

# Lateral ventricle chordoid meningioma presenting with inflammatory syndrome in an adult male: A case report

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**Abstract.** Chordoid meningioma (CM) is a rare type of intracranial tumor. Intraventricular CM presenting with inflammatory syndrome is also rare. Meningioma is uncommonly accompanied by fever. The present case report documents a 28-year-old male who was admitted to the Affiliated Taian City Central Hospital of Qingdao University (Taian, China) with a 7-day history of unexplained fever and a 3-day history of progressive headache, which was accompanied with blurred vision in the right eye. Laboratory findings revealed an inflammatory condition with increased C-reactive protein levels, elevated erythrocyte sedimentation rate and moderate leukocytosis. MRI also revealed a lesion located in the right lateral ventricle. Subsequently, the tumor was excised through the right transtrigone lateral ventricle route and the tumor was then completely removed. H&E staining revealed characteristic cords of meningeal epithelial cells embedded in a prominent myxoid background, with numerous lymphocytes and plasma cells surrounding the tumor. Immunohistochemical analysis indicated focal positive staining for epithelial membrane antigen and S100, and negative staining for glial fibrillary acidic protein. Following pathological examination, the tumor was identified to be a CM. During the early postoperative course, the clinical symptoms disappeared and the hematological values returned to normal. No evidence of tumor recurrence was observed after 24 months of follow-up. To the best of our knowledge, the present study was the second to report the case of an adult patient with lateral ventricle CM presenting with inflammatory syndrome and it was the first case in an adult male.

## Introduction

Chordoid meningioma (CM) was first described by Kepes *et al* (1) and is a rare type of meningioma. It accounts for <0.5% of all meningioma types (2). CM has an aggressive clinical course and is classified to be a grade II meningioma according to the World Health Organization (WHO). The recurrence rate of the disease has been reported to be high, especially following a subtotal resection (2-7). CM is characterized by the presence of chordoma-like histopathological features and lymphoplasmacytic infiltration (2,8). With abundant chordoid elements, CMs are composed of epithelioid or spindle cells, forming cords or nests, in a pale and basophilic mucoid matrix (2,4,6). The tumor cells are diffusely positive for epithelial membrane antigen (EMA) and vimentin (9). The most frequent symptoms of CM were headache or dizziness, followed by visual or auditory impairment, limb weakness, and epilepsy (10,11).

Ventricular meningiomas account for 0.5-5% of all intracranial meningiomas (12,13). However, intraventricular CM is rare, with only 18 cases reported in the English-language literature as of 2022 (6,10,11,14-19). Inflammation is uncommon in the majority of cases of ventricle CM (17). To date, to the best of our knowledge, only two cases of lateral ventricle CM presenting with inflammatory syndrome have been reported (15,17), comprising a male pediatric patient (age, 11 years) and an adult female patient (age, 37 years). Therefore, the present case report documents the first case of lateral ventricle CM with inflammatory syndrome in an adult male.

## Case report

A 28-year-old male was admitted to the Affiliated Taian City Central Hospital of Qingdao University in January 2020 with a 7-day history of fever of unknown cause (body temperature, 37.5-38.3°C) and a 3-day history of progressive headache, with paroxysmal blurred vision in the right eye. The patient was a resident of Taian City, Shandong Province, North China, without any prior history of hematological or infectious diseases, or any family history thereof. Physical examination indicated no remarkable findings, whilst neurological examination revealed no deficits. In addition, no visual acuity impairment or visual field defect was recorded during the preoperative examination.

