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Case Report

Primary optic nerve sheath schwannoma: A case report and literature review ☆

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ABSTRACT

Schwannomas are benign tumors of the peripheral nervous system that arise from Schwann cells. Intracranial schwannomas most commonly arise from the vestibulocochlear nerve, followed by the fifth nerve. However, optic nerve sheath schwannomas are very rare due to the lack of Schwann cells on it. A few exceptional cases of optic nerve sheath schwannoma have been described in the literature. In this article, we report a rare case of primary optic nerve sheath schwannoma in a 48-year-old woman who presented with progressive visual loss and left-sided proptosis. Magnetic resonance imaging revealed a well-demarcated intraconal orbital mass with cystic appearance and rim enhancement. The diagnosis of optic nerve schwannoma was proposed and confirmed histologically after surgical resection.

Introduction

Schwannomas account for 8% of all primary intracranial tumors. They most commonly arise from the vestibulo-cochlear and trigeminal nerves [1]. Their intraorbital location accounts for 2%-5% of all operated intraorbital tumors [1]. Schwannomas may be isolated or associated with neurofibromatosis type 2 [2]. Their diagnosis is challenging and, in the absence of known fibromatosis, is based on MRI data. Treatment is exclusively surgical.

Case report

A 48-year-old female patient with no past medical history, especially no personal or family history of neurofibromatosis, presented with periorbital pain, and visual loss in the left eye associated with grade II exophthalmia, which had been progressing for 3 years. The lesion gradually increased in size and visual acuity gradually decreased. Ophthalmic examination revealed proptosis of the left eye, and orbital movements were restricted with nonreducible, axial exophthalmia. Visual acu-

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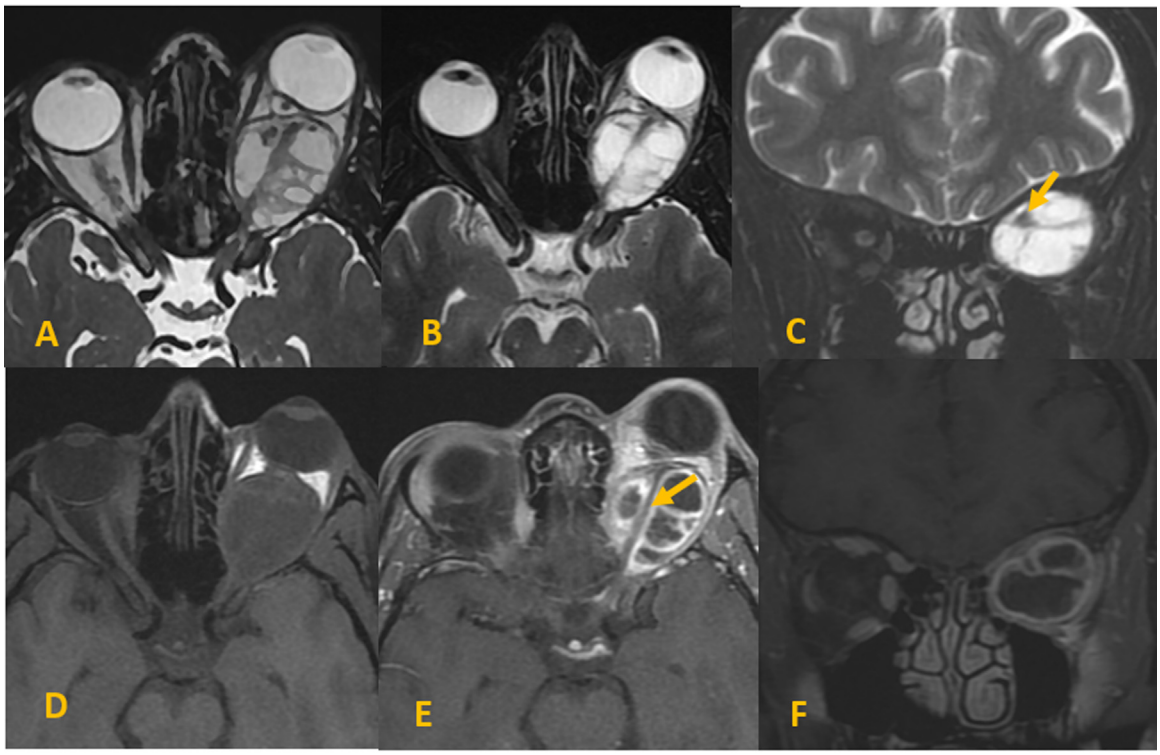

