Case Report

A Rare Case of Primary Heterotopic Meningioma of Nasal Cavity presenting with Right Eye Proptosis

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Abstract

Meningiomas are slow growing intracranial neoplasm of central nervous system accounting for 15% of all intracranial tumors. Rarely it appears extracranially, mostly in the head and neck region, and specifically in the sinonasal tract with variable clinical presentation. It can mimic various other malignancies which are common at a particular location. We present a case of a 26 year old female with progressive proptosis of the right eye and intermittent headache without any vision impairment for about three years. Radiological features showed an ill-defined solid-cystic lesion epicentred in right nasal cavity suggesting olfactory neuroblastoma. Histomorphological and immunohistochemistry assessment favoured a diagnosis of Transitional meningioma, WHO grade I. Awareness of primary heterotopic meningioma will help in early diagnosis and appropriate management of the patient, particularly while evaluating a case of sino-nasal mass.

Keywords:

Heterotopic meningioma, immunohistochemistry, nasal cavity, proptosis

Introduction

Meningiomas are slow growing intracranial neoplasm of central nervous system arising from arachnoid cap cells.[1] It accounts for 15% of all intracranial tumors.[2] Rarely (1-2%), it arise extracranially, most often in the head and neck region, and specifically in the sinonasal tract. An analysis of 146 cases of primary extracranial meningiomas by Rushing et al showed scalp skin (40.4%) as the most common site followed by ear and temporal bone (26%), and sinonasal tract (24%).[3] These ectopic meningiomas occur in sites without meningeal cover and based on their connection with the central nervous system, can be divided into primary and secondary types. Primary heterotopic meningioma refers to lesions without associations between the tumor and the intracranial structures, cranial nerve foramen or vertebral canal. They can occur in the nose, middle ear, mouth, parotid gland and neck.[4] Secondary heterotopic meningiomas are somehow connected to meninges. We report a case of primary heterotopic transitional

meningioma of the nasal cavity presenting with proptosis. Extracranial meningiomas of the sinonasal tract are often misdiagnosed as olfactory neuroblastoma, undifferentiated carcinoma, paraganglioma, melanoma or schwannoma.[3] This case report aims to present this unusual site and presentation of heterotopic meningioma, the awareness of which is needed while evaluating a mass in the nasal cavity.

Case Report

A 26 year old female presented in our hospital with progressive proptosis of the right eye and intermittent headache for about three years. Vision in both the eyes were normal and light reflexes present.

Plain CT of the orbit showed an ill-defined lobulated heterogenous solid-cystic lesion measuring approximately 58x44x65mm epicentred in right nasal cavity, breaching the medial wall of the right orbit and extending into it. There was also breach in the right cribriform plate and floor of the anterior cranial fossa with extension into the neuroparenchyma. Radiological features suggested olfactory neuroblastoma. Frontal craniotomy and tumour decompression was done and the tissue was sent for histopathologic examination. Post operative CEMRI revealed ill-defined lobulated, heterogeneously enhancing solid-cystic lesion measuring 33x32x47mm epicentered in the superior olfactory recess of right nasal cavity, eroding the cribriform plate and floor of anterior cranial fossa with intracranial extension and involvement of the right frontal lobe. Right orbital extraconal extension was seen. (Figure 1).

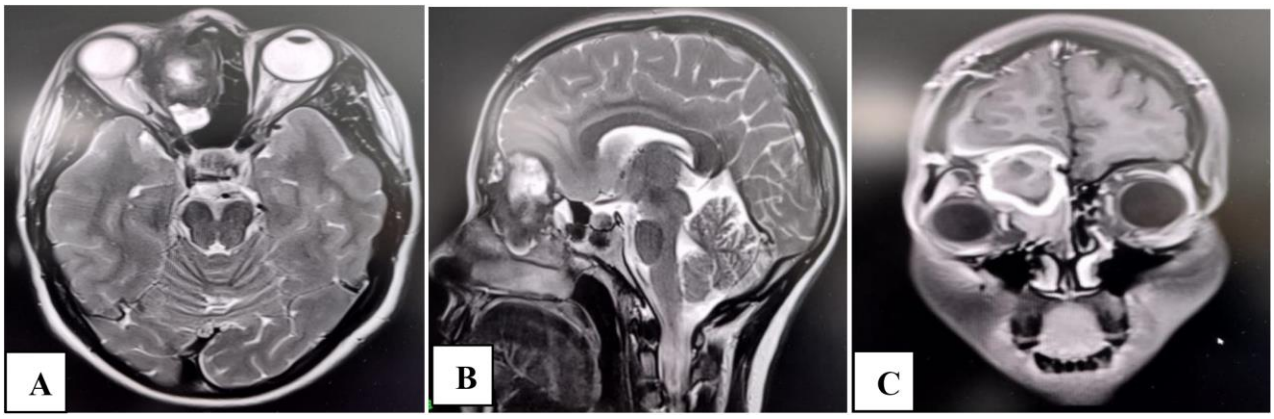

Figure 1: (A-C) Contrast enhanced T2-weighted axial (A) and sagittal (B), T1-weighted axial (C) slices show a heterogenous enhancing mass in the right nasal cavity extending into the orbit and breeching the cribriform plate and extending into anterior cranial fossa.