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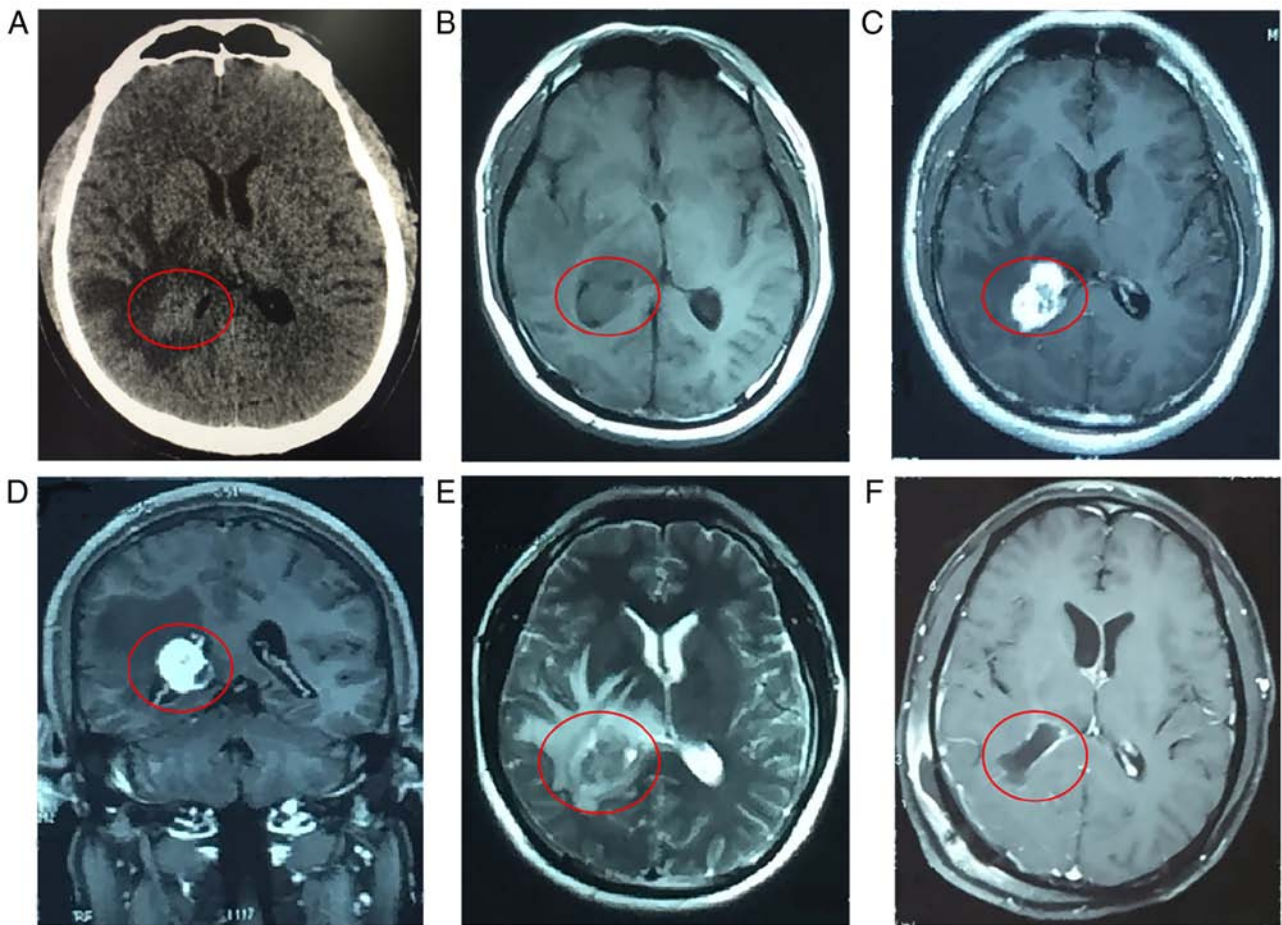


Figure 1. CT and MRI images of the tumor. (A) Preoperative CT scan of the head revealing a 3.5-cm diameter tumor (red oval) located in the right lateral ventricle. (B) Preoperative T1-weighted axial MRI displaying an isointense lesion (red oval). (C) Preoperative T1-weighted gadolinium-enhanced axial MRI and (D) coronal MRI depicting a homogeneously contrast-enhanced mass (red oval). (E) Preoperative T2-weighted axial MRI images displaying the hyperintense tumor (red oval) surrounded by brain edema. (F) Postoperative contrast-enhanced MRI image displaying the right posterior horn of the lateral ventricle (red oval) after complete removal of the tumor with Simpson grade I resection.

