical, cystic lesion and has a dumbbell shape. It does seem to involve the internal carotid artery. The lesion is hypo-intense on T1 weighted MRI, and hyperintense on T2 and STIR sequences. Contrast enhanced images are not available. The lesion does not seem to be a meningioma, lymphoma or metastasis. Such lesions are not common at this patient's age. Schwannomas are slow growing tumors that can develop from the myelin sheaths of peripheral nerves. They can affect the

orbit and involve the intraconal or extraconal space or the orbital apex. They can conform to the surrounding structures and can assume a dumbbell shape if they extend into the cranial vault within the superior orbital fissure. They are isointense to the brain on T1 and hyperintense on T2 and can show homogenous enhancement. This lesion could be a schwannoma arising from the oculomotor nerve. However, contrast enhanced images have not been provided. Another possibility is a neurofibroma. Other cystic processes should also be considered.

The clinical and imaging features do not suggest an inflammatory lesion. However, infectious process such as tuberculoma cannot be entirely excluded although cavernous sinus tuberculoma is exceedingly rare.

The imaging features do not meet the criteria for a vascular lesion such as a capillary or cavernous hemangioma, caroticocavernous fistula or other compressive lesions such as a subperiosteal hematoma, mucocele and fibrous dysplasia.

Detailed laboratory testing for infection and inflammation is mandated in every orbital apex lesion. This should include inflammatory markers namely erythrocyte sedimentation rate and C reactive protein, complete blood count with differential, complex metabolic panel and autoimmune work-up including anti-nuclear antibody and anti-neutrophil cytoplasmic antibody. In this case, one should consider testing for tuberculosis with purified protein derivative or quantiFERON-TB Gold test and a chest X ray. Lumbar puncture may also be required. Suspicion for sexually transmitted diseases remains low in this case.

Biopsy of the lesion may be the most definitive way to establish the diagnosis. A multidisciplinary team involving a neurosurgeon, ENT surgeon and oculoplastic surgeon may be needed for surgical excision of the lesion. Surgical approach should be undertaken with the understanding that the patient may be left with diplopia post-operatively. It is also possible that she may have improvement.

The diplopia can be temporarily be overcome by patching one eye. However, for a more definitive treatment, prisms may be required. Surgical correction for misalignment may be required eventually. She may need more than one surgery which should be undertaken only when the misalignment has been stable over a period of 6 to 9 months. This patient should be periodically evaluated at the neuro-ophthalmology or pediatric ophthalmology clinic.

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Conflicts of Interest

There are no conflicts of interest.

SUGGESTED READINGS

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