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(CASE REPORT)

Chordoid Meningioma Along the Sphenoid Wing: A Case Report and Brief Review of the Literature

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Abstract

Chordoid meningiomas (CMs) are rare WHO grade 2 CNS meningiomas that exhibit aggressive behavior and a high rate of recurrence. Diagnosis of CMs can be challenging, as they closely mimic other distinct tumors, such as true chordomas, chondrosarcomas, or chordoid gliomas. An accurate diagnosis requires a careful combination of MRI characteristics, immunohistochemistry (IHC) analysis, and review by an experienced neuropathologist.

We report a 49-year-old patient with a six-month history of progressive frontal headaches, intermittent blurred vision in the right eye, and dysphasia. Examination revealed subtle papilledema, mild anomic aphasia, and a right superior quadrantanopia. Brain magnetic resonance imaging (MRI) showed a heterogeneous 4.2 × 3.8 cm extra-axial mass near the left sphenoid wing with broad dural attachment, homogeneous enhancement, and marked peritumoral edema.

Because of proximity to the cavernous sinus, the patient underwent subtotal tumor resection (Simpson grade IV). Histologic examination showed cords of epithelial tumor cells in a highly cellular myxoid stroma, with frequent mitoses and focal brain invasion. IHC was positive for SSTR2A (somatostatin receptor) and EMA and negative for brachyury, supporting CM and excluding chordoma. Molecular testing identified an NF2 mutation and a DNA methylation profile consistent with CM. The patient received intensity-modulated radiation therapy after surgery. A local recurrence developed at 26 months and was treated with stereotactic radiosurgery. At 36-month follow-up, the tumor size remained stable. This case highlights the diagnostic challenges, differential diagnosis, and management of this rare tumor and includes a brief review of the literature.

Keywords: Chordoid meningiomas; Aggressive; Dural tail; Supratentorial region; Local recurrence; Intensity-modulated radiation therapy; Castleman-like

1. Introduction

Meningiomas are the most common type of primary CNS tumors arising from arachnoid cells. [1] To date, 15 histological subtypes of meningiomas have been identified and have been categorized into three World Health Organization (WHO) grades. CMs are rare variants and are classified as WHO Grade II. [2] They are characterized by trabeculae of epithelioid

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cells within a mucoid stroma on histopathology. [3] Symptoms of meningiomas depend on tumor size and location. Symptoms include vision changes, focal neurological deficits, seizures, and headaches that are worse in the morning. [4]

Histologic assessment of CMs reveals eosinophilic cords of cells containing vacuolated cytoplasm within a myxomatous matrix. This meningioma variant includes distinct areas of chordoid morphology in addition to typical meningioma morphology. [5] Due to the predominance of chordoma-like histology, there is a high likelihood of incorrect diagnosis as chordoma. Therefore, precise histopathologic confirmation is essential for definitive diagnosis.

CMs are typically treated with maximum safe surgical removal, followed by adjuvant radiation therapy to prevent recurrence. While generally favorable, the prognosis is guarded; they are classified as WHO Grade II tumors, as they have a higher rate of recurrence compared to typical benign tumors. [6]

Given their low incidence and morphologic resemblance to chordoid neoplasms, CMs remain underdiscussed, with limited literature. We report a rare case of chordoid meningioma along the sphenoid wing to highlight its diagnostic and management challenges and the essential role of a multidisciplinary team approach for optimal diagnosis and management.

2. Case Presentation

2.1. Clinical Presentation & Initial Workup

A 49-year-old woman presents to her primary care physician with a six-month history of progressive, non-pulsatile, predominantly frontal headaches, episodic blurring of vision in her right eye, and mild word-finding difficulty noted by her husband. She denied nausea, vomiting, seizures, or focal weakness. She was otherwise healthy with no prior neurological history.