Echo scanning indicated no hepatosplenomegaly or lymphadenopathy. The patient did not receive any intervention prior to hospitalization. Laboratory findings revealed that the patient had inflammatory syndrome with a C-reactive protein (CRP) levels of 58 mg/l (normal range, 5-10 mg/l), an erythrocyte sedimentation rate (ESR) of 99 mm/h (normal range, 0-40 mm/h) and a white blood cell count of  $16.78 \times 10^9$  cells/l (normal range,  $4-10 \times 10^9$  cells/l). Routine blood examination indicated no anemia. In addition, since routine urine examination suggested no abnormalities, urinary tract infection was therefore excluded. A CT scan of the head [performed with a Toshiba 64-slice CT-scanner (Aquilion; Toshiba Medical Systems); imaging parameters: Voltage, 120 kV; tube current, 50 mA; scanning thickness, 1 mm] revealed a 3.5-cm diameter tumor located in the right lateral ventricle (Fig. 1A). In addition, T1-weighted MRI images [acquired with a 3.0-T Philips Ingenia MRI scanner (Philips Medical Systems); brain MRI with unenhanced and contrast-enhanced T1 weighted: Repetition time (TR), 673 msec; echo time (TE), 8.4 msec; T2 weighted: TR, 6,200 msec; TE, 123 msec], slice thickness, 5 mm] displayed an isointense lesion that was homogeneously enhanced following gadolinium administration (Fig. 1B-D). On T2-weighted MRI scans, the lesion appeared hyperintense and was surrounded by brain edema (Fig. 1E). A preoperative

chest CT scan was also performed, revealing no abnormalities. The patient provided written informed consent. As this was a single case investigation, formal approval from the local ethics committee was not required for this study.

Following hospitalization for 3 days, the lesion was excised through a right transtrigone lateral ventricle approach. The tumor was removed by Simpson grade I resection (Fig. 1F). Antiemetic (ondansetron, 4 mg i.v.) was given as needed. 5% glucose dissolved in normal saline was used as nutritional support. The patient was given appropriate dehydration (mannitol, 150 ml twice a day for 5 days) due to postoperative cerebral edema. During the postoperative period, despite the absence of antibiotics, the patient exhibited gradual improvement of the symptoms and remained afebrile, accompanied by a decline in the ESR and CRP levels in addition to the white blood cell count. On day 3 after surgery, the headache and blurred vision disappeared. The laboratory values returned to normal 11 days after surgery (Table I) and the patient was discharged the following day.

Histopathological examination of the excised tumor was performed as follows: Tissue specimens were fixed in a 10% buffered formaldehyde solution and embedded in paraffin wax. Sections of all specimens ( $3 \mu\text{m}$  thickness) were cut and stained with haematoxylin and eosin (H&E) according to a standard



Table I. Changes in laboratory findings prior to and after the operation.

Laboratory findings	Erythrocyte sediment rate, mm/h (normal range, 0-20)	C-reactive protein, mg/l (normal range, 5-10)	White blood cell count, $\times 10^9/l$ (normal range, 4-10)
Day 1 after admission	58	99	16.78
Day 1 post-OP	31	78	17.29
Day 3 post-OP	17	46	8.13
Day 11 post-OP	2	5	5.02

OP, operation.

