Case Report

Primary extracranial meningioma of maxilla- A case report

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Abstract

Meningiomas represent one of the commonest benign neural tissue neoplasms, but it is an extremely rare finding in the oral cavity. It tends to arise from the arachnoid cap cells pertaining to dura matter of the Meninges, the choroid plexus, or the brain stem. Herein, we report the case of a 36-year-old male patient, who visited our Department with the chief complaint of facial swelling associated with asymmetry. Based on thorough clinical, radiological, histopathological and immunohistochemical analyses, a diagnosis of Primary Extracranial Meningioma (PEM) of the Maxilla was made. Later, the tumor was surgically removed, and on follow up, the patient is doing well. Our presented case is extremely unique-it represents a PEM involving the oral cavity.

Keywords: EMA, immunohistochemistry, meningothelial, primary extracranial meningioma

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INTRODUCTION

Meningiomas represent the most common benign tumor of the Central nervous system (CNS). They originate from meningothelial cells of arachnoid cap cells occurring in mid and late adult life.[1] Usually, meningiomas are intracranial, but occasionally they appear extracranially as a direct extension of their intracranial counterpart (secondary extracranial meningioma) or very rarely as a true extracranial tumor (primary extracranial meningioma), the most common site for this being the orbit.

True extracranial meningioma occurring in oral cavity is extremely rare. Due to their rarity, they can be misdiagnosed. To the best of our knowledge, only 20 cases of extracranial intraoral meningioma have been reported in literature till date.[2] Our presented case is the 21st instance of a PEM occurring in the oral cavity-involving maxilla.

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CASE HISTORY

A 36-year-old male patient reported to the Department of Oral pathology and Microbiology in Burdwan Dental College and Hospital, West Bengal, India with the complaint of a painless swelling of the right side of face present since 2 years causing facial asymmetry [Figure 1]. Intraoral examination revealed the presence of a large fleshy mass involving the whole of the palate extending to the buccal side also. The overlying mucosa was normal in appearance [Figure 2]. Medical and family histories of the patient were nonsignificant.

Contrast enhanced Computed tomography(CECT) of face showed a large expansile lytic lesion with thinning of surrounding bony wall arising from right maxilla with no intraorbital connection but with compression of the nasal septum. Foci of calcification are also

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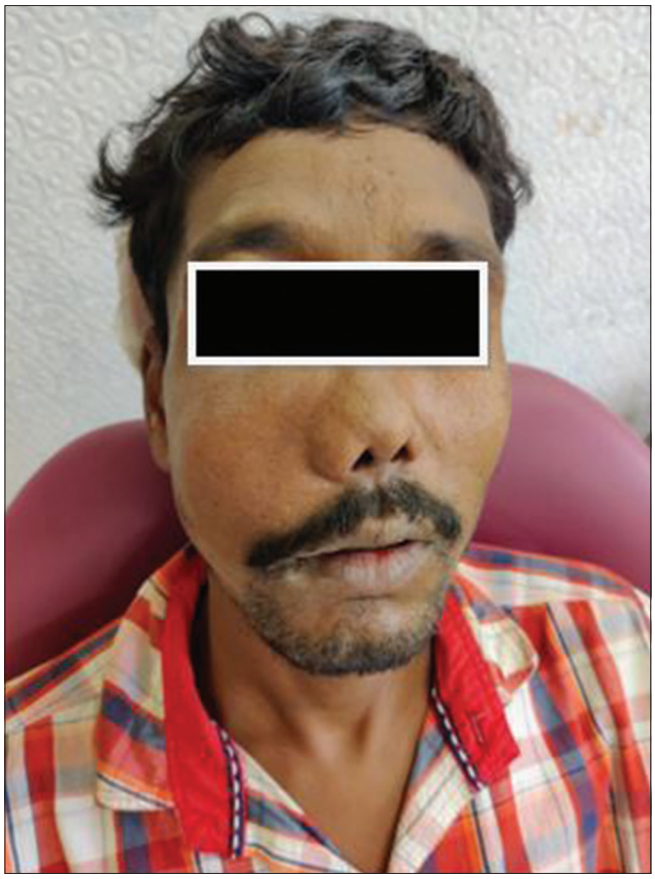

Figure 1: Extra oral image showing swelling on the right side of the face causing marked facial asymmetry

present [Figure 3]. A provisional diagnosis of soft tissue sarcoma and minor salivary gland tumor was made.

