CASE REPORT

Spontaneous globe rupture in a patient with anaplastic spheno-orbital meningioma

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SUMMARY

Anaplastic meningioma is rare and has always been linked to a grave prognosis. We present a case of unilateral anaplastic spheno-orbital meningioma (SOM), which was complicated by spontaneous globe rupture and invasive recurrences. A 55-year-old female presented with extreme non-axial proptosis in the right eye with lateral globe displacement a month post-radiotherapy following the fourth surgery for the right SOM. Compressive optic neuropathy led to non-perception of light (NPL) in the right eye, while the left eye vision remained at 6/6. Refusing evisceration, the patient experienced spontaneous right globe rupture after 1 month. Brain magnetic resonance imaging showed an advanced exophytic right infiltrative SOM. Bevacizumab therapy failed, necessitating a fifth surgery. Six months postoperatively, SOM infiltration caused NPL in the left eye. The patient's general health declined, leading to death after 3 months. SOM is a blinding condition that can be associated with high morbidity and a poor prognosis for survival.

INTRODUCTION

Meningiomas are primary intracranial neoplasms arising from arachnoid cap cells and constitute about one-third (37.6%) of all primary brain tumours, with an incidence rate of 8.81 in 100 000.[1-2] They are typically slow-growing tumours which can be invasive. SOM is a unique subset of meningioma that arises from the sphenoid ridge with an orbital extension, accounting for 2 to 9% of all intracranial meningioma and only 2% of all orbital lesions.[3-4] The vast majority of SOMs are benign. Anaplastic SOMs are rare and potentially manifest itself with an aggressive clinical course. We reported an advanced presentation of anaplastic SOM with spontaneous globe rupture, followed by invasive recurrences, leading to bilateral vision loss and finally loss of life.

CASE PRESENTATION

A 55-year-old female was diagnosed with right SOM at the age of 40. The patient has no other medical comorbid and had four previous surgeries for SOM with histological transformation from World Health Organisation (WHO) grade I to grade III. Her right proptosis significantly reduced after each debulking surgery and a prior course of radiotherapy (Table I). However, her right visual acuity (VA) had declined from 6/6 to non-perception of light (NPL) after 5 years of disease onset due to tumour compression into the right optic nerve. She presented with right severe proptosis 1 month after undergoing radiotherapy following her fourth surgery. This occurred around 3 months after the fourth surgical procedure (Figure 1A). The proptosis rapidly worsened with a complete inability for eyelid closure and eye irritation. Examination showed right severe non-axial proptosis, dry ocular surface with intraocular pressure (IOP) of 22 mmHg and total restriction of right ocular motility (fifth recurrence). Left eye examination was unremarkable. The patient was counselled for evisceration but opted for moist chamber, intensive lubrication and IOP lowering agents. She defaulted her subsequent appointment and only presented after 1 month, complaining of serous eye discharge for 3 days. She also had intermittent generalised headaches, nose block and anosmia, but denied having trauma to the right eye, reduced hearing, shortness of breath, loss of appetite or loss of weight.

Examination showed a huge orbital mass occupying the right periorbital area, with some lobulations medially. There was a spontaneous right globe rupture with no visible anterior segment structure, leaving the posterior sclera exposed, surrounded by severe conjunctiva chemosis (Figure 1B-C). The residual posterior eyeball structure was distorted and was pushed laterally by the orbital mass. There was a reduced sensation on the right ophthalmic and maxillary distributions of the trigeminal nerve. Left eye VA remained 6/6 with normal anterior segment, intraocular pressure, fundus findings and ocular motility. Other neurological and systemic examinations were normal.

Magnetic resonance imaging (MRI) brain and orbit revealed marked tumoral growth of the SOM with lobulated exophytic component. The mass extends to the ethmoid and sphenoid sinuses, right cavernous sinus, anterior and medial cranial fossae causing mass effect on the anterior temporal and inferior frontal lobes (Figure 2A). The patient was put on Gutt

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Table I: Patient's clinical course in chronological order prior to presentation

