



Case Report

Giant chondrosarcoma of the falx in an adolescent: A case report

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ABSTRACT

Background: Intracranial chondrosarcomas are slowly growing malignant cartilaginous tumors that are especially rare in adolescents.

Case Description: A 19-year-old woman with no medical history presented with symptoms of intermittent facial twitching and progressive generalized weakness for 6 months. The patient's physical examination was unremarkable. Imaging revealed a large bifrontal mass arising from the falx cerebri, with significant compression of both cerebral hemispheres and downward displacement of the corpus callosum. The patient underwent a bifrontal craniotomy for gross total resection of tumor. Neuropathologic examination revealed a bland cartilaginous lesion most consistent with low-grade chondrosarcoma. Her postoperative course was uneventful, and she was discharged to home on postoperative day 3.

Conclusion: This is an unusual case of an extra-axial, non-skull base, low-grade chondrosarcoma presenting as facial spasm in an adolescent patient.

Keywords: Adolescent, Chondrosarcoma, Falx cerebri, Intracranial, Pediatric

INTRODUCTION

Intracranial chondrosarcomas are rare, slowly growing, malignant cartilaginous tumors first described in 1899. They are thought to originate from embryonal cartilage or meningeal fibroblasts,^[1,2,6,9] account for an estimated 6% of all skull base tumors, and rarely originate at other sites.^[1,4] They are especially rare in children and adolescents. Reported clinical manifestations include headache, dizziness, cranial nerve palsies, limb weakness, seizures, and even coma.^[1,2,6] There is sparse literature on the diagnosis and management of intracranial chondrosarcomas that arise away from the skull base. To the best of our knowledge, this is the first report of an intracranial chondrosarcoma in an adolescent patient presenting as bilateral facial spasm.

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CASE DESCRIPTION

History and physical examination

A 19-year-old female with no medical history presented to the emergency department complaining of intermittent facial twitching and progressive generalized weakness for 6 months. She noted that each episode of facial twitching lasted several minutes and would self-resolve. She described her weakness as “heaviness” in all extremities such that occasionally she was unable to lift her arms above her shoulders. She endorsed bilateral frontal headache alleviated by ibuprofen, and “fullness” in both ears, more severe on the right. She denied visual symptoms, dizziness, tingling in the extremities, pain, or difficulty with ambulation. The initial physical examination was unremarkable; the patient was neurologically intact.

Imaging

The initial non-contrast computed tomogram of the head showed a large, bifrontal, coarsely calcified extra-axial mass that scalloped the inner table of the skull and displaced the cerebral hemispheres circumferentially [Figure 1a]. Cerebral angiography revealed no significant arterial feeding vessels or tumor blush. Magnetic resonance imaging (MRI) revealed a 7.6 × 5.5 × 6.2 cm bifrontal mass arising from the falx cerebri. The mass abutted on the right side of the superior sagittal sinus and right high convexity dura [Figure 1b-d], displaced both cerebral hemispheres inferolaterally, and displaced the corpus callosum and pericallosal vessels inferiorly. There was minimal edema. She was subsequently started on levetiracetam 2000 mg/day to prevent further preoperative events.

Surgical treatment

Surgery was recommended to alleviate the mass effect and obtain a tissue diagnosis. Given the size of the tumor and the possible pathologies considered, initial biopsy was not performed, and maximal safe resection was planned. The patient underwent bifrontal craniotomy for resection of tumor. The lesion was extremely firm and not amenable to ultrasonic aspiration or typical cautery and suction techniques. Monopolar loop cautery was utilized to progressively debulk the lesion piecemeal so that the capsule could be defined from the surrounding compressed brain. Calcifications were removed *en bloc*, as they could not be divided sharply or with cautery. There was no brain invasion, and the tumor was removed completely.

Pathology

Pathologic examination revealed a low-grade cartilaginous neoplasm. The lesion was composed of an encapsulated

bland chondroid tumor with a thin fibrous capsule with minimal nuclear atypia, no perceived increase in nuclei within the lacunar spaces, and no identifiable mitotic figures.

