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Primary Intranasal Meningioma a Rare Case with Review of Literature

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Abstract:
Meningiomas of sinonasal tract are a rare entity. Primary sinonasal meningiomas often pose difficulty in diagnosis because of their infrequent occurrence and clinically they appear to be nasal polyp. The final diagnosis rests on the histological examination. Extra cranial meningiomas are rare and tend to be more aggressive. Here a case of primary sinunasal meningoma with extension into the nasal cavity is presented and despite of its aggressive behavior it was benign. The importance of complete surgical extirpapion is undoubted and results in a good survival rate. Histological grading of the tumour is not crucial in predicting the rate of recurrence. The paucity of reported cases is the evidence for its rarity.

Keywords: *Meningioma _ intra nasal Extracranial*

1. Introduction

Meningioma is a well-recognized tumor of the central nervous system (CNS) that typically arises in proximity to the meninges. These neoplasms are more common in females during the middle decades of life and account for 24–30% of primary intracranial tumors [1]. Most commonly extra-neuraxial, meningiomas are found overlying the surface of the brain or at the skull base [2]. Uncommonly, meningiomas occur in intraventricular [3, 4], intraparenchymal [2, 5], or intraosseous locations. In rare instances (\2%), they appear as an extracranial tumor, most often in the head and neck region, and specifically in the sinonasal tract [6]. Extra cranial meningiomas of the sinonasal tract are rare tumours and are frequently misclassified, resulting in inappropriate clinical management. Occurrence of primary extracranial meningiomas is probably related to the transformation of embryonic arachnoid cell remmants of ectopic meningocytes derived from pluripotent mesenchymal cells. Due to many clinical, topographical, radiological and surgical factors the histology is not solely decisive for the prognosis of the meningiomas. [7] A genuine extracranial primary meningioma is to be confirmed after a CT scan, to rule out intracranial mass or any underlying bony erosion of the skull base. FNAC of the lesion can be deceptively mistaken and final diagnosis is usually made on the basis of histological examination of the excised mass. This extremely uncommon tumour justifies reporting an additional case, at the same time reviewing previous literature.

2. Case Report

A forty two old male presented at the Department of ENT, Government medical college Srinagar with complaints of, nasal obstruction for 1 year and an episode of epistaxis, nasal discharge, ptosis and lateral rectus palsy 5 days back.

Patient was having nasal obstruction since one year but was ignoring but seeked medical attention because of epistaxis, nasal discharge. Patient was of average built with mild ptosis and left lateral rectus palsy.

On anterior rhinoscopy purulent polypoidal fleshy mass was seen completely filling the left nasal cavity pushing the septum to the right. Mass does not shrink on application of the decongestant, did not bleed on touch and on probing mass had attachment high up in the nasal cavity. Posterior Rhinoscopy revealed mass peeping through the choana.

Visual acuity and movements of the right eye were normal

The patient did not show clinical signs of neurofibromatosis. Haemoglobin was 11.4g%, routine biochemistry and coagulation profile was normal.

MRI showed large soft tissue intensely homogenously enhancing mass lesion in the sinunasal cavity involing B/L sphenoid. , left ethmoid, upper nasal cavity and extending into left parasellar region. [Fig $1_{a,}1_{b]}$

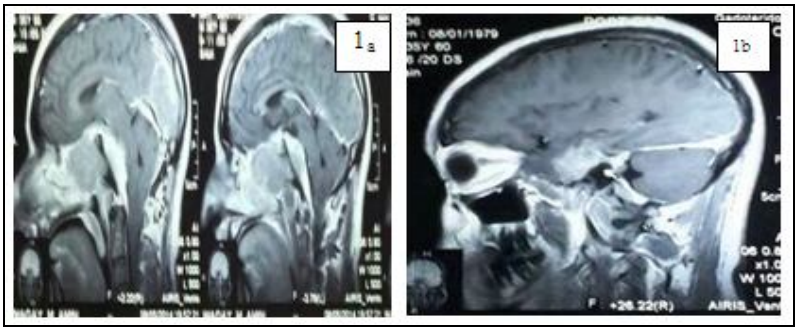

Figure 1(a) & Figure 1(b)

Patient was operated under General Anaesthesia on 20-1-05 by Lateral Rhinotomy approach. Moore's incision was given medial to the medial canthus of the (left) eye extending till (left) Nasolabial fold. Mass was seen to be coming out from the ethmoid sinus and extending till lamina papyracea and cribriform plate. Mass was rubbery, friable in consistency and it was removed in piecemeal. The sinonasal cavity showed expansion but the walls were intact and there was also intracranial extension. The nasal septum was repositioned back to its place. The wound was closed in layers and nasal cavity was packed with medicated ribbon gauze. Post operative period was uneventful and mass of was sent for Histopathological examination.

