Meningiomas of sinonasal tract are rare entities. A case of primary extracranial meningioma of the right ethmoid sinus is reported. A 55-year-old female patient presented to the Department of ENT with symptoms of visual disturbance and swelling of right eye since 6 months, progressive nasal obstruction on the right side since 3 months with few episodes of epistaxis. Clinical examination showed a solid mass occupying right nasal cavity. Imaging studies showed a well-defined mass lesion with central cystic and sclerotic margins, in the region of right ethmoid sinus, causing mass effect on surrounding structures and displacing the medial rectus muscle with alteration of course of the optic nerve. Transnasal endoscopic removal of the mass was done in-toto, and histopathological studies including immunohistochemistry proved it to be a psammomatous meningioma.

**Keywords:** Ethmoid, Extracranial, Meningioma

**INTRODUCTION**

The sinonasal tract rarely shows tumors such as Meningiomas. Primary extracranial (ectopic, extracalvarial) meningiomas of the nasal cavity, paranasal sinuses, and nasopharynx are rare. They constitute <2% of all meningiomas. These are frequently misclassified, leading to their inappropriate clinical management. Meningioma is a common intracranial neoplasm with a variety of histomorphologic growth patterns, which are usually easily recognized. The most common localizations of extracranial meningiomas are the skull bones as well as scalp, nose, orbit, paranasal sinuses, middle ear, neck, and skin. The paranasal sinuses most commonly found to be affected are the frontal, maxillary, ethmoid, and sphenoid sinuses. Occurrence of primary extracranial meningiomas is probably related to the transformation of embryonic arachnoid cell remnants of ectopic meningocytes derived from pluripotent mesenchymal cells. A genuine extracranial primary meningioma is to be confirmed after a computed tomography (CT) scan, to rule out intracranial mass or any underlying bony erosion of the skull base to exclude secondary extracranial meningioma. Fine needle aspiration cytology of the lesion can be deceptively mistaken, and final diagnosis is usually made on the basis of histological examination of the excised mass.

**CASE REPORT**

A 55-year-old female patient presented at the ENT Outpatient Department (OPD) with the complaints of progressive visual disturbances (including irritation, double vision, and watering) and swelling of the right eye for 6 months along with nasal obstruction for 3 months with intermittent epistaxis since a month. To begin with, 6 months back the patient slowly developed swelling of right eye with diminution of vision. The patient then developed progressive obstruction of the right nasal cavity which eventually involved the left side as well. A month before reporting to the ENT OPD patient had few episodes of epistaxis from the right nostril. Gradually, the swelling spread over the side of the nose filling the right nasofacial furrow. This was accompanied by protrusion of the right eye (Figure 1). On clinical examination, the patient was of average build and nutrition with proptosis of the right eye with restricted movement to the medial side. The ophthalmologic assessment showed a protrusion measuring 22 mm (using Hertel’s exophthalmometer) with a vision of 6/36 in the right eye and 6/18 in the left eye. There was...
no relative afferent pupillary defect in either of the eyes and normal color vision bilaterally. Anterior rhinoscopy revealed a pinkish solid mass filling the right nasal cavity with the septum grossly pushed to the left obstructing the left nasal cavity. The mass bled on touch and did not shrink on application of decongestants. Posterior rhinoscopy revealed a mass filling the nasopharynx.

The post contrast CT scan image showed evidence of well-defined mass lesion measuring 4.3 cm × 3.1 cm × 2.9 cm with a central cystic component (10-15 HU) and a well-defined sclerotic margin (500-600 HU) in the region of the right ethmoid sinus. The mass was causing mass effect on the surrounding structures, displacing nasal septum to left, medial maxillary wall laterally, anterior wall of sphenoid posteriorly, medial orbital wall laterally, pushing the medial rectus muscles, and altering the course of the optic nerve with near total obliteration of the right nasal cavity. There was no evidence of bony destruction. Retained mucus secretions were noted in the right maxillary, frontal, and sphenoid sinuses (Figure 2). No intracranial connection or extension was noted.

