A RARE TYPE OF EXTRACRANIAL MENINGIOMA-CASE REPORT

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Abstract:
Objective - Meningiomas arising from paranasal sinuses are rare, less than 40 cases have been reported in the literature. This objective is to report a patient with a rare type of meningioma arising from the ethmoidal sinus presenting with proptosis and nasal obstruction and highlight the treatment modality. Method - A twenty five year-old male patient with the complaints of nasal obstruction and proptosis and dimness of vision on the right eye with the diagnosis of primary extracranial meningioma of the paranasal sinuses. MRI showed a irregular, ill-defined, heterogenously enhancing lesion involving right side maxillary, right anterior posterior ethmoidal and frontal sinuses. Result - Total surgical resection by a facial translocation approach was done and patient recovered with no morbidity. Conclusion - Extra cranial meningioma accounts only 2 of all meningioma. The most common localizations of extracranial meningiomas are the skull bone, scalp, nose, orbit, paranasal sinuses, middle ear, neck and skin. The most frequent chromosomal aberration are monosomy 22, and Von Recklinghausen neurofibromatosis (NF-I). Friedman et al on reviewing the literature, found fewer than 150 cases of extracranial meningiomas of the head neck reported. Only 31 of these involved the paranasal sinuses. Clinical presentation is usually with nasal obstruction, headache, epistaxis, hyposmia anosmia, nose deformity, proptosis, orbital pain, blurring of vision, diplopia. The diagnosis of these tumors is difficult. It is part of the differential diagnosis of squamous cell carcinoma, angiofibroma (specially in men), esthesioneuroblastoma, sarcoma and lymphomas. Recent advances in skull base surgery enable radical resection of these tumors. Long term follow up is necessary to monitor for recurrences.

KEY WORD:
meningioma, extracranial, paranasal sinuses
**Introduction:**
25 year old male patient presented to us with h/o proptosis for the past three months accompanied with dimness of vision over the right eye and nasal obstruction. There was no history of headache, vomiting, seizures, weakness of limbs, nasal regurgitation, facial pain, facial numbness. There was no significant personal or family history of similar complaints. On examination patient was apparently normal with no significant findings on general examination and no neurological changes in higher function and cranial nerve function except for the visual acuity which was 6/36 in the right eye. Patient had no papilledema on fundoscopic examination. His spinomotor system and cerebellar functions were also normal. All basic investigations were within normal limits. Radiological evaluation was done and the CT and MRI revealed an irregular ill-defined heterogeneously enhancing lesion right maxillary, anterior and posterior ethmoidal and right frontal sinuses with displacement of septum to the opposite side and a thickened lamina papyracea displacing the orbit laterally.

**CORONAL CT-PNS**

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University Journal of Surgery and Surgical Specialties
The paranasal aspect was exposed by bony cuts the first of which extended from the infraorbital foramen to the nasal cavity and the second from infraorbital foramen to the ethmoid and the third from nasal cavity to ethmoid.

**BONY CUTS ALONG THE INFRAORBITAL FORAMEN**
With adequate visualization of the tumour, the lesion was debulked and completely removed. Adequate hemostasis was obtained and an inflated Foley’s was used for obliteration of the space in the maxillary sinus.
Post operatively patient had no complications and completely recovered with significant improvement of visual acuity in the right eye to 6/18 and the histopathological examination revealed the lesion to be psammomatous meningioma (grade I).

Psammomatous meningioma-HPE
Discussion
Meningiomas account for around 10-15% of all intracranial tumours and extracranial meningiomas amounting to 1-2%.

The most common localizations of extracranial meningiomas are the skull bone, scalp, nose, orbit, paranasal sinuses, middle ear, neck and skin with about 40 described cases of extracranial paranasal meningiomas in literature, and its diagnosis is difficult to be achieved. According to Petruillonis et al., tumors of epithelial origin such as angiofibroma and haematological such squamous cell carcinoma, neurogenic origin such as esthesioneuroblastoma, odontogenic tissues such as sarcoma and ameloblastoma, vascular origin such as as lymphoma must be part of the differential diagnosis. The classification of extracranial meningiomas as described by Hoye et al. is

**PRIMARY:**
Ectopic, without any connection either to foramen of a cranial nerve or to intracranial structures (Primary). Extra cranial extensions of a meningioma arising in a neural foramina (Primary).