Date	Age	Clinical presentation	Clinical status	Management	Histopathology
2015 (Aug)	49	Right proptosis VA RE 6/6, LE 6/6, No RAPD IOP BE normal BE full ocular motility	First presentation	RE lubrication Right craniotomy and tumour debulking (30/10/2015) - First surgery - Simpson grade 4 - Proptosis resolved	WHO grade I meningioma
2018 (May)	52	Recurrent right proptosis VA RE 6/7.5, LE 6/6 RE RAPD positive Grade 1 IOP BE normal BE full ocular motility	First Recurrence	RE lubrication Radiotherapy 60Gy 30 fractions (24/6-3/8/2018) - Proptosis improved	NA
2020 (Oct)	54	Worsening right proptosis Loss of right eye vision VA RE NPL, LE 6/6 RE RAPD positive grade 4 IOP BE normal Ophthalmoplegia with restriction on temporal, superior and inferior gaze	Second recurrence	RE intensive lubrication Right craniotomy and tumour debulking (17/11/2020) - Second surgery - Simpson grade 4 - Proptosis improved	WHO grade II meningioma
2021 (Jan)	55	Worsening right proptosis VA RE NPL, LE 6/6 IOP BE normal Ophthalmoplegia with restriction on temporal, superior and inferior gaze	Third Recurrence	RE intensive lubrication Right craniotomy and tumour debulking (16/2/2021) - Third surgery - Simpson grade 4 - Proptosis improved	WHO Grade II meningioma
2021 (Jul)	55	Worsening right proptosis Headache Nose block and loss of smell VA RE NPL, LE 6/6 IOP RE 22 mmHg, LE normal Ophthalmoplegia with restriction on all directions of gaze	Fourth Recurrence	RE intensive lubrication Gutt Latanoprost 0.005% nocte RE Right craniotomy and tumour debulking, combined with endoscopic excision of tumour and skull base reconstruction (12/8/2021) - Fourth surgery - Simpson grade 4 - Proptosis improved	WHO Grade III meningioma

BE: Both eyes; IOP: Intraocular pressure; LE: Left eye; NPL: Non perception of light; RE: Right Eye; VA: Visual acuity; WHO: World Health Organisation

chloramphenicol four times a day, and swab culture and sensitivity from the right residual globe showed no growth. A course of seven cycles of intravenous bevacizumab 10 mg/kg 2 weekly was given for treatment-refractory SOM. However, repeated MRI brain after completing bevacizumab showed a marked increase in size of the spheno-orbital mass, fungating beyond the right bony orbit (Figure 2B). The mass infiltrated the right middle and superior nasal meatus, bilateral posterior ethmoidal sinuses, right maxillary sinus, anterior cranial fossa and right temporal fossa, resulting in white matter oedema and midline shift.

The patient opted for another craniotomy and tumour excision after multi-disciplinary team discussions were held. A preoperative-tumour embolization was done prior to the surgery, with approximately 90% of the tumour with the feeders mainly from the right internal maxillary artery and right facial artery were embolised. Bifrontal craniotomy and tumour excision were carried out. Maximum safe resection surgery (Simpson grade 4) was done, to remove the soft tissue component that occupied the entire intraconal cavity, along with the excision of the intradural tumour that was attached to the right sphenoid wing and extradural component that infiltrated the anterior and posterior ethmoid sinus, as well as the sphenoid sinus. Histopathology showed epithelioid neoplastic cells with prominent nucleoli, ill-defined cytoplasmic borders and brisk mitotic figures ranging from 14- to 7/10 hpf with aberrant form identified which consistent with anaplastic meningioma, CNS WHO grade III (Figure 3A-C).

During an ophthalmology review at 6 months after the fifth tumour debulking surgery, the patient mentioned that her left eye VA had gradually deteriorated over a period of 1 month. Examination revealed right anophthalmia with a clean and well-healed surgical wound. The left eye VA was NPL with pale disc on the left eye on fundus examination. Left eye IOP was 12 mmHg and there was no demonstrable left proptosis or restriction in ocular motility. Neuroimaging confirmed the recurrence of the tumour (sixth recurrence), extending into the left orbit, encasing and compressing the left optic nerve (Figure 2C). The patient opted for conservative management, but ultimately became bedridden due to disease progression, leading to her demise 3 months later.

