Giant tumour of the pterygopalatine fossa

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Abstract Neurilemmomas are benign and slow growing tumours. Neurilemmomas of the pterygopalatine fossa are rare. In this paper we present a case of schwannoma arising in the pterygopalatine fossa with extension into infratemporal fossa and floor of the orbit. A transantral approach was used for excision of the tumor following which the defect in the floor of the orbit was reconstructed with the help of a temporoparietal flap.

Keywords Schwannoma · Neurilemmoma · Pterygopalatine fossa · Infraorbital nerve

Introduction

Schwannomas are well differentiated benign tumours arising from the Schwann cells of the nerve sheath with well defined border. They constitute 25–45% of all the head and neck tumours [1]. Schwannoma of the head and neck may develop from many nerves, i.e. cranial or spinal, the most common being the cranial nerves. Here we discuss a case of infraorbital nerve schwannoma presenting as a tumor of the pterygopalatine fossa. Schwannomas arising from the infraorbital nerve are very rare and very few cases are reported in literature. Our patient was treated in 2 stages, first stage comprising surgical excision by ENT surgeon and the second stage, reconstruction of the defect by reconstructive surgeon.

Fig. 1 CT scan showing extension of tumour into the orbital floor

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Fig. 2 Sagittal section of MRI scan showing the tumour occupying the pterygopalatine fossa

Fig. 3 Coronal section of the MRI scan showing the Tumour extension into the floor of the orbit

Fig. 4 Axial section of MRI scan showing bowing of the posterior wall of maxillary sinus

Fig. 5 Intraoperative picture showing the origin of tumour from the infraorbital nerve

Fig. 6 Excised specimen

Fig. 7 Showing the temporoparietal flap being brought to the floor of the orbit through a tunnel between the skin and the zygomatic arch

Case Report

A 22-year-old male presented to our department with complaints right sided nasal obstruction for 6 months, diminished vision in the right eye for 3 months associated with diplopia and numbness over the right side of the face. He had temporary relief of his symptoms with nasal drops. He had no history of trauma, nasal bleed, anosmia or any other long standing illness, fever or weight loss. Neither did he have any history of otalgia, otorrhoea or dental complaints in the recent past. Anterior rhinoscopy revealed thick mucoid discharge in the floor of the nose on the right side with narrowing of air way.

Otoneurological examination showed blunting of the right corneal reflex with decreased facial sensation over the right infraorbital region. Rest of the cranial nerve examination was normal. Diagnostic nasal endoscopy showed a bulge in the middle meatus with medialized uncinate pro-

cess, hypertrophied bulla and thick secretions in the middle meatus on the right side. Ophthalmological examination revealed enophthalmos, central atrophic changes in the retina and superior rectus palsy suggestive of optic nerve compression along with partial atrophy and centrocaecal scotoma in the right eye. His vision was 9/6. Injection Methylcobalamine was started to prevent further degenerative changes. Plain CT of the paranasal sinuses showed a well defined lobulated soft tissue mass in the right pterygopalatine fossa with superior extension into the floor of the orbit (Fig. 1). Plain and contrast enhanced MRI to assess any intracranial extension showed hyperintense mass on both T1 and T2 weighted images with right orbital extension but no intracranial extension (Figs. 2, 3). The bowing forwards of the posterior wall of the maxillary sinus i.e Holman Miller sign was positive (Fig. 4). Due to hyperintensity of the lesion a pre operative angiography was done. The tumour was found to be relatively avascular with minimal feeders from

branches of the internal maxillary artery and the same was embolised. A joint consultation with reconstructive surgeon was made and it was decided to defer the reconstruction of the orbital floor, till histopathology report was available. 48 hours post embolization excision of the tumour was carried out via transantral approach.