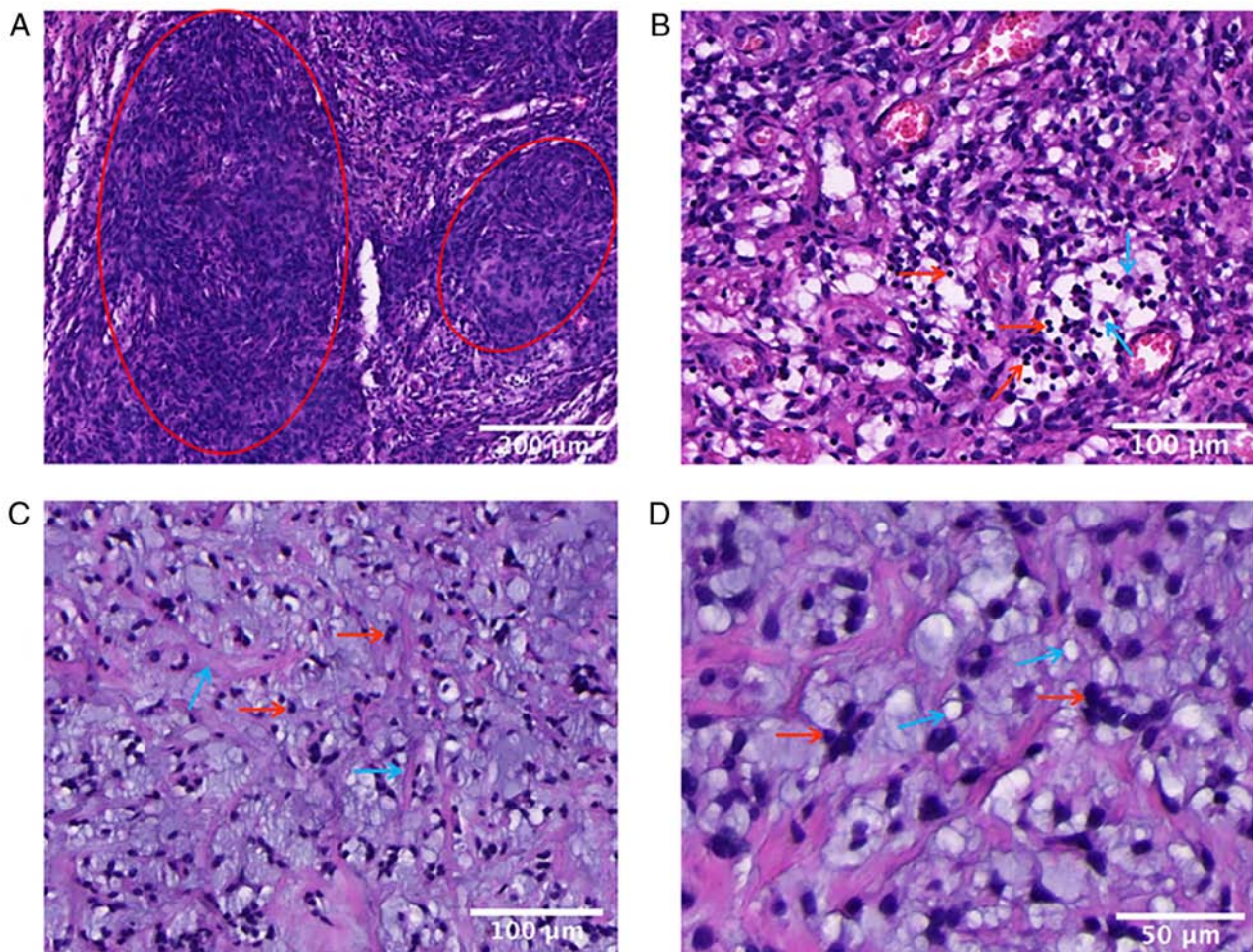


Figure 2. Histological analysis of tissue sections. (A) Tissue section including typical meningeoma-like areas (red oval; scale bar, 200  $\mu\text{m}$ ; H&E staining). (B) A growing number of lymphocytes (red arrow) and plasma cells (blue arrow) are visible around the tumor. (C) Typical meningeoma cells (red arrow) embedded in mucinous stroma (blue arrow) are visible (scale bars, 100  $\mu\text{m}$ ; H&E staining). (D) Typical meningeoma cells (red arrow) accompanied by coexisting areas of spindle-like cells embedded in mucinous stroma with cytoplasmic vacuoles (blue arrow) are present (scale bar, 50  $\mu\text{m}$ ; H&E staining).

protocol. After H&E staining, a camera was used to acquire color images of the H&E staining. The histological analysis revealed meningotheelial cells arranged in characteristic cords (Fig. 2A), with a large number of lymphocytes and plasma cells around the tumor (Fig. 2B). In particular, the tumor cells were embedded in a prominent myxoid background (Fig. 2C and D). Immunohistochemical analysis was then performed as follows: Sections of freshly excised tissue were fixed in 4% neutral buffered formalin, embedded in paraffin and sectioned at

5- $\mu\text{m}$  intervals. To retrieve antigens, sections were then placed in sodium citrate buffer and heated in a pressure cooker for 3 min. Endogenous peroxidase was blocked with 3% hydrogen peroxide for 10 min at room temperature. Sections were incubated with antibody diluted in PBS at 4°C overnight and then with the HRP-conjugated secondary antibody at 37°C for 30 min. Slides were visualized by using 3,3'-diaminobenzidine, counterstained with light hematoxylin, dehydrated and mounted with coverslips. Immunohistochemical staining



