

Primary Sinonasal Psammomatous Meningioma : A Rare Case Report

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Abstract: Meningiomas are one of the commonest tumour of central nervous system. Meningioma is the most common nonglial intracranial neoplasm, arising from meningiocytes (arachnoid cap cells) of arachnoid granulation tissue. These neoplasms are most likely to be diagnosed in females, during the middle decades of life. They are rarely found in children. These tumors are rarely found extracranially. Primary extracranial meningiomas do not have connections to dura or any other intracranial structure, while secondary extracranial meningiomas have dural extension and/ or metastasis. Primary extracranial meningiomas (less than 1% of all meningiomas) frequently occur in head and neck region (2%). Males showing predominance . Most commonly found in sinonasal tract , temporal bone, ear and in the scalp. This paper presents a rare case of primary sinonasal meningioma in a 25 years old male patient presenting with nasal obstruction and proptosis of left eye. Treatment comprises of surgical excision with lateral rhinotomy approach. Histologically meningioma was further categorized as psammomatous subtype. There are very few cases of primary sinonasal meningioma reported in literature justifying its rarity.

Keywords: Extracranial, Meningioma, Sinonasal, Psammomatous.

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I. Introduction

Meningiomas are diverse set of tumors arising from meningiocytes (arachnoid or meningotheial cells) capping the arachnoid villi or pacchionian granulations^[1] and constitutes 15-20% of all intracranial tumors^[2]. Less frequently, meningiomas occur in intraventricular^[3,4], intraparenchymatous^[5,6] or intraosseous locations. Primary extracranial meningiomas do not have connections to the dura or any other intracranial structure, while secondary extradural meningiomas, refers to those intradural meningiomas with extradural extension and/or metastasis. In rare occasions these tumors are found extracranially [less than 2%], primarily in head and neck region and specifically in the sinonasal tract^[7]. Primary extracranial meningiomas have also been found in the ear, nose, parotid, neck, mandible, pharynx and skin^[8]. Intracranial meningiomas with extracranial expansion, in places such as orbit, mid ear, nasal cavity, nasopharynx and paranasal sinus^[9,10,11,12]. Intracranial meningiomas more commonly occur in female patients, whereas male patients are more likely to have extracranial meningiomas^[13]. The pathogenesis of primary extracranial meningiomas remain unclear. It is postulated that development of primary extracranial meningiomas is probably related to transformation of ectopically located arachnoid cell rests. Sometimes an extracranial meningioma can occur as an extension of small intracranial meningioma through skull base foramina or diploic space. Meningiomas might also arise from metaplasia of undifferentiated mesenchymal cells^[14]. Imaging modality for detection of possible intracranial extension and involvement of adjacent structures is mandatory before surgery. FNAC can sometimes be deceptively mistaken as in this case, so histological examination of the excised mass is essential in confirming a definite diagnosis. Owing to benign nature of the tumour primary method of treatment is extensive surgical resection, as incomplete removal may lead to recurrence.

II. Case Report

A 25 years old male patient attended ENT OPD of Gauhati Medical College and hospital, Guwahati, Assam with left sided nasal obstruction and left eye proptosis from last 14 years. Nasal obstruction was gradual in onset, progressive in nature, associated with loss of sense of smell, and 1 episode of epistaxis. Left eye proptosis was not associated with double vision or diminished vision. However patient complained of occasional watering from left eye without any redness of eye or itching. In general examination patient was of average built and did not showed clinical signs of neurofibromatosis. The neurological examination shows no cranial nerve deficit. In anterior rhinoscopy a pale pinkish polypoidal fleshy mass was seen occupying the entire left

nostril upto the anterior end of inferior turbinate and pushing the septum to opposite side. On probe test the mass was found to be firm, non tender, insensitive and did not bleed on touch. Probe could be passed all around the mass but superior attachment of mass could not be identified. Posterior rhinoscopy revealed mass peeping in the nasopharynx through left choana. Visual acuity and movements of both the eyes were in normal limit. Blood investigations were normal.

FNAC came out to be inconclusive showing only inflammatory changes. In imaging modalities contrast enhanced CT scan was advised which revealed a well defined heterogeneously enhancing expansile lesion with cystic areas and ground glassing within, epicentered in the nasal cavity and left maxillary sinus with extension to ethmoid sinuses and projecting to nasopharynx. No intracranial extension was noticed. Contrast enhanced MRI also reported similar findings. Patient was operated under general anesthesia by lateral rhinotomy approach. Mass involving the maxillary antrum, anterior ethmoid air cells and left nasal cavity was removed. Floor and medial wall of maxillary antrum were curetted. Mass was rubbery in consistency, friable and it was removed by scooping out in piecemeal. The walls of sinonasal cavity showed expansion but were intact, there was no intracranial extension noted. The nasal septum was repositioned in midline. Left maxillary antrum and left nostril were packed with medicated ribbon gauze and hemostasis was achieved. Post operative period was uneventful. Histopathological report showed lesion displaying tumour cells arranged in sheets and vague whorling pattern having discrete round or oval nuclei, inconspicuous nucleoli and eosinophilic cytoplasm. Extensive psammoma bodies were noted confirming the diagnosis of psammomatous meningioma. Follow up showed no recurrence at 3 months and 6 months post operatively.

