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Spinal chordoid meningioma in a child: A case report and review of the literature

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Abstract. As an uncommon subtype of meningioma, chordoid meningioma (CM) of the spinal canal is extremely rare. There have been only two reported cases of intraspinal CM in the literature, and this lesion has not been previously reported in the pediatric age group. To the best of our knowledge, the present study reports the first case of spinal chordoid meningioma in a pediatric patient. A 12-year-old female presented with a 3-month history of progressive numbness and weakness in the right-side limbs, and intermittent pain in the neck and right shoulder. Spinal magnetic resonance imaging (MRI) revealed an intraspinal lesion at the C2-3 level with irregularly heterogeneous enhancement. The patient underwent a C2-3 laminotomy. Due to adhesion to the dura and proximity to the vertebral artery, the tumor was partially removed intraoperatively. The post-operative course was uneventful and the symptoms were apparently relieved. The patient experienced recurrence 5 years subsequent to surgery. MRI revealed an extradural regrown tumor at the C2-5 level. Partial removal combined with radiotherapy was performed. However, the patient experienced progression of tetraplegia and succumbed to severe pneumonia and respiratory failure 5 months subsequent to the second surgery. In the present study, the clinicoradiological findings and treatment outcome of this rare entity are discussed, in addition to a review of the relevant literature. Spinal CMs should be included in the differential diagnosis of intraspinal tumors of the pediatric spine. Multidisciplinary treatment, consisting of total surgical removal and adjuvant radiotherapy, should be considered due to the aggressive nature of this abnormality and the risk of long-term recurrence.

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Introduction

Meningiomas are common tumors in the central nervous system and have a large variety of histopathological appearances. The majority of the variants are slow growing and follow a benign clinical course without significant differences in prognosis. However, certain subtypes of meningioma exhibit aggressive clinical behavior, including atypical, chordoid, clear cell, papillary, rhabdoid and anaplastic subtypes (1).

Chordoid meningiomas (CMs), which were recorded as a novel subtype of meningioma in the 1993 World Health Organization (WHO) classification of tumors (2), are rare histological variants of meningioma that have been reported to predominantly occur intracranially (3). CMs account for 0.5-1.0% of all meningiomas and are more common in younger patients (<18 years). These lesions demonstrate unique clinical associations and prognostic implications (4). Histopathologically, CM tumors are composed of epithelioid or spindle cells arranged in cords or nests in a basophilic mucoid matrix (2). Gross total resection is the treatment of choice for CMs, however, in the event of residual or recurrent CMs, postoperative radiotherapy is considered the standard treatment (2,3). Although CM is considered a benign tumor (WHO Grade II), this rare entity demonstrates a more aggressive nature with a higher recurrence rate than that of typical meningiomas (WHO Grade I). However, the overall prognosis of CM is relatively good with a three-year survival rate of 93.4% (4,5).

Spinal meningiomas comprise almost 46% of all primary spinal cord tumors, and they are extremely uncommon in children (5). CM of the spinal canal is also extremely rare, and to the best of our knowledge, there have been only two cases of intraspinal CM reported in the literature (6,7). The first case, reported by Couce *et al* (6), was a 28-year-old woman with a CM at the C2 level who underwent gross total resection of the tumor and exhibited good recovery 5 years after surgery. The second case, presented by Ibrahim *et al* (7), was that of a 26-year-old man who harbored a C2-3 intradural and extramedullary chordoid meningioma. The patient underwent gross total resection and exhibited no neurological deficits 2 months after surgery.

The present study reports a case of spinal CM of the cervical region in a 12-year-old girl. To the best of our knowledge, the present study is the first case of pediatric spinal CM

to be reported in the literature. Written informed consent was obtained from the patient's family.