However, because cartilaginous tumors of the falx cerebri are very rare and the distinction between chondroma and low-grade chondrosarcoma is challenging, an outside pathologist was consulted for a second opinion. This pathologist noted that although the diagnosis was difficult in this case, due to the presence of moderate cellularity, chondrocyte crowding, mild nuclear atypia, frequent binucleation, patchy necrosis, and myxoid change, a

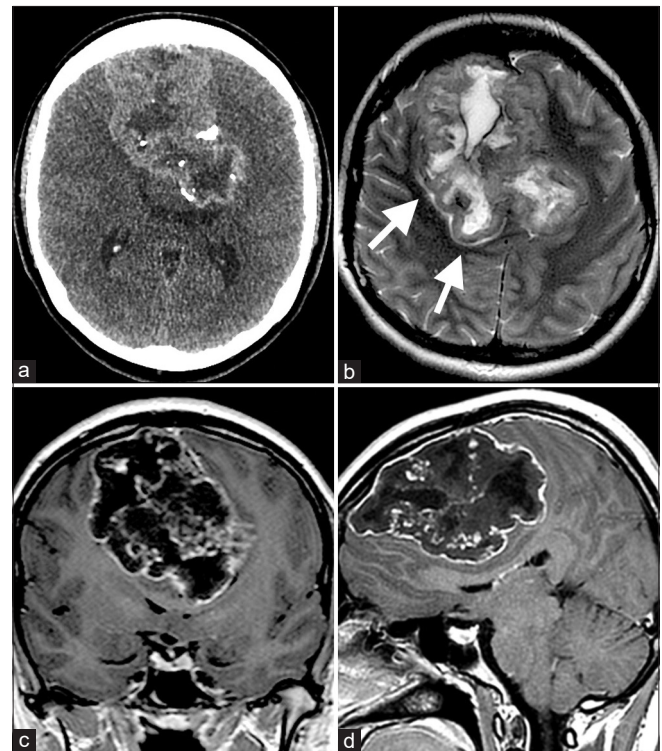


Figure 1: (a) Non-contrast axial CT section demonstrates a large, well-defined, partially calcified midline mass that displaces each cerebral hemisphere laterally and displaces lateral ventricles posteriorly. There is no evidence of cerebral edema. (b) Non-contrast axial T2-weighted MRI displays heterogeneous internal texture of the mass. Myelinated cerebral white matter shows normal signal intensity with no evidence of cerebral edema. Clear, cerebral spinal fluid-containing cleavage plane (arrows) between medial surface of the right hemisphere and central mass confirms that the mass is extra-axial. (c and d) Contrast-enhanced T1-weighted MRI in coronal (c) and sagittal (d) planes shows heterogeneous enhancement along the surface and within the interstices of the tumor. There is marked compression of corpus callosum and lateral ventricles and a central vector of mass that displaces the brain downward through tentorial incisura and displaces cerebellar tonsils downward through foramen magnum. The nearly linear contour of the left side of mass and of the medial border of the left cerebral hemisphere again suggests that the mass arises from midline falx.

diagnosis of low-grade chondrosarcoma was most likely [Figures 2-4].

Postsurgical course

The patient's postoperative course was uncomplicated. Postoperative MRI confirmed a gross total resection with no significant ischemia or edema. She was discharged home on postoperative day 3 with home physical therapy. On the first day at home, she experienced a 3-minute episode of facial spasm. Her dose of levetiracetam was increased from 2000 mg/day to 3000 mg/day. Since this change, she has done well with no complaints. At the patient's 2-month postoperative follow-up, her only complaints were mild headaches, difficulty sleeping, and some fatigue. At 4-month follow-up, she was completely asymptomatic, and she is currently in the process of attempting to wean off levetiracetam. On-going surveillance imaging for recurrence is planned.

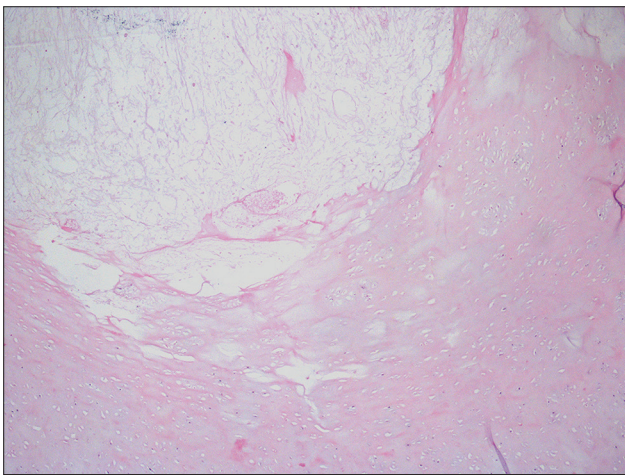


Figure 2: H&E at ×4 showing myxoid change and patchy necrosis.

