Intraventricular chordoid meningioma in a child: fever of unknown origin, clinical course, and response to treatment

Case report

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The authors present the case of an 11-year-old boy with an intraventricular chordoid meningioma, which is a rare presentation of prolonged fever of unknown origin due to a rare tumor in a rare location. The fever resolved after excision of the lesion. Subsequent imaging revealed recurrence at 1 year. After a repeat excision and fractionated radiotherapy, the patient has remained disease free 5 years after the first surgery. Very few cases of intraventricular chordoid meningioma have been reported to date. The pathological features and clinical course are described. A review of the literature describing management options for this tumor type, recently found to have a higher recurrence rate, is described herein. (http://thejns.org/doi/abs/10.3171/2012.9.PEDS11398)

Key Words • chordoid meningioma • radiotherapy • oncology • Castleman disease • fever of unknown origin

Meningiomas account for approximately 15%–20% of all primary intracranial tumors. Chordoid meningioma is a rare variant of meningioma, comprising less than 0.5% of all meningiomas, and is characterized by chordoma-like histopathological features with lymphoplasmacytic infiltration. The occurrence of this tumor within the ventricle is extremely rare, with only a few cases reported to date.14 Chordoid meningiomas are often associated with a systemic inflammatory syndrome, which dramatically improves after tumor removal.1 Few case reports exist of chordoid meningiomas presenting with fever due to a systemic inflammatory syndrome in adults.3,7 Castleman disease, first described by Benjamin Castleman in 1956, is an atypical lymphoproliferative disorder that includes generalized adenopathy, fever, hepatosplenomegaly, dysgammaglobulinemia, and elevated sedimentation rate.

Case Report

History and Examination. This 11-year-old boy presented with FUO lasting for 1 year, which was treated with different courses of antibiotics. The patient complained of poor school performance, and 3 weeks prior to admission he developed headache and vomiting. No other neurological symptoms were reported. Findings from the physical examination were unremarkable. Laboratory and radiological investigations for FUO were significant for mild anemia, high erythrocyte sedimentation rate (72 mm/hr), and elevated C-reactive protein level (147 mg/dl). During hospitalization, the patient developed meningismus, and a lumbar puncture revealed a CSF glucose level of 38 mg/dl, a protein level of 299 mg/dl, and a CSF cell count of 67 cells/mm3 with a differential count of 90% mononuclear cells. Results of all bacterial cultures of CSF, blood, and urine were negative, and the patient was empirically started on antitubercular treatment by a pediatrician suspecting possible tubercular meningitis. Subsequently, the patient experienced multiple brief episodes of sudden loss of awareness with irrelevant speech and occasional speech arrest, suggestive of seizures. A brain MR image showed a uniformly enhancing intraventricular lesion in the left trigonal region with significant edema, mass effect, and midline shift (Fig. 1).

Initial Operation. The lesion was excised via a transsulcal approach to the trigone of the left lateral ventricle. During the postoperative course, the patient showed gradual improvement and remained afebrile with a decline in erythrocyte sedimentation rate and C-reactive protein levels. On the 7th postoperative day, he developed right-sided focal motor seizures with secondary generalization and a transient postictal right hemiparesis, which resolved within...
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24 hours. A CT scan showed CSF trapped in the temporal horn, with significant mass effect and midline shift.

Postoperative Course. On follow-up after discharge from the hospital, the boy recovered well without any residual speech or motor deficits. He finally completed his schooling and was able to join high school.

Histopathological Examination. Histopathological examination showed chordoid morphology with focal areas of meningothelial pattern (Fig. 2a). The cells were arranged in chords and lobules, separated by fibrovascular septae in a mucoid stroma (Fig. 2b). Dense lymphoplasmacytic infiltrates with perivascular cuffing were present (Fig. 2c). The myxoid matrix stained bright blue with Alcian blue stain done at pH 2.5 (Fig. 2d). There were no areas of brain invasion or necrosis. On immunohistochemical staining, the majority of tumor cells were strongly positive for EMA and vimentin (Fig. 2e and f) and negative for GFAP and CD34. The Ki 67 proliferation index was less than 3% in most cellular areas. A histological diagnosis of chordoid meningioma (WHO Grade II) was made.

Additional Treatment. Adjuvant therapy was initially withheld given the gross-total resection and the patient’s young age. After 5 months, the patient developed severe symptoms of increased intracranial pressure due to a trapped CSF collection in the temporal horn. Over the next 2 months, the patient required 2 CSF shunting procedures, after which he remained neurologically stable. At 6 months, his condition again deteriorated due to migration of the shunt catheter, and a contrast-enhanced CT scan revealed an enhancing subependymal nodule within the temporoparietal horn of the left lateral ventricle (Fig. 3).

A repeat surgery was performed. The tumor nodule was identified under MRI guidance and was excised totally. Histopathological examination of the tumor nodule was consistent with chordoid meningioma recurrence (Fig. 3d).

Because of the early recurrence after gross-total resection and the WHO Grade II histology, we decided to treat the lesion with focal 3D radiotherapy, delivering 45 Gy in 25 fractions with a 9-Gy boost in 5 fractions using 15 MV and 6 MV x-rays. Of note, the boy suffered further episodes of recurrent trapped CSF collections in the temporal horn, requiring a total of 2 shunt revisions.