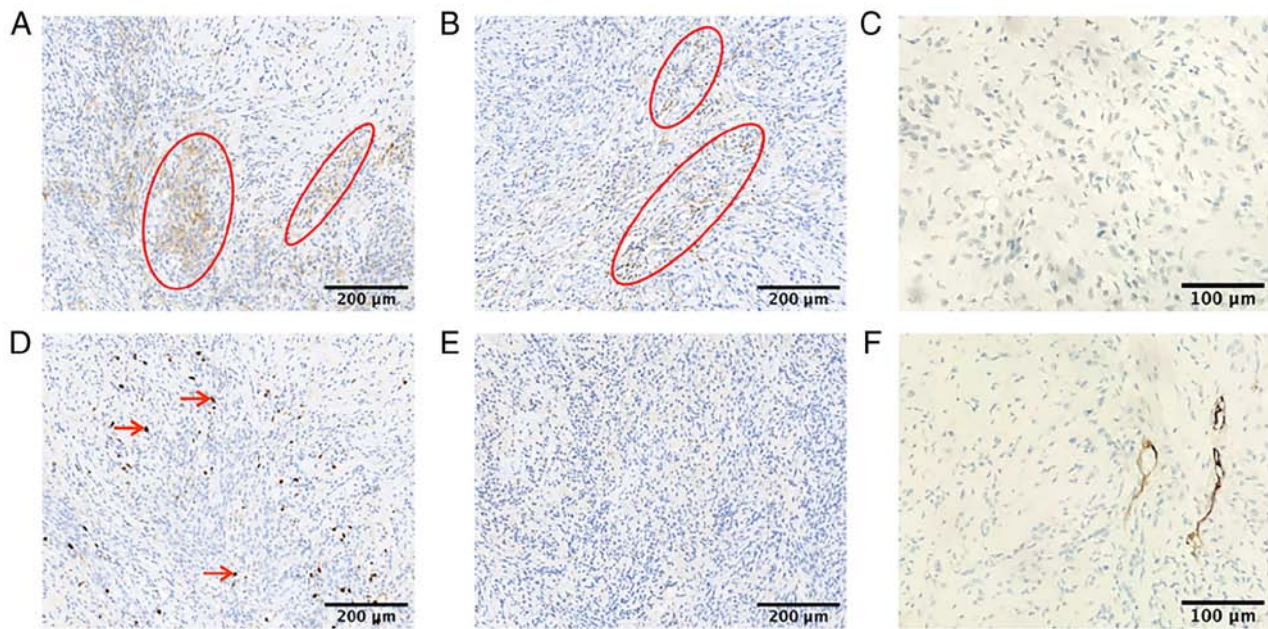


Figure 3. Immunohistochemical analysis. (A) Focal cells (red oval) positive for epithelial membranous antigen. (B) Focal cells (red oval) positive for S100 protein (scale bars, 200  $\mu$ m). (C) Negative staining for thyroid transcription factor-1 is presented (scale bar, 100  $\mu$ m). (D) The Ki-67 proliferation index was <10% (red arrows, Ki-67 positive cells). (E) Negative staining for glial fibrillary acidic protein (scale bars, 200  $\mu$ m). (F) Tumor cells were negative for CD34 (brown) staining (scale bar, 100  $\mu$ m).

was performed with the Envision technique using monoclonal antibodies (Maixin Biological Technology) against EMA (1:50 dilution; cat. no. Kit-0011), glial fibrillary acidic protein (GFAP; 1:100 dilution; cat. no. MAB-0769), S100 (1:300 dilution; cat. no. Kit-0007), transcription termination factor 1 (TTF-1; 1:100 dilution; cat. no. MAB-0677), CD43 (1:200 dilution; cat. no. MAB-0892) and Ki-67 (1:300 dilution; cat. no. MAB-0672) as primary antibodies. The Elivision Super kit (Maixin Biological Technology) was used for secondary antibody incubation and the DAB plus kit (Maixin Biological Technology) was used to develop immunostaining. Appropriate positive and negative controls for each antibody were run in parallel. The Ki-67-stained sections were evaluated by an experienced neuropathologist (HY). All counts were performed at a magnification of 9,400 (field size, 0.16 mm<sup>2</sup>) and five viable fields from the area of maximum labelling were chosen for counting. Distinct nuclear Ki-67 staining of the tumor cells was recorded as positive. The Ki-67 labelling index (Ki-67 LI) was calculated as the percentage of Ki-67-positive tumor cells in the evaluated area. Vascular components and haematogenous cells were excluded, and evaluated areas also excluded necrotic, degenerate and poorly preserved areas. As presented in Fig. 3, immunohistochemistry revealed focal cells with positive staining for EMA (Fig. 3A) and S100 protein (Fig. 3B). In addition, the Ki-67 proliferation index was <10% (Fig. 3D), whilst the tumor cells were negative for GFAP, TTF-1 and CD34 (Fig. 3C, E and F). This pathological examination led to the diagnosis of grade II CM according to the WHO.

