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A Rare Case of Primary Atypical Meningioma of Nasal Cavity: Case Report and Review of Literature

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Abstract

Background: Meningioma is a tumour of central nervous system that typically arises in proximity to meninges. Extracranial primary atypical meningioma of sinonasal tract is rare.

Methods: We discuss the clinical, radiological, histological presentation of an elderly female with primary atypical meningioma of nasal cavity which we excised via endocopic endonasal approach.

Results: There was no recurrence up to 18 months of follow-up after endoscopic excision.

Conclusion: Extracranial primary atypical meningioma should be kept in mind as one of the differential diagnosis of nasal mass. Histopathological diagnosis along with immuno histochemistry should be used for definitive diagnosis.

Keywords: Atypical meningioma; Extracranial meningioma; Primary meningioma; Sinonasal meningioma

Abbreviations: WHO: World Health Organization; CECT: Contrast Enhanced Computed Tomography

Introduction

Meningioma is a tumour of central nervous system that typically arises in proximity to meninges. It is common in females and during middle decade of life. Rarely in <2% of the cases, meningiomas appear extracranially, especially in head and neck regions, sinonasal tract, ear and temporal bone and scalp [1]. Usually histopathological and immuno histochemistry examination is diagnostic. WHO (World Health Organization) has classified meningioma into 3 grades- benign, atypical and anaplastic [2]. Some studies has shown 30% of meningiomas to be atypical [3]. Here we are presenting a case of primary atypical nasal meningioma.

Case Report

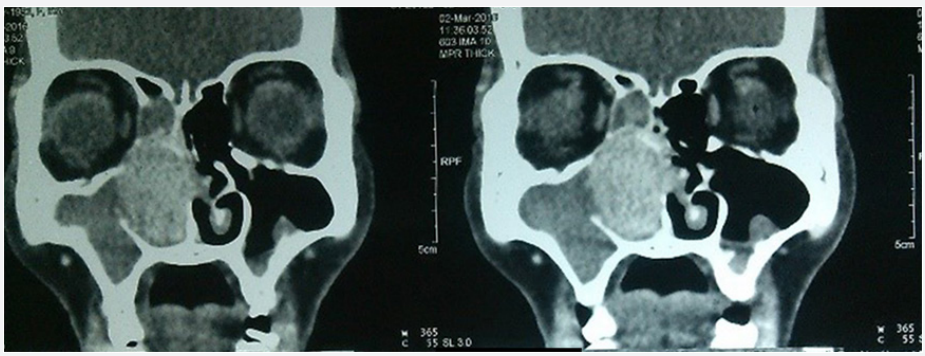

Figure 1: CECT Nose & PNS coronal cut showing heterogeneously enhancing mass in right nasal cavity, likely centered in middle turbinate with extension into right ethmoid sinus superiorly and destruction of nasal septum with extension of the mass to contralateral side.

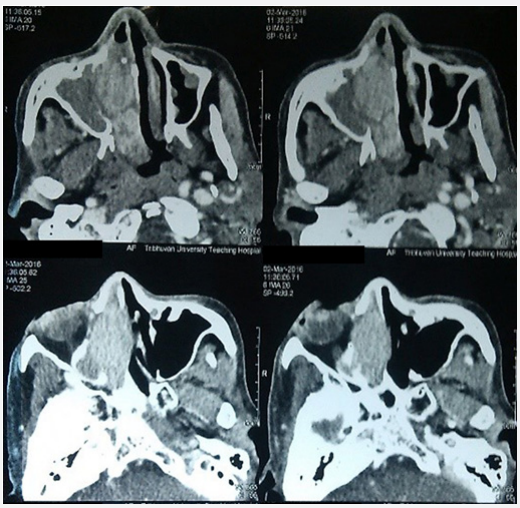

Figure 2: CECT Nose & PNS axial cut showing heterogeneously enhancing mass in right nasal cavity, likely centered in middle turbinate with extension into right ethmoid sinus superiorly and choana and sphenoid sinus posteriorly with erosion of adjacent bone.