Fig 1. Pre operative photos

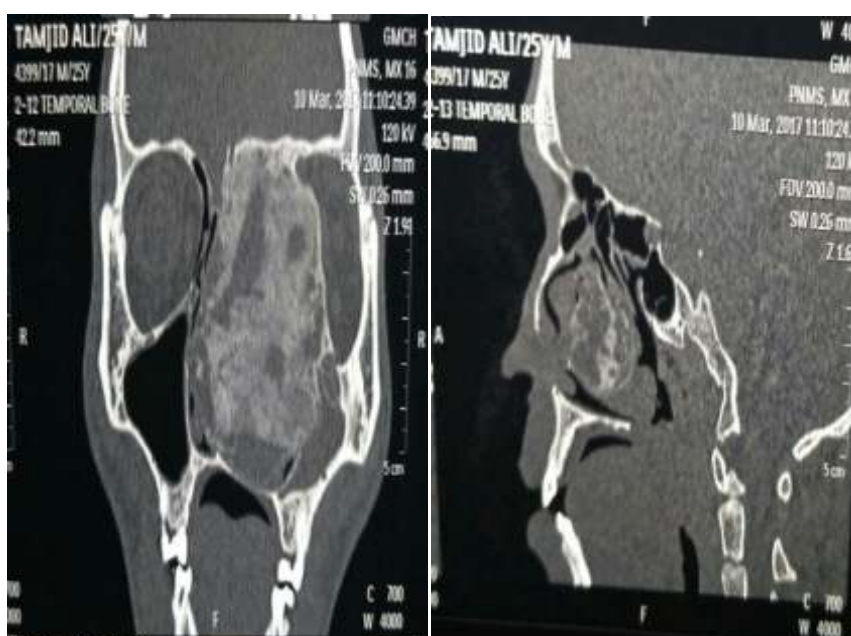


Fig 2. CT Scan images

Fig 3. MRI Images

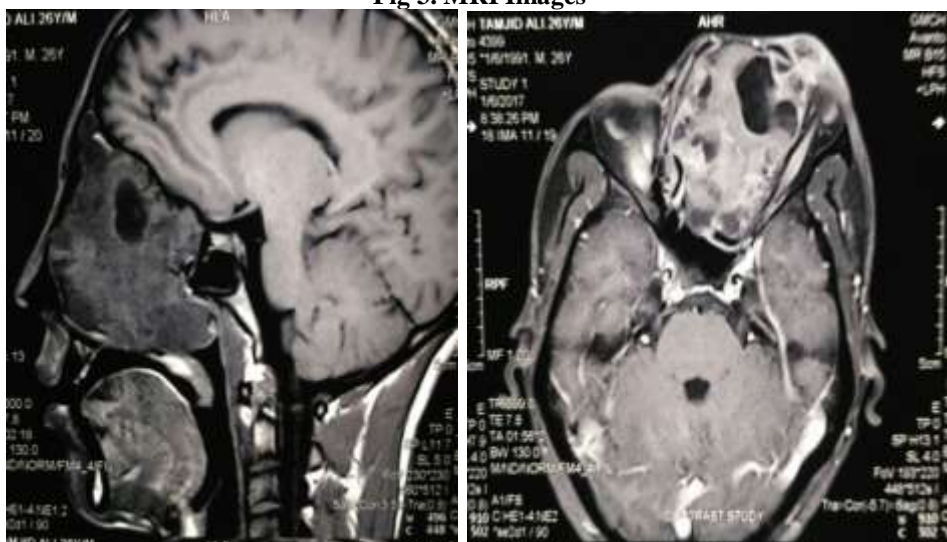
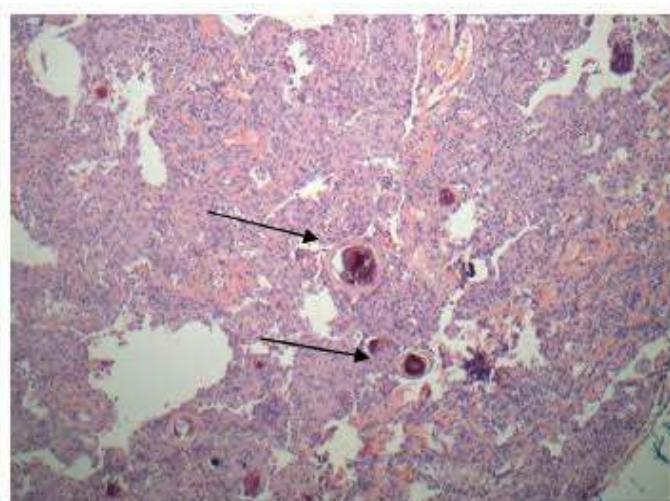