A follow-up was conducted 24 months after the operation. At 24 months postoperatively, the patient had recovered well and no longer suffered from fever, headache or blurred right-eye vision. The patient did not experience any complications or additional symptoms associated with the disease and the tumor exhibited no signs of recurrence. Finally, the

laboratory values were in their normal ranges at 24 months after discharge. Therefore, the patient received no further treatment, such as adjuvant radiotherapy.

Since the symptoms of discomfort disappeared after tumor resection. The last follow-up date was January 10, 2022, the patient was able to have a normal life and he was satisfied with the operation.

## Discussion

CM is an atypical meningioma classified as a grade II meningioma according to the WHO (20). It has been previously reported that following the subtotal resection of CM, there is a high tendency for recurrence (21). CM has a higher incidence rate in middle-aged patients (3,18,22), and is a type of meningioma that predominantly exhibits areas with chordoma-like features (17). A chordoma-like appearance and a meningothelial cell-like pattern in certain areas of the tumor form the basis for the diagnosis of CM (21). Histopathologically, CMs are composed of fusiform-shaped epithelioid cells with eosinophilic cytoplasm arranged in 'chains and cords' patterns, in a basophilic extracellular matrix (23). The pathological characteristics of CMs are relatively comparable with several other tumor types, such as chordoid glioma, chordoma, papillary ependymoma and epithelioid hemangioendothelioma. Chordoid glioma exhibits strong positivity for GFAP expression (22). Chordomas originate from embryonic notochordal remnants and are commonly located in the midline along the spinal axis, and typically exhibit positive immunohistochemical staining for cytokeratins, S100 and EMA (24), whilst the expression of nuclear brachyury is considered a hallmark for the diagnosis. By contrast, papillary ependymomas exhibit finger-shaped structures, which are lined with a single layer of cuboidal cells with smooth surfaces and positivity for GFAP expression (25). Intracranial epithelioid

hemangioendothelioma is characterized by the presence of epithelioid tumor cells with prominent intracytoplasmic vacuolation and typical disposition in the cords or nests in a myxoid stroma. These endothelial cells tend to express the CD34 marker strongly (26). Therefore, cytological features accompanied by histological examinations are beneficial for the diagnosis of CM.

The first cases of CM in children and adolescents were reported by Kepes *et al* (1), which indicated that CM in the seven pediatric cases was commonly associated with systemic symptoms, such as iron-refractory hypochromic/microcytic anemia, hepatosplenomegaly, bone marrow plasmacytosis and dysgammaglobulinemia (Castleman syndrome), but not with fever. However, the cause of this systemic inflammation in patients with CM remains elusive. Denaro *et al* (27) suggested that the prominent lymphoplasmacellular infiltration in CM may be the cause. In previous case report, IL-6 was detected in samples derived from patients with CM, suggesting that the manifestation of inflammation may be associated with the excessive production of pyrogenic cytokines (15,28). The present case report of CM in an adult male patient provided histopathological evidence of chronic lymphocytic infiltrates. Removal of the mass, which was able to eliminate the source of pyrogenic cytokines, was able to explain why the fever immediately subsided in this patient. Fever may be one of the manifestations of tumors with inflammatory cell infiltrations (2,8). Considering that in the present case, the expression of pyrogens was not measured, their association with systemic inflammation could not be verified. It is possible that the pyrogens are secreted by the tumor itself, which is supported by the alleviation of these manifestations after tumor resection. Therefore, the febrile state may be largely caused by the tumor itself.

The paroxysmal blurred vision disappeared on day 3 after the operation. In the present case, the cause of blurred vision was difficult to explain. However, it may be due to the fact that CM is able to compress a part of the optic radiation.