A 63years female presented to outpatient department with chief complaints of right nasal obstruction and intermittent epistaxis for 6 months. Nasal obstruction was insidious in onset, gradually progressive and complete since 3 months. Epistaxis was intermittent, 1 episode in a month, spontaneous, 250-500ml in amount in each episode. There was no history of any systemic diseases. Examination of nasal cavity revealed pinkish fleshy mass occupying whole of the right nasal cavity which bled easily on touch. CECT (Contrast Enhanced Computed Tomography) of nose and paranasal sinuses (Figures 1 & 2) showed heterogeneously enhancing mass in right nasal cavity, likely centered in middle turbinate with extension into right ethmoid sinus superiorly and choana and sphenoid sinus posteriorly with erosion of adjacent bone. There was destruction of nasal septum with extension of the mass to contralateral nasal cavity which was features suggestive of inverted papilloma.

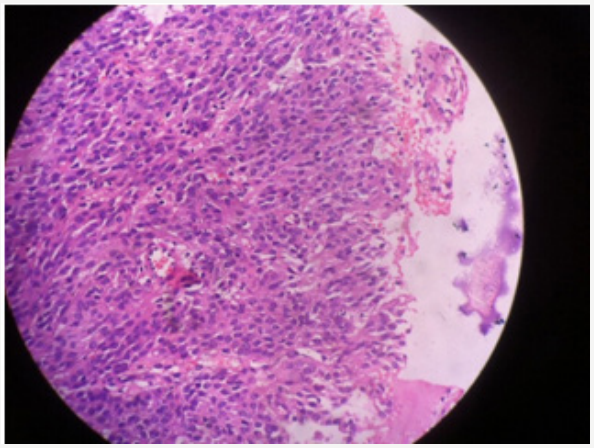

Figure 3: Histopathological examination suggestive of atypical transitional meningioma, WHO grade 2.

To confirm the diagnosis biopsy was taken from the mass. Histopathological examination (Figure 3) revealed unencapsulated tumor composed of tumor cells arranged in sheets with focal whorling pattern in subepithelium. The tumour cells were epithelioid to spindle shaped with monomorphic round nuclei, moderate amount of eosinophilic cytoplasm and indistinct cell borders. The chromatin was fine with occasional intranuclear inclusions and many prominent nucleoli. Mitotic figures were 4 per 10 high power fields. No necrosis and pigment were seen. Hence histopathological examination was suggestive of atypical meningioma. Due to prominent nuclei and S100 positivity, HMB-45 was also done which came out to be negative hence malignant melanoma was ruled out.

Patient was admitted and was planned for internal maxillary artery embolization but due to tortuosity of the right common iliac artery, catherization was difficult, so the procedure was abandoned. She underwent endoscopic excision of the mass. Per operatively there was pinkish, vascular, friable mass occupying right nasal cavity, extending anteriorly up to anterior end of middle turbinate, posteriorly occupying right choana and extending to left choana (Figure 4). Superiorly mass was attached to the roof of ethmoid and inferiorly to the floor of right nasal cavity. No destruction of septum was seen as suggested by CECT. Post- operative histopathological examination showed neoplastic tissue lined by ciliated pseudostratified columnar epithelium with some of the areas showing ulceration and necrosis.

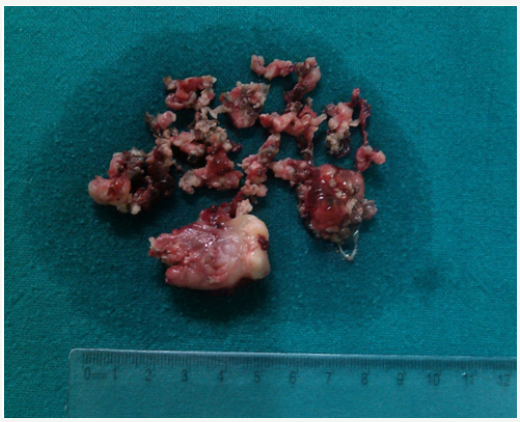

Figure 4: Meningioma after endoscopic excision.

