Fibrocartilaginous Mesenchymoma of the Spine

A Case Report

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Abstract

Case: A healthy, 19-year-old woman was incidentally found to have a large, destructive tumor of T11 without neurologic symptoms. Biopsy demonstrated fibrocartilaginous mesenchymoma (FCM). The patient was treated with resection including subtotal corpectomy and T8-L1 fusion with use of cage and allograft strut construct. The patient remained without recurrence over 3 years of follow-up.

Conclusion: FCM arising from the spine is a rare tumor, of which this is the sixth report. FCM affects primarily young adults and is benign but locally aggressive, requiring complete excision to prevent recurrence.

Fibrocartilagenous mesenchymoma (FCM) arising from the spine is an exceptionally rare tumor that can be locally destructive with potential for recurrence and compression of neural elements. FCM was identified by Dahlin et al.[1] in 1984 among a series of 15,000 primary bone tumors at the Mayo Clinic. Among that series, 5 FCMs were identified among patients aged 9 to 25 years. Since then, only 5 have been reported in the spine. This rare tumor tends to affect younger individuals and has been identified in a patient as young as 19 months old[2,3]. Pain is the most typical presenting symptom[2].

Histologically, FCM is characterized by benign-appearing cartilage "islands" surrounded by bland spindle cell stroma, with cartilage cells arranged in a sheet-like formation similar to epiphyseal plates. FCM most commonly affects the long bones, but may involve the axial skeleton, including the pelvis, ribs, and spine. Radiographically, FCM lesions may appear lytic on plain films and computed tomography (CT). The matrix may have "popcorn calcifications" or a "ring and arc" appearance consistent with its chondroid nature. There may be a sclerotic rim as it is typically slow-growing. On MRI, FCM typically demonstrates significant contrast uptake and tends to be hypointense on T1 sequencing and hyperintense on T2, although hypointensity on both T1 and T2 has been described[4-6]. Given its potential for significant destruction and recurrence, it has been recognized that complete excision is required[1,2,5,7,8].

In this report, we present the sixth case ever reported of FCM of the spine. This patient had a thoracic FCM that was treated with a gross total resection, including a T11 subtotal vertebrectomy and T8-L1 posterior spinal instrumented fusion. She remains highly functioning and tumor-free over 3 years after resection.

The patient was informed that data concerning the case would be submitted for publication, and she provided consent.

Case Report

A 19-year-old, previously healthy woman presented to the emergency department for right lower-quadrant abdominal pain worsening over 3 days. On examination, the patient had full strength and normal sensation in all extremities. No long tract signs were present, and she ambulated with a normal gait. Rectal tone was normal. Radiographs of the thoracic spine demonstrated loss of the normal left pedicle contour at T11 with mild vertebral body wedging (Fig. 1). CT revealed a lytic, expansile, and stippled lesion involving the left pedicle, posterior elements, and a large part of the T11 vertebral body, resulting in significant cortical destruction. A sclerotic bony rim around the lesion was noted (Fig. 2). On magnetic resonance imaging (MRI), the lesion was hypointense on T1-weighted sequences and isointense on T2-weighted sequences (compared with bone) with heterogeneous contrast enhancement. The tumor measured 4.4 cm anterior-posterior, 3.9 cm transversely, and 5.9 cm cranial-caudal. It extended into the epidural space, causing cord compression without cord signal change. Our initial radiologic differential was broad and included both malignant and benign/benign aggressive lesions, including chondrosarcoma, aneurysmal bone cyst, and giant cell tumor. Desmoplastic fibroma was also included in the

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Keywords spinal tumor; fibrocartilaginous mesenchymoma; Dahlin tumor

