Case Report

Chordoid Meningioma, Part of a Multiple Intracranial Meningioma: A Case Report & Review

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Abstract

Chordoid meningioma, classified as atypical meningioma according to the World Health Organisation (WHO) classification, is a rare subtype, which represents only 0.5% of all meningiomas and is associated with a high incidence of recurrence. Multiple intracranial meningiomas are rare in non-neurofibromatosis patients. We present a female patient with both of these rare types of meningioma. The patient presented with two concurrent intracranial meningiomas, with one a meningotheliomatous subtype and the other a chordoid meningioma. Given the wide array of histological differential diagnoses in chordoid meningioma, immunohistochemistry has a significant role to play in differentiating them. Recurrence in chordoid meningioma can be generally predicted based on the extent of resection, the percentage of chordoid element, and proliferation indices.

Keywords: *meningotheliomatous meningioma, multiple meningiomas, benign meningioma, immunohistochemistry, recurrent brain tumor*

Introduction

Meningiomas, which account for 30% of primary intracranial neoplasms, are known to have a broad spectrum of clinical manifestations and distinct multiple histological subsets. Chordoid meningiomas (CM) belong to a rare subset of meningiomas, which have regions of histological patterns similar to chordomas. CM are associated with a high likelihood of recurrence and represent only 0.5% of all meningiomas (1). Multiple intracranial meningiomas (MIMs) are defined as at least two spatially separated meningiomas occurring at the same time, or more than two meningiomas arising sequentially from two clearly distinct regions. MIMs are rare in non-neurofibromatosis (NN) patients (2). We present an NN patient who presented with two concurrent intracranial meningiomas, where one was a purely meningotheliomatous subtype and the other was a CM.

Case Report

A 38-year-old female patient initially presented with a progressively worsening frontal headache of three years duration with no other neurological deficits. The patient had no history of seizures. Clinically, the patient had no features suggestive of neurofibromatosis. A computerised tomography (CT) scan of the brain revealed two distinctive contrast-enhancing masses over the right sphenoid wing and the left frontoparasagittal area measuring 5.8 × 5.2 cm and 2.0 × 2.0 cm, respectively. Subsequently, the patient underwent magnetic resonance imaging (MRI) of the brain with gadolinium (Figure 1). A diagnosis of right sphenoid wing meningioma (RSWM) and left frontal meningioma (LFM) was made. Craniotomy and debulking of the RSWM only were performed in view of its huge size and significant mass. Total excision of the tumour was not achievable due to its close relationship with major vessels. The LFM was left untouched. An immediate post-operative contrasted CT scan of the brain revealed a residual RSWM measuring 2.7 × 1.3 cm. Laboratory analysis of the tumour specimen identified it as a meningotheliomatous meningioma.

Two years later, on follow up, the patient reported having a recurrent headache and episodes of breakthrough seizures. An MRI of the brain with gadolinium was performed. This showed that the LFM had increased in size to 3.4 × 2.3 cm. In addition, a grossly enlarged residual tumour was noted in the right sphenoid wing within the suprasellar and pre-pontine cistern (Figure 2). Recraniotomy and debulking of

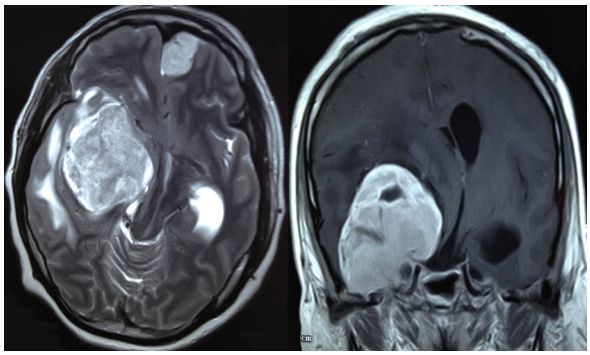

Figure 1: Results of MRI of the brain with gadolinium. Axial view on the left and coronal view on the right, showing the heterogeneously enhanced mass over the right sphenoid wing and the homogenously enhanced mass on the left frontoparasagittal. Note the right sphenoid wing encasing the right internal carotid artery and the optic nerve.

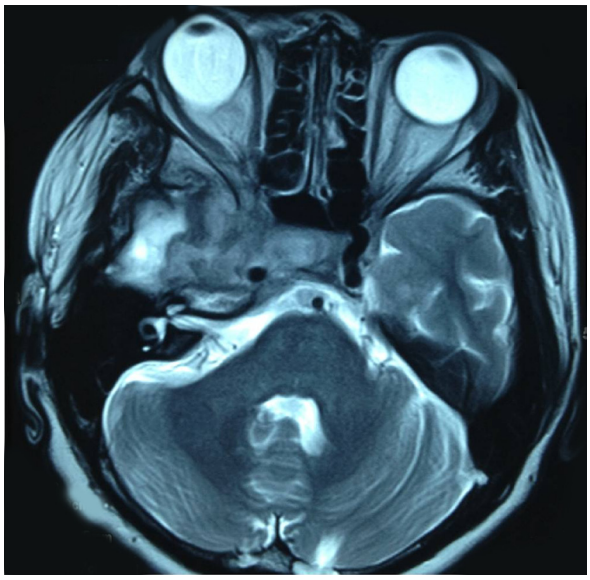

Figure 2: T2-weighed MR image of the brain two years later, showing the grossly enlarged residual tumour in the right sphenoid wing, which had infiltrated the right orbit. It infiltrated the right orbit through the widened right optic canal, as well as the right superior orbital fissure. Note the tumour tissue encasing the right internal carotid artery, cavernous sinus, and trigeminal nerve roots V1, V2, and V3. Erosion of the greater wing of the sphenoid and temporal bone is also visible.

the bilateral tumour was performed, and the LFM was totally resected. However, only debulking was done for the RSWM.