An incisional biopsy was performed from the representative area of the growth [Figure 4a]. The cut section of the tissue showed a firm, fleshy mass with a glistening and gritty consistency [Figure 4b]. On microscopy of H and E stained section, presence of meningothelial cells with round to oval nucleus and poorly defined cellular membrane, interlacing bundles of spindle-shaped cells, whorling pattern, psammoma bodies, and ossification were found with no evidence of increased mitoses or tumor cell necrosis [Figure 5]. On Immunohistochemistry (IHC), the tumor cells expressed vimentin, epithelial membrane antigen (EMA), and Progesterone receptor (PR) positivity as well as S-100, SOX-10, CD34, SMA, Desmin, and beta-catenin negativity. Ki-67 labelling index was at 2–3%. So, the overall features were suggested toward Meningioma [Figure 6a-c].

Magnetic resonance imaging (MRI) (plain and contrast) of brain and paranasal sinus was done to rule out any intracranial connection. MRI of Para nasal sinus (PNS) revealed the presence of heterogeneously enhancing

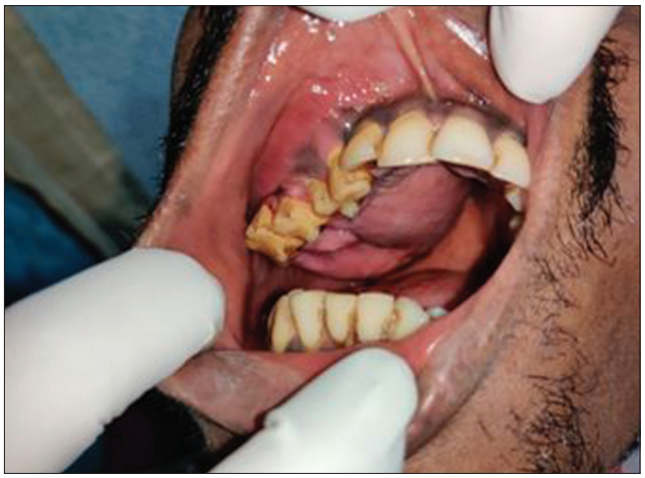

Figure 2: Intraorally, a large, fleshy, expansive growth involving right side of palate with extension towards the buccal aspect

well-defined mass of 76 mm × 64 mm × 77 mm dimension at maxillary sinus region – superiorly extending to the right orbit, inferiorly to the oral cavity involving the masticatory space causing compression over nasopharynx and right orbit. Brain MRI showed no connection to the dura mater that reinstates this case to be an exclusively primary extracranial one [Figure 7].

DISCUSSION

Primary extracranial meningioma (PEM) is an exceedingly rare tumor especially in oral cavity. Till now to the best of our knowledge, only 20 cases have been reported,[2] with most cases presented in mandible involving female patients. Hence, we assume our case of a 36-year-old male patient with lesion involving maxilla and hard palate to be a truly rare case.

The pathophysiology of PEMs has been associated with defects of cell migration from the neural crest. However, several plausible mechanisms have been postulated: origin from arachnoid cells of nerve sheaths or blood vessels while emerging from the skull foramina, arachnoid granulations getting detached and entrapped in extracranial sites, arachnoid islets getting displaced due to intracranial hypertension and finally, from undifferentiated mesenchymal cells like fibroblasts, Schwann cells, or a combination of these.[3] However, in our case, no history of tooth extraction or increased intracranial pressure could be discerned.