The histopathological appearance was consistent with transitional type having whorled clusters of cells in tight groups amd elongated cells with abundant collagen between them.[fig $2_{a,}2_{b,}2_{c].}$

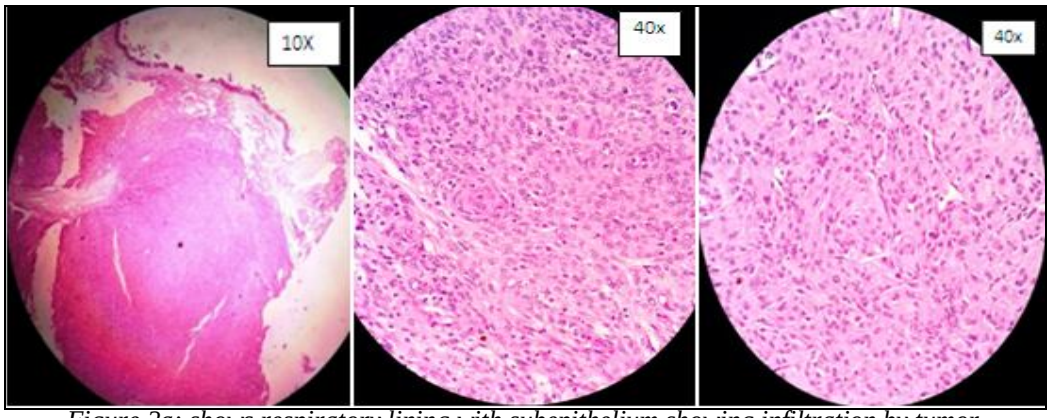

Figure 2a: shows respiratory lining with subepithelium showing infiltration by tumor.
Figure 2b and 2c: whorls and clusters of tumor cell

3. Discussion

Extra cranial primary meningioma is notably a tumour of rare occurrence with an incidence of 0.9% as compared to 2% seen in the case of secondary meningiomas.[8] Such a tumour occurring in the paranasal sinus is believed to arise from arachnoid 'cap cells' or meningocytes, which have migrated in the nerve sheath and become detached during development.[9] The classification system of Hoye[10] encompasses the major etiologies proposed in the development of extracranial meningiomas

- Extracranial extensions of a meningioma with an intracranial origin (Secondary).
- Extracranial extensions of a meningioma arising in a neural foramina (Primary).
- Ectopic, without any connection either to foramen of a cranial nerve or to intracranial structures (Primary).
- Extracranial metastasis from an intracranial meningioma (Secondary).

Our case belonged to the third group as there was no clinical or radiological evidence of an intracranial lesion. The current WHO classification distinguishes 3 grades of meningioma the typical or benign type (Grade I), the atypical with frequent mitosis (Grade II) and the anaplastic type with invasion (Grade III). However Zulch[7] stated that it is not the grading which is most crucial in the rate of recurrence of meningiomas, but primarily the completeness of extirpation.

Meningiomas are benign in so far as they do not metastasize, but they often show a predilection for local permeation of crevices and foramina, whilst pressure necrosis may result in spread from one cavity to other.[11] In our case benign nature of the tumour was evident from the slow growth of the tumour and non involvement of the orbit or any cranial nerves.

CT scan or MRI precisely gives information of the extent and invasion of the tumor and is imperative in the diagnosis. However, the final diagnosis depends on the histological examination. Hyperostosis of surrounding bone a classic finding in meningioma was also noted in our case.

76% of tumours have progesterone receptors96% have somatostatin receptors, 89% have Epidermal growth factor receptors, 19% have estrogen receptors. Hence the use of Tamoxifen and RU-486 (antiprogesterone) are under study. Characteristic and most frequent chromosomal aberration in meningiomas is monosomy 22, which however, has been shown not to be relevant to the prognosis.[12]

Radical surgical resection remains the primary mode of treatment and is correlated with a good prognosis. Efficacy of adjunctive radiotherapy after surgery is not established.[13] Radiotherapy is used in unresectable malignant meningioma or recurrent meningioma where surgery is not feasible. Newer techniques in treatment include proton irradiation and stereotactic radiosurgery with gamma knife. The diagnosis and management of this tumours is 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.

4. Summary

- A case of primary sinunasal meningioma is presented that is rare.
- It is often confused with Nasal polyp and final diagnosis rests of the histological examination of the excised mass.
- CT scan is imperative in diagnosis, extent of the disease and to rule out intracranial extent of the mass.
- Complete surgical removal is the definite treatment and so also the good prognostic indicator.

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