Fig. 1 – Intraconal process around the left optic nerve (yellow arrow) with low signal on T1-weighted ponderation (D), high signal on T2-weighted images (A, B, and C), and rim enhancement after intravenous gadolinium administration (E and F). The orbital mass caused grade II exophthalmia.

ity was 8/10 (right) and 0/10 (left). No other abnormalities were found in the clinical examination of the patient. Magnetic resonance imaging (MRI) revealed an intraconal process centered on the left optic nerve with low signal on T1-weighted images, hypersignal on T2-weighted images, cystic appearance, and thin rim enhancement after intravenous gadolinium administration. The orbital mass caused grade II exophthalmia (Fig. 1). There was no intraorbital fat infiltration. The diagnosis of optic nerve schwannoma was suspected based on MRI data. An orbitotomy was performed with partial excision of the tumor due to heavy adherence to the optic nerve, and histopathologic examination confirmed the diagnosis of schwannoma. One year later, clinical examination showed regression of proptosis and no residual pain, but no improvement in visual acuity. Orbital MRI showed a small retro-orbital residual process with regression of exophthalmia (Fig. 2).

Discussion

Orbital tumors are common and associated with various etiologies. They have always been a challenging diagnosis for radiologists. Tumors of neurogenic origin are rare in the orbit, accounting for approximately 1% of all orbital tumors [3]. Orbital schwannomas are rare, accounting for approximately 2%-5% of orbital tumors. However, only a few case reports of orbital schwannomas are intraconal. According to the recent literature, only 6 cases of optic nerve schwannoma have been reported [4]. These tumors are most seen in adulthood with a median age of presentation of 40 years [3]. They are localized, usually unilateral, and rarely malignant, and their diagnosis is suggested by MRI. Their treatment is exclusively surgical [1].

Optic nerve schwannoma (ONS) is unusual because of the absence of Schwann cells. The possible origin of ONS is ectopic neural crest Schwann cells and perivascular Schwann cells accompanying sympathetic nerves innervating orbital blood vessels. The exact site of nerve origin may not be identifiable even intraoperatively [4].

The clinical features of ONS are not specific. Like other orbital tumors, they are mainly exophthalmia, orbital pain, and visual acuity disturbances. Diplopia may also be a common complaint. Invasion or compression of parasympathetic fibers may cause a pupil size change. Optic nerve atrophy or papilledema and choroidal folds are also common findings [1].

On imaging, ONS appears as a solitary orbital retrobulbar mass that is predominantly smooth, ovoid, and well-circumscribed. The location of ONS is most commonly in the superior orbit in the direction of the nerve. Its long axis is usually anteroposterior. On CT scan, the tumor is isodense or slightly hyperdense compared to the brain and often shows homogeneous or heterogeneous moderate to marked contrast enhancement after intravenous contrast injection [1]. On MRI, schwannomas commonly show low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Homogeneous contrast enhancement has been re-