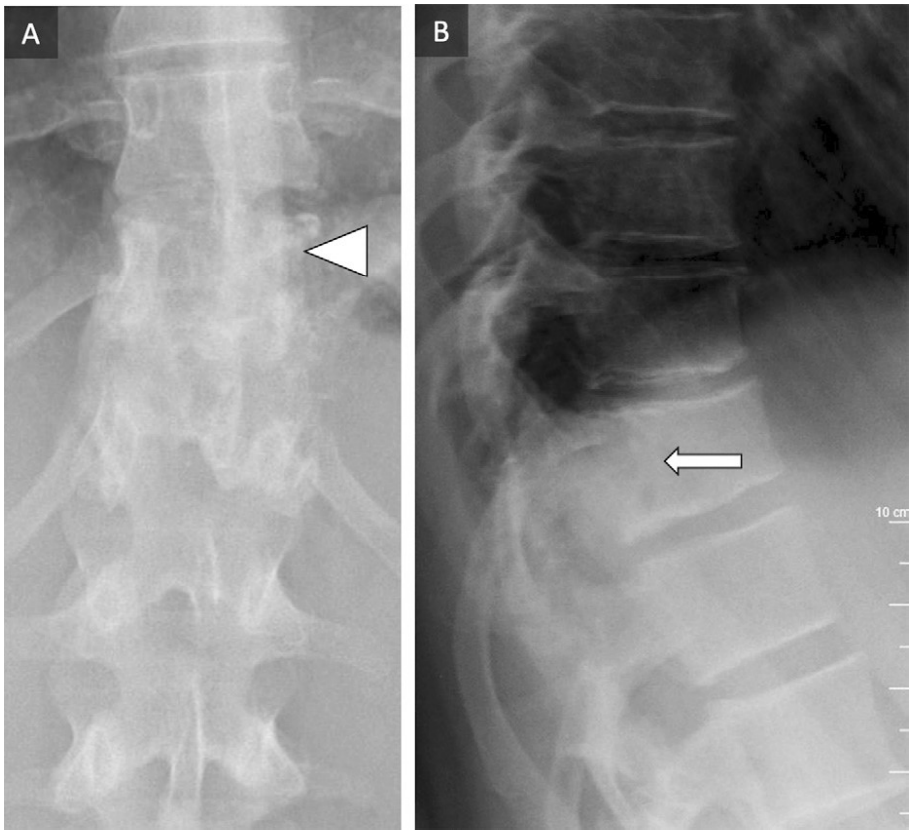

Fig. 1

Standing anteroposterior (**Fig. 1-A**) and lateral (**Fig. 1-B**) radiographs of the thoracic spine on initial presentation. There is loss of the left T11 pedicle contour that would normally be seen on anteroposterior radiograph (panel A; arrowhead) and wedging of the T11 vertebral body with destruction of the posterior elements (panel B; arrow). Mild focal kyphosis at T11 is present.

initial radiographic differential diagnosis; however, it is not typically identified in the spine. Other lesions included on the radiologic differential, although considered less likely, included osteosarcoma and chordoma. Further workup, including CT of the chest, abdomen, and pelvis, revealed no other tumor or metastatic disease. The patient's flank pain spontaneously resolved after initial workup, making the tumor most likely an incidental finding.

The core needle biopsy demonstrated nodules of bland hyaline cartilage with plate-like internal architecture and endochondral ossification, imbedded in a mild to moderately cellular spindle cell stroma. There was mild atypia of some of the chondrocytes, and no significant atypia or mitotic activity was observed within the collagenous stroma. (Fig. 3). This was felt to favor FCM over fibrocartilaginous dysplasia (FCD), enchondroma, or a low-grade chondrosarcoma. Considering FCM is exceedingly uncommon, the biopsy was repeated, and slides were circulated to an outside pathologist at another large, academic medical center . The diagnosis was confirmed, noting the above findings and adding that the specimen contained a hypocellular to moderately cellular collagenous stroma composed of spindle cells with a storiform pattern in the more cellular areas. The diagnosis of FCM was made with these histological findings paired with clinical imaging of an expansile, lytic, and focally calcified mass arising from the posterior elements and extending into the canal, with a well-circumscribed sclerotic margin. *IDH1* and *IDH2* gene mutation analysis by next-generation sequencing was negative. These mutations can contribute to tumorigenesis and have been described in chondrosarcoma as well as other malignancies[9].