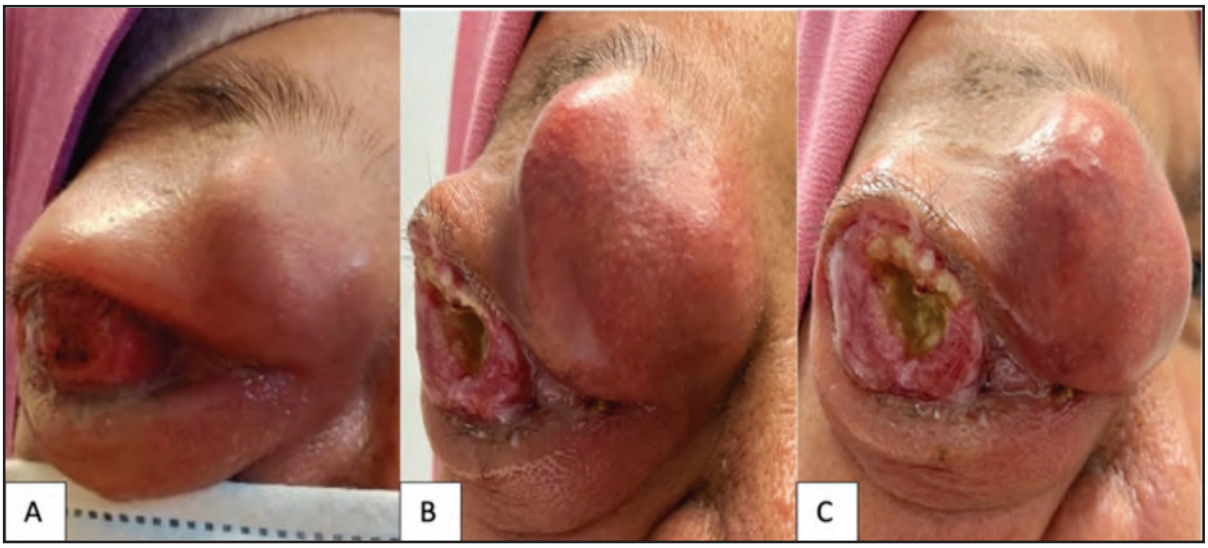

Fig. 1: Patient's photo (A) at presentation of fifth recurrence shows right severe non-axial proptosis with exposure keratopathy, (B) anterior view and (C) lateral view shows spontaneous globe rupture with worsening exophytic component of the right orbital mass

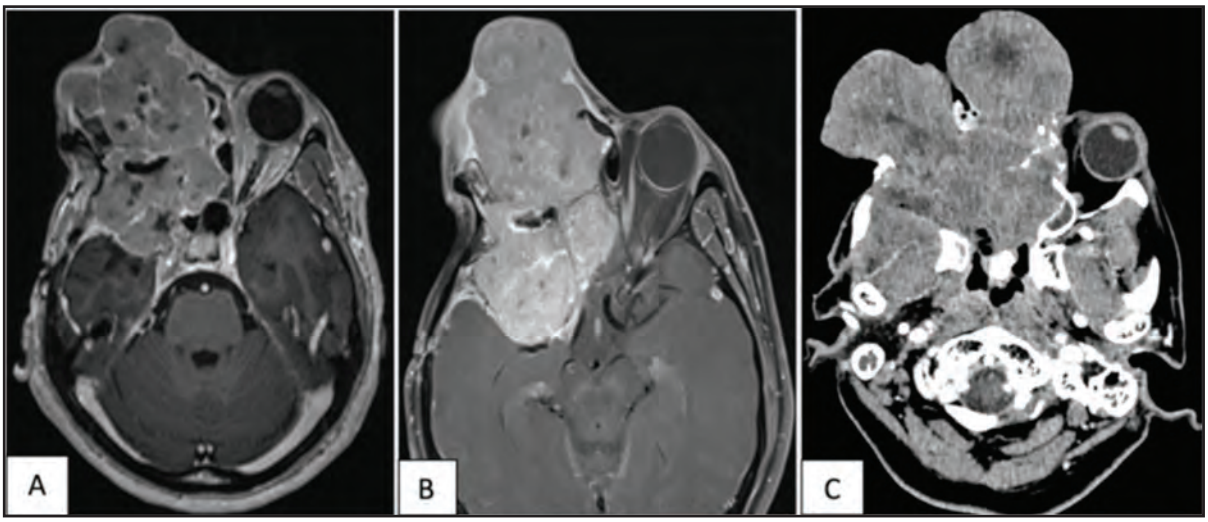

Fig. 2: Spheno-orbital meningioma in axial contrast-enhanced T1-weighted MRI reveals (A) marked tumoral growth with lobulated exophytic component at fifth recurrence. The mass extends to the ethmoid and sphenoid sinuses, right cavernous sinus, anterior and medial cranial fossae causing mass effect to the anterior temporal and inferior frontal lobes, (B) marked infiltrative tumour recurrence with heterogenous exophytic component, worsening locoregional and intracranial extension despite of a course of bevacizumab therapy prior the fifth surgery, (C) axial computed tomography (CT) image reveals further progression of infiltrative exophytic tumour recurrence in the right orbit, nasal cavity and paranasal sinuses with increasing degree of intracranial extension to bilateral anterior and right middle cranial fossae at sixth recurrence

