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CASE REPORT Open Access

An unusual indication of maxillectomy—a case presentation of sphenoid wing meningioma

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Abstract

Background Sphenoid wing meningiomas (SWMs) are named because of their site of origin which is in the vicinity of the sphenoid wing. They are further classified into globoid type and en plaque (aka spheno-orbital meningiomas). These tumours are known for their difficult management because of their invasion to various neurovascular structures and bones. Surgical removal of these tumours is challenging. Complete surgical excision of the tumour is not always guaranteed; hence, intentional incomplete removal sometimes is mandatory to reduce postoperative morbidity. The purpose of this case report is to emphasize and describe the unexplored role of total maxillectomy in the excision of the extensive skull base tumours involving the orbit and infratemporal fossa.

Case presentation In this case report, we are presenting a case of a 50-year-old male diagnosed a case of atypical sphenoid wing meningioma with orbital and paranasal sinus extension who was previously operated on by extracranial approach and presented with the recurrence. Here, we are providing insight and surgical management of this case using total maxillectomy as an approach to access the intracranial approach.

Conclusion Maxillectomy with orbital exenteration can be used as a successful surgical approach to manage skull base lesions and intracranial tumours without any evident post-op complications.

Keywords Maxillectomy, Spheno-orbital meningioma, Paranasal sinus, Orbit

Background

Meningioma term was first described by Harvey Cushing to describe the tumours which arise from the pachymeningeal coverings of the brain and spinal cord [1]. These are the most common primary intracranial tumours with an incidence of 2.3–8.3 in 100,000 [2]. Meningiomas are mostly benign, but the atypical and anaplastic variants are more aggressive with a propensity of recurrence and worse outcomes [3]. SWMs are a type of meningiomas arising from the sphenoid wing and has a tendency to spread to the orbit and paranasal sinuses. Surgical excision of the tumours, SWMs, is challenging. Here, we are describing the management of this tumour using total maxillectomy as an approach to access the intracranial part. Maxillectomy is the surgical excision of the maxillary bone either partial or complete depending on the extension of the tumour, which can be both benign and malignant, also in some non-neoplastic conditions like invasive fungal sinusitis. The management of a patient planned for maxillectomy is multidisciplinary involving the maxillofacial surgeon, otolaryngologists, radio-oncologist, reconstructive surgeon, prosthodontist, speech pathologists and clinical psychologist, depending on the extent of the surgery [4].

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Case report

A 50-year-old male presented to the department of neurosurgery with chief complaints of headache, loss of vision, proptosis of left eye and fullness of lateral aspect of the skull (fronto-temporal region) with weakness of the bilateral upper and lower limbs for the last 6 months (Fig. 1 A). The patient was known with a case of temporal meningioma, operated in a private hospital in April 2020; post-operative biopsy suggested atypical meningioma grade 2. The patient was referred to the radiation oncology department where he was advised to take radiotherapy, 27 cycles, but after 8 cycles, the patient was referred to GB pant for the new-onset symptoms. The patient was apparently well till 6 months following which the patient developed above presenting symptoms. There was no history of seizure or loss of consciousness and vomiting. Higher cerebral functions were normal. Contrast-enhanced MRI of the brain and PNS was done (Fig. 2), and it showed a well-defined extra-axial lobulated mass lesion of size 10 × 11 × 10 cm involving the left temporal lobe, frontal lobe, orbit, maxillary sinus, sphenoid sinus and nasal cavity with complete deformity and loss of normal anatomy of orbit. There was complete encasement of the internal carotid artery and anterior and middle cerebral artery. A referral was sent to the ENT department for the surgical excision in collaboration with the neurosurgery team. After reviewing the contrast-enhanced MRI scans of the brain, there was a large destructive soft tissue

