Optic Nerve Sheath Meningocele: a case report and review of the literature

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Swiss Journal of Radiology and Nuclear Medicine - SJORANM GmbH - Rosenweg 3 - CH-6340 Baar - Switzerland

Abstract

Optic nerve sheath meningocele (ONSM) is a rare condition with only a few cases reported in the medical literature. The etiology is unknown. The condition is characterized by an expansion of the cerebrospinal fluid space surrounding the optic nerve, without associated inflammation or the presence of orbital or cerebral neoplasms at the apex of the orbit. The condition is characterized by the absence of specific symptoms, with the most common being blurred vision and retro-orbital pain. We present the case of a young patient who was admitted to the emergency department at an external hospital. A clinical examination revealed painless right exophthalmos. No additional neurological symptoms were observed. A Computed Tomography (CT) scan and Magnetic Resonance Imaging (MRI) revealed an ONSM.

Keywords: Optic nerve; Sheath, Meningocele; CT; MRI

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Introduction

ONSM is a rare condition consisting of dilation of the nerve sheath that creates a cavity filled with cerebrospinal fluid (CSF) around the optic nerve without any underlying pathology (1, 2, 3). In 1918, Bane described primary dilation of the optic nerve sheath and referred to it as an optic nerve dural sheath cyst. Since then, a few cases have been reported (4, 5, 6).

One theory suggests that the difference in osmotic gradient between the cerebral and perioptic subarachnoid spaces leads to fluid accumulation. Another theory suggests that congenital narrowing of the optic or craniorbital junction may lead to accumulation in the perioptic subarachnoid space (2).

There are no characteristic symptoms associated with optic nerve sheath meningocele. However, blurred vision and retrobulbar pressure are common symptoms (4). An MRI of the orbits is often used to diagnose the condition, as it easily reveals a dilated optic nerve sheath filled with CSF.

Due to the rarity of ONSM, there is currently no consensus on the optimal therapeutic approach.

Case Report

A 16-year-old patient was admitted to the emergency department of an external hospital with painless right exophthalmos. No history of head trauma or other neurological

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deficits was detected. A CT head was performed. The images revealed a well-defined hypodense cystic mass located just behind the right eye, with evidence of displacement of the extraocular muscles. There was no evidence of bone expansion or erosion (Fig. 1)

MRI demonstrate the presence of an expansive cystic process, measuring 19 x 23 x 21 mm, with well-defined by a thin capsule, located in intraconal space of the right orbit. It has a close anatomical relationship with the optic nerve, which causes anterior compression, deformity, and displacement of the eyeball. In addition, the MRI showed signal characteristics equivalent to those of cerebrospinal fluid in all sequences. No significant anomalous enhancement was observed in the walls or within the lesion after the paramagnetic contrast (gadolinium) intravenous injection (Fig. 2).

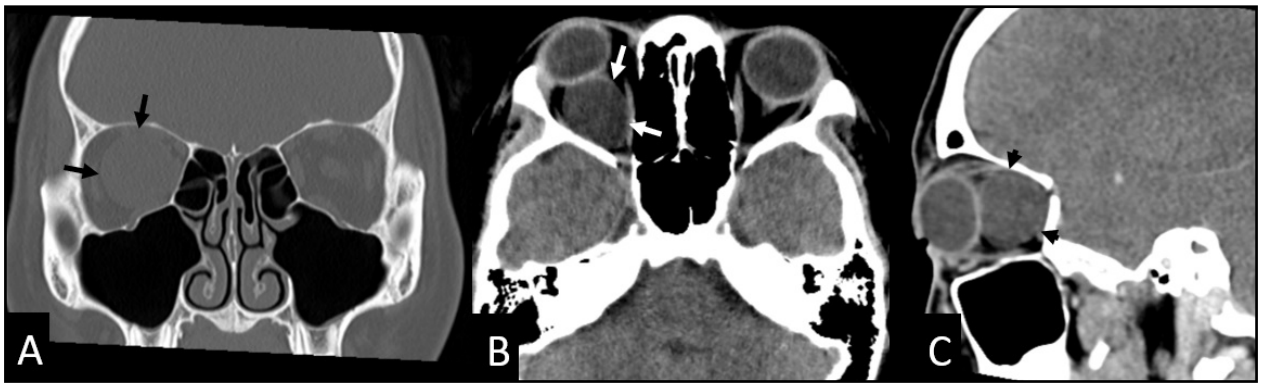

Figure 1: CT: well-defined intraconal cystic mass without bone compromise (black arrows); homogeneous hypodense mass with evidence of extraocular muscle displacement (white arrows), without enhancement after intravenous contrast injection (black arrowheads) A. bone window, B. soft tissues window, C. soft tissues window+contrast.

Discussion

ONSM is a rare condition involving dilation of the nerve sheath. This dilation creates a cavity filled with CSF around the optic nerve without any underlying pathology such as inflammation, orbital, or brain neoplasia at the apex of the orbit. (1, 5). Bane was the first to describe optic nerve sheath dilation in 1918, referring to it as an optic nerve dural sheath cyst (4). Garrity et al. described an "optic nerve sheath meningocele" in a historical article on 13 patients. Since then, a few cases have been documented in the literature (1, 3, 4, 6, 8).

One theory suggests that the difference in osmotic gradient between the cerebral and periorbital subarachnoid spaces leads to fluid accumulation. This assertion is further substantiated by Lunari et al., that documented elevated protein levels in the CSF of an optic nerve cyst (7). Another theory suggests that a congenital narrowing of the optic or cranio-orbital junction may lead to CSF

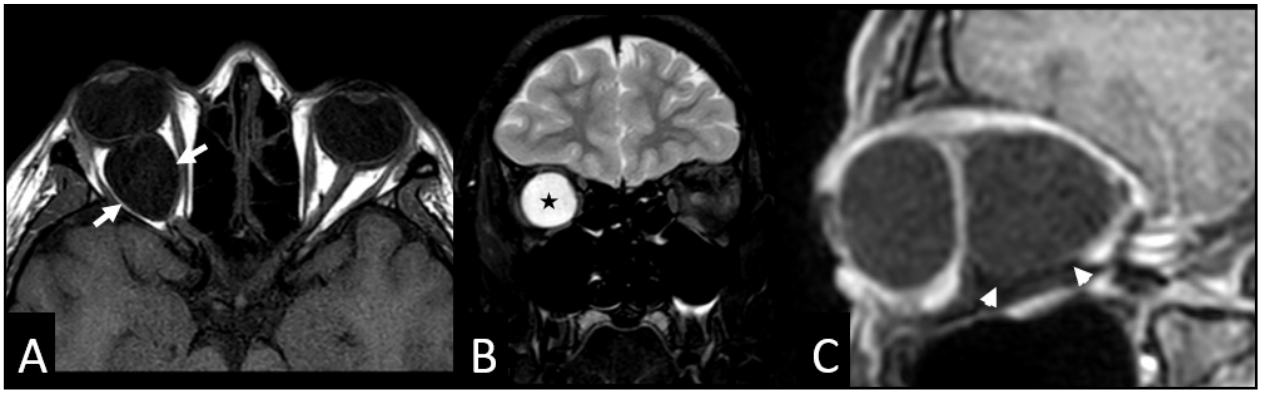

Figure 2: MRI: A) hypointense on T1 WI retroorbital mass with anterior displacement of the globe; B) hyperintense on T2 WI identical signal to cerebrospinal fluid (black star); C) No evidence of enhancement after intravenous contrast injection (white arrowheads)

accumulation in the perioptic subarachnoid space (2, 3).
Enlargement of the optic nerve or optic nerve sheath complex may be indicative of an apical neoplastic mass, such as a meningioma, vascular hamartoma, glioma, neurofibromatosis, von Hippel-Lindau disease, hemangioma, intracranial hypertension or skull-orbital fracture (3, 4, 5).

ONMS does not present with characteristic symptoms. However, blurred vision, headache, retrobulbar pressure, and retroorbital pain are common symptoms (2, 4).
Typically, a MRI scan of the orbits is used to diagnose the condition, as it allows for visualization of a dilated optic nerve sheath filled with CSF (5, 8). Some T2-weighted coronal images show a “bull´s eye” appearance, representing the expanded CSF spaces around the optic nerve (3). The use of fat suppression techniques is an effective method for ruling out the presence of intraorbital tissue lesions and optic nerve compression (4).
Furthermore, MRI provides a more detailed differential diagnosis of ONSM, including other optic nerve tumor lesions, such as gliomas or meningiomas, especially cystic ones (2, 3, 4, 6).

Due to the rarity of ONSM, there is currently no consensus on the optimal therapeutic approach. Treatment is customized to meet each patient's specific needs (1). In some cases, medical therapy with oral acetazolamide has shown positive results (3). In cases where no improvement has been observed with medical treatment, surgical decompression is the preferred option, especially when vision loss is progressive (1, 5, 6, 7, 8). In situations where progress is minimal or nonexistent, observation may be a valuable approach (2).

Conclusion

ONSM is a rare disorder with no characteristic symptoms, characterized by the collection of cerebrospinal fluid in the subarachnoid space of the intraorbital portion of the optic nerve. The etiology of this condition remains unclear, underscoring the necessity for comprehensive diagnostic imaging. Among the available imaging modalities, MRI is regarded as the preferred study to guide treatment, which can range from conservative therapy to complex surgical repair. Therefore, it is essential to implement individualized treatment protocols to optimize patient outcomes.

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Declarations
Consent for publication: The author clarifies that written informed consent was obtained and the anonymity of the patient was ensured. This study submitted to Swiss J. Rad. Nucl. Med. has been conducted in accordance with the Declaration of Helsinki and according to requirements of all applicable local and international standards. All authors contributed to the conception and design of the manuscript, participated in drafting and revising the content critically for important intellectual input, and approved the final version for publication. Each author agrees to be accountable for all aspects of the work, ensuring its accuracy and integrity.
Competing interests: None.
Funding: No funding was required for this study.

Conflict of interest:

The authors declare that there were no conflicts of interest within the meaning of the recommendations of the International Committee of Medical

Journal Editors when the article was written.

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