She had no prior radiation exposure, no family history of neurofibromatosis 2 (NF2) or other hereditary tumor syndromes, and no history of malignancy. She was a non-smoker. Because her work involved prolonged screen time, her visual symptoms were initially attributed to screen exposure.

Ophthalmology examination was reported as follows: Vital signs were within normal limits. Neurological exams revealed subtle right-sided papilledema on fundoscopy, mild anomic aphasia, and a right visual field defect (right superior quadrantanopia). Cranial nerves II–XII were otherwise intact. Motor and sensory exams were unremarkable. No meningismus (meningism) was present. Papilledema with visual field loss suggested a space-occupying lesion and raised intracranial pressure, warranting urgent neuroimaging.

2.2. Neuroimaging & Differential Diagnosis

MRI of the brain with and without gadolinium demonstrates a 4.2 × 3.8 cm extra-axial, lobulated mass along the left sphenoid wing with broad dural attachment and a dural tail sign. The lesion was T1 isointense, T2 mildly hyperintense, and showed intense homogeneous enhancement. Significant peritumoral edema displaced the left temporal lobe, and no intratumoral hemorrhage or restricted diffusion. CT showed focal calcification. MR spectroscopy revealed elevated choline (Cho) and reduced N-acetylaspartate (NAA), indicating rapidly dividing tumor cells and the displacement of healthy brain tissue.

Differential diagnosis included meningioma, solitary fibrous tumor, dural metastasis, lymphoma, chordoma, and schwannoma. Meningioma was the leading diagnosis considering dural tail and homogeneous enhancement, but other differentials had to be ruled out.

2.3. Tumor Board Discussion, Preoperative

The case was presented at the multidisciplinary neural tumor board (neurosurgery, neuroradiology, neurology, radiation oncology, and neuropathology). Imaging favored a WHO grade 1–2 meningioma. Given the mass effect, peritumoral edema, and progressive symptoms, surgical resection was recommended as first-line management. Tumor location along the sphenoid wing with proximity to the cavernous sinus, middle cerebral artery branches, and carotid encasement were noted on preoperative angiography. Simpson grade I/II resection was unlikely to be achievable without significant vascular risk; gross total resection was the goal, but subtotal resection was anticipated.

2.4. Pathology, Intraoperative & Final Diagnosis

Frozen section intraoperative consultation favored meningioma. The excised tumor was rubbery to gelatinous, gray-tan, and lobulated. Dural attachment with focal osseous invasion was noted. It was an incomplete resection (Simpson grade IV, residual cavernous sinus component).

Microscopic examination showed tumor cells arranged in cords and clusters within a prominent myxoid-to-mucinous stroma, a characteristic feature of CM. The cells were epithelioid, with eosinophilic cytoplasm and vacuolization resembling physaliphorous cells of chordoma. Focal areas of more typical meningioma were interspersed. Atypical histomorphologic features included increased cellularity, prominent nucleoli, focal sheet-like growth, and an elevated mitotic index of 4 mitoses per 10 high-power fields. No necrosis was identified. (Figure 1 A, B, C, D) Focal brain invasion was present at the resection margin. Table 1 summarizes the IHC reaction of the tumor cells.

Table 1 Immunohistochemistry (IHC) reaction of the tumor cells

Marker	Result	Significance
EMA (epithelial membrane antigen)	Positive	Confirms meningothelial lineage
Vimentin	Positive (diffuse)	Mesenchymal marker, expected
SSTR2a (somatostatin receptor)	Positive	Diagnostic & therapeutic target
S100	Focal positive	Seen in the chordoid variant
Brachyury (T-box gene)	Negative	Excludes chordoma
GFAP	Negative	Excludes glial origin
Ki-67 (MIB-1 index)	8%	Elevated; supports WHO grade 2
Progesterone receptor (PR)	Negative	Associated with a higher grade

Molecular studies detected the NF2 mutation (loss of chromosome 22q). TERT promoter mutation was absent. CDKN2A/B deletion was not identified. Methylation profiling was consistent with chordoid meningioma, methylation class MC-CH.

