Gurjinder Kaur, OD, Carolyn Graeber, MD

Title: Intracavernous epidermoid cyst: A rare cause of Sixth Nerve Palsy

Abstract: To describe a case report of an intracavernous epidermoid cyst causing chronic abducens (6th nerve) palsy.

I. Case History:
25-year old obese Hispanic female presents as a new patient who was referred by her primary care physician for an ocular evaluation.

The patient presents with symptoms of diplopia and a large right eye turn. She reports that since birth her right eye intermittently turned inward multiple times and she was able to straighten it out with a few blinks and rubs. Her right eye, however, permanently “turned in” about 6 months ago. She also reports headaches that occur occasionally and are localized to the temporal region of her head. She denies all other ocular or visual complaints.

The patient presents with no past ocular history or instillation of any ocular medications or drops. She has a positive medical history for Non-insulin dependent Diabetes Mellitus for 5 years. The patient is unsure of her Hemoglobin A1C or last blood sugar readings but states that her blood sugar is “controlled” with 500 p.o. Metformin taken daily. The patient is unremarkable for any other ocular or medical conditions as well as the use of all other medications.

II. Pertinent Findings:
*Visual Acuity with correction, OD: 20/20-2 with left head turn, OS: 20/20
*Intraocular Pressures: 11 OD, 14 OS using Goldmann Applanation Tonometry
*Cover Test without correction revealed an incomitant deviation: Distance and Near in Primary Gaze: Esotropia > 50 D, Distance and Near in Right gaze: Esotropia of 18 D, Distance and Near in Left Gaze: Esotropia of 90 D
*Dry Auto Refraction was done revealing findings of -0.25-1.00x135 OD and plano OS. The patient was currently wearing glasses with the following Rx: -0.50sph OD and plano OS.
*Upon slit lamp exam evaluation, lids and lashes had mild blepharitis on the upper eyelids of both eyes. There was no ptosis or retraction present of either lid. All other findings including conjunctiva, sclera, iris, cornea, anterior chamber, and lens were unremarkable in both eyes. There was no neovascularization of the iris in both eyes. *Upon pupil testing, there was no afferent pupillary defect in either eye and both pupils were equal, round, equally reactive to light, and accommodated well.
*Dilated fundus examination revealed 0.2/0.2 cup to disc ratio in both eyes with unremarkable posterior examination findings except mild vitreous floaters in both eyes. No signs of papilledema or optic atrophy were noted. No signs of diabetic retinopathy or clinically significant macular edema were present in the retina. The periphery was unremarkable with no signs of retinal tears, holes, or detachments. No extraocular motor
restrictions were noted when examining the retinal periphery with binocular indirect ophthalmoscope.

Impression and Plan:
Esotropia of the right eye. MRI of the head and orbits with and without contrast was recommended to rule out intracranial and intra-orbital causes of sixth nerve palsy. The patient was scheduled to return to office in 6 weeks with MRI results.

Radiology reports revealed: 2.5 x 1.5cm focus of signal alteration along the right aspect of the cavernous sinus extending into the preptontine cistern most compatible with an epidermoid cyst.

III. Differential diagnosis:
Before sending our patient out for imaging, our differentials included an intracranial tumor compressing the abducens nerve, diabetic related vascular impingement on cranial nerve six, and Thyroid Eye Disease. Myasthenia Gravis is a disorder of neuromuscular transmission recognized clinically by varying muscle weakness, which is characterized by worsening with fatigue. Grave’s disease restrictive myopathy was one of our differentials because it is a common cause of spontaneous diplopia in adults. These patients would have a positive forced duction, which means movement is limited in the direction of inflamed muscle with Q-tip globe push. Also, the MRI of the orbits will reveal an enlarged or inflamed extra ocular muscle. Our leading diagnosis was an intracranial tumor that would be evident upon MRI of the head.

Microvascular cranial nerve palsy was not at the top of our list even though our patient has history of non-insulin dependent diabetes. This is because our patient was only 25 years old and vascular related incidents are more common in adults over the age of 40. It was important to send our patient out for an MRI of the head and orbits due to the risk of a mass compressing on the abducens nerve. Based on the images of our patient’s MRI, our differentials include arachnoid and dermoid cysts. An arachonoid cyst is a cerebrsospinal (CSF) filled sac that does not communicate with the ventricular system. On MRI, it does not restrict on diffusion weighted imagining and follows CSF on all MR sequences. A dermoid cyst is a benign, ectopic, squamous epithelial cyst containing dermal elements, including hair follicles, sebaceous and sweat glands. On imaging, it is T1 hyperintense, which means it is bright on T1. Dermoid cysts are usually located along the midline of the brain, whereas epidermoid cysts are located off the midline.