Case report

A 12-year-old girl presented to the outpatient clinic of Beijing Tiantan Hospital (Beijing, China) with a 3-month history of progressive numbness and weakness in the right-side limbs, and intermittent pain in the neck and right shoulder. The patient did not experience any bowel or bladder symptoms. Neurological examination revealed that muscle tone was increased in the right lower limb, and muscle power was grade 4/5 in the right upper and lower limbs, which was classified using the Medical Research Council grading system (8). Deep and superficial sensation in the right leg and superficial sensation in the right arm were reduced. The right lower limb also exhibited increased deep tendon reflexes and a positive Babinski sign. Laboratory data was within the normal limits and the medical history was unremarkable.

Pre-operative magnetic resonance imaging (MRI) of the cervical spine demonstrated an intraspinal extramedullary lesion at the C2-3 level on the right side of the spinal canal and neural foramina, measuring 11x21x26 mm at the maximal dimensions (Fig. 1). The tumor demonstrated isointensity on T1-weighted images and mixed hyperintensity on T2-weighted images. Contrast-enhanced MRI of the lesion revealed irregularly heterogeneous enhancement subsequent to gadolinium administration. The tumor severely compressed the spinal cord and the cord was displaced to the right portion of the spinal canal. The lesion extended through the enlarged C2-C3 neural foramen on the right side as an exophytic paravertebral component, compressing and partially encompassing the right vertebral artery. Abnormal flow void signal, peritumoral edema and associated syringomyelia were not observed. The brain MRI findings were normal. Other imaging work-ups, including bone scanning and ultrasonography of the abdomen, provided no novel information. According to the location and MRI features of the tumor, the lesion was pre-operatively diagnosed as schwannoma.

The patient underwent a C2-C3 laminotomy, which was performed using the posterior midline approach. The dura was extremely tense and a longitudinal incision was made in the center of the dura. The intraoperative findings revealed that the tumor possessed a complete capsule and was yellow in color, firm in texture and moderately vascular. The tumor was firmly attached to the ventrolateral surface of the dura. The spinal cord was displaced to the left side of the thecal sac, and the spinal cord was severely compressed. The expansile mass extended through the right widening C2-C3 neural foramen and encompassed the adjacent nerve rootlet. Due to the adhesion of the lesion to the dura, and the proximity of the lesion to the right vertebral artery, a gross total resection was challenging to achieve. The intradural contents of the tumor were removed and, due to tumor infiltration, the affected nerve rootlet was transected under spinal evoked-potential monitoring, based on the protection of spinal functions. The effects of cord compression discontinued, while the exophytic paravertebral component remained residual (Fig. 2).

On histopathological examination, the tumor was found to be composed of strands and cords of polygonal cells with eosinophilic cytoplasm, several of which demonstrated vacuolation. The majority of the tumor cells contained mucin-rich chordoid elements embedded in the abundant myxoid matrix. The immunohistochemical examinations revealed that the tumor cells were positive for the expression of vimentin and epithelial membrane antigen (EMA), but did not express desmin, S-100 or myogenin. In total, ~20% of cells were positive for Ki-67. All these findings were consistent with the diagnosis of CM (WHO Grade II) (Fig. 3).

The post-operative course was uneventful and the sensory deficits were apparently relieved. Due to the aggressive nature of lesions with high Ki-67 expression and the risk of tumor recurrence, adjuvant radiotherapy was strongly recommended. However, additional treatment for the residual spinal tumors was refused and the patient was discharged from Beijing Tiantan Hospital 2 weeks subsequent to the procedure. Gradual improvement in the strength of the limbs was noted during the follow-up. Regular follow-up MRI was performed, and the residual tumor demonstrated no regrowth or metastasis 3 years subsequent to the surgery. However, the patient experienced recurrence 5 years subsequent to the procedure.

The patient presented with severe pain in the neck and right shoulder that had recurred 2 months prior to admission to the Department of Neurosurgery of Beijing Tiantan Hospital. An immediate MRI revealed an extradural well-defined tumor that demonstrated marked enhancement at the C2-5 level and involved the right paraspinal region and adjacent neural foramen, measuring 70x30x17 mm at the maximal dimensions (Fig. 4). A C2-5 laminectomy was performed using a posterior approach. The tumor was resected in sections and partial removal was achieved using microsurgical techniques. Histopathological examination confirmed the recurrence of CM. Following the surgery, the patient received one course of radiation therapy. External beam radiotherapy was administered to the surgical site at a total dose of 40 Gy over 16 fractions. However, the patient did not respond well to the treatment. The pain worsened 1 month subsequent to surgery and the patient experienced a progression of tetraplegia, with rapid enlargement of the residual tumor. The patient succumbed to severe pneumonia and respiratory failure 5 months subsequent to the second surgery.