After discussion with the patient and multidisciplinary tumor board, we proceeded with a gross total resection. Using a standard posterior midline incision, the thoracolumbar spine was exposed, and posterior instrumentation was placed T8-L1. The posterior elements of T10-T12 were removed pedicle-to-pedicle, and a corpectomy of T11 was performed through a left lateral extracavitary approach. Approximately 70% of the vertebral body was removed, as was the gross tumor. Although adherent to the dura, the tumor was completely resected without durotomy. An irrigating ultrasonic diamond tip burr was used to cauterize the rim of bone remaining anteriorly and on the right. A titanium Harms cage was filled with allograft and placed from T10 to T12, rods were placed, and structural allograft struts were placed posteriorly. The patient tolerated the procedure without complication. Postoperative radiographs are shown in Fig. 4. Final surgical pathology was comparable with the previous biopsies and consistent with FCM. Also within this cartilaginous lesion was a

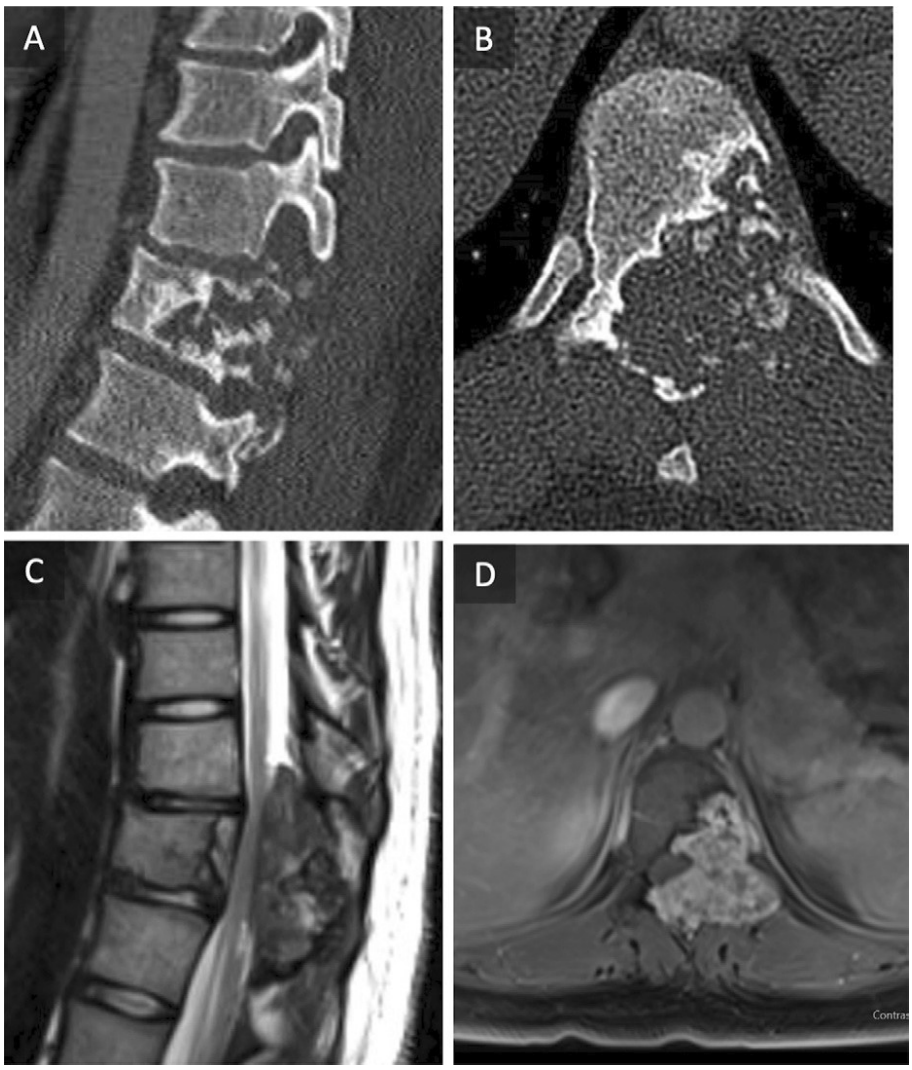

Fig. 2

Advanced imaging of the tumor including sagittal (**Fig. 2-A**) and axial (**Fig. 2-B**) CT cuts demonstrating a large lytic lesion at T11 with destruction of the posterior elements, left pedicle, and left-sided vertebral body. A sclerotic bony rim around the lesion is noted, as are calcifications within the tumor body in a "popcorn" or "ring and arc" pattern. T2-weighted MRI (**Fig. 2-C**) demonstrates a heterogenic hypointense mass with extension into the canal resulting in severe canal stenosis, with enhancement shown on the T1 postcontrast axial imaging (**Fig. 2-D**). The tumor measured 4.4 cm in the anterior-posterior direction, 3.9 cm transversely, and 5.9 cm in the cranial-caudal direction. CT = computed tomography, and MRI = magnetic resonance imaging.