DISCUSSION

SOMs are defined as primary en plaque tumours of the lesser and greater sphenoid wings. It is uniquely categorised as invasive tumour characterised by pathological hyperostosis and a widespread, carpet-like soft-tissue growth at the dura that may invade the orbit, optic canal, superior orbital fissure and other critical neurovascular structures.[3,5] SOMs are more common in women than in men (6:1 ratio), particularly in middle age of onset with incidence increases with age.[6] Anaplastic SOMs are rare, consist of 1 to 2% of all meningioma, which approximately half of this originate de novo, while the other 50% are the result of anaplastic transformation of lower-grade meningioma.[7]

Risk factors for anaplastic meningioma include childhood exposure to radiation and genetic mutation for NF2.[2] Our patient had none of the clinical features of neurofibromatosis type 2 including the absence of plaque-like skin lesions and hearing loss. Additionally, there was no family history, and genetic testing has excluded the genetic mutation for NF2. She had a triad of classic presentations of SOM, including proptosis, vision loss and restricted ocular motility, similar to other previously reported cases.[3-5] She developed headache, nasal block, anosmia and anaesthesia along the right ophthalmic and maxillary distribution of the trigeminal nerve as sequelae of further tumour progression, as similarly found in other cases.[1,7] Spontaneous globe rupture in SOM is

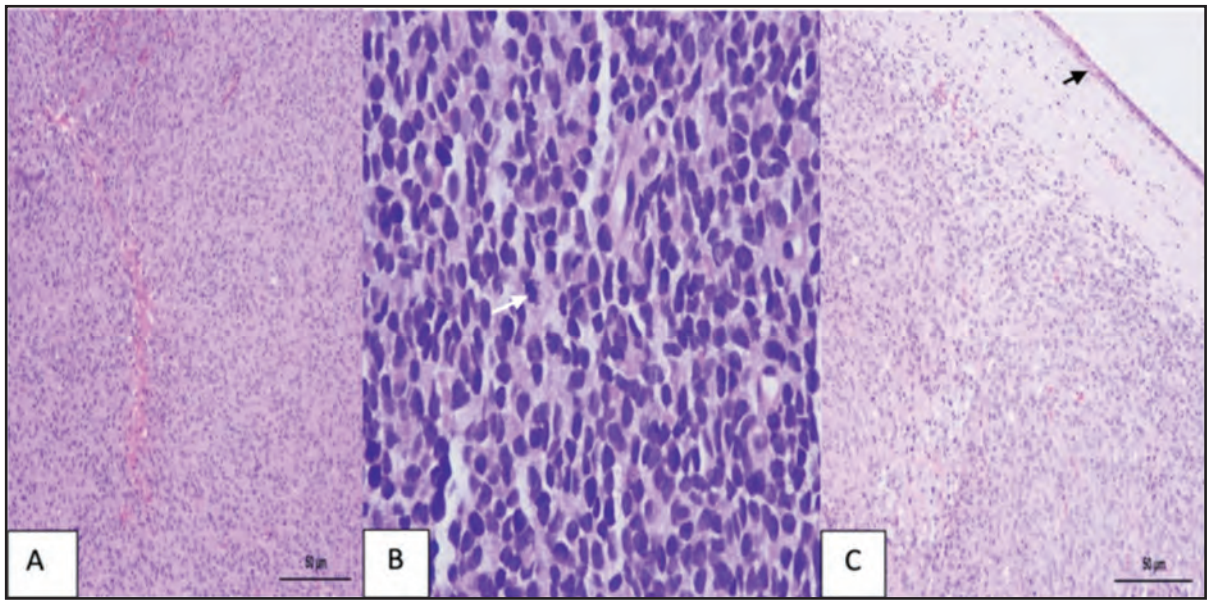

Fig. 3: Histopathology showed features of anaplastic meningioma. (A) Tumour cells arranged in diffuse sheath with interlacing fascicles infiltrating the bony trabeculae and adjacent connective tissue. Extensive areas of geographical tumour necrosis seen. (B) The epithelioid tumour cells display markedly pleomorphic nuclei and prominent nucleoli with ill-defined cytoplasmic borders. Mitotic figures are easily seen (indicated by white arrow). (C) Tumour with overlying respiratory type epithelium (indicated by black arrow). No whorling pattern, psammoma bodies or macrocalcifications observed

