Primary nerve repair following resection of a neurenteric cyst of the oculomotor nerve

Case report


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Neurenteric cysts are rare congenital lesions of endodermal origin occurring in the spinal canal and infrequently in the posterior cranial fossa. They are mostly encountered in the lower cervical and upper thoracic spine, frequently associated with structural anomalies of the adjacent vertebra and spinal cord. Neurenteric cysts are believed to result from dysgenesis of the notochord during the third week of embryonic development, leading to persistence of endodermal elements along the craniospinal axis.

We report a case of a neurenteric cyst of the oculomotor nerve in a 3-year-old child. This is the fourth reported case of a neurenteric cyst involving the oculomotor nerve. In our case, complete resection of the neurenteric cyst and primary oculomotor nerve repair were performed.

Abbreviation used in this paper: GFAP = glial fibrillary acidic protein.

Case Report

History and Examination. An 8-month-old female presented initially with sudden onset, complete right third cranial nerve palsy. She had no other neurological symptoms and was otherwise in good health. An MR imaging study demonstrated some irregularity in the region of the right third cranial nerve. She made a spontaneous partial recovery after several months, with the pupil remaining dilated and unreactive. At 3.5 years old she presented again with a recurrent complete right third cranial nerve palsy. Repeat MR imaging showed a cystic mass lesion in the right ambient cistern, with compression of the anterolateral aspect of the brainstem. The patient underwent a craniotomy, complete excision, and a primary third cranial nerve repair. While there have been 3 reported cases of neurenteric cysts arising from the oculomotor nerve, this is the first documented case with a primary nerve repair.

Key Words • neurenteric • endodermal cyst • oculomotor nerve • nerve repair • peripheral nerve

This article contains some figures that are displayed in color online but in black and white in the print edition.
of persisting complete right third cranial nerve palsy the patient underwent surgical intervention. A stereotactic right frontotemporal craniotomy and microsurgical excision of the cyst were performed. Intraoperatively, the cyst had completely destroyed a section of the oculomotor nerve. After resection of the cyst, primary oculomotor nerve repair was performed by end-to-end anastomosis with 10-0 nylon suture (Ethilon). Pathological investigation showed the lesion to be a neurenteric cyst (Fig. 2).

On follow-up examination at 7 months there had been considerable and ongoing improvement in her third cranial nerve function. She was able to completely open her right eye with minimal ptosis that was of no aesthetic or functional concern to the family. While she did not complain of diplopia, this is likely due to established amblyopia in her right eye. The pupil remained unreactive and dilated, but she was able to adduct her eye slightly past the midline, and it was believed that she might benefit from strabismus surgery in the future.

Discussion

Third cranial nerve palsies are rare lesions in children,

with different origins than in adult populations. Congenital third cranial nerve palsies are frequently attributed to adverse intrauterine events or perinatal injury during delivery. Postnatal trauma, infections, and migraine are common causes of third cranial nerve palsies in children,

while intracranial aneurysms and brain tumors are more likely causes in adults.

Neurenteric cysts arise most frequently in the lower cervical and upper thoracic levels, and constitute 0.3%–0.5% of all spinal cord tumors. Although neurenteric cysts may be intramedullary, most (90%) occur as intradural extramedullary lesions located ventral to the spinal cord and are associated with vertebral and spinal cord abnormalities.

There is a male predominance with these lesions, and they typically present in the first or second decade of life. Intracranial neurenteric cysts are very rare lesions, occurring mostly in the posterior cranial fossa anterior to the brainstem or within the fourth ventricle. There have been only 3 previ-
Primary nerve repair after resection of a neurenteric cyst

Neurenteric cysts have variable signal intensities on MR imaging depending on their protein content; most are usually slightly hypointense relative to CSF on T1-weighted images and hyperintense on T2-weighted images. While peripheral enhancement of the cyst with Gd is unusual, our case demonstrated rim enhancement. Most neurenteric cysts are ovoid or lobulated and sharply demarcated, with variable size from a few millimeters to several centimeters.

Advancements in microsurgical peripheral nerve repairs have lead neurosurgeons to apply these techniques in cranial nerve reconstruction. Because the facial nerve is the most common cranial nerve damaged, it has thus been studied widely. Reconstruction of the facial nerve can often restore at least partial function, with 83% of patients having improved tone and some degree of voluntary facial motion in 1 study. Posttranssectional aberrant reinnervation is well documented in facial nerve reconstruction, leading to mass motion, synkinesis, and partial reinnervation. In facial nerve reconstruction, the most important factor is tension-free anastomosis, with end-to-end anastomosis preferred to an interpositional graft. Aberrant reinnervation is associated with the time needed for axonal tips to cross the injured zone, grow down the nerve, reach the muscle target, and restore the neuronal reinnervation of motor end plates.

End-to-end anastomosis is more limited for intracranial portions of cranial nerves due to limited length reserves compared with peripheral nerves that may be mobilized and stretched, with end-to-end repair not possible for intracranial nerve deficits greater than a few millimeters.

There is increasing literature reported on oculomotor nerve reconstruction with many commonalities to facial nerve repairs. Aberrant reinnervation is problematic, with difficulties achieving coordinated motion of extraocular muscles. In animal studies recovery of extraocular motility occurred in all animals without aberrant phenomena at the level of the orbital fissure, while with more proximal transections the degree of recovery and aberrant phenomena varied greatly. Iwabuchi et al. and Sekhar et al. showed partial recovery of function following reconstruction of the oculomotor nerve, with greatest improvement in the levator palpebrae superioris and medial rectus muscles. Our case demonstrated a similar outcome with significant improvement in eye opening and adduction, with less satisfactory outcome in vertical gaze and pupillary reactivity.

Although surgical intervention in neurosurgical cysts should endeavor to achieve complete resection, in one-third of cases the cyst is adherent to adjacent neurovascular structures, making complete resection potentially dangerous. Simple aspiration is uniformly discouraged due to unacceptable recurrence rates; therefore, subtotal resection is often advocated to avoid further neurological deterioration. However, subtotal resection has a greater recurrence rate than total resection, thus potentially requiring further surgical intervention and risk of progressive and/or new neurological deficits. In pediatric cases with a complete deficit we advocate that complete resection be attempted to reduce the risk of recurrence and to minimize any further neurological deficits caused by recurrent cysts and repeated operations.

Reconstruction of the oculomotor nerve may not achieve binocular vision, but the improvement in the levator palpebrae superioris and medial rectus muscles leads to an aesthetically acceptable eye. We recommend that repair of the oculomotor nerve ought to be attempted as the ptosis in a complete third cranial nerve palsy is the most difficult part to treat surgically, while the dilated pupil can be treated with a contact lens, and strabismus surgery may be necessary at a later time to gain a midline pupil.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following: Conception and design: Dexter. Acquisition of data: Turner. Analysis and interpretation of data: Turner. Drafting the article: Turner. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Turner.

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