Posttreatment Course. Eventually, the ventriculoperitoneal shunt was removed, and the patient has remained shunt free for the past 3 years. He has no focal sensorimotor or speech deficits and subsequently completed high school and has joined a vocational music school. He has remained disease free as documented on MRI at 56 months after the second excision and radiotherapy (Fig. 4).

Discussion

The term “chordoid meningioma” was coined by Kepes et al. in 1988 to describe a meningeal tumor accounting for only 0.5% of all meningiomas. These lesions are found more commonly in young patients and only rarely in children. The neoplasm is a variant of menin-
A chordoid meningioma that predominantly shows areas resembling a chordoma intermixed with more typical meningioma areas. To our knowledge, at least 81 cases of chordoid meningiomas have been reported in various locations in the nervous system. The largest series of 42 patients, published by Couce et al., described only 2 cases in children. The vast majority of chordoid meningiomas are supratentorial, although occasional posterior fossa and rare spinal locations have been reported. Systemic effects such as fever or hematological abnormalities, such as Castleman syndrome, may be absent or present. An intraventricular location for the tumor is very rare, with only 2 cases reported to date in adults. Fever was the presenting symptom in both instances. None of the 7 pediatric cases described by Kepes et al. had fever. More recently, other brain tumors, including clear cell meningioma, have been reported in association with Castleman syndrome in the adolescent population, indicating that this presentation is not exclusive to chordoid meningiomas. Sato et al. summarized the occurrence of Castleman syndrome in relation to brain tumors in 14 individuals (chordoid meningiomas in 11 and a clear cell meningioma in 1).

The morphological differential diagnosis of chordoid meningioma includes chordoma, myxoid chondrosarcoma, metastatic mucinous carcinoma, and chordoid glioma. Chordomas show a strong positivity for cytokeratin, EMA, and often S100 protein staining. Chordoid gliomas are GFAP positive and EMA negative. Chordoid meningiomas, in contrast, are positive for EMA and vimentin, but only a minority stain positive for cytokeratin and S100 proteins. Ki 67–positive staining for these tumors has been reported to range from 0.4% to 11.4% (mean 5.2%).

To date, chordoid meningiomas have been reported to occur in both the pediatric and adult populations. The systemic inflammatory syndrome has been reported to occur in children and young adults up to 30 years of age. The absence of systemic inflammatory syndrome in adults with chordoid meningioma along with ultrastructural features prompted some authors to suggest that these tumors may be only a morphological variant of meningioma in adults, whereas in children this may represent a true meningioma subtype.

The cause of the systemic inflammatory syndrome in patients with a meningioma also remains unclear. Different pathophysiological mechanisms proposed include the characteristic lymphocytic infiltration and interleukin-6. The initial clinical finding of meningeal irritation in our patient and the mononuclear CSF pleocytosis may support this theory. Excision of the mass would have eliminated the source of pyrogens, leading to immediate resolution of fever. It is possible that the persistent and problematic trapping of the temporal horn of the lateral ventricle may have also resulted from prolonged intense inflammatory changes along the ventricle wall. In retrospect, an endoscopic solution for the loculated CSF trapping could have been attempted, possibly averting multiple shunt revisions.
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The aggressive behavior and tendency for early recurrence is well documented. In particular, the proliferation index and extent of resection is generally accepted as one of the most important prognostic factors for WHO Grade II meningiomas. On follow-up in 36 of the 42 cases of chordoid meningioma reported by Couce et al., 14 patients (39%) experienced 1 or more recurrences between 1.8 and 16 years postoperatively (mean 5.6 years). All but one recurrent tumor had initially been subtotally resected. A possible explanation for the high rate of recurrence of chordoid meningiomas could be related to the mucoid quality of the stroma, which, like that of chordoma, both mechanically facilitates tumor spread and makes it difficult to achieve complete resection, thus leading to subsequent recurrence. Extensive spread along the neuraxis has been described. In this particular case, the location deep to eloquent cortex was additionally problematic. Periodic imaging after surgery is therefore indicated, regardless of the extent of resection.

Although the role of adjuvant radiation therapy in the postoperative management of atypical meningiomas is still unclear due to the smaller number of atypical meningiomas and the lack of prospective controlled studies, literature exists that is in favor of the use of adjuvant radiotherapy in patients who have undergone subtotal resection. With respect to chordoid meningiomas, several case reports in favor of adjuvant radiotherapy, at least upon recurrence, in the management of chordoid meningioma exist. In this particular case, radiotherapy was initially avoided because of the child’s age. However, due to histologically proven symptomatic recurrence in a relatively eloquent location, it was decided to irradiate the tumor bed to help avoid local recurrence. The disease-free state at 56 months after surgery and radiation therapy for this recurrent lesion may suggest a role for postoperative adjuvant therapy in preventing local recurrence. Song et al. reported on a similar intraventricular chordoid meningioma that was managed with complete excision alone and was recurrence free on the 1-year MRI study. Although the role of radiotherapy remains controversial, we wish to report this case, demonstrating long-term recurrence-free survival (56 months) after excision of a recurrent tumor nodule and local irradiation as a management option in children with recurrent WHO Grade II meningiomas.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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