Histopathological examination revealed the tumor cells in sheets, vague whorls and short fascicles intervening in between bony trabeculae along with multiple foci of psammomatous calcification. The cells were monomorphic, epithelioid to spindled, with indistinct cell border, ovoid nucleus with fine chromatin, inconspicuous nucleoli and eosinophilic cytoplasm (Figure 2). Mitotic activity was sparse (0-1/10 high power field) and necrosis was not seen. Immunohistochemistry showed diffuse and weak EMA immunoreactivity in the tumor cells. Synaptophysin, chromogranin A and S100p were negative. Nuclear labeling with Ki-67 was almost negligible (<1%) (Figure 3). A diagnosis of Transitional meningioma, WHO grade I was given based on the histomorphology and immunoprofile.

Following the diagnosis, the mass was excised and the patient is currently on follow-up for 6months which has been uneventful

so far.s

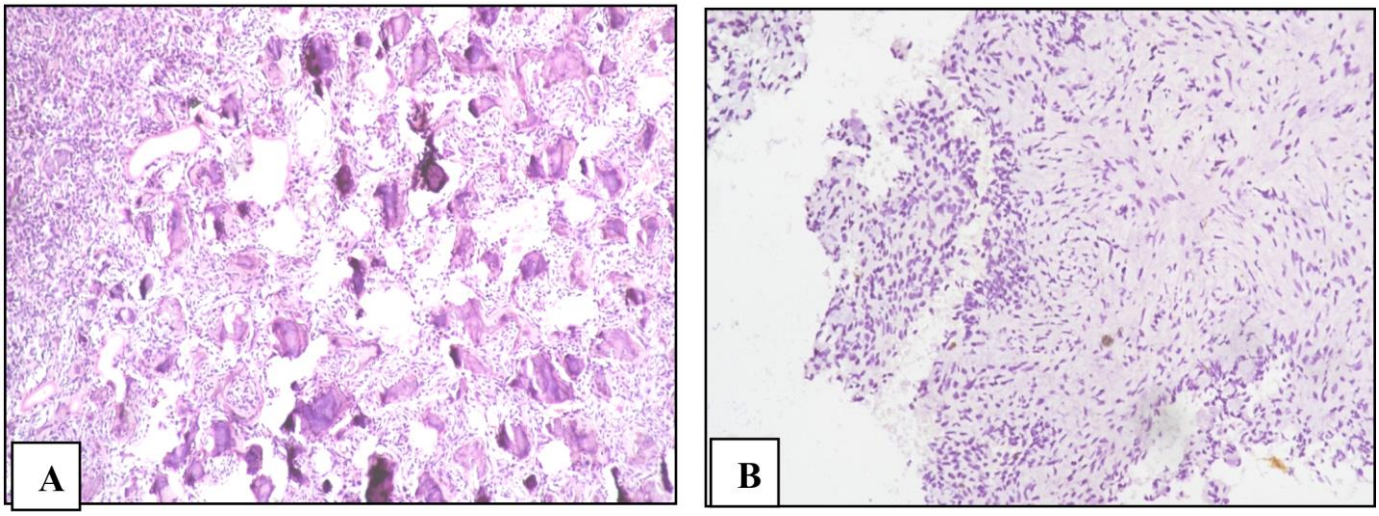

Figure 2: (A-B) Histopathological examination shows tumor cells with psammomatous calcification (A), tumor cells arranged in sheets, vague whorls and short fascicles.

