

Chordoid Meningioma with Extensive Lymphocytic Infiltration Forming Lymphoid Follicles: A Case Report

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Abstract

Background: Meningiomas, the most common primary central nervous system (CNS) tumors, include 15 distinct subtypes reflecting its broad morphological spectrum. Most subtypes have a benign clinical course and correspond to CNS World Health Organization (WHO) grade I. Chordoid meningiomas are a rare subtype (0.5% of all meningiomas) with relatively aggressive behavior and are classified as WHO grade II.

Case presentation: We present a case of a 20-year-old man who presented to the emergency room with convulsions. A right parietal cerebral mass with peritumoral edema and a significant mass effect was discovered by magnetic resonance imaging. There was no systemic manifestations and hematological tests were normal. He underwent surgery and a histopathological examination of the excised tumor revealed the diagnosis of chordoid meningioma, WHO grade II. In addition, there was dense lymphocytic infiltration forming lymphoid follicles.

Conclusion: Chordoid meningioma may be associated with extensive lymphoid infiltration with follicle formation in the absence of systemic or hematologic manifestations.

Keywords: Chordoid, Lymphoid follicles, Meningioma

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Introduction

Meningiomas are the most common primary central nervous system (CNS) tumors ¹. The risk of meningioma increases with age with a median age at diagnosis of 66 years. Across all ages, the incidence of grade I meningioma is 2.32 times greater in women than in men, with the greatest risk differential (3.28) seen before menopause and decreasing thereafter. The incidence is significantly higher in Black people than in White people ².

The presentation of meningioma is in general non-specific and depends on its location and how it affects the different areas of the brain and CNS vasculature.

For example, neurological deficits are the usual presentation in skull base meningiomas. On the other hand, convulsions are a frequent presentation for non-skull base lesions. Overall, the most common presenting symptoms are headache, cranial nerve deficits, convulsions, cognitive changes, and weakness. In 9% of cases, meningioma is asymptomatic ³.

Meningioma is a single category in the WHO's Central Nervous System 5th edition, with 15 distinct subtypes reflecting its broad morphological spectrum. Most subtypes have a benign clinical course and correspond to CNS WHO grade I ².

Grade I meningiomas include the angiomatous, fibrous, lymphoplasmacyte-rich, meningothelial,

metaplastic, microcystic, psammomatous, secretory, and transitional subtypes which are characterized by the presence of < 4 mitoses per 10 HPF³. Grade II meningiomas include the atypical, chordoid and clear cell subtypes. Atypical meningioma is an intermediate-grade meningioma with increased mitotic activity (4–19 mitoses per 10 HPF), brain invasion, and/or at least three of the following: high cellularity, small cells with a high N/C ratio, prominent nucleoli, sheeting (uninterrupted patternless or sheet-like growth), and foci of spontaneous (non-iatrogenic) necrosis³. Grade III meningiomas include the anaplastic, papillary and rhabdoid subtypes. Anaplastic (malignant) meningioma is a high-grade meningioma with overtly malignant cytomorphology (anaplasia) that can (1) resemble carcinoma, high-grade sarcoma, or melanoma, (2) display markedly elevated mitotic activity (20 or more mitotic figures in 10 consecutive HPF) (3) harbor a TERT promoter mutation; and/or (4) have a homozygous CDKN2A and/or CDKN2B deletion^{2,3}.

Chordoid meningioma is categorized as grade II by the WHO due to reports of aggressive behavior and high propensity for rapid recurrence unless a complete surgical resection is obtained⁴. The chordoid subtype was first described by Kepes et al⁵. Chordoid meningiomas make up only 0.5% of meningiomas, which makes them extremely rare^{6,7}. The tumor cells resemble chordoma cells^{5,8}. Many cases of chordoid meningiomas have been associated with microcytic normo- or hypochromic anemia, or Castleman's disease in early age groups^{7,9}. Chronic inflammatory infiltrates may be significant but are typically patchy when present⁹.

Case report

A 20-year-old male presented in the emergency room with convulsions. His laboratory investigations were normal. A cerebral right parietal, partially cystic and partially solid mass with peritumoral edema and significant mass effect was detected in magnetic resonance imaging (Figure 1). The pathology

department received the tumor after it had been excised.

