Symptomatic Oculomotor Nerve Cyst in a 3-Year-Old Child: Case Report With Emphasis on Surgical Management

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BACKGROUND AND IMPORTANCE: Third nerve palsies in the pediatric population are most commonly caused by trauma, tumors, or vascular abnormalities. Cystic oculomotor nerve neuropathies, however, are rare. We report the case of a symptomatic cyst along and within the oculomotor nerve, which has not been described previously.

CLINICAL PRESENTATION: Here, we report a case of a 3-yr-old girl presenting with a progressive painless oculomotor nerve palsy. A magnetic resonance imaging revealed a cystic formation along the cisternal and cavernous course of the nerve. Due to lack of alternative treatment options, surgery was offered. Intraoperative direct nerve stimulation allowed for identification of a non-functional part of the cyst wall and open fenestration and biopsy were executed. Histopathology revealed neuritis. Serology was negative for various pathogens. The oculomotor palsy rapidly resolved. At a follow-up 5 yr after surgery, the girl is asymptomatic and the cisternal part of the cyst remains collapsed.

CONCLUSION: This is the first report of a symptomatic cyst along and within the oculomotor nerve treated effectively with open fenestration and decompression highlighting the importance of intraoperative neuromonitoring in cranial nerve surgery. Uncertainty remains regarding the etiology of this disease.

KEY WORDS: Oculomotor nerve, Central nervous system, Cysts, Case report

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Third nerve palsies in the pediatric population are most commonly caused by trauma, tumors, or vascular abnormalities.[1,2] Cystic oculomotor nerve neuropathies, however, are rare. Only a small number of cases have been reported in literature, mostly either of arachnoid or endodermal etiology.[3,4] We report a case of a symptomatic cyst along and within the oculomotor nerve, which has not been described previously.

CLINICAL PRESENTATION

Patient Information

A 3-yr-old otherwise healthy girl developed a gradually progressive ptosis followed by mydriasis and diplopia over a course of 2 mo. No extraocular symptoms or complaints were noted. The physical examination was otherwise unremarkable. The patient's legal guardians gave full consent to publish this case.

Diagnostic Assessment

Ophthalmologic assessment confirmed a pain-free complete right-sided oculomotor palsy with mydriasis, ptosis, loss of reaction to light or accommodation, and restricted adduction. A magnetic resonance imaging (MRI) was ordered showing a complex cystic lesion along the course of the oculomotor nerve (Figure 1). The cisternal aspect was T1 hypointense and T2 hyperintense. This most prominent part was continuous with a smaller T2 isointense component extending into the cavernous sinus. Nodular contrast enhancement in the para-clinoidal cyst wall was noted. There was no diffusion restriction. Computed tomography revealed no bony abnormalities.

Surgical Intervention

Given the undetermined nature of the lesion and the patient being symptomatic, surgery was recommended. The child was positioned supine, and the head fixed at 45° to the left side using

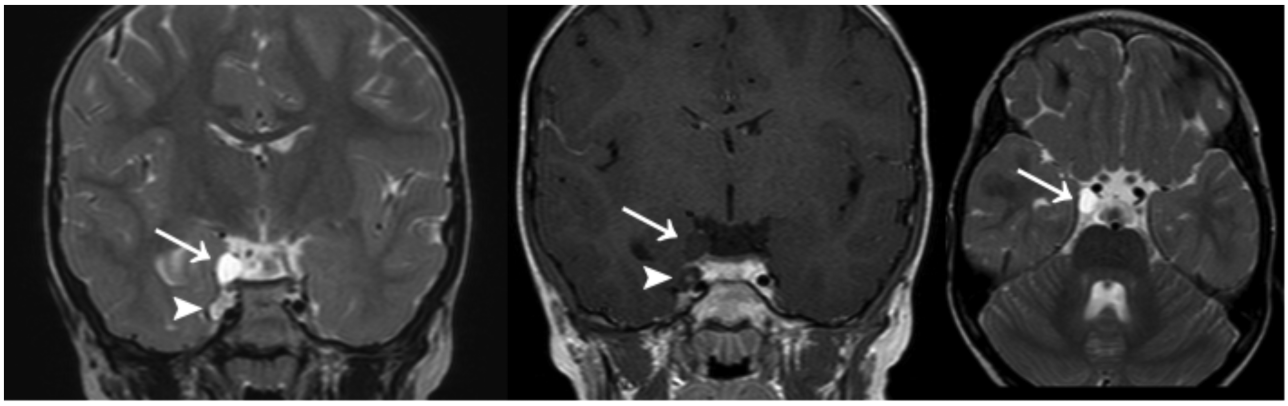
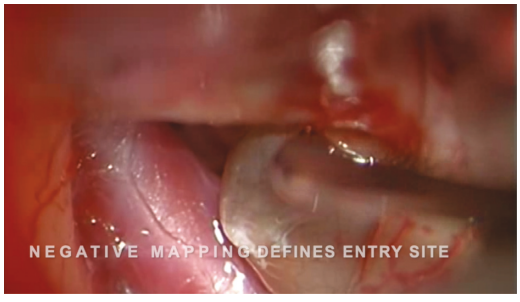