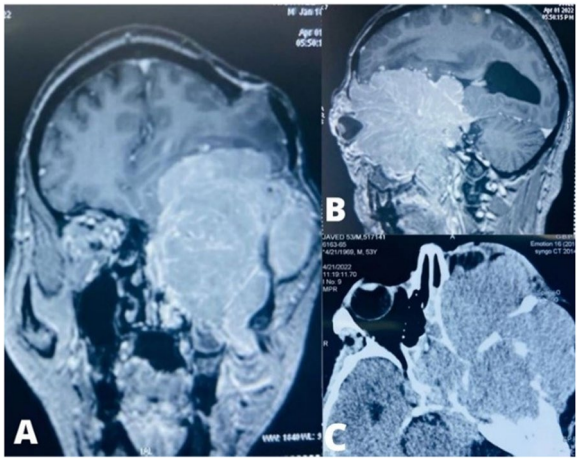

Fig. 2 Contrast-enhanced MRI of the brain with brain and paranasal sinus. **A**, **B** Coronal and sagittal post-contrast T1W images of the head reveal a large lobulated solid heterogeneously enhancing mass centred in the region of the left sphenoid wing extending into the sphenoid sinus, sella turcica, left ethmoid and maxillary sinuses, left nasal cavity and left orbit with involvement of extraocular muscles and optic nerve, resulting in proptosis and deformation of the left globe. Intracranial extension into the frontal and temporal regions is also seen. **C** Axial non-contrast CT of the head reveals the mass to be homogeneously hyperdense to grey matter and shows bone destruction in the region of the body and left greater and lesser wings of the sphenoid

Fig. 1 The clinical picture of the patient. **A** Proptosis with large lobulated swelling of the left temporal region. **B** Modified Weber-Fergusson incision marked. **C** Specimen after excision of the tumour (total maxillectomy with orbital exenteration). **D** Reconstruction of the skull base. **E** Post-operative image after suturing

mass involving the nose, ipsilateral maxilla and orbit, with major intracranial extension into the temporal and frontal lobes with sphenoid extension. The patient was planned for surgical excision under general anaesthesia by a combined approach involving a modified Weber Fergusson Dieffenbach approach with lateral extension for removal of the nose and paranasal sinus extension of the tumour and craniotomy for intracranial removal of the tumour. Skin incision marked and flaps raised to expose the part of the tumour. Total maxillectomy along with the orbital exenteration was done (Fig. 1 B, C). This exposed the whole intracranial extension of the tumour. There was direct wide-field exposure to remove the intracranial part of the tumour by transorbital approach. Some intracranial tumour was left behind going into the substance of brain tissue. After the removal of the tumour, the skull base was reconstructed in layers using DuraGen, facia lata and the bone flap (Fig. 1 D). The maxillary defect was packed with gauze, and skin flaps were repaired in layers (Fig. 1 E). Post-operatively the patient was shifted to the neurosurgery ICU, and a lumbar spinal fluid drain was inserted with a 45° elevation of the patient's head. The patient was tracheostomized on post-operative day 2, and on POD 3, patient's GCS was E3M5V4. The post-operative biopsy came out to be an atypical transitional meningioma, grade 2 (Fig. 3). The patient was discharged on post-op day 15 with tracheostomy in situ and referred to the radiation oncology department for a future course of action. Patient was lost to follow-up since then.

Discussion

Meningiomas are the most common primary intracranial tumours with an incidence of 2.3–8.3 in 100,000 [5]. Atypical meningiomas are aggressive with high chances of recurrence. Standard of care includes surgical resection followed by adjuvant irradiation partially resected atypical meningiomas. Chemotherapy and other medical therapies are available as salvage treatment once standard options are exhausted; however, the efficacy of these agents remains limited [6]. SWMs account for 11–20% of intracranial meningiomas, classified as "globoid" and "en plaque" tumours. The term spheno-orbital meningiomas (SOM) has been used to describe en plaque meningiomas that arise at the greater sphenoid wing and extend to the orbit with hyperostosis or bone invasion. SWMs has higher mortality, disability and recurrence rate due to their intricate relationship with the cranial nerves and orbital structures. Atypical meningioma involving the orbit and extending to the maxilla, infratemporal fossa is not reported in the literature. The extension of SWMs to the extracranial structures is directly proportional to morbidity and mortality [7, 8]. The surgical approaches described in the literature to excise SWMs included classic pterional, extended pterional approaches and pterional-orbitozygomatic approaches. The three approaches were mainly chosen according to tumour location, invasion and size. The classic pterional approach was the most commonly used surgical