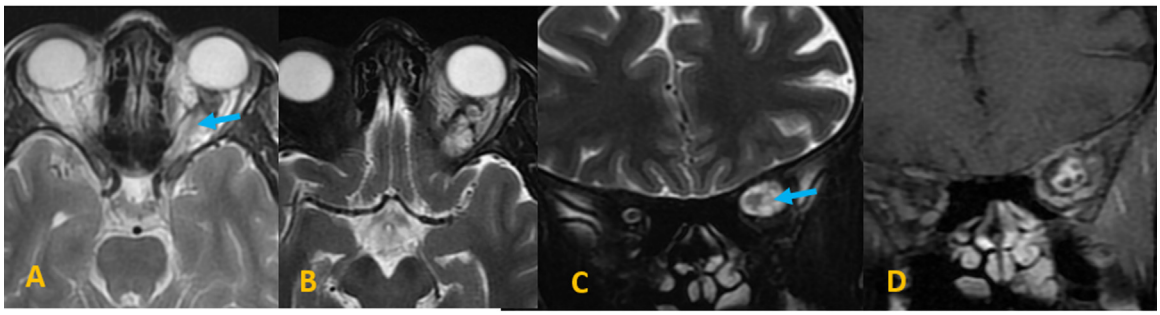

Fig. 2 – Orbital MRI after one year of follow-up. A small retro-orbital residual process (blue arrow) with high signal on T2 weighted ponderation (A, B, and C) and a heterogeneous enhancement (D).

ported. ONS may appear as a well-defined lesion with mixed components [5,6]. In our case, the tumor has a cystic appearance with a high signal on T2 images and peripheral rim enhancement. This cystic appearance may be due to a mucinous or microcystic component, hyaline degeneration of the tumor, poor vascular supply and tumor necrosis, or tumor hemorrhage followed by blood resorption [5]. Because of its extreme rarity, ONS could be misdiagnosed on imaging for more common lesions such as optic meningioma, hemangioma, or glioma [7]. Histopathologic examination is required to confirm the diagnosis [1].

Complete surgical excision of these tumors usually results in a definitive cure without recurrence. In our case, complete excision was not possible because of the strong adhesion to the retrobulbar part of the optic nerve. Malignant transformation is possible, especially in children. In such cases, rapid progression and pain are signs of malignancy. In addition, optic nerve schwannomas with heavy nerve attachment have a poor visual prognosis [5].

Conclusion

Optic nerve schwannomas are very rare and have no specific clinical or radiologic appearance. They are often difficult to diagnose and can be confused with other intraorbital tumors. However, radiologists should be aware of this diagnosis, especially when it presents as a solid or cystic mass around the optic nerve with heterogeneous enhancement. Histologic confirmation is always required, and surgical management is the rule if symptomatic.

Patient consent

Informed consent was obtained from the patient to publish this case report and accompanying images.

REFERENCES

[1] Barhmi I, Mahdoufi R, Khallouq A, Tatari M, Abada R, Roubal M, Mahtar M. Uncommon presentation of orbital schwanomma: a case report. Int J Surg Case Rep 2016;26:173–5. doi:10.1016/j.ijscr.2016.07.045.
[2] Leruez S, Gohier P, Menei P, Milea D. Schwannome du nerf optique. J Franç Ophtalmol 2013;36(3):49–53. doi:10.1016/j.jfo.2012.01.014.
[3] Hemalatha AL, Vani D, Giripunja M, Bharathi M, Geetanjali S. Rare retro-orbital intraconal occurrence of benign schwannoma - a case report. J Clin Diagn Res 2013;7(12):2964–5. doi:10.7860/JCDR/2013/6827.3810.
[4] Sharma A, Singh D, Saran R. Primary optic nerve sheath schwannoma: a case report. Br J Neurosurg 2021;8:1–3. doi:10.1080/02688697.2020.1869181.
[5] Kashkouli MB, Abdolalizadeh P, Jafari S, Shahrzad S, Karimi N. Is primary optic nerve sheath schwannoma a misnomer? Report of two cases and literature review. Orbit 2019;38(5):419–23. doi:10.1080/01676830.2018.1545239.
[6] Leruez S, Gohier P, Menei P, Milea D. Schwannome du nerf optique. J Franç Ophtalmol 2013;36(3):49–53. doi:10.1016/j.jfo.2012.01.014.
[7] Ramey WL, Arnold SJ, Chiu A, Lemole M. A rare case of optic nerve schwannoma: case report and review of the literature. Cureus 2015;7(4):e265. doi:10.7759/cureus.265.