Pathological findings

The specimen's overall evaluation revealed a well-defined mass with dimensions of 4x4x3 cm with a rubbery grayish white and gelatinous cut section. According to histological analysis, the tumor consisted of focal solid areas resembling meningothelial meningioma and areas of epithelioid tumor cells with eosinophilic cytoplasm within the mucoid matrix stained positively for Alcian blue. In the cellular areas, the mitotic count was 4-5 mitosis per 10 high-power fields. There was a remarkably dense lymphocytic infiltration forming lymphoid follicles (Figure 2).

According to immunohistochemical analysis, the tumor cells had positive epithelial membrane antigen (EMA) staining and the tumor's Ki-67 proliferative index was 5%. Glial fibrillary acid protein (GFAP) and progesterone receptor were negative in the tumor cells (Figure 3). Consequently, chordoid meningioma, WHO grade II, was the final diagnosis.

Discussion

Chordoid meningioma is a variant of meningioma mimicking chordoma and is made up of trabeculae and cords of epithelioid cells (or, less frequently, spindle cells) in a stroma that is myxoid-rich^{10,11}. Mucicarmine, periodic-acid-Schiff, and Alcian blue are used to stain this myxoid stroma¹¹.

Even in the absence of enhanced mitotic activity, brain invasion, or other unusual criteria, chordoid meningiomas are classified as a WHO grade II variant because they are associated with greater rates of recurrence^{4,11}.

Earlier publications linked chordoid meningioma to Castleman-like syndrome due to reports describing refractory microcytic hypochromic anemia in patients with chordoid meningioma and this was explained by an assumed systemic reaction to peritumoral lymphoblastcellular infiltrate^{12,13}. On the other hand, a case series of chordoid meningioma suggested that there is no connection between it and systemic manifestations⁶.