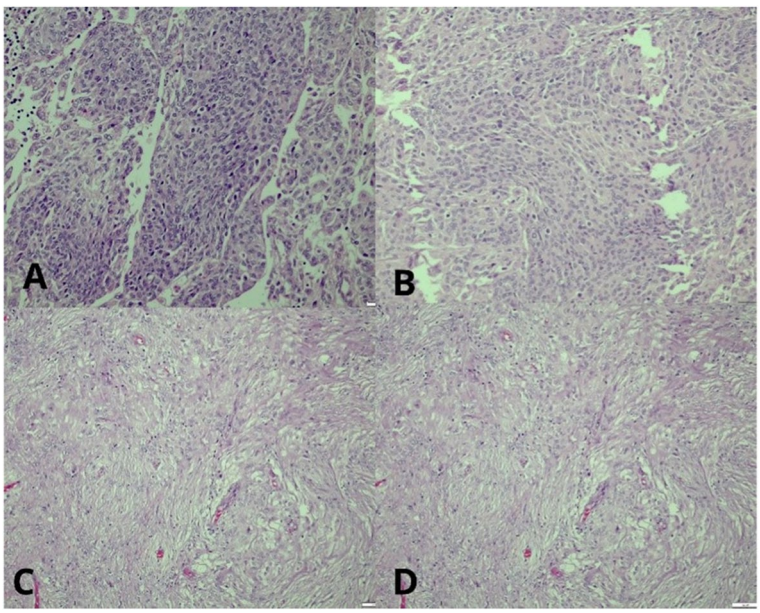

Fig. 3 Image showing the H&E-stained sections of the specimen. **A**, **B** Focal increased cellularity (20×) and sheeting (10×) are seen. **C**, **D** Tumour cells arranged in fascicles, whorled in diffuse sheet (10×) and mitosis including atypical mitosis seen (10×)

approach. Extended pterion approach was mainly chosen for large SWMs with skull base bone invasion or medial SWMs. Sometimes, the intentional incomplete surgical excision is necessary to get an improved postoperative quality of life [9]. There is an upcoming role of 3D fusion imaging in SWMs as a tool for presurgical evaluation, to predict the extent of tumor resection and its outcome [10].

The extent of surgical resection of the maxilla depends on the nature of the tumour, the site of the maxilla involved and the extent of its local spread. Skull base tumours have been considered hopelessly incurable in the past, which is now manageable with the advent of medicines and various skull base approaches, which is anterior, posterior and lateral. These approaches have augmented the surgical access of the nasal and ear tumours with intracranial extension or vice versa. Subtotal or total maxillectomy gives exceptional exposure or wide-field view of the middle compartment of the skull base from the roof of the sphenoid to the 5th cervical vertebrae, exposing the skull base between each Eustachian tube and the carotid canals, and provides ease of access to control haemorrhage [11]. The maxillary tumours can create devastating effects on the patients' aesthetics and functional well-being as they are not always limited within the boundaries of the maxilla and can involve adjacent structures like the hard and soft palate, skin and orbital contents which may have to be sacrificed [12]. The multidisciplinary approach is commendable, and this relates primarily to the experience and skill of the skull base surgeon, the availability of the operating room and radiotherapeutic equipment, as well as a mutual working relationship among specialty surgeons. The wide external exposure also helps in reaching the areas with very limited in-depth apex and internal exposure and also in the reconstruction of the defect like in this case the skull defect reconstruction was also done with ease. The chances of complete tumour removal are also more with the wide-field exposure. If the tumour removal is incomplete, perhaps a palliative result may be achieved with the additional assistance of radio-chemotherapy. The duration to make a surgical access is also less compared to the other aggressive approaches. The external facial scar falls in natural facial crease lines and is cosmetically acceptable. The limitations of this approach are the risk of exposure to the skull and meninges to nasal and pharyngeal flora that may lead to severe complications along with poor cosmesis due to ectropion and upper lip scarring. An appropriate antibiotic coverage and careful watertight closure of the soft tissues in the operative defect, reinforced by coverage with or without fat, bone or cartilage grafts, have eliminated these complications [13].

Conclusion

Maxillectomy with orbital exenteration can be used as a successful surgical approach to manage atypical sphenoid wing meningiomas without evident post-op complications.

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Authors' contributions
IS—conception and design of work. RS—conception and design of work. AJ—acquisition and analysis. AG—acquisition and analysis. PN—draft revision. The authors read and approved the final manuscript.

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Availability of data and materials
The datasets used and/or analysed during the current study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
Institutional Ethical committee (Maulana Azad Medical College and Associated Hospitals LNH, G.I.P.M.E.R, G.N.E.C) clearance taken. Ethical committee number – F.1/IEC/MAMC/No. 578 (05/05/2022).

Consent for publication
Written informed consent for publication of the patient's clinical details and images was obtained from the patient's next of kin.

Competing interests
The authors declare that they have no competing interests.

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