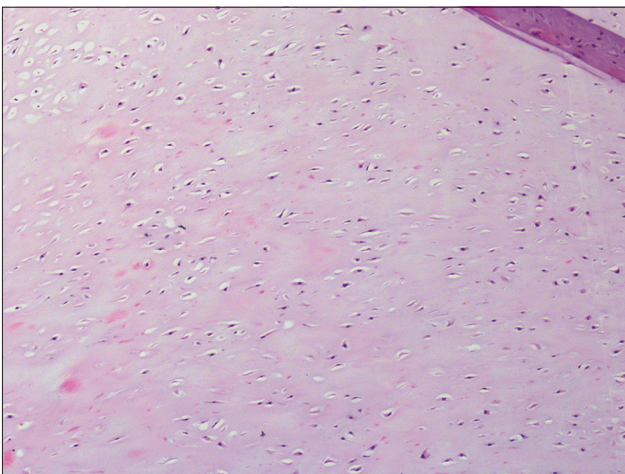


Figure 3: H&E at ×10 showing focal increased cellularity.

DISCUSSION

In older children and adolescents with a new meningeal neoplasm, the leading diagnostic consideration is meningioma. Differential diagnoses that are more common in pediatric than adult patients include teratoma, other congenital dysplastic tumors, hemangiopericytoma, and, rarely, massively exophytic oligodendroglioma.^[1]

Comprehensive literature search revealed eleven distinct cases of intracranial chondrosarcoma arising from the falx in patients under 25 years old; six of which cases were pediatric or adolescent patients (<18 years old).^[1-9] In the majority of pediatric patients, focal seizures with limb weakness were the presenting symptoms.^[1,4,6,9] Rarer presentations included optic disc swelling and abducens paresis.^[3,8] One case reported a hard scalp mass that was grossly visible on external examination and that gradually increased in size over the 3 months before presentation.^[6]

Intracranial chondrosarcoma is difficult to distinguish from other meningeal neoplasms on the basis of clinical presentation and preoperative imaging. In one case, contrast-enhanced MRI demonstrated a heterogeneous lesion with perifocal edema presumed to be a meningioma or hemangiopericytoma preoperatively.^[3] In another case, the lesion demonstrated patchy calcifications on MRI.^[6]

Chondrosarcomas show a distinct pathology that is subclassified as mesenchymal or myxoid. The majority of falcine and parasagittal chondrosarcomas are mesenchymal.^[1,6] The neuropathology of the low-grade chondrosarcoma in this case is consistent with other reports of low-grade chondrosarcomas in both adult and pediatric patients.^[3,8] These tumors are typically cartilaginous tissue showing low to moderate cellularity, individual cells surrounded by a basophilic matrix, rare mature hyaline cartilage and blood vessels. Mitotic events are typically rare, although most cells are binucleated.^[1,6]

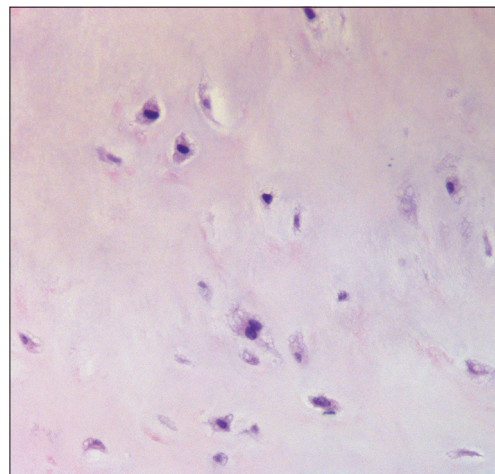


Figure 4: H&E at ×40 showing a binucleate cell and mild atypia.

Gross total surgical resection is the ideal treatment of intracranial chondrosarcomas. Postoperative radiotherapy is controversial, as some researchers express concern that radiation may increase the risk for malignant transformation.^[2,6,8] The prognosis of these tumors has been reported variably in the literature, with mesenchymal type pathology carrying a particularly poor prognosis.^[9] In one review of 20 adult and pediatric patients, nine were reported to have died after a median 4-month survival, and five had recurrence after surgery at a median of 21 months.^[6]

CONCLUSION

The present report describes a pathologically confirmed intracranial, extra-axial low-grade chondrosarcoma projecting from the falx cerebri in an adolescent patient presenting as facial spasm ultimately thought to be due to substantial tumor mass effect. To the best of our knowledge, this is the first reported case of chondrosarcoma in an adolescent with these specific presenting symptoms. Her prognosis is deemed overall favorable given that the pathology was low grade. Close surveillance with serial imaging over the next several years is warranted.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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