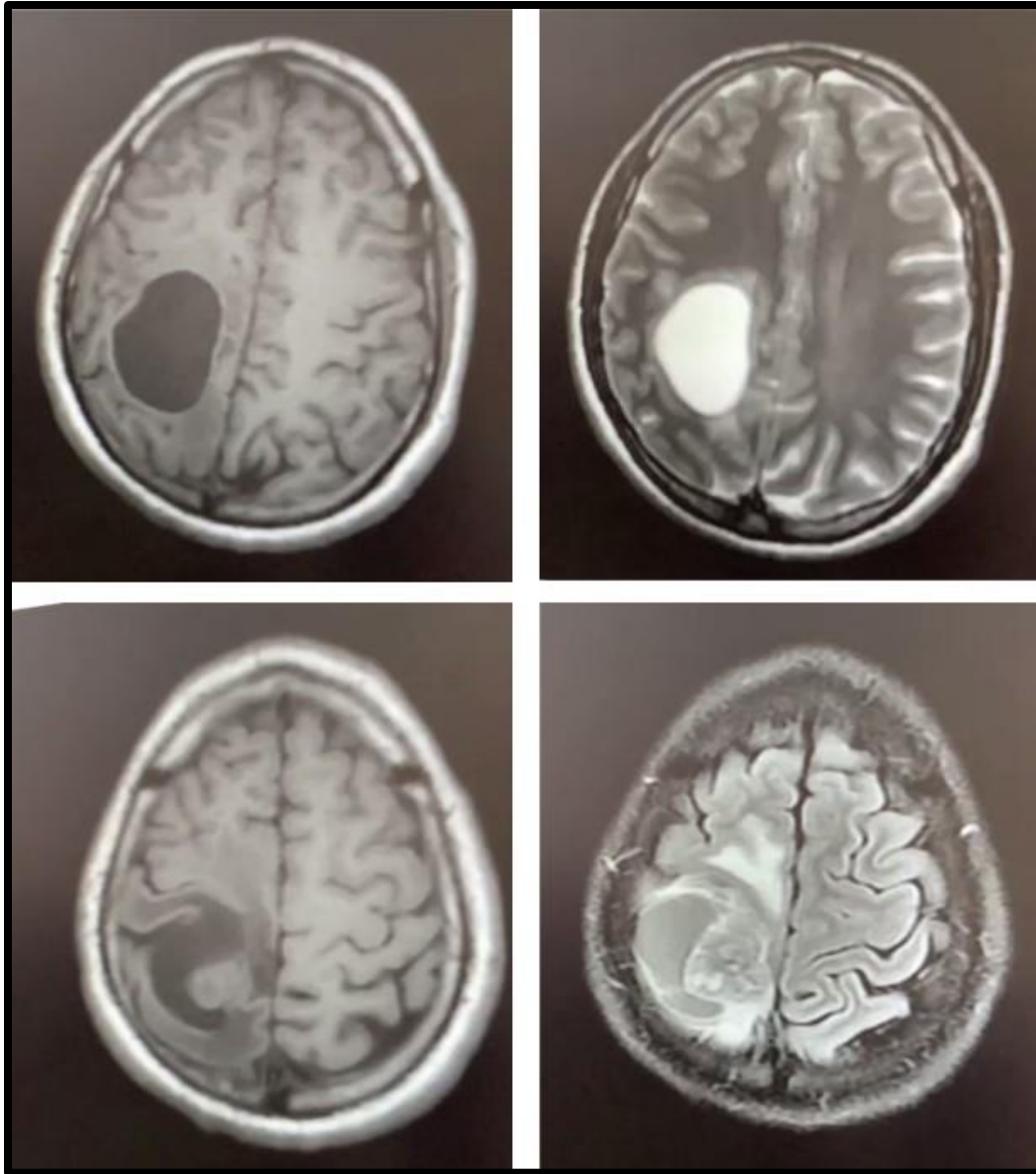


Figure 1: Magnetic resonance imaging showing right parietal well defined heterogenous extra-axial mass

Despite ultimate relationships to Castleman's disease, recent reports have not confirmed this association ^{4,14}. Chronic inflammatory infiltrates may be significant in chordoid meningioma but are typically patchy when present ⁹. It can be associated with lymphoid follicles ¹¹. This lymphoid infiltration is not a required histological finding for the diagnosis of chordoid meningioma, but it has been linked to exclusively childhood chordoid meningiomas ^{7,15}. In our case, however, lymphoid cell infiltration was

significant with lymphoid follicle formation and there were no systemic or hematological problems.

The microscopic diagnosis of chordoid meningioma must be made based on the tumor's histological similarity to a chordoma as well as the classic whorls of meningioma which should be discernible even in focal areas. Moreover, confirmation by immunohistochemistry is required to rule out the possible differential diagnoses such as chordoma, chordoid glioma and extra-skeletal myxoid chondrosarcoma.

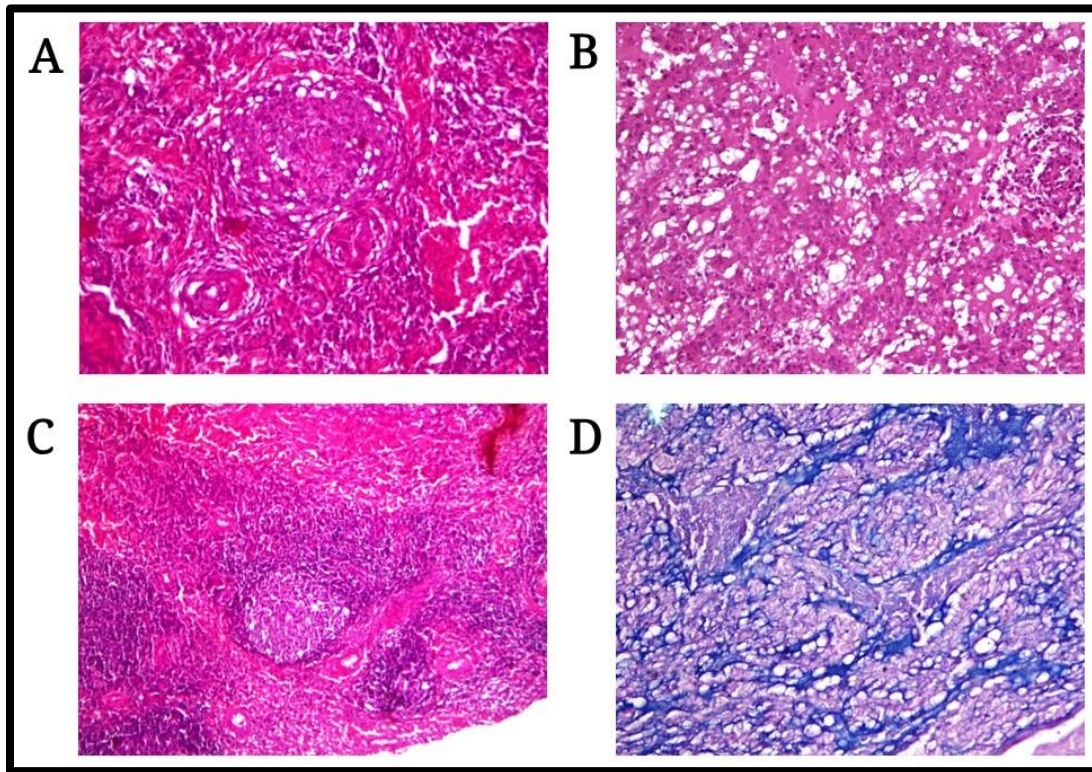


Figure 2: A) Solid area with meningotheelial meningioma morphology [H&E x400], B) Chordoma like areas, showing epithelioid tumor cells with eosinophilic cytoplasm within a mucoid matrix [H&E x200], C) Extensive lymphoid follicles [H&E x200], D) Mucoid matrix stained blue [Alcian blue x200]

