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CASE REPORT

Presumed Pediatric Isolated Oculomotor Nerve Schwannoma - A Case Report

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Abstract: This report presents a healthy 2.5-year-old child exhibiting headache, ptosis, exotropia, and left mydriasis. Initial neuroimaging, including computed tomography (CT), computed tomography angiography (CTA), and standardized magnetic resonance imaging (MRI), failed to identify any lesions; however, high-resolution MRI revealed an ovoid mass adjacent to the left proximal oculomotor nerve within the superior cavernous sinus. This case underscores the necessity for advanced imaging techniques and a thorough diagnostic approach to enhance understanding of this rare pediatric condition. Moreover, it highlights the limited documentation of pediatric oculomotor schwannomas, leading to an inadequate understanding of their diagnosis and management, and emphasizes the need for enhanced awareness and research to establish effective diagnostic protocols, particularly utilizing advanced neuroimaging techniques.

Keywords: headache, peri-tumoral edema, third nerve palsy, high-resolution MRI, steroids

Introduction

Schwannomas are benign slow-growing tumors originating most commonly from Schwann cell layer of the vestibular portion of the vestibulocochlear nerve and rarely from the lower distal cranial nerves, comprising approximately 6% to 8% of all intracranial tumors.[1] Among the various types of schwannomas, isolated pediatric oculomotor nerve schwannomas are exceedingly rare, particularly when not associated with neurofibromatosis.[2] When associated with neurofibromatosis type 2 (NF2), it occurs typically alongside trigeminal nerve schwannomas.[3] The first documented case of an isolated oculomotor nerve schwannoma was reported by Kovacs in 1927, with a limited number of cases subsequently described in the literature,[4,5] therefore their presenting features, and optimal treatment strategies, remain to be fully elucidated.[4]

These lesions may present with characteristics of complete oculomotor nerve palsy, which typically includes a non-reactive, dilated pupil, complete ptosis, and ophthalmoparesis affecting elevation and adduction.[6] In primary gaze, the affected eye may be positioned in abduction, with slight depression and intorsion, commonly referred to as the "down and out" position.[7]

Case Presentation

We report a case involving a previously healthy 2.5-year-old child who presented to our Pediatric Emergency Department with new-onset ptosis of left eye and headache. The child's parents denied any history of trauma or insect bites. Upon physical examination, the child exhibited ptosis, exotropia, and mydriasis in the left eye (Figure 1). An ophthalmological assessment revealed exotropically deviated and abducted left eye with a relative afferent pupillary defect, without any signs of nystagmus. Upon the slit lamp examination, the anterior segment was found to be unremarkable and with no evidence of abnormal pathological findings. Unfortunately, we were unable to assess visual acuity due to the child's lack of cooperation. Neurological examination demonstrated otherwise intact functional cranial nerves, with no evidence of cerebellar signs.

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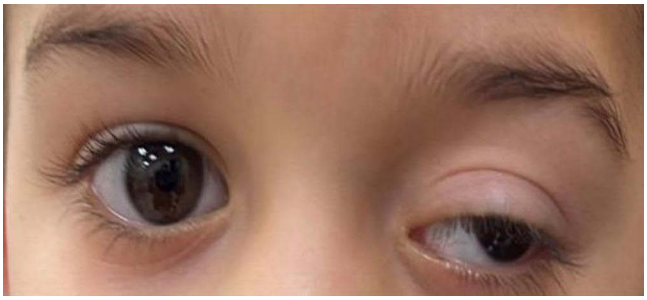

Figure 1 Initial presentation of the child with ptosis, exotropia and mydriasis in his left eye.

The child was admitted to our ophthalmology department and underwent a series of comprehensive evaluation, which included a contrast-enhanced head CT, lumbar puncture, abdominal ultrasound, and both immunological and viral panels, all yielding normal results. CTA revealed no evidence of aneurysms in the anterior or posterior communicating arteries. Subsequently, a standard MRI was conducted; surprisingly, this examination also did not reveal any lesions. During the hospitalization, the decision was made to perform a high-resolution MRI with a thinner slice thickness of 0.5 to 1 mm. This advanced imaging technique successfully identified an ovoid mass measuring 5 mm in diameter., located in the third cranial nerve within the superior aspect of the cavernous sinus, cranial to the internal carotid artery (ICA). MRI was conducted using a 3T MRI scanner, increasing the signal strength of a traditional MRI and providing high quality imaging and a better visualization of the lesions. On T1-weighted images, the lesion exhibited an isointense signal, further detailed the lesion (Figure 2). High-resolution 3D T2-weighted MRI, obtained with a fine 0.5 mm slice thickness, revealed the lesion as hyperintense and homogeneous (Figure 3). Orbital MRI was also requested but revealed no detected anomalies (Figure 4).