FIGURE 1. *Preoperative imaging showing cisternal (arrow) and cavernous (arrowhead) parts of the cystic right oculomotor nerve on coronal (left image) and axial (right image) T2-weighted MRI. Note the nodular contrast enhancement within the cavernous part on coronal T1-weighted post contrast sequence (center image).*



VIDEO. *Intraoperative video demonstrating application of direct nerve stimulation and identification of a non-functional entry zone into the oculomotor nerve cyst.*

a pediatric head holder. To allow for intraoperative monitoring of third nerve function, a 20 mm oculomotor bipolar needle electrode (Inomed) was inserted into the ipsilateral inferior rectus muscle. A pterional craniotomy was raised. Following a subfrontal approach and partial Sylvian fissure split, the lesion was exposed in a microsurgical fashion. A small non-functional entry site was identified under continuous direct nerve stimulation with a monopolar probe up to 0.5 mA at 3Hz. The cyst was punctured for decompression and fluid collection. A region of the cyst wall which mapped negatively for function was resected using a diamond knife, creating a fenestration for prevention of recurrence (Video). Larger parts of the cyst wall were closely adherent to splayed nerve fibers as deduced from further mapping. Due to lack of a clear surgical plane no further efforts of resection were made. Aspirate and parts of the cyst wall were sent for neuropathology.

Histopathological Findings

Fluid drained from the cyst contained monocytes and lymphocytes. The cyst wall showed axons. A clear surface epithelium was not identified. Reactive changes and CD3 positive T-lymphocytes were encountered. Ki67/MIB1 proliferation index was 5%. The findings were consistent with the histopathological diagnosis of neuritis (Figure 2).

Perioperative Course

As pathology was consistent with a neuritis, viral and bacterial infections were ruled out after negative serology for Cytomegalo virus (CMV), Epstein-Barr virus (EBV), Herpes simplex virus (HSV), Varizella-Zoster virus (VZV), *Bartonella henselae*, *Borrelia burgdorferi*, Chlamydia, and mycoplasma. The girl was discharged a week after surgery. At that time, ptosis had completely resolved. The anisocoria was still present but the girl was again able to adduct her eyeball over 20° across midline.

Follow-up and Outcome

One year after the operation, diplopia had resolved completely and slight anisocoria was still present. At 5 yr after surgery, the only clinical finding was a sluggish pupil constriction in response to light. Figure 3 demonstrates collapse of the cisternal part with the cavernous part still being present. At 10 yr after surgery, symptoms had resolved completely.

DISCUSSION

The first documented case of a cyst causing oculomotor palsy was published in 1980 by Lesser et al.[5] The authors found an arachnoid cyst that was fenestrated and symptoms resolved accordingly. Over the following decades, further reports were published on arachnoid cysts compressing the oculomotor nerve.[4,6,7] A second etiology to consider is an endodermal or neurenteric cyst. This congenital and benign lesion is thought to arise from a failure of dissolution of the transient neurenteric canal.[8] Cox et al[3] recently provided a comprehensive literature review of endodermal cysts affecting the oculomotor nerve. A