Discussion

CM is a rare variant of meningioma that is characterized by epithelioid cord-like tumor cells in the myxoid stroma (4,9). CM has been recognized as an aggressive tumor of meningothelial origin and is considered to be a grade II lesion, in addition to clear cell and atypical meningioma, due to a high rate of recurrence, particularly following subtotal resection. Currently, >120 cases of CM have been reported in the literature, and these cases account for 0.5-1.0% of all meningiomas (5,10). The neuraxis may be involved at any level with a significant predilection for the supratentorial location and this rare entity occurs more commonly in young adult patients in comparison with other subtypes of meningiomas. Compared with typical meningiomas, the diagnosis and treatment of CMs is critical for providing accurate

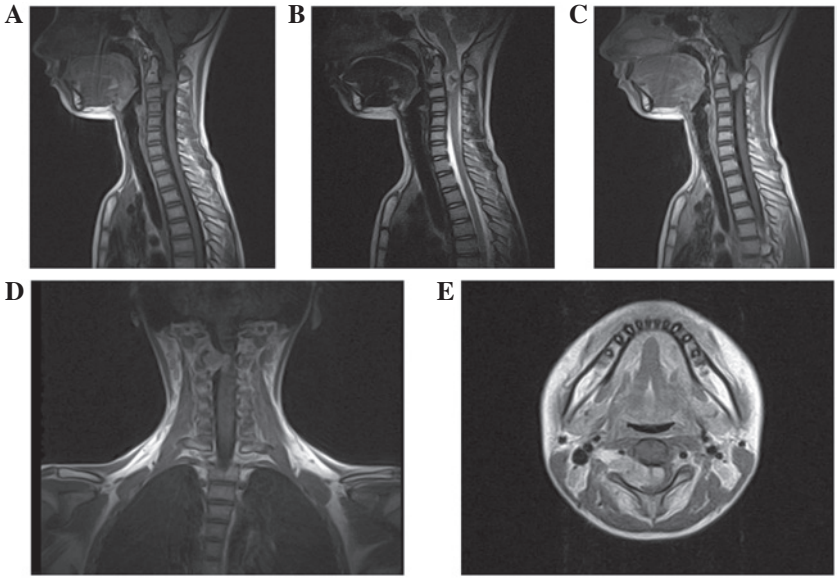

Figure 1. Pre-operative MRI revealing an intraspinal extramedullary lesion at the C2-3 level with (A) isointensity on the sagittal T1-weighted image and (B) mixed hyperintensity on the sagittal T2-weighted image. Gadolinium-enhanced (C) sagittal, (D) coronal and (E) axial T1-weighted MRI revealing that the mass extended through the enlarged C2-C3 neural foramen, compressing and partially encompassing the right vertebral artery, with irregularly heterogeneous enhancement. MRI, magnetic resonance imaging.

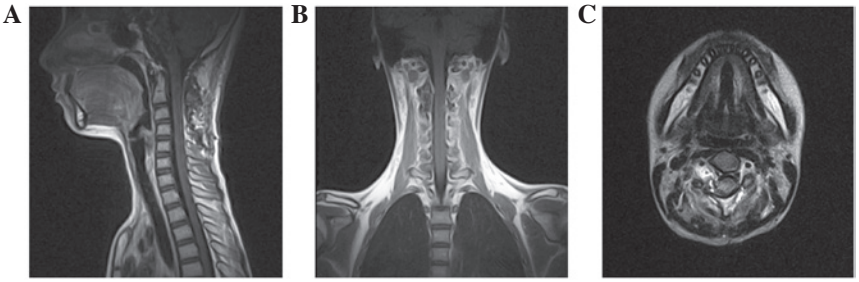

Figure 2. Gadolinium-enhanced (A) sagittal, (B) coronal and (C) axial T1-weighted magnetic resonance images revealing the achievement of cord decompression and an exophytic paravertebral residual component.