Histopathological finding of a meningioma can be varied. There are several histological subtypes; the most common being meningotheliomatous (63%) followed by transitional (19%), fibrous (13%), and psammomatous (2%).[4]

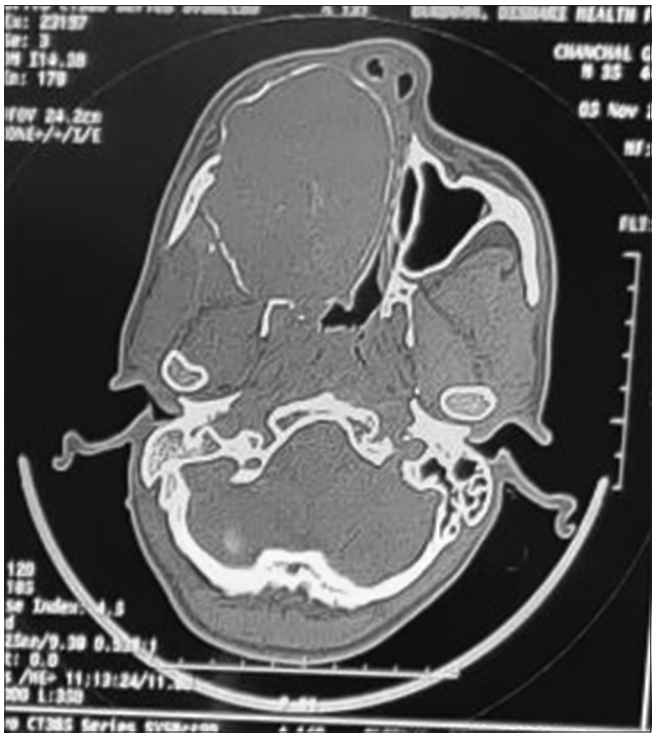

Figure 3: CECT of face showed a large expansile lytic lesion with thinning of surrounding bony wall arising from right maxilla with no intraorbital connection but with compression of the nasal septum with few calcific foci

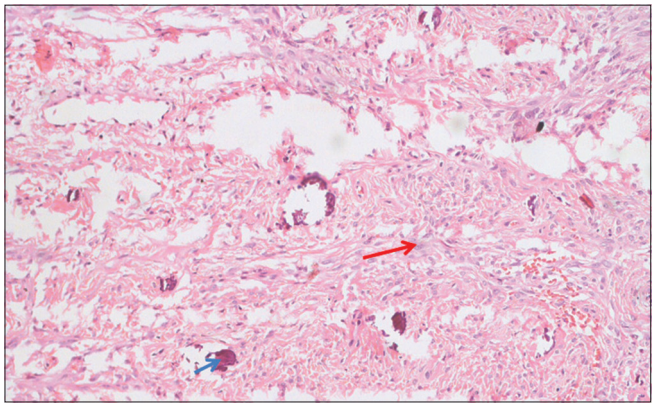

Figure 5: H&E stained section (×10 magnification) showed the presence of sheets and aggregates of meningothelial cells, (red arrow) possessing round to oval nuclei and indistinct cell membranes, with interlacing bundles of spindle cells. Occasional areas of psammoma bodies, were represented by basophilic calcifications (blue arrow)

Transitional meningioma contains elements of both meningothelial and fibrous meningioma. Meningothelial variant contains cells with round to oval nucleus having poorly defined cellular membrane and they form concentric whorls, whereas fibrous variant contains interlacing bundles of spindle shaped cells. As ours is a case of transitional (mixed) meningioma, we found a combination of both patterns, along with psammoma bodies which develop either from calcium deposits in degenerating tumor