Gross total resection should be the goal for the surgical management of primary CM, since the extent of resection is considered to be the most significant prognostic factor for the treatment of CM (7). In addition, the extent of the resection remains to be the most effective predictor of long-term tumor control rate (22). Since the risk of recurrence in patients with CM is high, careful follow-up should be recommended for each patient. When Simpson grade  $\geq$ II resection cannot be achieved, adjuvant radiotherapy is recommended after surgery (11,29,30). In the present case, the immediate resolution of fever post-surgery may be associated with the removal of the tumor, which may be the main source of pyrogens.

Of note, the present study has certain limitations. The expression of IL-6 or that of other types of pyrogens was not determined. Furthermore, although not necessary for the diagnosis, molecular analysis of the tumor, such as detection of 2p deletion and DNA methylation profiling in meningioma cells, could've been useful for verifying the presence of known molecular features of this tumor or for identifying possible novel molecular alterations. However, the appropriate equipment for detecting this was not available in the hospital of the present case.

Intraventricular CM presenting with inflammatory syndrome is exceptionally rare. To the best of our knowledge, the present case report was the second to report a case of lateral ventricle CM presenting with inflammation in an adult,

but it was the first case in a male adult patient. The tumor was removed by Simpson grade I resection, leading to the immediate resolution of fever and inflammatory syndrome.

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## Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

## Authors' contributions

SZ, GW and XC performed the operation. YH acquired data and drafted the manuscript. YZ interpreted clinical findings and revised the manuscript. HY completed the pathological diagnosis. SZ and YH checked and confirm the authenticity of all the raw data. All authors have read and approved the final version of the manuscript.

## Ethics approval and consent to participate

Not applicable.

## Patient consent for publication

The patient consented to the use of their clinical information/data and images for the purpose of research and to their publication.

## Competing interests

The authors declare that they have no competing interests.

## References

1. Kepes JJ, Chen WY, Connors MH and Vogel FS: 'Chordoid' meningeal tumors in young individuals with peritumoral lymphoplasmacellular infiltrates causing systemic manifestations of the Castleman syndrome. A report of seven cases. *Cancer* 62: 391-406, 1988.
2. Couce ME, Aker FV and Scheithauer BW: Chordoid meningioma: A clinicopathologic study of 42 cases. *Am J Surg Pathol* 24: 899-905, 2000.
3. Tena-Suck ML, Collado-Ortiz MA, Salinas-Lara C, García-López R, Gelista N and Rembao-Bojorquez D: Chordoid meningioma: A report of ten cases. *J Neurooncol* 99: 41-48, 2010.
4. Lin JW, Ho JT, Lin YJ and Wu YT: Chordoid meningioma: A clinicopathologic study of 11 cases at a single institution. *J Neurooncol* 100: 465-473, 2010.
5. Lui PC, Chau TK, Wong SS, Lau PP, Tse GM, Thomas TM and Ng HK: Cytology of chordoid meningioma: A series of five cases with emphasis on differential diagnoses. *J Clin Pathol* 60: 1024-1028, 2007.
6. Epari S, Sharma MC, Sarkar C, Garg A, Gupta A and Mehta VS: Chordoid meningioma, an uncommon variant of meningioma: A clinicopathologic study of 12 cases. *J Neurooncol* 78: 263-269, 2006.
7. Choy W, Ampie L, Lamano JB, Kesavabhotla K, Mao Q, Parsa AT and Bloch O: Predictors of recurrence in the management of chordoid meningioma. *J Neurooncol* 126: 107-116, 2016.

