



## Hyperostotic Sphenoid Wing Meningioma with Proptosis: A Rare Case Report with Literature Review

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### ABSTRACT

**Background:** We present a case of proptosis with headache, decrease in vision and orbital pain where patient underwent sphenoid-orbital decompression, and excision of mass, resulting in significant improvement in her vision with complete resolution of proptosis, headache and orbital pain. This study was done to diagnose and evaluate clinical outcome of hyperostotic sphenoid wing meningioma with orbital involvement after surgery.

**Case:** A 40 year-female presented with severe headache, right eye swelling, periorbital pain and decreasing in vision for two years, and right eye proptosis more severe for three months.

**Observations:** She underwent right frontotemporal craniotomy with removal of hyperostotic sphenoid wing and excision of meningioma after magnetic resonance imaging demonstrated a right sphenoid wing meningioma extending to orbital and middle cranial fossa. She had improvement of her symptoms post-operatively.

**Conclusion:** Surgical decompression of orbit with total resection of meningioma and sphenoid wing hyperostosis can result in significant improvement in the vision and proptosis.

**Key words:** Hyperostotic, Meningioma, Outcome, Proptosis, Sphenoid wing and Orbit.

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## INTRODUCTION

The sphenoid wing meningioma (SWM) accounts for 14%-18% of all intracranial meningioma (Sandalcioglu et al, 2005; MacCarty,1972) and three to six times more common in female (Schick et al, 2006). Sphenoid wing meningiomas are associated with approximately 40% of bony hyperostosis (Scarone et al, 2009). Sphenoid wing, lateral and superior orbit and anterior clinoid process are the common sites of bony hyperostosis (Terrier et al, 2018). The visual function deterioration, proptosis, and cosmetic deformity can be caused by bony hyperostosis and mass effect of intracranial meningioma (Oyaet al, 2011; Terrier et al, 2018). Surgical total resection or radiotherapy or combination of both are treatment options. There are few studies that describe the restoring of visual function and relieving associated clinical symptoms from mass effects on the orbit (Tamura et al, 2016; Schick et al, 2006). We report a case of right

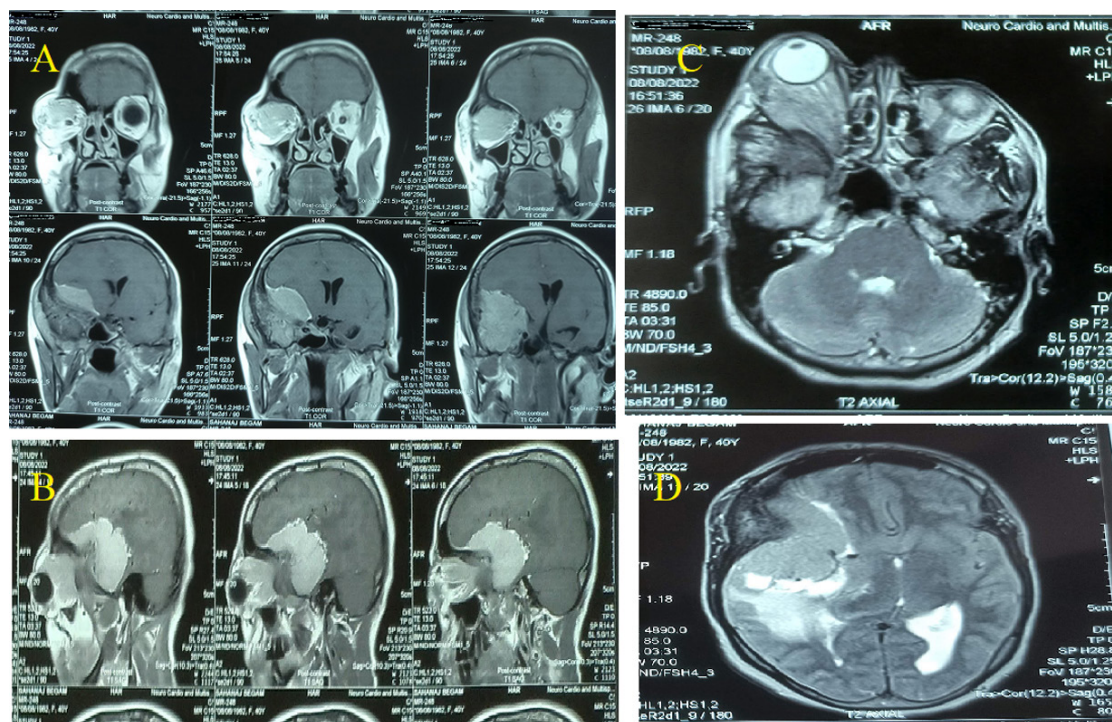
large sphenoid wing meningioma with proptosis where patient underwent gross total excision of the tumour and removal of bony sphenoid-orbital hyperostosis.