Subepithelium showed edema and hemorrhage with prominent vessels with lumen occluded by thrombus. Stroma was composed of tumour cells arranged in lobules, nests and sheets. These tumour cells were polygonal to oval having moderate amount of eosinophilic to clear cytoplasm and indistinct borders. Nucleus was oval with fine chromatin andprominent nucleoli. Some areas also showed spindling with oval nucleaus arranged in short fascicles. Intranuclear inclusions and grooving were also identified. Mitotic figures were 1-2 per 10 high power fields. Tumor necrosis was not identified. Features were suggestive of atypical transitional meningioma, WHO grade 2.

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Discussion

Meningiomas are non-glial tumours of the central nervous system, representing 24-30 % of all intracranial neoplasms. They arise from the arachnoid cap cells (meningocytes) which are derived from the neural crest. These tumours have a predilection for middle aged and elderly females [1]. They have been reported to occur extracranially in only 1-2 % of cases. 20% of extracranial meningiomas are secondary extensions of intracranial tumours [4]. Primary extracranial meningiomas with no direct communication with the intracranial region are extremely rare. Histologically primary extracranial meningiomas are identical to intracranial counterparts.

Different mechanisms which give arise to extracranial meningiomas like : arachnoidal cells are present in the sheaths of nerves or vessels where they emerge through the skull foramina, displaced pacchionian bodies become detached, pinched off, or entrapped during embryologic development in an extracranial location, traumatic event or cerebral hypertension that displaces arachnoid islets, origin from undifferentiated or multipotential mesenchymal cells, such as fibroblasts, Schwann cells, or a combination of these, perhaps explaining the diverse pathologic spectrum found in meningiomas [1,5,6]. They have been reported to occur in the sinonasal tract, cranial bones, middle ear, scalp and soft tissues of the face and neck and parotid gland. An analysis of 146 cases of primary extracranial meningiomas showed that majority originated from the skin and scalp (n = 59) followed by middle ear (n = 26) and sinonasal tract (n = 25). Other rare locations in the head and neck included the temporal bone, mandible, nasopharynx, parotid gland, orbit and neck [7].

Symptoms of these lesions depend on the anatomic site of involvement. Meningiomas involving the sinonasal tract may mimic sinusitis with patients presenting with nasal obstruction, anosmia, facial pain, nasal discharge and epistaxis [4,8]. Some author describes the average duration of extracranial sinonasalmeningioma to be 31.1months.Nasal endoscopy usually shows a firm reddish pink to grey mass in the nasal cavity which could be globular or lobulated, but well-circumscribed with displacement and without infiltration into the surrounding tissues [9]. Radiological finding are usually nonspecific and included clouding or opacification of the sinuses, bony sclerosis, and focal destruction of the surrounding sinusoidal or nasal cavity bony tissues [10].

In the current WHO edition (2007) grade I meningiomas (benign) are recognized by their histologic subtype and lack of anaplastic features. Grade II meningiomas (atypical) are defined by one or more of the following four criteria:

a) Choroid or clear cell histologic subtype,

b) Four to 19 mitoses per ten high-power field (HPFs),

c) Brain infiltration, and

d) Three or more of the following five histologic features

Small cell change, increased cellularity, prominent nucleoli, sheet-like growth, or necrosis. Grade III meningiomas (anaplastic/malignant) are defined by rhabdoid or papillary subtypes, a histological picture of frank malignancy resembling that of carcinomas, melanomas, or high grade sarcomas, or 20 or more mitosis per ten HPFs [2].

The differential diagnosis of sinonasal tracts meningiomas includes mucocele, olfactory neuroblastoma, carcinoma, hemangioma, sarcoma and angiofibroma [8,9]. Histopathology and immuno histochemistry are confirmatory. Meningiomas are strongly immunoreactive to Vimentin, EMA (Epithelial membrane antigen) and pancytokeratin. Besides these, some meningiomas also show positive reactions to CK7, S-100 protein, CAM5.2, Synaptophysin, CK20, GFAP, Ki-67 index (>1%), CD34, SMA, PCNA, Progesterone receptor, Estrogen receptor [1,7].