We decided to initiate administration of prednisolone at a dosage of 15 mg twice daily for seven days, followed by a tapering down of the dose to 3 mg for an additional week. On the day of discharge, the child exhibited clinical improvement, demonstrating the ability to open his left eye, along with a slight enhancement in ocular movements. The child has been transferred to another medical center, where he had Botox injections to the lateral rectus muscle.

This constellation of signs is attributed to lesions which compress the oculomotor nerve. Such pathology may compromise the parasympathetic fibers of the nerve, which supply the sphincter pupillae muscle. These parasympathetic fibers originate in the Edinger-Westphal nucleus of the midbrain and run on the superficial surface of the third nerve and are thus vulnerable to compressive lesions. As many of the underlying causes may be amenable to surgical treatment strategies, the presence of pupillary involvement in a third nerve lesion is termed as "surgical third-nerve palsy".[7] The differential diagnosis for a surgical third-nerve palsy associated with cavernous sinus lesion is wide and to conclude a final diagnosis, such as Schwannoma, histopathologic examination is necessary.

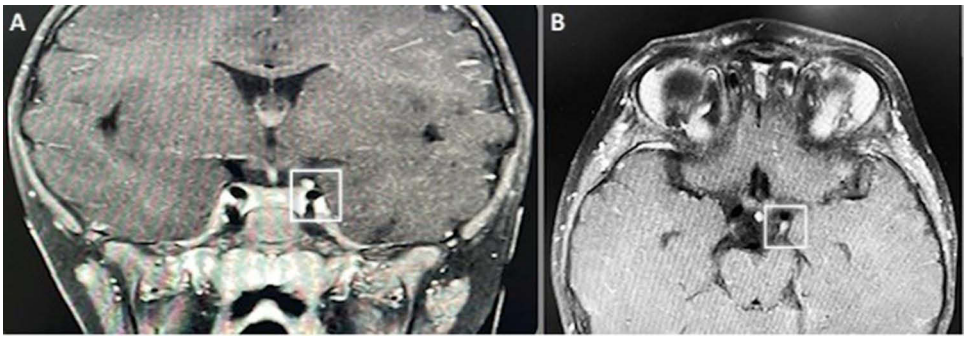

Figure 2 T1-weighted MRI with gadolinium contrast and fat saturation; (**A**) coronal View, (**B**) axial view, an ovoid isointense mass, approximately 5.5 mm in diameter (outlined in the white box), is located within the third cranial nerve at the superior aspect of the cavernous sinus, cranial to the internal carotid artery (ICA). The mass demonstrates homogeneous enhancement following gadolinium administration, consistent with a contrast-enhancing lesion. There is no significant mass effect on adjacent structures.

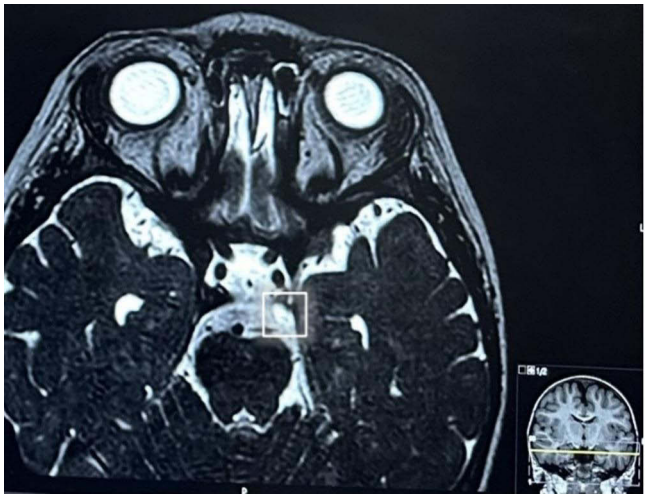

Figure 3 High-resolution 3D T2-weighted MRI sequence using the Vista protocol-axial view: an ovoid hyperintense mass, measuring approximately 5.5 mm in diameter (outlined in the white box) with diffuse enhancement.