## CASE REPORT

A forty-year-female presented in the outpatient department of neurosurgery with complaints of severe headache, decreased vision, right eye swelling and orbital pain for two years; and severely protruding of the right eye (proptosis) for three months (Figure 1). On examination, the visual acuity on the right eye was hand movement (HM) and left eye was 6/6. There was proptosis of the right eye and inadequate closure of both upper and lower eyelids. Extraocular movement was restricted. Conjunctival examination showed conjunctival chemosis along with circumcorneal congestion. On corneal examination, there was corneal ulcer with hypopyon about 3 mm, corneal haze, decreased corneal sensation and dryness of



**Figure 1: Patient presented with proptosis of right eye and decreased vision as well as right eye chemosis, conjunctival edema and exposure keratopathy.**

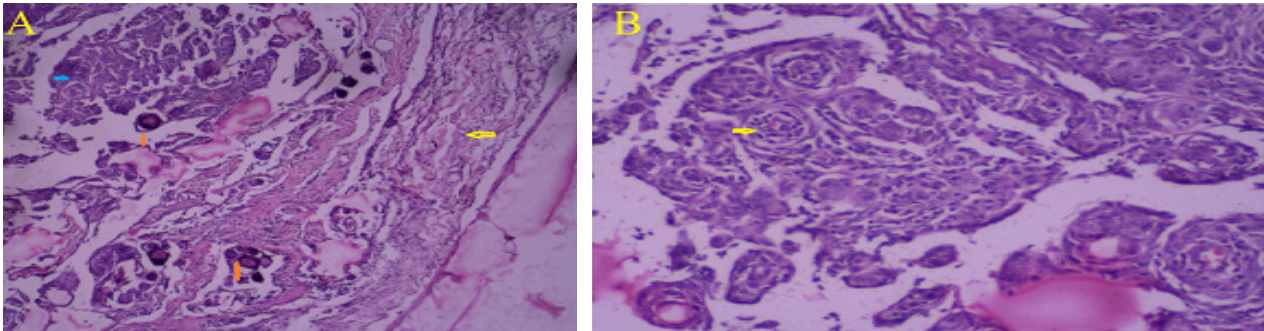


**Figure 2 (A, B): Magnetic imaging (MRI) T1 –weighted post contrasting image demonstrating enhancing tumor and optic nerve compression. (C, D): MRI T2 –weighted images demonstrating proptosis and optic nerve compression.**

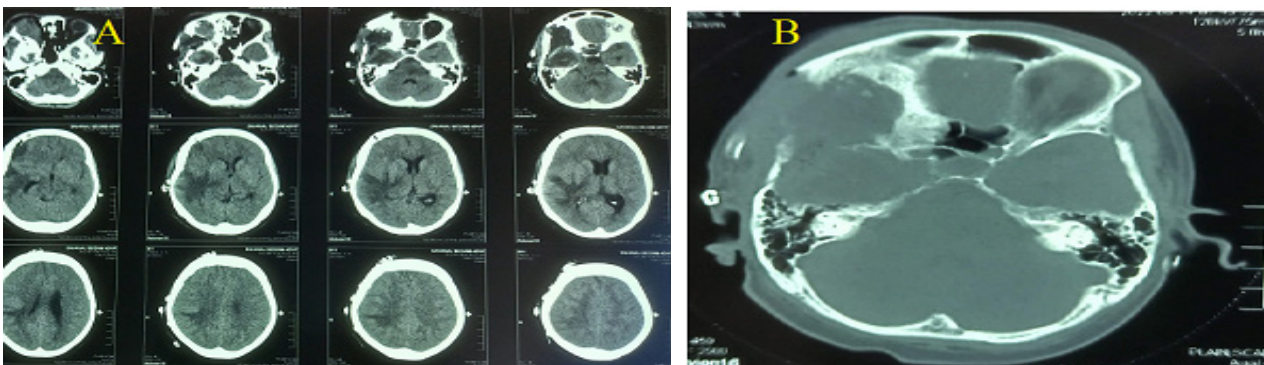
cornea. Pupil was round, regular and reactive to light but poor view of the fundus was due to corneal haze. After admission she was treated with frequent instillation of antibiotics drops, mydriatics, ointments, and lubricating eye drops. After one week of treatment there was improvement such as hypopyon started decreasing and cornea started getting clearer along with decrease in periorbital pain.