8. Di Ieva A, Laiq S, Nejad R, Schmitz EM, Fathalla H, Karamchandani J, Munoz DG and Cusimano MD: Chordoid meningiomas: Incidence and clinicopathological features of a case series over 18 years. *Neuropathology* 35: 137-147, 2015.
9. Mawrin C and Perry A: Pathological classification and molecular genetics of meningiomas. *J Neurooncol* 99: 379-391, 2010.
10. Jie D, Liu Z, He W, Wang S, Teng H and Xu J: Clinical features, radiological findings, and prognostic factors for primary intracranial chordoid meningioma. *Front Neurol* 13: 1002088, 2022.
11. Yang Y, Li D, Cao XY, Hao SY, Wang L, Wu Z and Zhang JT: Clinical features, treatment, and prognostic factors of chordoid meningioma: Radiological and pathological features in 60 cases of chordoid meningioma. *World Neurosurg* 93: 198-207, 2016.
12. Cushing H and Eisenhardt L: *Meningiomas, their classification, regional behaviour, life history, and surgical end results*. Springfield, IL, Charles C Thomas, 1938.
13. Rohringer M, Sutherland GR, Louw DF and Sima AA: Incidence and clinicopathological features of meningioma. *J Neurosurg* 71: 665-672, 1989.
14. Song KS, Park SH, Cho BK, Wang KC, Phi JH and Kim SK: Third ventricular chordoid meningioma in a child. *J Neurosurg Pediatr* 2: 269-272, 2008.
15. Arima T, Natsume A, Hatano H, Nakahara N, Fujita M, Ishii D, Wakabayashi T, Doyu M, Nagasaka T and Yoshida J: Intraventricular chordoid meningioma presenting with Castleman disease due to overproduction of interleukin-6. Case report. *J Neurosurg* 102: 733-737, 2005.
16. Wilson JL, Ellis TL and Mott RT: Chordoid meningioma of the third ventricle: A case report and review of the literature. *Clin Neuropathol* 30: 70-74, 2011.
17. Nambiar A, Pillai A, Parmar C and Panikar D: Intraventricular chordoid meningioma in a child: Fever of unknown origin, clinical course, and response to treatment. *J Neurosurg Pediatr* 10: 478-481, 2012.
18. Sadashiva N, Poyuran R, Mahadevan A, Bhat DI, Somanna S and Devi BI: Chordoid meningioma: A clinico-pathological study of an uncommon variant of meningioma. *J Neurooncol* 137: 575-582, 2018.
19. Wind JJ, Jones RV and Roberti F: Fourth ventricular chordoid meningioma. *J Clin Neurosci* 17: 1301-1303, 2010.
20. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, Ohgaki H, Wiestler OD, Kleihues P and Ellison DW: The 2016 world health organization classification of tumors of the central nervous system: A summary. *Acta Neuropathol* 131: 803-820, 2016.
21. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, Scheithauer BW and Kleihues P: The 2007 WHO classification of tumours of the central nervous system. *Acta Neuropathol* 114: 97-109, 2007.
22. Wang XQ, Mei GH, Zhao L, Li ST, Gong Y, Zhong J, Chen H and Jiang CC: Clinical features and treatment of intracranial chordoid meningioma: A report of 30 cases. *Histopathology* 62: 1002-1017, 2013.
23. Hasegawa S, Yoshioka S, Urabe S and Kuratsu J: Rapidly enlarging chordoid meningioma with abundant mucin production. *Neuropathology* 26: 438-441, 2006.
24. Shih AR, Cote GM, Chebib I, Choy E, DeLaney T, Deshpande V, Hornicek FJ, Miao R, Schwab JH, Nielsen GP and Chen YL: Clinicopathologic characteristics of poorly differentiated chordoma. *Mod Pathol* 31: 1237-1245, 2018.
25. Dohrmann GJ and Collias JC: Choroid plexus carcinoma. *J Neurosurg* 43: 225-232, 1975.
26. Ellis TS, Schwartz A, Starr JK and Riedel CJ: Epithelioid heman-gioendothelioma of the lumbar vertebral column: case report and review of literature. *Neurosurgery* 38: 402-407, 1996.
27. Denaro L, Di Rocco F, Gessi M, Lauriola L, Lauretti L, Pallini R, Fernandez E and Maira G: Pyrogenic cytokine interleukin-6 expression by a chordoid meningioma in an adult with a systemic inflammatory syndrome. Case report and review of the literature. *J Neurosurg* 103: 555-558, 2005.
28. Cassereau J, Lavigne C, Michalak-Provost S, Ghali A, Dubas F and Fournier HD: An intraventricular clear cell meningioma revealed by an inflammatory syndrome in a male adult: A case report. *Clin Neurol Neurosurg* 110: 743-746, 2008.
29. Prokopienko M, Wierzb-Bobrowicz T, Grajkowska W, Stępień T and Sobstyl M: Chordoid meningioma. Case report and review of the literature. *Niger J Clin Pract* 25: 1-4, 2022.
30. Tahta A, Genc B, Cakir A and Sekerci Z: Chordoid meningioma: Report of 5 cases and review of the literature. *Br J Neurosurg* 37: 41-44, 2023.



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