The LFM was a grade 1 meningotheliomatous meningioma according to the WHO's classification. Histopathology findings for the RSWM noted a section showing meningothelial cells. These were arranged in cords with a myxoid background, and the cells had uniform oval nuclei (Figure 3). There was no nuclear atypia or necrosis observed suggestive of a malignant tumour. Immunohistochemical staining was positive for Epithelial Membrane Antigen (EMA) and S-100 protein and negative for Glial Fibrillary Acidic Protein (GFAP). The MIB-1 proliferative index for this tumour was reported as 1–2%. The pathologist concluded that the RSWM was a CM.

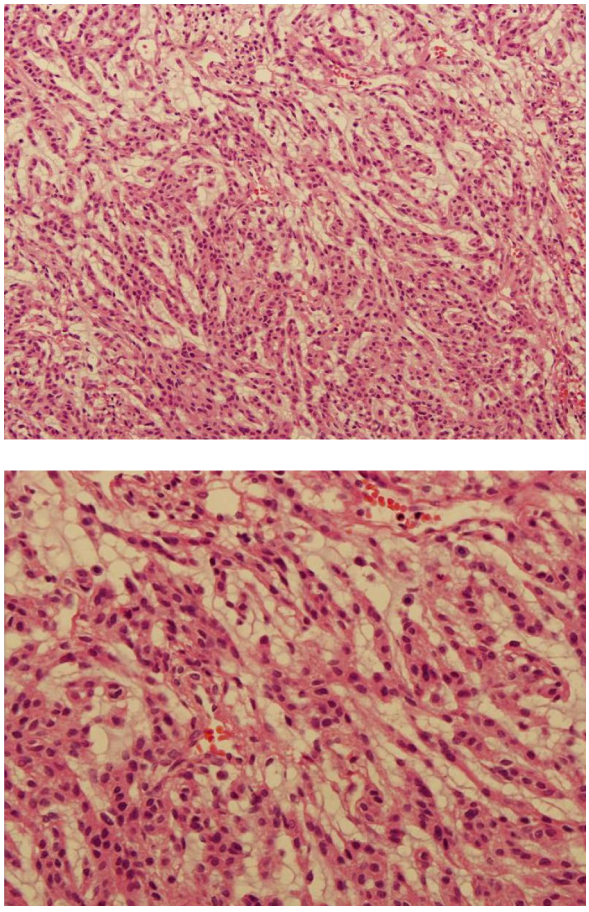

Figure 3: Histolopathology of the bottom sphenoid wing tumour, showing meningothelial cells arranged in cords with a myxoid background. (H&E stain 100× magnifications on the top and 400× magnification on the bottom).

Discussion

CMs are grade II atypical meningiomas according to the WHO's classification. Eighty-eight percent of CMs are large and supratentorial. Chordoid elements in these tumours can vary from 10 to 100% of the tumour (1).

CMs, despite being a rare variant, are often associated with peritumoural lymphoplasmacellular infiltration causing Castleman syndrome (3). In the largest-ever series of CMs reviewed, Couce et al. (1), examined 42 cases and reported no significant association with systemic or haematological abnormalities.

Histologically, CMs are characterised by strands and cords of meningothelial cells arranged in a mucinous stroma. Tumour cells in CM contain epithelial membrane antigen (EMA), D2–40, and focal S-100 protein (4). EMA cytoplasmic staining has been reported in 9 of 10 (90%) cases of CM (5). D2–40 immunostaining showed positivity in 80% of CM cases (5). Staining with S-100 exhibited focal, moderate staining in only 40% of CMs (5). GFAP and synaptophysin were reported to be negative for CM (4). In a report of 10 cases of CM, Tena Suck et al. (4), noted Ki-67 (MIB-1) proliferative index ranges from 6 to 9%, proliferating cell nuclear antigen (PCNA) Li ranges from 6 to 20%, and microvascular density ranges of 6–16%. The same study also noted that tumours that showed a recurrence were associated with a higher mean MIB-1 proliferative index compared with those without recurrence (4). In contrast to this study, our patient who had a tumour recurrence had a low MIB-1 proliferative index. Immunohistochemical findings can aid the differentiation of CM from its various pathological mimics such as chordoma and metastatic carcinoma (5,6).

As noted earlier, CM has been associated with high rates of recurrence. The major predictor in determining tumour recurrence is the extent of resection. Couce et al. (1), noted 42% of their CMs showed one or more recurrence, and 92% of those recurrent tumours were subtotally resected. In 85.7% of recurrent CMs, they also observed chordoid elements present in more than 50% of tumour tissue. In addition to these factors, proliferation indices have also been used to predict recurrence in meningioma. The Ki-67 proliferation index has been shown to exhibit a highly significant increase from benign to atypical and anaplastic meningiomas (7).

In relation to MIMs, initial reports quoted an incidence of 1–2% (8). However, this increased to 5.4 to 10.5% in different series with the advent of CT and MRI. MIMs are particularly rare in NN patients (8). There are two distinct hypotheses for its pathogenesis: i) an independently arising tumour and ii) the occurrence of a single transforming event, followed by spreading of the original clone of cells throughout the meninges (9).

As CM is a rare subtype of meningioma, it is important to be able to distinguish it from other histological differentials in view of the high likelihood of recurrence. A total resection can help to prevent the recurrence rate in this rare tumour subtype compared with a subtotal resection or debulking.

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Conflict of Interest

None.

Fund(s)

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