The final diagnosis was Chordoid meningioma, a CNS grade 2 tumor with atypical histomorphological features. Incomplete resection (Simpson grade IV) and focal brain invasion at the margin.

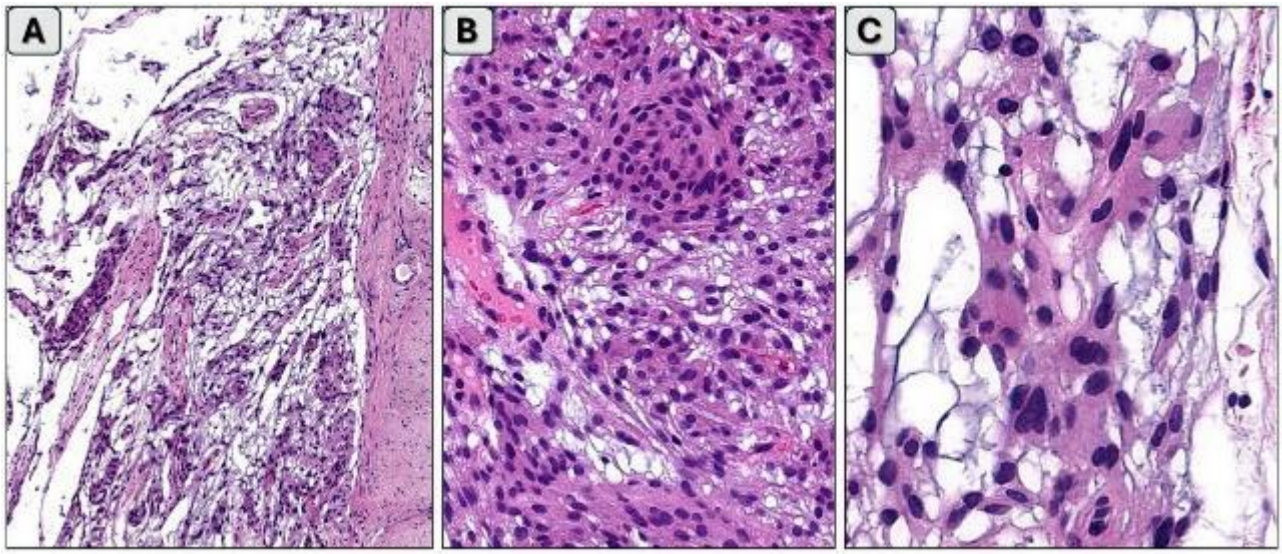
2.5. Tumor Board Presentation, Post-Operative Management

Post-operative MRI at 48 hours confirmed Simpson IV resection with residual cavernous sinus tumor cells. Given the grade 2 designation, atypical features, incomplete resection, and brain invasion, the multidisciplinary board recommends adjuvant radiotherapy. Observation alone was considered insufficient. SSTR2a positivity noted for future reference.

2.6. Treatment, recurrence & follow-up

The patient received adjuvant intensity-modulated radiotherapy (IMRT) to the tumor bed and residual disease during the first 2 months after surgery. Treatment was well tolerated, with only mild fatigue, and dexamethasone was gradually tapered. Surveillance MRI at 6 and 12 months showed stable residual disease with no significant interval change. Clinically, aphasia partially improved, although the visual field defect persisted.

At 26 months, MRI demonstrated a 2.1 cm nodular enhancement at the posterior margin of the prior resection cavity, consistent with early local recurrence, and PET-DOTATATE confirmed somatostatin receptor–avid disease. At 28 months, a second tumor board reviewed management options, including repeat resection, stereotactic radiosurgery, and enrollment in a systemic therapy trial. Because repeat resection carried high risk in the previously treated field, the team proceeded with stereotactic radiosurgery for the recurrent nodule. By 36 months, follow-up contrast-enhanced MRI showed decreased enhancement at the treated site. The patient remained neurologically stable, and MRI surveillance every 6 months was recommended with ongoing multidisciplinary follow-up.