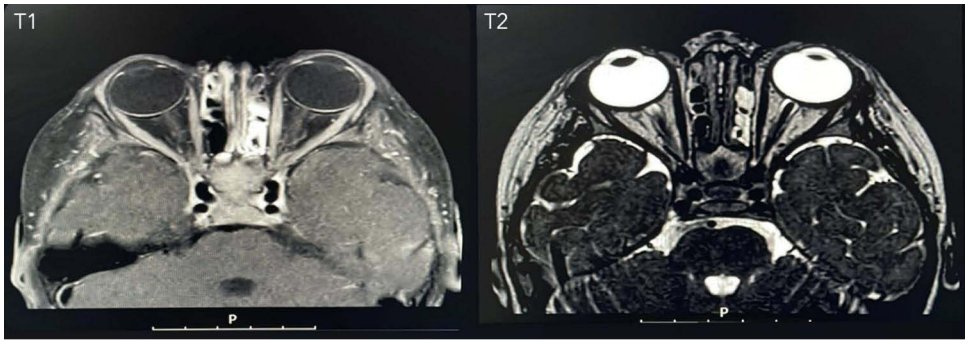

Figure 4 Orbital MRI - T1 and T2 weighted, axial view without gadolinium: both optic nerves are normal and symmetrical, with no abnormal enhancement. The bilateral eye globes and lenses are normal. The structures of the orbital cavities, including the extraocular muscles and eyeballs, are symmetric with no signs of proptosis.

Discussion

In our case, the constellation of signs is attributed to a lesion compressing the oculomotor nerve, resulting in complete third nerve palsy. The oculomotor nerve (cranial nerve III) has significant anatomical complexity during its transition from central to peripheral structures, complicating diagnosis and treatment. Measuring only 0.6 mm at its transition zone, it traverses critical areas, including the interpeduncular cistern, parasellar region, cavernous sinus, and orbital apex.[7]

Third nerve palsy in pediatrics is a relatively uncommon condition, with a variety of potential etiologies. While it is often congenital, arising from intrauterine or birth-related events, it can also develop as a consequence of postnatal factors such as trauma, infections (eg bacterial meningitis), inflammation, tumors or childhood migraines. Although aneurysm of the anterior or posterior communicating artery are rare, certain pediatric populations are at increased risk, including those with polycystic kidney diseases, coarctation of the aorta, or connective tissue disorders such as Ehlers-Danlos syndrome.[8] Additionally, infiltrative conditions such as leukemia or toxic effects related to chemotherapy may contribute to the development of third nerve palsy. Traumatic third nerve palsy typically results from severe, high-energy impacts of the head. The tumor is classified as benign and complete erosion of the encapsulated nerve is a rare occurrence.[9] However, increased growth of the tumor may result in permanent and irreversible cranial nerve damage, increased intracranial pressure (ICP) and at worst midbrain compression.[9] In most cases, oculomotor schwannomas tend to remain stable in size. Therefore, in instances of minimal progression of cranial nerve deficits, an active monitoring approach is often preferred.[10] This strategy has been applied in the case presented.

In this case, the presentation of a child with newly onset complete third nerve palsy, accompanied by a significant headache, heightened the suspicion of an intracranial tumor. Initially, we were unable to identify any lesions despite a comprehensive series of radiological examinations, which included contrast-enhanced head CT, orbital CT, CT angiography (CTA), and cranial MRI. Subsequently, we requested high-resolution MRI scans with a slice thickness of 0.5 mm, which revealed a lesion measuring approximately 5 mm in vertical diameter within the cavernous sinus, originating from the oculomotor nerve.

