Case Report/Series

Atypical orbital primary optic nerve sheath meningioma with severe disfiguring proptosis: an alternative surgical approach

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ABSTRACT

Primary optic nerve sheath meningioma is generally a benign tumor. In rare instances, however, the growth rate and intraocular and intracranial extensions can be highly aggressive, especially in children, leading to poor prognosis. Here, we reported a case of a 24-year-old woman who presented with left eye swelling for 3 years. This was associated with blurred vision, retrobulbar pain, and redness. On examination, the left eye was severely proptosed with complete ophthalmoplegia. Magnetic resonance imaging showed an extensive tumor occupying the whole left orbital cavity with a disfigured eyeball. However, no intracranial extension was observed. Interestingly, complete surgical excision was feasible via transconjunctival anterior orbitotomy without bone removal. The histopathological examination confirmed the diagnosis of optic nerve sheath meningioma. Adjunct radiotherapy was given. On a follow-up after 2 years, left enophthalmos with esotropia was observed.

KEYWORDS enophthalmos, esotropia, meningioma, ophthalmoplegia

Primary optic nerve sheath meningioma (PONSM) is a benign tumor originating from the meningothelial cap cells of the arachnoid villi. PONSM can develop at any location along the entire course of the optic nerve sheath with practically no mortality rate[1,2] and accounts for approximately 1–2% of all meningiomas and 5–10% of all orbital tumors.[3,4]

The optimal treatment strategy for PONSM remains controversial. Watchful waiting is recommended in cases with no vision loss, whereas radiotherapy is indicated when vision is affected. In cases of significantly impaired vision or blindness, disfiguring proptosis, or intracranial extension, surgery alone or in combination with radiotherapy is indicated.[2,5,6] Although benign, PONSM can exhibit aggressive tumor growth, causing intraocular extension, intracranial extension, and severely disfiguring proptosis, especially in children.[7–9] Various surgical approaches have been described for orbital PONSM requiring surgery. This case report described our surgical approach of transconjunctival

anterior orbitotomy, sparing the bone, optic nerve, and eyeball for extensive atypical orbital PONSM. The surgery was less invasive and achieved complete surgical excision in an aesthetically acceptable manner.

CASE REPORT

Informed consent was obtained from the patient before publishing this case report. A 24-year-old woman with no known medical illness presented with progressive left eye swelling for 3 years, associated with blurred vision, retrobulbar pain, and redness. She denied any history of trauma, loss of appetite, weight loss, or neurological deficits.

The patient's left eye was severely disfigured, proptosed (Hertel exophthalmometry >36 mm), and exposed (Figure 1, a and b). Visual acuity was counting fingers at one foot (CF1FT) with an intraocular pressure of 12 mmHg. Generalized conjunctival redness with signs of exposure keratopathy was also observed. The pupil was mid-dilated with a limited posterior segment view, partly due to severe keratopathy and a distorted eyeball. The left eye showed complete ophthalmoplegia. The right eye and other systemic examination results were unremarkable.

Complete blood count, renal function, and liver function were normal, and tumor markers (alpha-fetoprotein, carcinoembryonic antigen, CA 19-9, and CA 125) were not elevated. Contrast-enhanced magnetic resonance imaging (MRI) of the brain and orbit showed a large left intraconal orbital mass measuring 6.7 cm × 4.7 cm x 4.9 cm with a severely proptosed eyeball (50 mm). The mass showed mild homogeneous contrast enhancement. The extraocular muscles were stretched and displaced with no clear plane between them. The optic nerve was stretched and laterally deviated. Remodeling of the left lamina papyracea was also observed. No intracranial extension was observed (Figure 1c).

The patient underwent tumor removal via bone-sparing transconjunctival anterior orbitotomy. Intraoperatively, a 360-degree conjunctival peritomy was performed. All rectus muscles were isolated. A lateral canthotomy was performed with the disinsertion of the superior and medial rectus muscles to enable better access to the posterior orbit. The mass was separated from the recti muscles, posterior eyeball, and optic nerve before being excised into fragments until to the orbital apex. Total tumor excision was performed by sparing the eyeball, optic nerve, and rectus muscles. The superior and medial rectus muscles were reinserted with Vicryl 6/0 sutures, and the lateral canthotomy was opposed using Vicryl 7/0 sutures. The conjunctival peritomy was sutured using Vicryl 8/0 sutures (Figure 2).