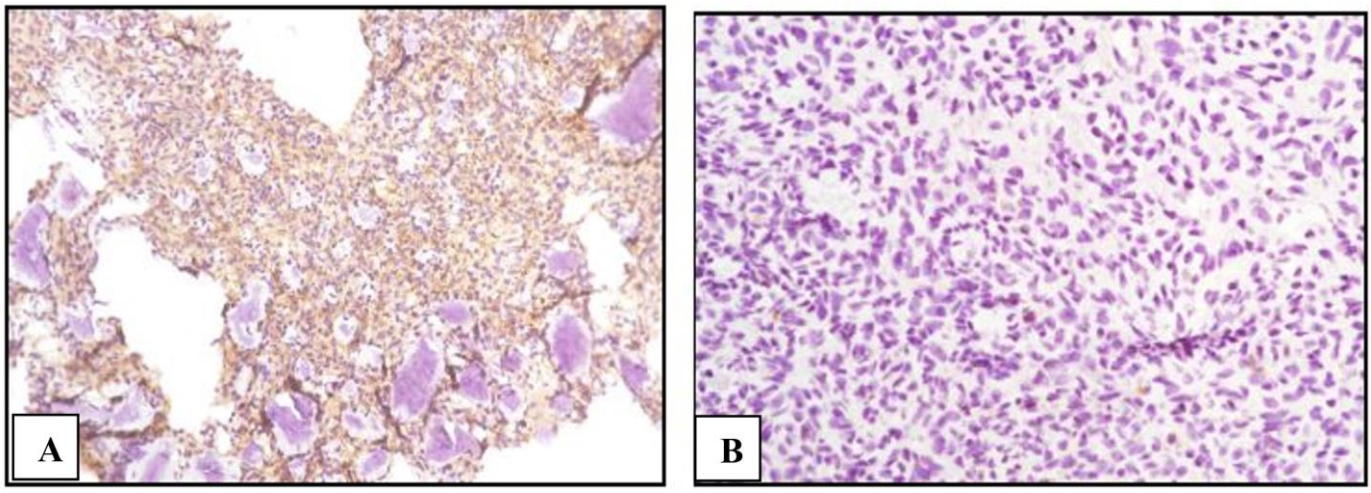

Figure 3: (A-B) Immunohistochemistry of the tumor cells showing positivity for EMA (A) and negligible Ki-67 activity (B).

Discussion

Meningioma is a non-glial central nervous system tumor that typically arises in proximity to meninges. Rarely (< 2% cases), they occur extracranially, especially in head and neck region, sinonasal tract, ear, temporal bone and scalp.[5] Twenty percent extracranial meningiomas are secondary extensions of the intracranial tumor.[6] Primary extracranial meningiomas without direct communication with the intracranial region are rare. There are different mechanisms leading to the development of the extracranial meningiomas. During embryogenesis, arachnoidal cells that are present in the sheaths of nerves or vessels emerge through the skull foramina, get displaced, pinched off or entrapped in an extracranial location. It can also occur following a traumatic event or cerebral hypertension that displaces the arachnoid islets, originated from undifferentiated or multipotent mesenchymal cells. Histologically, primary extracranial meningiomas are identical to intracranial counterparts.[7]

Meningiomas have a predilection for females and have bimodal age distribution with first peak in the second decade and second peak during fourth to seventh decades of life. Meningiomas involving the sinonasal tract may mimic sinusitis with patients presenting with the nasal obstruction, anosmia, facial pain, nasal discharge, and epistaxis. The differential diagnosis of sinonasal tract meningiomas includes mucocele, hemangioma, angiofibroma, olfactory neuroblastoma, undifferentiated carcinoma and sarcomas. Histomorphology and immunohistochemistry are confirmatory for the diagnosis. Meningiomas are strongly immunoreactive to vimentin, EMA and pancytokeratin.[5][7][8]

Maharjan et al reported a rare case of primary extracranial atypical transitional meningioma of nasal cavity in an elderly female with no recurrence upto 20 months of follow-up.[7] Gozgec et al reported a 44 year old female presenting with right-sided nasal obstruction and intermittent epistaxis which was clinically diagnosed as a bleeding nasal polyp. The mass was excised and was diagnosed as meningioma on histopathologic examination.[9] Xanwen Hu et al reported a 31year old male presenting with progressive nasal obstruction and polypoidal mass in nasal cavity which was diagnosed as ectopic fibrous meningioma on histopathology with no recurrence post excision.[4] Magdayao et al presented a case of a 53 year old male with right nasal obstruction and recurrent epistaxis with nasal polyp for two years. CT scan showed a large expansile right intranasal mass with dehiscence of the cribriform plate of ethmoid with extradural extension. A diagnosis of ectopic meningioma in the sinonasal tract was made on the punch biopsy and the patient underwent excision of the intracranial extension of the tumor along with intranasal meningioma. Follow up was uneventful.[10] Aiyer et al reported a case of primary extracranial meningioma in a 45 year old female with right nasal mass and nasal obstruction with proptosis for three years.[11]

In the present case, there was long standing proptosis and intermittent headache, together with a mass in nasal cavity as shown in radiology. The differential diagnosis included olfactory neuroblastoma and melanoma. Negative immunoexpression for synaptophysin, chromogranin A and S100p ruled out these closely mimicking tumors at that site.

In a study done in 163 cases, the most common histopathological subtype in primary extracranial meningiomas of head was meningothelial meningioma (53.4%) followed by transitional type (12.3%), psammomatous type (11.7%), and fibrous type (6.7%).[12] The present case was a transitional meningioma, grade 1 with negligible Ki-67 proliferative activity.

Meningiomas can present in a wide variety of sites, especially within the head and neck region. They behave as slow-growing neoplasms with a good prognosis, with longest survival associated with younger age, and complete resection.[3] Awareness of this entity in an unexpected location with unusual presentation will help to avoid potential difficulties associated with the diagnosis and management of these tumors.

Conclusion

Meningiomas are slow growing intracranial neoplasm with good prognosis in younger age and with complete resection. Rarely, they originate extracranially in unusual locations like the sinonasal tract with unusual presentation resulting in frequent misdiagnosis and inappropriate management. Awareness of primary heterotopic meningioma will help in early diagnosis and appropriate management of the patient, particularly while evaluating a case of sino-nasal mass.

Abbreviations
EMA- epithelial membrane antigen,
WHO- world health organization

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