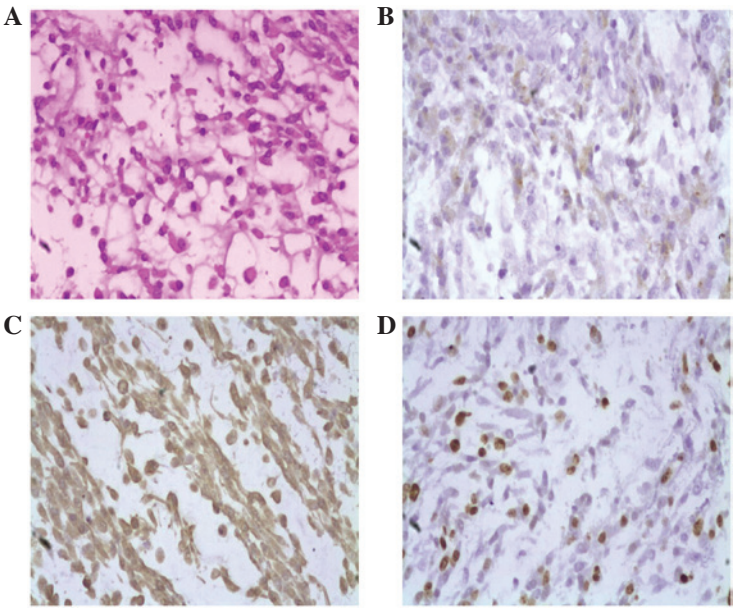

Figure 3. (A) Photomicrograph revealing the strands and cords of polygonal cells with eosinophilic cytoplasm and mucin-rich chordoid elements embedded in the abundant myxoid matrix (hematoxylin and eosin; magnification, x200). The immunohistochemical examinations reveal that the tumor cells are strongly positive for (B) epithelial membrane antigen (magnification, x200) and (C) vimentin (magnification, x200), and (D) 20% of cells are positive for Ki-67 (magnification, x200).

Table I. Summary of reported patients with spinal chordoid meningiomas.

Author, year (Ref.)	Age, years	Gender	Duration of initial symptoms	Tumor location	Symptoms	Treatment	Outcome
Couce *et al*, 2000 (6)	28	F	NA	C2	NA	GTR	Good recovery at 5 years
Ibrahim *et al*, 2005 (7)	26	M	NA	C2-3, IDEM	Cramping and weakness in lower limbs	GTR	Good recovery at 2 months
Present case	12	F	3 months	C2-3, IDEM + ED	Numbness and weakness of right-side limbs and intermittent pain in the neck and right shoulder	1st, PR; 2nd, PR + RTX	Recurrence at 5 years; succumbed 5 months following the second surgery

M, male; F, female; ED, extradural; IDEM, intradural and extramedullary; GTR, gross total removal; NA, not available; PR, partial removal; RTX, radiotherapy.

Figure 4. Gadolinium-enhanced (A) sagittal, (B) coronal, and (C) axial T1-weighted magnetic resonance images demonstrating an extradural well-defined regrown tumor with marked enhancement, compressing the cord ventrally at the C2-5 level and involving the right paraspinal region and adjacent neural foramen.

information for the treatment and prognosis of patients and establishing a reasonable schedule for outpatient follow-up.

The occurrence of spinal CMs is exceedingly rare. Since Couce *et al* described the first patient with spinal CM in 2000, only two cases of spinal CM have been reported in the English literature (6,7). Table I summarizes the clinical features of the previously reported patients and present patient with spinal CM. The age of all three patients at the time of presentation was <30 years, which is similar to the age of patients with intracranial CM, but different from the age of patients with intraspinal typical meningiomas, which mostly occur in patients 40-60 years old. All tumors arise from the cervical spinal regions, while intraspinal meningiomas are most frequently identified in the thoracic region of the spine. The symptoms of CM, including somatic pain, numbness and weakness in the limbs, are consistent with those of other common intraspinal tumors, such as schwannoma and neurofibroma. In the present patient, the duration between the onset of symptoms and presentation was 3 months. This is shorter than the typical duration for intraspinal meningiomas of 6 months, which is likely to reflect the aggressive nature and relatively rapid growth pattern of the tumor.