Her hemodynamic status remained stable intraoperatively with no oculocardiac reflex during manipulation of the extraocular muscles and eyeball. The estimated intraoperative blood loss was 700 ml, and she was transfused with one pint of packed cells. Postoperatively, her vision was static, with no signs of anterior segment ischemia. Postoperative eye appearance on Day 1 is shown in Figure 1, d and e. Histopathological examination confirmed the diagnosis of orbital meningothelial meningioma (PONSM) of the World Health Organization grade 1. Given the large, fast-growing tumor, adjunct radiotherapy was administered to the left eye. Repeated MRI scan of the orbit and brain 18 months post-surgery showed enophthalmos in the left eye with no evidence of recurrence or intracranial extension (Figure 1f). Two years after surgery, visual acuity in the left eye remained at CF1FT, with left eye enophthalmos and esotropia (Figure 1g).

DISCUSSION

Optic nerve sheath meningiomas commonly present with progressive, painless vision loss, or visual field defect.[10] Proptosis is seldom as the initial presentation. Typically, the proptosis ranges from 2–5 mm.[3] Although rare, severe or extreme proptosis may occur in atypical cases. Amoli et al[7] have histologically confirmed benign PONSM with aggressive behavior in a 20-year-old woman. The patient presented with severe proptosis with an intracranial and intraocular extension.

Surgery is indicated if PONSM causes blindness, disfiguring proptosis, or any intracranial extension.[5] However, bleeding from a highly vascularized tumor poses a significant challenge during the surgical excision of severe proptosis or a large tumor. Moreover, bleeding affects the visualization of the tumor and other structures, providing limited space for manipulation during surgery. In the present case, the estimated blood loss was 700 ml, and the patient required a transfusion of one pint of packed cells. The post-transfusion hemoglobin was 10.4 g/dl.

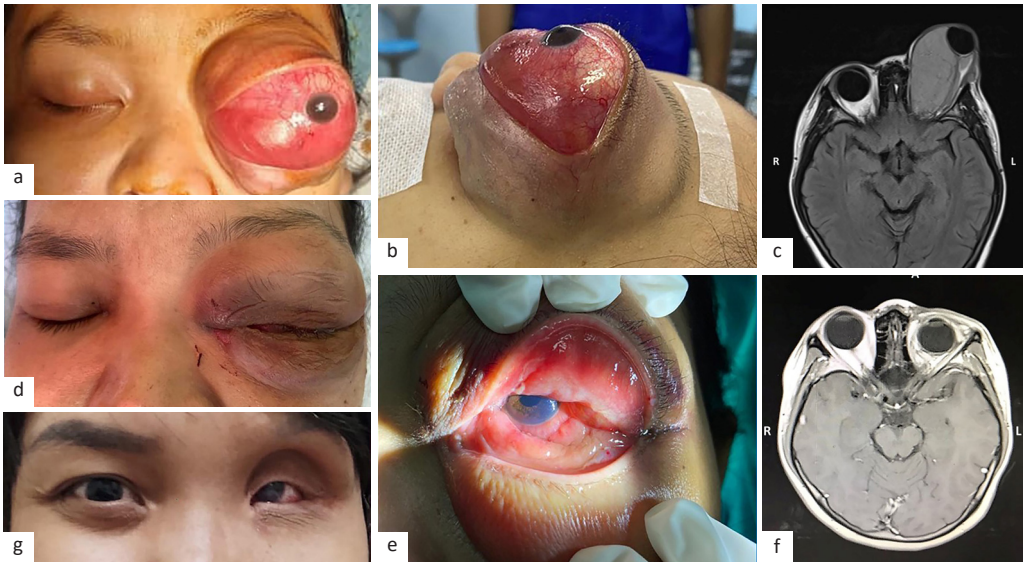

Figure 1. Pre- and postoperative ocular features. (a & b) Left eye showing severe disfiguring proptosis, lagophthalmos, and generalized conjunctival redness preoperatively; (c) MRI brain and orbit with contrast showing left orbital mass measuring 6.7 cm × 4.7 cm × 4.9 cm with severe left eye proptosis and distorted eyeball; (d & e) the eye appearance on postoperative Day 1; (f) MRI brain and orbit showing left eye enophthalmos with no evidence of recurrence or intracranial extension; (g) the eye appearance in 2 years postoperative. MRI=magnetic resonance imaging