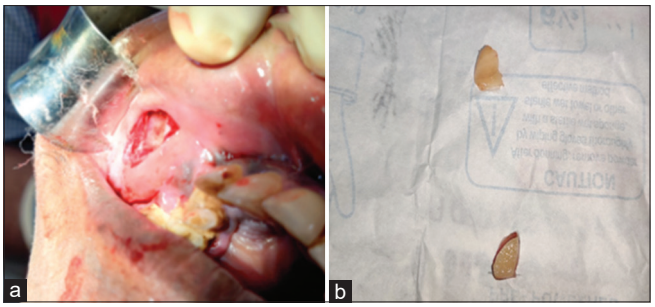

Figure 4: (a) Preoperative view during biopsy and (b) Cut section of the tissue showed a fleshy mass with glistening appearance and gritty consistency

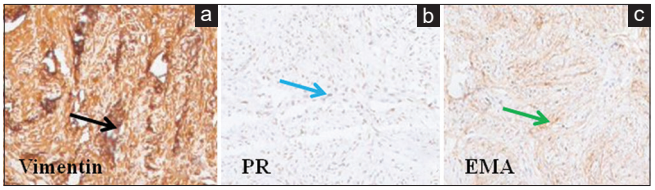

Figure 6: (a) IHC shows Vimentin positivity (indicated by black arrow) (b) IHC shows PR positivity (indicated by blue arrow) (c) IHC shows EMA positivity (indicated by green arrow)

cells, or from calcium deposited in the thickened hyalinized wall of blood vessels.[1] Also, ossification was noted.

According to the World Health Organization (WHO) criteria, meningiomas are classified into three grades. Grade I meningiomas are the most frequent (80–90%) and are considered as benign with a low risk of recurrence. Grade II meningiomas are less common (5–15%) and have a higher rate of recurrence, and grade III tumors are rare (1–3%) and are associated with poor overall survival rates along with increased risk of metastasis.[5]

The present case is of a transitional meningioma of WHO grade I with no evidence of increased mitosis or tumor cell necrosis.

However, given the primary extracranial location of the tumor, in this case, diagnosis cannot be made only based on histopathology, and sometimes can be confused with neurofibroma, Schwannoma, soft tissue perineuroma, paragangliomas, solitary fibrous tumor (SFT) etc. Hence, immunohistochemistry is used to differentiate between these tumors. Currently, the most commonly used IHC markers for the diagnosis of meningiomas are EMA and progesterone receptor (PR), CD34 and S100 etc. However, these markers have questionable sensitivities and specificities and may vary with regard to the grades or subtype of meningioma. Newer markers such as somatostatin receptor 2A (SSTR2A), signal transducer and activator of transcription 6 (STAT6), and

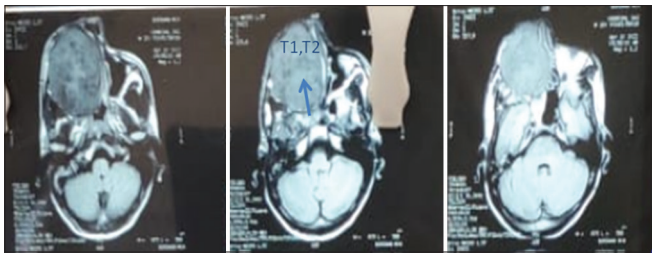

Figure 7: Axial section MRI of PNS revealed the presence of heterogeneously enhancing well defined mass at maxillary sinus region -superiorly extending to the right orbit, inferiorly to the oral cavity involving the masticatory space, indicated by blue arrow. No connection with the dura matter was there Sequences taken here-Axial Plane,T1, T2 weighed image

sex-determining region Y-box 10 (SOX10) have recently been shown to give better diagnostic insight than the said classic IHC markers.[6]

EMA is a classic IHC marker for meningioma. Almost 80% of meningiomas[7] are EMA positive. However, expression of EMA is negative in schwannoma and solitary fibrous tumor.

Meningiomas generally do not exhibit CD34 reactivity whereas CD34 positivity is characteristic of SFTs.[8] Also, SFT is EMA and PR negative, whereas meningiomas are EMA and PR positive.[9] Nuclear beta-catenin staining may be helpful as a diagnostic marker for SFT.