focus of tumor with a cartilaginous cap and underlying cartilaginous seams entering into subchondral bone, a feature resembling osteochondroma, but also typical of FCM.

At three-year follow-up, the patient remains neurologically intact and without pain. She is well-aligned clinically, with maintained alignment on radiographs. Follow-up surveillance MRI and CT scans are without recurrence now over 3 years postoperatively, with robust fusion through the cage and posterior allograft struts (Fig. 4).

Discussion

FCM is a rare tumor lacking malignant features and with no known potential for metastases. Among published FCM cases, those treated with incomplete resection have had the tendency to recur[1,2,5,8,10], necessitating complete resection to prevent recurrence.

Only 5 previous cases of FCM of the spine are reported in the English literature. In 1992, the first published case of spinal FCM described a 19-year-old man who presented with radicular leg pain from an L5 mass[10]. He was treated with a 2-stage L5 vertebrectomy, placement of a structural femoral allograft, and posterior instrumented fusion of L3 to the sacrum. The tumor resection was complicated by durotomy and profuse epidural bleeding because the tumor was very adherent to the dura. More recently, in 2010, Martínez-Lage et al.[4] reported a 9-year-old boy with FCM of the T12 vertebra. The patient presented with back pain and was noted to have mild thoracolumbar kyphosis without neurologic deficits.