**SECONDARY:**
Extra cranial metastasis from an intracranial meningioma (Secondary). Extra cranial extensions of a meningioma with an intracranial origin (Secondary). This classification is primarily based on the localization of the meningioma and the relationship of the intracranial moiety if present with special importance to the primary extracranial types which are also segregated on the basis of location. There are some considered mechanisms to explain physiopathology of primary extracranial meningioma: 1 - Presence of arachnoid cells in nerves or vases where these emerge from the Central Nervous System. 2 - Tissue Migration of meninges for extracranial areas during embryogenesis. 3 - Traumatic event or intracranial hypertension that dislocates arachnoid cells. 4 - Origin in indifferent mesenchymal cells About 20% of intracranial meningiomas have an extracranial extension including the skull, scalp (all cutaneous sites), orbit, upper airway involvement (nasal cavity, paranasal sinuses, nasopharynx), soft tissues, and ear and temporal bone. However, when the scalp, orbit, sinonasal tract, oral cavity, and soft tissues are excluded, the incidence decreases to less than 1%. hence it is imperative to consider the possibility of intracranial tumour. Meningothelial (77.4%) types of meningiomas are predominantly observed followed by the
occurrence of atypical (7.5%), psammomatous (4.1%), transitional, metaplastic, fibrous and angioblastic types. Immunohistochemical profiling includes staining for CK 7 seen in 21.7% of cases with this type of staining pattern helping in clinching the diagnosis of extracranial meningiomas. With regard to parasal meningiomas females are more commonly affected. The age group of predominant affliction is in the third to fourth decade. The most common chromosomal aberration is monosomy -22 and is often associated with NF-Type-1. Friedman et al on reviewing the literature found fewer than 150 cases of extracranial meningiomas of the head & neck reported. Only 31 of these involved the parasal sinuses: 10 ethmoidal sinus, 8 frontal sinus, 6 maxillary sinus, 2 sphenoidal sinus & 5 involved more than one sinuses. Most of these lesions have associated receptors the distribution of which is (76% of tumours have progesterone receptors, 96% somatostatin receptors, 89% Epi- dermal growth factor receptors, 19% estrogen receptors). Common clinical presentations include nasal obstruction, headache, epistaxis, facial pain, hyposmia/anosmia, nose deformity, proptosis, orbital pain, watery eyes, blurring of vision and diplopia. Investigative modalities are usually CT with contrast, MRI with contrast, MRI with MRA and MRV, and endoscopic biopsy. Surgical extirpation is the treatment of choice with radiotherapeutic modalities used for inoperable or malignant type of lesions. These tumours exhibit a high incidence of recurrence in atypical and malignant lesions and in cases of incomplete removal. In general, the prognosis of extracranial meningiomas appears to be excellent, with an overall median survival of 28 years which depends on the specific anatomical site, histologic type, tumor grade, gender, and age of the patient. The recurrence rate for meningiomas after total excision varies from 7% to 84% depending upon the number of years of follow-up. This is similar to intracranial meningiomas which have a recurrence rate of up to 20% and a mean survival around 7 years.

**Conclusion**

Meningiomas arising from parasal sinuses are rare, less than 40 cases reported in the literature. Recent advances in skull base surgery enable radical resection of these tumours and long-term follow-up is necessary to monitor for recurrences. Aim of presentation is to highlight the rarity of presentation and surgical options for extracranial meningiomas.

**References**:
1. Friedman CD, Constantino PD, Tietelbaum B, Berkold RE, Sisson GA. Primary extracranial meningiomas of the head and neck. Laryngoscope 1990; 100:41-8