When first described, CM was hypothesized to affect young patients that presented with one or more of the features of a systemic inflammatory disorder associated with Castleman syndrome, such as iron refractory microcytic anemia, hepatosplenomegaly, retardation of somatic and sexual development, and bone marrow dysfunction (4,11,12). However, in certain studies (5,13), the clinical manifestation of Castleman syndrome was not reported to be a prerequisite for the diagnosis of CM, and the three reported patients with spinal CM demonstrated no evidence of Castleman or inflammatory syndrome.

MRI is the modality of choice for the diagnosis of spinal tumors. Pre-operative differential diagnosis for intraspinal tumors is important when planning surgical strategies and determining the extent of the required resection to avoid overtreatment and unacceptable complications. However, the imaging features of CMs are indistinguishable from those of a typical meningioma in the pre-operative period due to the lack of a highly specific appearance. Thus, a definitive pre-operative diagnosis of CM based only on MRI was challenging in the present study, and a complete histopathological examination was required to differentiate the CM from other intraspinal lesions.

Detailed pathological features of CMs have been described in certain clinicopathological studies (4,5,7). Histologically, CM is characterized by cords and clusters of spindle and epithelioid cells embedded in a myxoid matrix, with vacuolated eosinophilic cytoplasm. The tumor cells are usually diffusely positive for the expression of EMA and vimentin. In certain cases of intracranial CM, the rapid enlargement of the tumor over a relatively short time period may be due to high mucin-producing activity (14). The histological differential diagnosis of CM should include the following: chordoma; chondrosarcoma; chordoid glioma; myxopapillary ependymoma; and clear cell meningioma.

Due to the rarity of CM, the most effective method of treatment remains ill-defined. When compared with typical meningiomas (WHO grade I), which typically exhibit a good clinical outcome and low risk of long-term recurrence following subtotal or total resection, the presence of a WHO grade II meningioma should indicate the requirement for a different management path, possibly including adjuvant radiotherapy, closer follow-up and a lower threshold for surgery in the event of recurrence. Since CM is histologically benign and usually well circumscribed, gross total removal using a microsurgical technique based on the protection of the spinal function should be attempted during surgical exploration. However, due to the adhesion of the tumor to the dura and proximity to the vertebral artery, the tumor in the present study was partially removed for mass effect relief to improve the myelopathic symptoms and avoid severe operative complications. This incomplete removal may have led to an increased risk of long-term recurrence and caused a poorer outcome compared with the outcome of the two previously reported cases, which were treated with total removal.

It is hypothesized that adjuvant treatments, such as radiotherapy or chemotherapy, may prevent tumor recurrence. In the event of subtotally resected intracranial CMs, post-operative radiotherapy is the standard treatment (10,14). Due to the aggressive growth pattern of spinal CMs and long-term fatal outcome in the present patient, post-operative early adjuvant radiotherapy and close follow-up investigation in adult and pediatric patients is recommended, although certain studies debate the benefit of post-operative adjuvant treatments in pediatric patients (2,7,15). The efficacy of adjuvant radiation therapy for controlling spinal CMs remains uncertain. In addition, other systemic therapies may be considered for unresectable or recurrent tumors, but additional investigation is required. Since the overall prognosis is uncertain and long-term recurrence or regrowth is likely, regular follow-up MRI is required.

The present study suggests that spinal CM should be added to the differential diagnosis of intraspinal tumors in the pediatric spine. Considering the aggressive nature of this rare entity and the risk of long-term recurrence, total surgical removal combined with adjuvant radiotherapy is recommended, although the overall prognosis remains uncertain. Additional investigation with long-term follow up and studies using larger sample sizes are required.

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