1A: Low-power view showing tumor cells arranged in cords and clusters within a prominent myxoid-to-mucinous stroma, attached to the dura (H&E X20). 1B**:** Intermediate power view showing focal interspersed areas of more typical meningioma (H&E X40). 1C**:** High-power view showing pleomorphic tumor cells, mitosis, and focal sheet-like growth. The cells are epithelioid, with eosinophilic cytoplasm and vacuolization (H&E X60).

Figure 1 Microscopic features of chordoid meningioma

3. Discussion

3.1. Background (History, epidemiology, risk factors, and WHO classification):

Chordoid meningioma (CM) was first described by Kepes et al. in 1988 as a meningioma with a chordoma-like histologic appearance and clustering of tumor cells. [7] Epidemiologically, CM primarily affects adults, with a mean age at presentation of approximately 47 years, which is nearly a decade younger than that of non-chordoid meningiomas. CM is rare and usually described as near-equal in males and females, though some recent series show a male predominance. [8,16]

While meningiomas in general are associated with several risk factors, specific data for CM is limited. However, based on broader meningioma epidemiology and molecular findings in CM, risk factors include older age, higher BMI, history of atopic conditions, and ionizing radiation exposure. [9]

In a minority of cases, particularly in children and adolescents, CM has been associated with Castleman-like systemic inflammatory manifestations, likely related to cytokine production such as IL-6; however, this association is uncommon and is not a required diagnostic feature [Kepes] The scientific and clinical consensus today is that while a legitimate link between pediatric chordoid meningioma and Castleman disease exists, it is rare and by no means a mandatory diagnostic feature.

CMs are rare tumors, with a reported incidence of up to 1% of all intracranial meningiomas. [4] The majority of reported cases are in adults. There is an association with genetic molecular mutations, as CM has *NF2, TRAF7, KLF4, SMO, AKT1*, and *SMARCB1* mutations that are common to non-chordoid meningiomas. [10]

CMs arise most frequently in the supratentorial region and rarely in infratentorial locations [10]. A significant number of CMs exhibit a high recurrence rate and progressive behavior. [11] WHO classifies CMs as WHO grade II given their aggressive tendency and high recurrence rate. [4,12]

3.2. Pathogenesis, Pathophysiology

The exact cellular origin and molecular pathogenesis of CMs involve complex genetic and epigenetic alterations. Like classical meningiomas, chordoid meningiomas are believed to arise from the meningothelial (arachnoid cap) cells of the meninges. [8] A study by Daoud et al reported that molecular profiling indicated that CM is heterogeneous. Approximately 50-60% of cases exhibit NF2 gene mutations or loss-of-function mutations on chromosome 22q. Other tumors in this category may feature mutually exclusive alterations in TRAF7, KLF4, AKT1, or VHL. [10] In the same

study, they found that CMs have a higher abundance of chromatin remodeling genes, including EP400, KMT2C, and KMT2D, compared to non-chordoid mutations. [10]

The pathophysiology of chordoid meningiomas is driven by their specific histological composition and extra-axial growth. The fluid-rich matrix creates a gelatinous consistency and contributes to its distinct, restricted diffusion characteristics on MRI. [13] Because CMs grow extra-axially, but inside the skull, they slowly compress and displace adjacent neurological structures. This compression, combined with the release of pro-inflammatory factors, frequently induces peritumoral vasogenic edema, causing the tumor to exert a mass effect disproportionate to its actual size. [4] CMs tend to have a higher proliferative index compared to Grade I meningiomas. They can exhibit brain invasion, elevated mitotic figures, and elevated Ki-67 labeling indices. Incomplete surgical removal allows remaining cells to rapidly repopulate and grow, making the extent of surgical resection the most critical prognostic factor for survival. [8]