Magnetic resonance imaging (MRI) of brain revealed right sphenoid wing meningioma extending to orbit, anterior and middle cranial fossa (Figure 2) for which she underwent right front temporal craniotomy with gross total

excision of tumor and removal of hyperostosis and decompression of orbital wall. World health organization (WHO) grade I meningioma was diagnosed by histopathology report (Figure 3). The patient had immediately improved in her symptoms such as headache, orbital pain and reduction in size of proptosis and later on the best corrected visual acuity was 6/12. On the day of discharge she was able to close eyelids and fundus examination was normal. Post-operative computed tomography (CT) head was done which showed complete resection of meningioma and edema with removal of hyperostosis bony part (Figure 4). Patient was discharged on the tenth day of surgery.



**Figure 3: Photomicrograph (A) H& E stain x 100 showing : yellow arrow-meninge, orange arrow-Psammoma bodies and blue arrow-whorls of meningo-endothelial cells, (B) H& E stain x 400 showing arrow-whorls of meningo-endothelial cells.**



**Figure 4: Post-operative CT head showing total resection of tumour (A) and resection of sphenoid bony hyperostosis, lateral and superior orbital wall, clinoid and floor of middle cranial fossa (B).**

## DISCUSSION

Sphenoid wing meningiomas with bony hyperostosis are rare meningioma which are also known as sphenoid-orbital meningioma that arise usually from sphenoid ridge and represent approximately 20% of all meningioma and 4% of orbital tumors (Bowers et al, 2016; Bonavolonta et al, 2013). These tumors may encroach anterior, middle cranial base, orbit and nasal sinuses (Sughrue et al, 2013; Nakamura et al, 2006). In our case report, the meningioma was located within dura which are also called as “thin carpet like” or “en-plaque” tumor which can also invade to cavernous sinus (Mariniello et al, 2013; Mironeet al, 2009). Hyperostosis sphenoid wing meningioma can spread through

extra-cranial, orbital, peri-orbit, intraconal space and temporalis muscle ( Rogers et al, 2015; Talacchi et al, 2014).

The clinical manifestation of hyperostosis sphenoid meningioma with proptosis mainly includes headache, orbital pain, visual deficit, exophthalmos and cosmetic deformity (Terrier et al, 2018). In our study, our patient had severe headache, right orbital pain along with right eye severe exophthalmos. Because of its location, most of the patients came with visual /or proptosis (Saeed et al, 2011) which can be caused by compression of hyperostotic bone and intradural meningioma. We believed that radiotherapy alone would not improve in rapid recovery in the visual deficit and proptosis along

with clinical symptoms. Radiation therapy has been used in the patients with meningiomas not manageable to gross total resection due to meningioma's location (Skull base, or involving optic nerve sheath or cavernous sinus) and medical comorbidity or at recurrences (Rogers et al. 2015; Bower et al 2013; Bloch et al 2012; Pollock et al 2013). Rapid decompression of optic nerve and orbital structures can only be achieved by surgery rather than radiation (Sughrue et al 2011, Wu et 2018). So total surgical resection of this meningioma with hyperostotic bone can improve visual function and associated symptoms because of its intraosseous nature and hyperostosis of bony orbit. Oya et al (2011) reported on 39 patients with sphenoid-orbital meningioma of which four patients had severe vision loss and two had blindness; and neither of patients with blindness had functional recovery of vision; despite two patients with severe vision loss of 12 months had functional recovery of vision to mild or no symptoms. Our patient had significant improvement in vision and clinical symptoms after orbital decompression with removal of tumor and hyperostotic

bone, similar to other studies (Freeman et al, 2017; Mirone et al, 2009; Heufelder et al, 2009). Pathology shows meningiomatous cell encroaching haversian canals so hyperostotic bone should be considered as one of the part of neoplastic process (Bikmaz et al, 2007). Therefore diagnosis of hyperostosis sphenoid wing meningioma with proptosis is determined by radiological and clinical features rather than histopathology report. However, a definite diagnosis was confirmed by histopathology report.

## CONCLUSION

We conclude that significant post-operative improvement in visual deficit and clinical symptoms associated with hyperostosis sphenoid wing meningioma can be achieved after decompression of optic nerve and orbital structures by gross total resection of hyperostotic bone and meningioma.



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