Fig 4. Histopathological image



Proliferation of meningothelial cells with psammomatous bodies (arrows), hematoxyllin-eosin , magnification 40X.

III. Discussion

Meningiomas are considered to be the most common non glial tumors of the brain and spine which constitute about 15% of all intracranial and 25% of all spinal tumours^[15]. Intracranial meningiomas are more common than extracranial tumours. There are two forms of extracranial tumours, a more common secondary meningioma (2%) and the rare primary meningioma (0.9%)^[16]. Here we have presented a case of this rare variety of primary extracranial meningioma as sinonasal mass. It is believed that such a tumour in the sinonasal region have arisen from extracranial growth from arachnoid cells within the nerve sheaths of cranial nerves and become detached during development^[17]. The pathogenesis of extracranial meningiomas suggested by Hoyer et al^[18] include four types

Type A – Involves direct extension from a primary intracranial tumour through the foramina of the base of the skull. (Secondary)

Type B – Involves extracranial growth from arachnoid cells within the sheaths of cranial nerves. (Primary)

Type C - Involves extracranial growth from embryonic rests of arachnoid without any apparent connection to the foramina of the skull base or cranial nerves. (Primary)

Type D – Involves distant metastasis from intracranial meningioma. (Secondary)

Majority of male patients show extracranial meningioma with age group ranging from 2nd to 7th decade.^[13] Patients mainly present with intranasal primary extracranial meningioma, as in our case patient presents with nasal obstruction, nasal mass, proptosis, epiphora, pain and occasionally epistaxis. The tumour may arise

from nose, maxillary sinus and frontal sinus^[9]. Ethmoid sinuses are less frequently involved but sphenoid sinuses are rarely involved.

According to recent WHO guidelines meningiomas are classified in to 3 grades :

Grade I – typical or benign type

Grade II – atypical with frequent mitosis

Grade III – anaplastic type with invasion

Fortunately 80% of extracranial tumours are benign^[18]. In our case benign nature of the tumour was evident from the slow growth and non involvement of the orbit or any cranial nerves. However grading is not most crucial regarding the recurrence of meningiomas but primarily the completeness of extirpation, as stated by Zulch^[19]. Paucity of cases reported in literature, little knowledge about their biological behavior and non specific symptoms makes their diagnosis challenging. According to one previous study^[20] differential diagnosis includes squamous cell carcinoma, esthesioneuroblastoma, sarcoma, ameloblastoma, angiofibroma and lymphoma.^[20] The radiographic findings of extracranial meningioma of the head and neck are not pathognomic. But detection of possible intracranial extension and involvement of adjacent structure is mandatory before preceding for surgery. Immunohistochemical studies can aid in diagnosis of an extracranial meningioma^[21,22]. They show positive expression of EMA(epithelial membrane antigen) and vimentin. Sometimes focally positive for S-100, keratin and CEA^[23,24]. Macroscopically tumours are of rubbery consistency, pale pink in colour, very friable and can be scooped out in piecemeal as in our case. Histologically meningiomas have further subtypes based on predominant cellular morphology such as meningothelial (syncytial), fibrous (fibroblastic), psammomatous, angiomatous and transitional (mixed). Most of the extracranial meningiomas of the paranasal sinuses have been reported to be syncytial variety^[25]. Primary mode of treatment remains extensive surgical resection, as it reduces the risk of recurrence. The role of post operative radiotherapy remains controversial^[26]. According to few literature radiotherapy is used in as a part of a palliative approach in elderly patients, unresectable malignant meningioma, recurrent or inoperable cases^[27].

IV. Conclusion

Primary extracranial meningioma of sinonasal region are quite rare. However it must be a part of differential diagnosis of sinonasal masses. The diagnosis of these tumours is often challenging owing to its rarity, lack of knowledge about its behavioural pattern, non specific symptoms and failure of FNAC to report it. Imaging modalities can show the extension of the tumour and useful for planning the surgery. Definitive diagnosis is by the histopathological examination of the excised specimen. Primary treatment is the complete surgical removal and also the good prognostic indicator.

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