a rare entity. Cases of spontaneous globe rupture in orbital tumours have been reported in Group E retinoblastoma and choroidal melanoma, presumed to be related to raised intraocular pressure.[8,9] Such cases have also been reported in orbital metastasis from breast cancer, with evidence of severe keratitis leading to corneal perforation.[10] Spontaneous globe rupture in our patient possibly resulted from a combination of reduced corneal sensation, severe proptosis, raised intraocular pressure and the severe infiltrative nature of the SOM, causing disruption of the globe's integrity. Others have also reported that patients with SOM may also experience various neurological symptoms, such as sensorineural hearing loss, facial palsy, seizures, cognitive impairment, limb weakness, vertigo or dizziness, ataxia and gait changes.[1-3,7] However, these symptoms were not observed in our patient.

The combination of extensive bony hyperostosis and orbital extension are distinct characteristic imaging features of SOM. The common sites for hyperostosis are typically the greater and lesser sphenoid wings, the clinoid process and the roof and lateral walls of the orbit. Hyperostosis is best visualised on a high-resolution CT scan. T1-weighed MRI with gadolinium enhancement and fat suppression is necessary to define the limits of the dural tail and intraorbital extension.[11] Neuroimaging in our patient showed progressive lesions of both components that worsen at locoregional with intracranial extension. The majority of SOMs are grade I WHO lesions (90%), typically regarded as benign and slow growing. A small percentage of SOMs are composed of WHO grade II lesions (borderline), whereas WHO grade III SOMs (malignant) are very rare.[6,12] In our case, the initial WHO grading of meningioma was grade I, then progressed to grade II, and finally to grade III, an anaplastic subtype that poses a more rapid disease recurrence with a more aggressive disease presentation.

Due to the anatomical complexity of this region in proximity to many critical neurological and vascular structures, SOMs present a surgical difficulty.[13] The extent of resection, is determined by the Simpson grade which is based on the surgeon's evaluation during the surgery with considerations that affect the overall safety of surgery. Gross total resection was defined as Simpson grade I, II and III, whereas subtotal resection is defined as Simpson grade IV.[11] Our patient underwent maximum safe resection surgery for each craniotomy and two courses of fractionated radiotherapy. Fractionated radiotherapy (at least 54 Gy given in 1.8 to 2.0 Gy per fraction) has been recommended for WHO grade III tumours, regardless of gross total or subtotal resection.[2,11] After the fourth craniotomy that confirmed WHO grade III tumour, our patient was immediately subjected to radiotherapy. Unfortunately, she developed the fifth tumour recurrence after 1 month completion of radiotherapy treatment. The tumour continued to grow aggressively despite targeted therapy with bevacizumab. Bevacizumab is a vascular endothelial growth factor (VEGF) inhibitor with antiangiogenetic properties. It disrupts the binding and signal transduction processes that are essential for tumour vascularisation, resulting in the regression of the tumour's blood supply.[14] Other targeted therapies such as α-Interferon, sunitinib, everolimus and somatostatin receptor agonists, have also been administered to patients with recurrent or progressive meningiomas that no longer respond to surgery or radiotherapy.[6] Although some successes were reported in several case reports, the use of targeted therapy for meningioma is under level C evidence, and limited efficacy is anticipated.[11]

Recurrence rate for SOM is as high as 33 to 59%.[13] Our patient had a total six recurrences within a period of eight years, with rapid and advanced recurrences more pronounced after being diagnosed with anaplastic meningioma. She succumbed to her illness after one and half years of histological finding of anaplastic transformation. Anaplastic meningiomas are aggressive tumours with a dismal prognosis with a median overall survival period of one and half years and 10-year overall survival rate of 0%.[2,7] Literature has shown that distant extracranial metastases may occur in 0.18% of patients with meningioma, commonly found in the lung, bone, liver, kidney and spine.[15] However, further confirmation of any metastasis was not done in our patient as her condition deteriorated and she became bedridden, and the family opted for conservative and palliative care.

CONCLUSION

Anaplastic SOMs are fast-growing tumours, highly aggressive and associated with a poor prognosis. It leads to considerable cosmetic deformity, visual impairment, high morbidity and ultimately fatality. Decision-making for the management of advanced SOM is challenging, and it must be tailored to each patient, taking into consideration its impact on daily life and the patient's wishes.[3]

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COMPETING INTEREST

No potential conflict of interest was reported by the author(s).

ETHICAL CLEARANCE

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No ethical clearance was required as this is a case report.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the legal guardian.

AUTHORSHIP

All authors attest that they meet the current ICMJE criteria for Authorship.

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