Both meningioma and Schwannoma are primary tumor of CNS. Schwannoma originating from neural crest cells shows S100 and SOX10 positivity but Meningioma is S100 and SOX10 negative. However, fibrous meningioma can occasionally show S100 positivity.

Ki-67 labeling index is the most important criteria for distinguishing anaplastic meningioma from those of benign meningioma.[10] It is also important to evaluate the proliferative activity of meningioma and thus its course, management and prognosis. Ki-67 scoring is more with grade III meningiomas followed by grade II meningiomas, while grade 1 meningiomas exhibit only 1–2% of Ki-67-positive cells.[10] Since in our case Ki-67 positivity is only 2–3%, this could be considered as grade I or benign meningioma.

Also positive expression of PR is related to lower grade or benign tumor and late recurrence.[11]

In our case of transitional meningioma, the tumor cells were EMA and PR positive, whereas S-100 protein, SOX-10, and CD 34 negative with a Ki-67 labeling index of 2–3%. Hence, after combining the results of histopathology and IHC, it could be inferred that this is a case of transitional meningioma of grade I with a good prognosis and chance of lower recurrence. After confirming the diagnosis, the patient was sent to the Department of Surgery, where he underwent partial maxillectomy along with surgical removal of tumor mass. Post 6 months follow up, the patient is doing well and is scheduled for fabrication of a maxillary obturator following maxillectomy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Haberland C. Clinical Neuropathology: Text and Color Atlas. LLC 386 Park Avenue South New York, New York: Demos Medical Publishing; 2007.
2. Amer HW, Hafed L, Ibrahim S, Shaker S. Case report: Rare site for intraoral meningioma F1000Research 2020;9:95.
3. Umana, GE, Scalia G, Vats A, Pompili G, Barone F, Passanisi M, *et al*. Primary extracranial meningiomas of the head and neck. Life 2021;11:942.
4. Bhat AR, Wani MA, Kirmani AR, Ramzan AU. Histological-subtypes and anatomical location correlated in meningeal brain tumors (meningiomas). J Neurosci Rural Pract 2014;5:244-9.
5. Boulagnon-Rombi C, Fleury C, Fichel C, Lefour S, Marchal Bressenot A, Gauchotte G. Immunohistochemical approach to the differential diagnosis of meningiomas and their mimics. J Neuropathol Exp Neurol 2017;76:289-98.
6. Ng J, Celebre A, Munoz DG, Keith JL, Karamchandani JR. Sox10 is superior to S100 in the diagnosis of meningioma. Appl Immunohistochem Mol Morphol 2015;23:215-9.
7. Maggio I, Franceschi E, Tosoni A, Nunno VD, Gatto L, Lodi R, *et al*. Meningioma: not always a benign tumor. A review of advances in the treatment of meningiomas. CNS Oncol 2021;10:CNS72.
8. Okada T, Fujitsu K, Ichikawa T, Mukaihara S, Miyahara K, Tanino S, *et al*. A strongly CD34-positive meningioma that was difficult to distinguish from a solitary fibrous tumor. Ultrastruct Pathol 2014;38:290-4.
9. Cheng L, Ni H, Dai Y. Intracranial solitary fibrous tumor mimicking meningioma: A case report. Medicine 2020;99:50:e23504.
10. Ghanghoria S, Sodhiya P, Niranjan V, Argal V, Ghanghoria A. Diagnostic and prognostic evaluation of meningioma by IHC markers- A study of 100 cases. J Med Sci Clin Res 2019;7:382-90.
11. Mnango L, Mwakimonga A, Ngaiza AI, Yahaya JJ, Vuhahula E, Mwakigonja AR. Expression of progesterone receptor and its association with clinicopathological characteristics in meningiomas: A cross-sectional study. World Neurosurg X 2021;12:100111.