IV. Diagnosis and Discussion:
A handful of cranial nerves as well as the internal carotid artery pass through the cavernous sinus. The cranial nerves that pass through the cavernous sinus are CN III (Oculomotor nerve), CN IV (Trochlear nerve), CN VI (Abducens nerve), CN V1 (Ophthalmic nerve), and CN V2 (Maxillary nerve), as well as the postganglionic sympathetic fibers. Any mass or cyst in the cavernous sinus can compress any of these nerves and cause cranial nerve palsy. The abducens nerve (6th nerve) is responsible for innervating the lateral rectus muscle. The lateral rectus muscle pulls the eye away from the nose. However, when the lateral rectus muscle is weak, the eye crosses inward
toward the nose resulting in an esotropia. In an abducens nerve palsy, the measured esotropia is larger at distance and in the gaze to the same side as the affected lateral rectus muscle. In our patient, the epidermoid cyst was large enough to compress the abducens nerve in the right cavernous sinus, causing a sixth nerve palsy resulting in an ipsilateral esotropia.

Intracranial epidermoids are congenital inclusion cysts. Symptoms depend on the location and effect on adjacent neovascular structures. Epidermoid cysts are lobulated, irregular, cauliflower-like masses with “fronds.” The most common symptom of these cysts is headaches. It presents between the ages of 20-60 years with peak at 40 years and is the most common congenital intracranial tumor. Epidermoid cysts grow slowly as epithelial component growth rate commensurate to that of normal epithelium. It does not have a preference for a particular gender, as it is seen in men and women equally. To differentiate different types of masses, we must look at different MRI sequences.

Upon review of the MRI sequences, our top differential diagnosis of a solid mass was luckily unlikely as our patient’s cyst presented as a fluid signal as opposed to a solid on the imaging. However, contrast is recommended to completely rule out tumor as a differential since tumor presentations can vary on T1 and T2 imaging. Contrast is important to enhance the mass making it easier to differentiate. One setback with our case is that contrast imaging was not done even though it was recommended by our referral. However, it is still very clear that the cyst in the images is fluid filled and not a solid mass as tumors would present.

On axial T2 imaging, fluid is bright. In our patient, the Axial T2 weighted image shows a T2 hyperintense mass in the right cavernous sinus, which extends to the prepontine cistern. On axial FLAIR imaging, bulk fluid is normally suppressed. In our patient, axial FLAIR images shows that the lesion does not suppress on FLAIR, and therefore is likely not an arachnoid cyst since an arachnoid cyst will suppress. On axial T1 imagining, fat, protein, and blood is bright. Our patient’s axial T1 images shows that the lesion is not bright and therefore it is not likely a dermoid cyst since dermoid cysts contain fat. Lastly, on axial diffusion images, anything that restricts water diffusion is bright (for example, stroke, highly cellular components.) Our patient’s axial diffusion images show that the lesion is bright (therefore restricts diffusion) and thus is likely an epidermoid cyst. Our patient’s cyst is T2 hyperintense, restricts diffusion, and does not suppress on FLAIR, which are all classic findings of an epidermoid cyst.

V. Treatment and Management:

Treatment of epidermoid cysts includes microsurgical resection, but recurrence is common if incompletely removed. Upon resection, it is unknown if our patient will have full recovery of the abducens palsy. However, a literature search reveals that epidermoid cysts are very rare in the cavernous sinus. In one particular case report, Kuroi et. al describe a 41 year old male with a left abducens palsy. On MR, this patient was found to have an epidermoid cyst in the left cavernous sinus. After resection, the patient had complete cure of the abducens palsy. Another article reveals that vast majority of patients with cysts invading or compressing the cavernous sinus present with an excellent functional recovery after surgery independent of the extent of tumor resection.
VI. Conclusion:
In adults, the top three most common etiologies for a six nerve palsy are undetermined, neoplasm, and vascular. In children, the most common etiologies include trauma and neoplasm. It is important to remember that the age of the adult matters in accessing the condition at hand. Adults aged 15-40 are most likely to have a neoplasm or mass-related condition. It is less likely that this age group will have vascular related cranial nerve palsy. Therefore, imaging these patients is very crucial. Adults over the age of 40 are more likely to have a vascular related etiology such as from diabetes or hypertension. It is important to watch these patients over the course of 3 months for improvement. If no improvement is made at 3 months, these patients must also be imaged. Lastly, giant cell arteritis should be a top differential in patients over the age of 60.

Bibliography:
