Unilateral papilledema revealing an intracanalicular optic nerve sheath meningioma

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ABSTRACT

A 43-year-old female is referred to our clinic for evaluation of an incidentally discovered unilateral papilledema in her right eye of unknown cause. She had no past ocular or medical history. Examination revealed a corrected-distance visual acuity of 20/20 in both eyes. Posterior segment examination showed an elevated right optic nerve head with blurred edges. Ultrasonography failed to reveal optic disc drusen. Visual field testing showed enlargement of the blind spot in the right eye. Magnetic resonance imaging of the brain and orbit was done and revealed enlargement of the intracanalicular and prechiasmatic part of the right optic nerve and a diagnosis of a presumed right intracanalicular optic nerve sheath meningioma was made. She was managed conservatively and several years later remains asymptomatic.

ARTICLE HISTORY

Received 12 May 2018
Accepted 25 May 2018

KEYWORDS

Magnetic resonance imaging; optic canal meningioma; intracanalicular optic nerve sheath meningioma; primary optic nerve sheath meningioma; unilateral papilledema

Photo essay

A 43-year-old female is referred to our clinic for evaluation of an incidentally discovered unilateral papilledema in her right eye of unknown cause. She had no past ocular or medical history. Examination revealed a corrected-distance visual acuity of 20/20 in both eyes. Color vision was 12/12 in both eyes using Ishihara plates. Pupillary examination was normal. External ocular examination and ocular motility were normal. Anterior segment examination was unremarkable. Posterior segment examination of the right eye revealed an elevated optic nerve head with blurred edges. The left optic nerve head was completely normal. B-scan ultrasonography was done and showed elevation of the right optic nerve head with no evidence of disc drusen. Visual field testing revealed an enlarged blind spot in the right eye and a normal visual field in the left eye. Magnetic resonance imaging (MRI) of the brain and orbit was done, which revealed enlargement of the intracanalicular and prechiasmatic part of the right optic nerve, (Figure 1) and a diagnosis of a presumed right intracanalicular optic nerve sheath meningioma (ONSM) was

made. The patient was managed conservatively due to the absence of visual complaints and following several years of follow-up remains asymptomatic.

ONSMs are the second most common optic nerve tumors following gliomas.¹ They can be primary or secondary. In primary ONSM, the tumor arises from the orbital part, or less commonly from the canalicular part, of the dural sheath of the optic nerve. In secondary ONSM, they arise from the dura on the planum sphenoidale and spread within the optic nerve sheath. Most commonly, meningiomas will spread locally in the subarachnoid space; however, direct tissue invasion may occur.² They are most common in middle-aged females with a reported mean age of presentation of 42.5 years.¹ Meningiomas arising within the optic canal are rare and represent a clinical and radiological diagnostic challenge.³ Diagnosis is usually delayed in these cases, and several reported cases have been initially misdiagnosed as optic neuritis.^{3,4} This however did not occur in our case, perhaps due to the absence of visual complaints. Clinically, there is progressive visual loss with optic disc edema or atrophy, and some patients develop optociliary shunts.^{3–5} Visual field defects include central scotoma, enlargement

Figure 1. MRI imaging of the brain and orbit. (A) T1-weighted coronal image at the level of the optic canal showing an enlarged right optic nerve (white arrow). (B) T1-weighted coronal image showing enlargement of the prechiasmatic part of the right optic nerve (white arrowhead) (C) T1-weighted axial image showing mild enlargement of the right optic nerve (black arrow). (D) T2-weighted axial image showing an increased fluid signal around the right optic nerve (black arrowhead).

of the blind spot, and generalized depression.⁵ Radiological diagnosis is best achieved with high-spatial-resolution MRI of the cranium and orbit with contrast enhancement and fat suppression.³ On MRI, ONSM appears isointense to the optic nerve on T1- and T2-weighted images with the presence of en-plaque or nodular tumor growth.^{3,4,6} Enlargement of the intraorbital optic nerve may also be present with a surrounding fluid signal on T2-weighted MRI.⁶ Management by observation and follow-up is the standard of care for ONSM with no visual affection; however, some have advocated a more aggressive early approach since the tumor is slowly growing and can lead to

complications if left untreated.^{7,8} Interventions are in the form of stereotactic fractionated radiotherapy or surgery.^{7,8}

Acknowledgment

None.

Ethical approval

This report was approved by Cairo University research ethics committee and followed the tenets of the Declaration of Helsinki.

Funding

None.

Conflicts of interest

None.

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