Studies have shown the slow growth of the tumour, hence surgical extirpation without the necessity of adjuvant therapy as treatment of choice but complete excision was not always possible due to complex anatomy of nasal cavity and paranasal sinuses [1,7]. Complete extirpation of these masses may be accomplished by endoscopic, open or a combination of both approaches. Radiation therapy has been suggested to improve survival in meningiomas of central nervous system but role of radiotherapy in extracranial meningiomas hasn't been studied yet. In our case endoscopic surgery was done; and microdebrider along with monopolar cautery was used to excise tumour in piecemeal due to vascular nature of the mass. In cases with high risk tumour locations, stereotactic surgery has shown improved results. Studies have found tumours ranging from 1 to 8cm with average of 3.5cm, usually infiltrative to bone of the sinus or nasal cavity. However surface epithelium was not ulcerative or penetrated [1].

There was no recurrence up to 18 months of follow-up after endoscopic excision in our case. Usually recurrence occurs in the same anatomic site as the primary lesion and depending on the time interval, it may be distinguished from residual disease. Additional surgery if clinically feasible is advisable as radiation therapy doesn't always result in a clinical response. None of any clinical, radiographic or pathologic features have correlated with patient outcome in the literature. Prognoses of extracranial meningioma are excellent with only limited recurrence. Joseph et al has found recurrence rate for atypical meningioma within two years to be 28% compared to 9.3% in benign meningioma while Rushing et al has found recurrence to be 17.8 % within few months of initial surgery. Similarly Thompson LDR has also found similar recurrence rate of 20% ranging from a few weeks to 21years after initial presentation. A 5-year disease free survival rate 66.9% to 82.1% and 10year disease free survival rate 54.6% to 78.6% [1,7].

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Conclusion

Primary sinonasal atypical meningioma is a rare condition. It should be kept in mind as one of the differential diagnosis of nasal mass. Histopathological diagnosis along with immunohistochemistry should be used for definitive diagnosis. Excision can be done by endoscopic endonasal approach.

Acknowledgement

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References

1. Thompson LDR, Gyure KA (2000) Extracranial Sinonasal Tract Meningiomas: A Clinicopathologic Study of 30 Cases With a Review of the Literature. Am J Surg Pathol 24(5): 640-650.
2. Louis DN, Ohgaki H, Wiestler OD, Webster KC, Peter CB, et al. (2007) The 2007 WHO Classification of Tumours of the Central Nervous System. Acta Neuropathologica 114(2): 97-109.
3. Backer-Grøndahl T, Moen BH, Torp SH (2012) The histopathological spectrum of human meningiomas. International Journal of Clinical and Experimental Pathology 5(3): 231-242.
4. Friedman CD, Costantino PD, Teitelbaum B, Berktold RE, Sisson GA (1990) Primary extracranial meningiomas of the head and neck. Laryngoscope 100(1): 41-48.
5. Chang CY, Cheung SW, Jackler RK (1998) Meningiomas presenting in the temporal bone: the pathways of spread from an intracranial site of origin. Otolaryngol Head Neck Surg 119(6): 658-664.
6. Batsakis JG (1984) Pathology consultation. Extracranial meningiomas. Ann Otol Rhinol Laryngol 93(3 pt 1): 282-283.
7. Rushing EJ, Bouffard J-P, McCall S (2009) Primary Extracranial Meningiomas: An Analysis of 146 Cases. Head and Neck Pathology 3(2): 116-130.
8. Swain RE, Kingdom TT, DelGaudio JM, Muller S, Grist WJ (2001) Meningiomas of the paranasal sinuses. Am J Rhinol 15(1): 27-30.
9. Thompson LDR, Gyure KA (2000) Extracranial sinonasal tract meningiomas. Am J Surg Pathol 24(5): 640-650.
10. Lee KF, Suh JH, Lee YE, Berry RG (1979) Meningioma of the paranasal sinuses. Neuroradiology 15(1): 27-30.

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