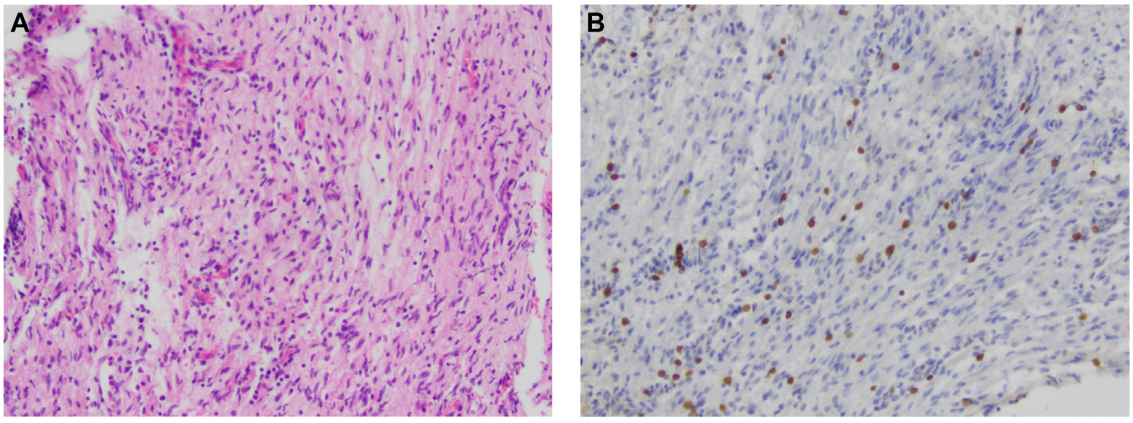


FIGURE 2. *Hematoxylin and eosin staining* **A** *of cyst wall showing myelinated and unmyelinated axons with infiltration of CD3 positive T-lympocytes* **B**. *Positive neurofilament stain (not shown) further proved the existence of axons within the cyst wall.*

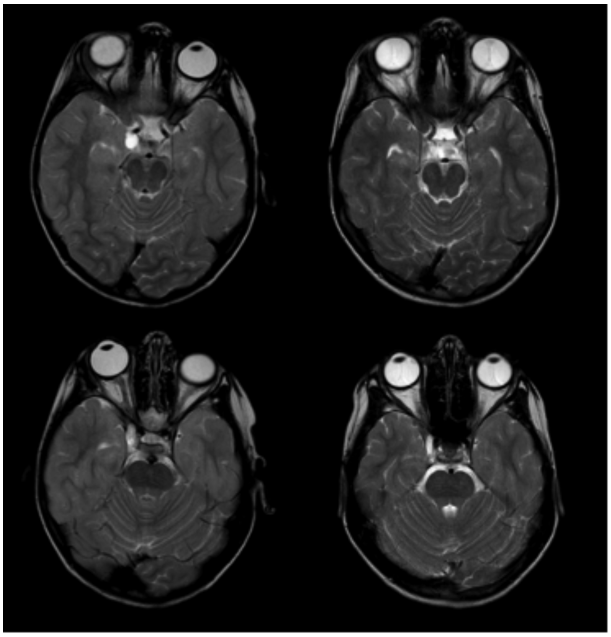


FIGURE 3. *Direct comparison of cisternal (upper row) and cavernous (lower row) parts of the cystic right oculomotor nerve on axial T2-weighted MRI before (left column) and 5 yr after surgery (right column). Note the bulbar realignment reflecting complete resolution of diplopia.*

condition most commonly found symptomatic in the first decade of life.[3]

The cyst presented in the case above did resemble an endodermal cyst on multiparametric MRI; however, tissue diagnosis was consistent with neuritis. Also, the recent review provided by Cox et al[3] clarifies that in most patients, the cyst can be at least partially resected. For functional reasons, this was not possible in the present case. With this in mind, the cyst was most likely to be intrinsic to the oculomotor nerve. In summary, this is the first description of an idiopathic intrinsic cyst to the oculomotor nerve.

Whether lymphocytic infiltration of the biopsy specimen was a result of chronic compression or directly related to an infectious disease remains unclear. Cranial neuropathies can be a result of various infections.[9-11] Still, the girl tested negative during a comprehensive serology workup as described above. The literature lacks reports on infectious cranial neuropathies of cystic nature.

Importantly, this is a unique case with unknown natural history. Thus, uncertainty remains concerning treatment indications. Cyst fenestration is a well-accepted mode of treatment for symptomatic cases if resection is not deemed safe for functional reasons. A recent report states that for recurrent cases, a cysto-subarachnoid shunt may be a feasible option for large cysts[3] but would obviously not been of value for the present case.

Contemporary cranial nerve surgery requires neuromonitoring. As clear recommendations on intraoperative electrophysiology for the oculomotor functions are lacking, we used a pragmatic approach with direct bipolar nerve stimulation and EMG monitoring of an oculomotor muscle. We felt this approach to be very helpful for identifying functional regions of the cyst wall. Despite histopathology showing functional tissue, ie, axons in the biopsied cyst wall, the patient's symptoms went on to recover fully within the following years.

CONCLUSION

Cystic lesions of the third cranial nerve are rare. When operating on symptomatic patients, intraoperative neuromonitoring is key to allow for a maximal safe procedure. This holds especially true for cases in which the cyst cannot be resected for functional reasons. Here we present the first case report of

an intrinsic cyst within the third nerve, successfully treated via fenestration after mapping for nerve function.

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Disclosures

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

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