3.3. Comparative Analysis of Our Case with Existing Literature

The present case shared several features with the reported clinicopathologic profile of CM, while also illustrating features that complicate management at the skull base. Our patient, a 49-year-old woman without anemia, dysgammaglobulinemia, or constitutional symptoms, resembled the adult sporadic pattern described by Couce et al. and later institutional series rather than the original pediatric Castleman-associated phenotype. [14,15]

Radiologically, the left sphenoid wing location, dural attachment, dural tail, calcification, and visual symptoms were consistent with skull-base meningioma behavior, and the patient's age and tumor size were close to those reported in systematic and institutional series. [15,16] In contrast to the sphenoid-wing case reported by Matyja et al., which lacked peritumoral edema and remained recurrence-free after Simpson grade I resection, our case showed significant edema, incomplete Simpson grade IV resection because of cavernous sinus involvement, and recurrence at 26 months. [17] This clinical course was concordant with Choy et al., who found that subtotal resection and elevated MIB-1/Ki-67 were major predictors of recurrence, and with Zhang et al., who reported improved progression-free survival after gross-total resection and adjuvant radiotherapy. [15,16]

The NF2 alteration and PR negativity in our case also contrasted with more favorable molecular and immunophenotypic patterns described in the recent comparative series by Ren et al., reinforcing the biologic heterogeneity of this rare WHO grade 2 subtypes. [18]

4. What have we learned from This Case?

This case highlights the need to consider CM when a seemingly typical dural-based sphenoid wing mass is associated with disproportionate edema, progressive visual symptoms, papilledema, language impairment, or imaging evidence of nearby neurovascular involvement that may limit safe resection. Although conventional imaging strongly favored meningioma, definitive diagnosis required histopathology and IHC, particularly identification of epithelioid cells arranged in cords and clusters within a myxoid stroma and exclusion of chordoma by negative brachyury reaction. It also underscores that WHO grade 2 behavior may reflect not only histologic subtype but also brain invasion, increased mitotic activity, elevated Ki-67, loss of PR expression, NF2 alteration, and residual disease.

From a management perspective, maximal safe resection remained the foundation of treatment. However, aggressive pursuit of Simpson grade I resection was not appropriate when cavernous sinus and vascular involvement created unacceptable morbidity. In this setting, early multidisciplinary planning, adjuvant radiotherapy, somatostatin receptor–directed imaging, and long-term MRI surveillance were clinically actionable. The recurrence at 26 months despite adjuvant therapy supported the need for close follow-up rather than reassurance based solely on temporary radiographic stability.

Abbreviations

- Chordoid meningiomas (CMs);
- Immunohistochemistry (IHC);
- Intensity-modulated radiation therapy (IMRT)

5. Conclusion

This case illustrates the diagnostic and therapeutic complexity of a rare CM arising along the sphenoid wing, where imaging typical meningioma masked a biologically higher-risk WHO grade 2 tumors. It also highlights the practical

challenges of skull-based surgery and the recurrence risk associated with subtotal resection. Residual cavernous sinus disease, an elevated Ki-67 index, brain invasion, PR negativity, NF2 alteration, and subsequent local recurrence underscored the need for integrated radiologic, histopathologic, molecular, and multidisciplinary assessment.

For the medical community, this case reinforces the importance of maximal safe resection, individualized adjuvant therapy, and prolonged surveillance, even when early postoperative imaging appears stable.

Compliance with ethical standards

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Disclosure of conflict of interest

All authors make the following declarations:

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Statement of ethical approval

This work did not involve any studies on human or animal subjects performed by the authors. It was a retrospective review of archival pathology material collected during routine clinical care, and all data were fully de-identified before analysis.

Data access statement

All relevant data are included in the paper.

Author contributions

All authors contributed equally to producing this manuscript.

Statement of informed consent

The patient was lost to follow-up, and all attempts to reach the patient were unsuccessful. Therefore, the paper has been sufficiently anonymized to maintain patient confidentiality.

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