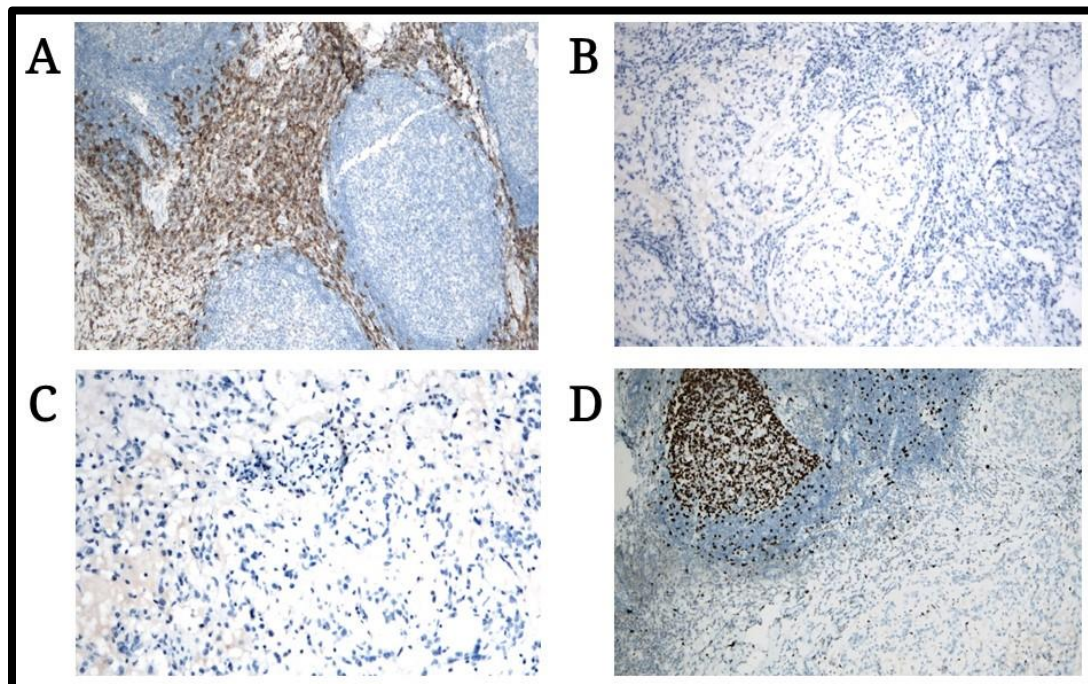


Figure 3: Immunohistochemistry revealing: A) Positive tumor cells for epithelial membrane antigen [x200], B) Negative GFAP [x200], C) Negative progesterone receptors [x200], D) KI67 proliferation index in 4-5 % of tumor cells highlighting the active germinal center [x200]

Unlike chordoid meningiomas, chordoma occurs in extradural midline structures like the clivus and sellar region. Chordoid glioma tumor cells are positive for GFAP in addition to the intra-axial location¹⁶.

Primary intracranial malignancies with extra-skeletal myxoid chondrosarcoma (EMC) features are extremely rare. Diagnosis depends on fluorescent in-situ hybridization (FISH) testing for EWSR1 break-apart signals¹⁷.

Conclusion

Chordoid meningioma may be associated with extensive lymphoid infiltration and follicle formation even if not associated with systemic or hematologic manifestations.

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Authors' contribution

Conception & Design: Both authors; Acquisition, analysis, or interpretation of data: FYF; Drafting the manuscript: FYF; Revising the manuscript: SMT; Approval of the final version of the manuscript: Both authors; Agreement to be accountable for all aspects of the work: Both authors.

Conflict of interest

The authors declare that they have no conflict of interest to disclose.

Data availability

Included in the manuscript.

Ethical considerations

All procedures performed in the study were in accordance with the ethics standards of the Institutional Research Committee and with the 1964 Helsinki declaration and its later amendments.

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Study registration

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