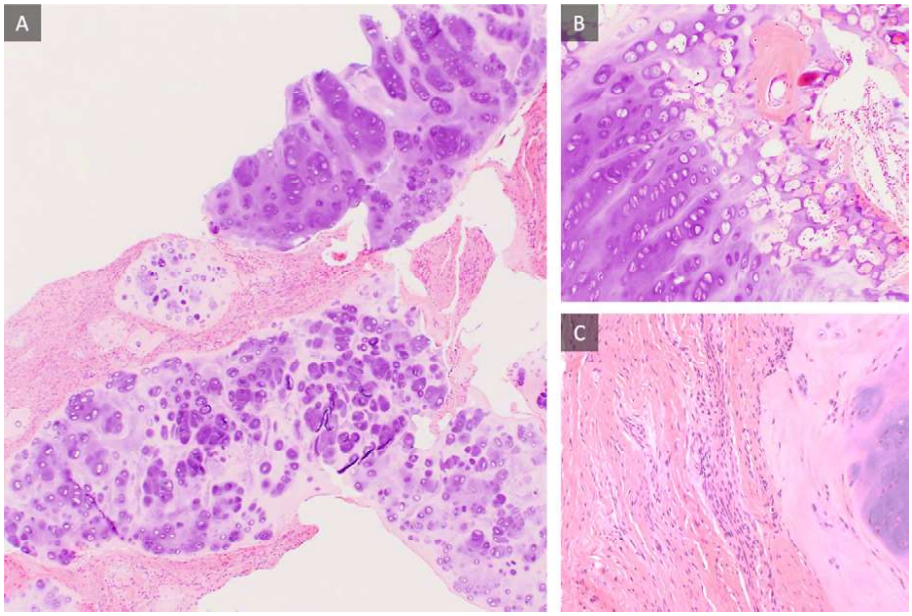

Fig. 3

Core needle biopsy of the T11 tumor with histopathology diagnostic of FCM. A low-power image of the core is shown (**Fig. 3-A**), demonstrating islands of hyaline cartilage with benign features surrounded by a spindle cell stroma. High-powered images demonstrate the stacked, plate-like chondral cells resembling the physeal architecture (**Fig. 3-B**), a hallmark of FCM. Overall, FCM demonstrates a moderately cellular stroma without significant atypia (**Fig. 3-C**). Both biopsy specimens and final surgical pathology demonstrated no evidence of malignant features. FCM = fibrocartilaginous mesenchymoma.

Radiographs and CT demonstrated tumor involvement of the posterior elements of T12 with wedging of the vertebral body and focal kyphosis. MRI demonstrated slight cord compression. The patient underwent gross total resection and posterior spinal fusion from T10 to L2. The remaining cases of spinal FCM are reported in 2 case series with limited clinical and surgical data. Bulychova et al.[2] included a 22-year-old man with a tumor involving the posterior elements of L4. The patient was treated with an intralesional excision and had no recurrence with 10 years of follow-up. Gambarotti et al.[5] included FCM of L3 in an 18-year-old woman and of L4 in a 13-year-old boy. Both patients presented with pain and were treated with *en bloc* resections. No disease recurrence was noted with 10- and 3-year follow-up, respectively. Of note, these reports provide limited surgical data; it is uncertain whether a reported "*en bloc*" resection was truly *en bloc* without violating the tumor capsule or simply a gross total resection. Although descriptions of the specific resection techniques vary among these 6 cases, they all report a gross total resection at minimum.

After initial biopsy in our case, a wide histopathologic differential diagnosis was considered, including enchondroma, FCD, and low-grade chondrosarcoma, all of which can include bland hyaline cartilage cells with few mitotic figures. Because a biopsy is a limited sampling of the tumor, final diagnosis is only made once the surgical specimen is examined. There has been some debate as to whether FCM is a distinct entity from FCD because both lesions have overlapping features[11]. Gambarotti et al.[5] performed a molecular identification of genetic mutations among their series of 26 cases and identified no *GNAS, IDH1/2,* or *MDM2* amplifications (which are characteristic of FCD), concluding that FCM may have its own distinct molecular identity.

With the addition of our case, there are a total of 6 cases of FCM of the spine now published in the extant literature. Notably, there has not been a report of recurrence in the spine in any of these cases. Currently, a true *en bloc* resection may not be warranted for this slow-growing, locally destructive tumor because it has little to no proven malignant potential and has not recurred in the previous spine cases reported. However, given its potential for local recurrence, gross total resection is recommended. In addition to the need for gross total resection often requiring an extensive exposure (e.g., lateral extracavitary) with a corpectomy or vertebrectomy, the tendency of this tumor to adhere to the dura can further contribute to the technically challenging nature of these resections. ■