High-resolution MRI utilizes a smaller field of view (FOV), enhancing detail in targeted regions and providing clearer images of small and complex structures such as the brainstem, cranial nerves, and cavernous sinus.[11] This precision is crucial for detecting millimeter-sized tumors, particularly in cases involving schwannomas and microadenomas. Such imaging is especially beneficial in pediatric cases requiring precise localization of abnormalities.[12] In addition to high-field MRI, Diffusion Tensor Imaging (DTI) is an advanced imaging technique providing valuable quantitative metrics, such as fractional anisotropy (FA) and mean diffusivity (MD), for evaluating nerve fiber axons and fascicles.[13–15] DTI is a non-invasive imaging modality, offering significant advantages in pediatric cases where minimizing radiation exposure is paramount.[16] Its ability to assess the integrity and microstructure of nerve fibers makes DTI an invaluable tool in evaluating both normal and pathological conditions.[17] Consequently, DTI plays a crucial role in detecting subtle pathological changes within peripheral nerves, enhancing diagnostic accuracy and supporting clinical decision-making.[18] Although not utilized in our case, DTI could potentially enhance diagnostic accuracy when assessing nerve pathologies, including oculomotor schwannomas, by offering insights about the differentiation between different types of nerve sheath tumors.[12]

In our case, the suspicion of oculomotor schwannoma was based on high-resolution MRI findings, as a biopsy was not performed for confirmation. The absence of ionizing radiation and noninvasiveness make MRI a particularly advantageous tool when imaging infants.[19] Although DTI was not used, we believe its ability to non-invasively assess nerve structure and function by providing complementary quantitative data makes it a promising tool for future diagnoses. This is especially true in cases where radiological features suggest nerve pathologies like Schwannomas in complex anatomical locations, where surgical access for biopsy is challenging. DTI could enhance diagnostic confidence and guide treatment strategies for nerve-related conditions.

In our case, the clinical presentation of a child with newly onset third nerve palsy, accompanied by a significant headache, heightened the suspicion of an intracranial lesion. Initial imaging studies, including contrast-enhanced head CT, orbital CT, CT angiography (CTA), and cranial MRI, failed to identify any abnormalities. However, a high-resolution MRI with a slice thickness of 0.5 mm eventually revealed a 5 mm lesion located in the cavernous sinus, originating from the oculomotor nerve. This emphasizes the importance of using high-resolution imaging techniques in detecting small, millimeter-sized tumors, especially in complex regions such as the brainstem and cavernous sinus, where smaller lesions can be easily missed with standard imaging protocols.

The child demonstrated substantial improvement following treatment with steroids. The decision to initiate steroid therapy in our patient was based on the potential anti-inflammatory effects and the possibility of reducing peri-tumoral edema. This response to steroids is intriguing and warrants further investigation into the role of inflammation in the symptomatic presentation of oculomotor nerve schwannomas.[20] Botulinum toxin injection is an effective adjunct therapy, by preventing the contracture of the antagonist lateral rectus muscle, facilitating improved ocular alignment and enhancing visual function.[21] Long-term follow-up is essential. Regular ophthalmological assessments, including visual acuity testing, ocular motility evaluation, and repeat neuroimaging, are necessary to monitor tumor growth and manage neurological deficits.[22]

Conclusion

Isolated oculomotor schwannoma in the pediatric population is a rare phenomenon. When suspected based on clinical presentation such as third nerve palsy or signs of increased intracranial pressure it is essential to conduct a thorough series of tests to rule out any congenital or acquired conditions. Upon referral for radiological evaluation, a high-resolution MRI in three phases, both pre- and post-Gadolinium injection,[23] should be performed. To date, there are no specific treatment guidelines available due to the complex anatomical location of these tumors and the generally unfavorable outcomes and prognosis following neurosurgical intervention.

Abbreviations

CT, computed tomography; CTA, computed tomography angiography; DTI, diffusion tensor imaging, FA, fractional anisotropy, ICA, internal carotid artery; ICP, increased intracranial pressure; MD, mean diffusivity; MRI, magnetic resonance imaging; NF2, neurofibromatosis; T, Tesla.

Ethical Approval

Only one case was described not requiring further approval from our Ethics Committee (ZIV Medical Center, Safed).

Consent

Parental written informed consent was obtained for the publication of this case report including the images.

Acknowledgments

In a memory for our beloved eye technician Arie Orr who passed away in a young age and left us alone.

Disclosure

The authors report no conflicts of interest in this work.

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