For the management, a transnasal endoscopic approach was used which revealed a firm bony-cystic mass in the right nasal cavity extending to posterior ethmoid, sphenoid sinuses, and bilateral choanae with a completely pushed nasal septum to the left. Complete resection of the mass was done, and the sample was subjected to histopathological evaluation.

Histopathology showed a tumor composed of round to oval cells with pale-staining nuclei arranged in whorls and sheets. A good number of psammoma bodies with bony trabeculae were noted suggestive of psammomatous meningioma (Figure 3). Immunohistochemical studies showed positive vimentin and epithelial membrane antigen status.

Postoperatively, a retraction of 4 mm was noted on the third post-operative day, and vision remained unchanged. There is no evidence of recurrence at 6 months follow-up.

DISCUSSION

Meningiomas are benign, slow-growing tumors, usually intracranial; they represent between 13% and 26% of all central nervous system tumors. They usually appear between the 4th and the 6th decade of life, with a clear predominance of women (75% of cases). If the origin is independent, they are considered primary and if a direct communication with the intracranial region is present, they are considered secondary. Most extracranial meningiomas are secondary since up to 20% of the intracranial cases present with an extracranial extension. The mainly affected areas of extracranial meningiomas are the cervico-facial regions such as scalp (40.4%), ear and temporal bone (26%), and sinonasal tract (24%). Histologically, primary extracranial
meningiomas do not differ from intracranial. The origin of primary extracranial meningioma is still unclear; however, there are several hypotheses that these tumors could arise from arachnoid cells existing in nerve sheaths or from ectopic arachnoid cells entrapped extracranially or arise from pleuripotential mesenchymal cells. The classification system of Hoye encompasses the major etiologies proposed in the development of extracranial meningiomas:

1. Extracranial extensions of a meningioma with an intracranial origin (secondary)
2. Extracranial extensions of a meningioma arising from neural foramina (primary)
3. Ectopic, without any connection either to foramen of a cranial nerve or to intracranial structures (primary)
4. Extracranial metastasis from an intracranial meningioma (secondary).

The reported case belongs to category three as no intracranial extensions were noted on CT scan. In general, prognosis following surgical resection is good (72% at 5 years). The diagnosis and management of this tumor are particularly important because of their infrequent occurrence and unpredictable clinical behavior. A clear understanding of etiology and appropriate diagnostic and management principles helps to overcome the challenges posed by primary extracranial meningiomas. Clinical features of sinonasal tract meningiomas reported in the literature are non-specific and include nasal mass, nasal obstruction, nasal discharge, epistaxis, sinusitis, headache, and less frequently visual changes. The reported case showed all the features of nasal mass along with visual changes. Visual symptoms are considered to be related to the mass effect and compression of the optic nerve, thus tumor removal and decompression is essential to improve visual acuity, although blindness does not always improve with removal of the tumor as was the case with our patient, who had no improvement in the vision postoperatively.

CT scan or magnetic resonance imaging can give precise information of the extent and invasion of the tumor and is imperative in the diagnosis. Hyperostosis of surrounding bone is a classic finding in meningioma.

The only curative treatment for meningiomas is complete resection that includes meticulous removal of the attachment of the tumor to avoid tumor recurrence. To achieve complete resection, the endoscopic approach offers some advantages. It provides a wider surgical field, close-up and multi-angled visualization, and minimal invasiveness. Furthermore, direct observation of the roof of the sphenoid sinus allows precise exposure of the attachment. Recent advancements of endoscopic skull base surgery allow removing the cranial base safely and dealing with cerebrospinal fluid leakage. We should take into consideration removing not only tumor bulk but also the affected bone to achieve complete resection of the disease and prevent recurrences.

CONCLUSION

A case of primary nasoethmoidal meningioma is presented which is often confused with nasal polyp and final diagnosis rests on the histological examination of the excised mass. CT scan is imperative in diagnosis, knowing the extent of the disease and to rule out the intracranial extent of the mass. Complete surgical removal is the definite treatment and with recent advances, endoscopic removal proves to be a safe option for complete resection.

ACKNOWLEDGMENTS

We would like to express gratitude to the Department of Ophthalmology & Department of Pathology, JSS Medical College & Hospital, Mysore for their support and contribution.

REFERENCES


Source of Support: Nil, Conflict of Interest: None declared.