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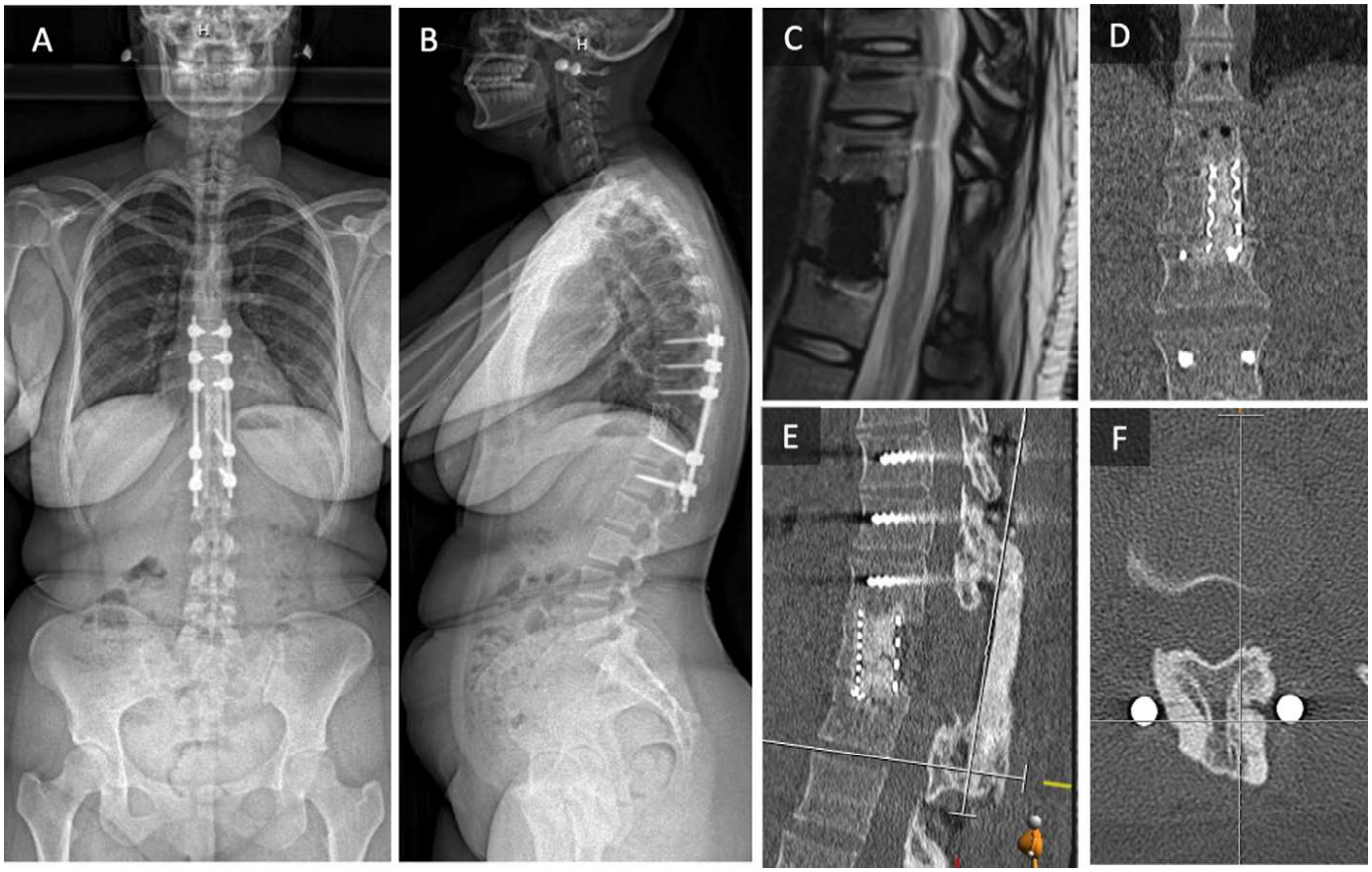

Fig. 4

Postoperative standing anteroposterior (**Fig. 4-A**) and lateral (**Fig. 4-B**) radiographs after tumor resection, including T11 corpectomy with cage placement and T8-L1 posterior instrumented fusion. The patient is now over 3-year status-post tumor resection and spinal fusion without signs of complications. Sagittal T2-weighted MRI demonstrates decompression of the spinal cord without evidence of recurrence (**Fig. 4-C**). Coronal (**Fig. 4-D**), sagittal (**Fig. 4-E**), and axial (**Fig. 4-F**) CT cuts 2 years postoperatively demonstrate solid fusion through the anterior cage as well as incorporation of the posterior allograft strut. CT = computed tomography.

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