Using Weber Fergusson incision on the right side the skin flaps were raised and the pyriform aperture was identified. After exposing the anterior wall of the maxilla, partial maxillectomy was performed by removing the medial wall as well as the medial half of the anterior wall of the maxillary sinus. The posterior wall of the maxillary sinus was seen thinned out and a thin shell of bone was seen covering the tumour. This thin shelf of bone was peeled out and the tumour was exposed. The tumour was found to be arising from the infraorbital nerve (Fig. 5) and hence the nerve had to be sacrificed to remove the tumour. Frozen section of the tumour showed spindle cells with no increased area of mitosis. By blunt dissection the tumour was removed in toto (Fig. 6). It was seen extending into the infratemporal fossa as well as the pterygopalatine fossa. Superiorly the floor of the orbit was thinned out. Post operatively patient had enophthalmos with sinking of the right globe. There was minimal improvement in vision in terms of counting fingers. Reconstruction of the floor of the orbit was carried out on the 8th post operative day using a pedicled temporoparietal flap based on the superficial temporal artery. A vertical incision was made over the scalp 2 cms anterior and superior to the ascending limb of the helix. After elevating the temporalis muscle and the periosteum the temporoparietal bone was exposed after identifying the superficial temporal artery. Bone flap was tunnelled underneath the skin over the arch of the zygoma and brought to the floor of the orbit (Fig. 7). The previous Weber Fergusson incision was reopened to position the bone flap and reconstruct the floor of the orbit and it was supported with antibiotic soaked gauze pack.

The patient recovered well in the immediate post operative period with stable position of the eye but still had restriction of the upward gaze which improved only after a month. Wound healed well and his vision was 6/6 at the end of one month. The patient is on regular follow up. Histopathological examination revealed spindle cells arranged in clusters i.e Antoni A type cells.

Discussion

Trigeminal neurilemmomas- schwannomas- may originate from any part of nerve roots or branches. However, those from extracranial branches of the nerves are less frequent and neurilemmomas in pterygopalatine region and infratemporal fossa are rare [2, 3]. Infraorbital nerve schwannomas form 0.2–0.4% of all the extracranial schwannomas. The tumour originating from ophthalmic, maxillary and mandibular branches of the trigeminal nerve may occupy the

infratemporal fossa, pterygopalatine fossa, orbita, paranasal sinuses and nasopharynx. Patients with infraorbital schwannomas can present with facial pain, sensory parasthesia and diminished corneal reflex. Trismus is noted if the third division of the trigeminal nerve is involved. Extension of tumour into the cavernous sinus can cause palsy of the III, IV and VI cranial nerves. Extension of the tumour into the pre pontine cistern can involve VII, VIII & X cranial nerves. The related symptoms are headaches, proptosis, loss of vision, exophthalmos, facial pain, hearing loss and impaired sensation. Radiological evaluation involves CT scan and MRI scan of the paranasal sinuses. MRI shows low intensity signals on T1 weighted images and high intensity signals on T2 weighted images. Angiography helps to rule out the possibility of a vascular lesion. Schwannoma is a histopathological diagnosis and can show Antoni type A or Antoni type B pattern. Our patient had Antoni type A, spindle shaped arrangement of cells in clusters. Immunohistochemical markers like S-100 may be positive in 50–70% of the patients. Iwai et al [3] employed orbitozygomatic infratemporal fossa approach in the removal of neurilemmoma that originated in pterygopalatine fossa and extended to the orbital apex and the cavernous sinus. Menard et al [4] used laterofacial approach to remove neurilemmoma extending from the pterygopalatine fossa to the parotid region. Pollock et al [2] have reported frontotemporal-infratemporal approach and lateral orbitotomy-frontal craniotomy approach in two trigeminal neurilemmoma cases with infratemporal and middle cranial fossa extension and in one with orbital apex location, respectively. Cansiz et al [5] reported transantral- pterygoidmalar approach which is a modification of degloving approach. With recently developed surgical techniques, almost all benign tumours are now successfully treated.

Conclusion

This case is being reported for its rarity. There has been no definite surgical approach described for the excision of infraorbital nerve schwannomas. A high degree of suspicion is required to diagnose these tumours as the clinical signs are not in proportion with the size of the tumour as evidenced in our case. The difficulties encountered in reconstruction of the defects postoperatively can be challenging.

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