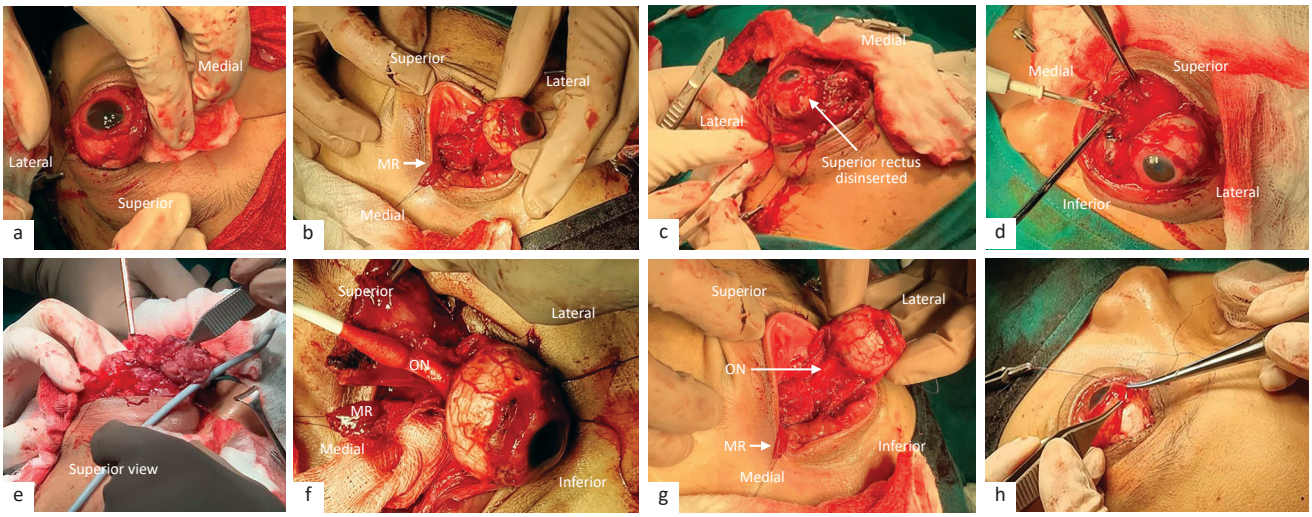

Figure 2. Surgical procedures. (a) Conjunctival peritomy; (b–d) disinsertion of superior and MR; (c) lateral canthotomy; (d & e) separation of the tumor from the ON, recti muscles, and the eyeball with excision of the tumor in fragments; (f & g) the eyeball, ON, and recti muscles were spared; (h) closure of conjunctiva. MR=medial rectus muscle; ON=optic nerve

Separation and manipulation of the extraocular muscles and eyeball from the tumor have a high risk of inducing an oculocardiac reflex during surgery, which was not observed in the present case.[11] If not done meticulously, tumor separation from the eyeball, and more from the optic nerve to achieve complete surgical excision, will further compromise postoperative visual acuity. Postoperative loss of visual acuity occurs in 94% of cases due to a compromised pial vascular plexus, direct damage to the optic nerve intraoperatively, or incomplete tumor resection with recurrence.[3] In the present case, no worsening of visual acuity was noted postoperatively.

While treating orbital PONSM, the primary objective is to achieve complete tumor excision without compromising vision or aesthetic components. Many surgical approaches have been described for tumor excision, such as anterior and lateral orbitotomy and

endoscopic endonasal and transcranial approaches, with or without sparing the optic nerve.[12–15] In many instances, mutilating exenteration has been performed for large tumors with severe proptosis.[7,12,14] However, this procedure typically disfigures the patient, which could lead to depression and low self-esteem.[16,17] In the present case, we decided to perform transconjunctival anterior orbitotomy with sparing of the optic nerve and eyeball. The procedure was successful, thus enabling the avoidance of disfigurement following exenteration.

In conclusion, radical surgical interventions, such as exenteration, may not always be the only option in treating orbital PONSM with severe disfiguring proptosis and blindness. Other options should be considered, particularly in young patients. Follow-up of a patient with an orbital tumor is warranted to detect early recurrence. Our experience in managing this case is worth sharing to benefit the ophthalmic fraternity by enhancing surgical treatment options in orbital PONSM.

Conflict of Interest

The authors affirm no conflict of interest in this study.

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