14-Year-Old Boy With Mild Antecedent Neck Pain in Setting of Acute Trauma: A Rare Case of Benign Fibrous Histiocytoma of the Spine

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Abstract

A 14-year-old boy presented to the emergency department with neck pain after a football accident. Imaging revealed a discrete, expansile, lytic, radiolucent mass extending anterior from the left C2 vertebral body. The differential diagnosis for this mass is broad and includes both benign and malignant lesions. A thorough history and physical examination, along with selective imaging and a tissue sample, are essential in making the correct diagnosis.

We report a case of benign fibrous histiocytoma (BFH) in the cervical spine of a pediatric patient, present the diagnostic dilemma involved in diagnosing BFH, and review the literature to compare the characteristics of BFH with those of other benign bone lesions. To our knowledge, this is only the second reported case of BFH in the cervical spine of a pediatric patient.

Benign fibrous histiocytoma (BFH) is a rare, well-recognized, primary skeletal tumor accounting for approximately 1% of all benign bone tumors. Spinal involvement is exceedingly rare with only 11 cases reported in the literature.[1,2] We present a case of BFH located in the cervical spine of a pediatric patient that was successfully treated with curretage through an anterior surgical approach, along with a review of the literature and appropriate management concerning BFH of the spine.

Case Report

A 14-year-old boy was tackled while playing football and noticed immediate neck pain and subjective paresthesia in the upper extremities. Examination revealed a nontender spine (cervical, thoracic, lumbar) and normal strength and range of motion in all extremities. Sensation was diffusely intact, long tract signs were absent, and gait was normal. On questioning, the patient endorsed mild antecedent neck pain but denied prior history of any trauma. Neck pain did not radiate and was slightly worsened by activity but was mostly intermittent and random. As the neck pain was very mild and was not interfering with daily activities, the patient had not sought care before presenting to the emergency department. He had no pertinent past medical or surgical history.

The patient presented with a computed tomography (CT) scan of his head and cervical spine and a magnetic resonance imaging (MRI) scan of the cervical spine. A magnetic resonance angiography (MRA) scan of the neck was ordered after his arrival.

Axial and sagittal CT (**Figures 1A, 1B**) showed a 1×1.2-cm discrete, expansile, lytic, radiolucent mass extending anterior from the left C2 vertebral body. The mass appeared to abut the left vertebral artery foramen. The cortical bone surrounding the lesion was thin but uniform. Sagittal and axial T1-weighted MRI (**Figures 2A, 2B**) showed the discrete, expansile, homogenous lesion with the same intensity as normal bone marrow. Sagittal and axial T2-weighted MRI (**Figures 2C, 2D**) showed a discrete, expansile, homogenous lesion with primarily high signal intensity. Sagittal short tau inversion recovery (STIR) MRI (**Figure 2E**) again showed the lesion with primarily low inten-

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sity. Given the close proximity of the lesion to the vertebral foramen, MRA was ordered; it showed the lesion was not interfering with the vertebral artery (**Figure 2F**).

The tumor's location, in the left anterior aspect of the C2 vertebral body, was not conducive to percutaneous biopsy for establishing tissue diagnosis, so the decision was made to surgically excise the lesion. A left-sided anterior incision was made 2 fingerbreadths inferior to the jaw line in a neck crease. A head and neck surgeon assisted with dissection. Dissection was carried down through the skin, subcutaneous tissue, and platysma on to the anterior part of the spine medial to the carotid sheath. Superior thyroid nerve and vessels and superior laryngeal nerve were identified and preserved. Fluoroscopy confirmed correct location at C2. The tumor was easily visualized, and the outer shell broke easily with palpation. Gentle curettage was necessary when removing the tumor off the vertebral artery. A portion of the specimen was sent during surgery for frozen section, which showed infrequent mitotic figures and no other findings concerning for malignancy. No instability was created after curettage and excision of the tumor, so no grafting or instrumentation was necessary.

Grossly, the tumor was pale tan and firm. Histologic examination with hematoxylin-eosin staining revealed a bland spindle-cell neoplasm that focally involved bone. A storiform pattern was present. The cells had scant cytoplasm and oval to elongate nuclei with tapered ends. Significant nuclear pleomorphism was not seen. The stroma was loose, with focal myxoid change. Benign multinucleated giant cells were present. Mitotic activity was infrequent (**Figures 3A–3D**). Two attending pathologists reviewed the case material and the frozen and formalin-fixed specimens independently and concurred with the diagnosis of BFH. In addition, the case was reviewed at the surgical pathology consensus conference; the reviewers agreed on BFH, and additional studies were deemed unnecessary.

Given the patient's complete clinical picture, the differential diagnosis included nonossifying fibroma (NOF), eosinophilic granuloma (EG), BFH, fibrous dysplasia, giant cell tumor (GCT), aneurysmal bone cyst (ABC), and osteoblastoma (OB).

Discussion

BFH is an extremely rare bone lesion, accounting for only 1% of all surgically managed bone tumors; not counting the present case, only 11 spine cases have been reported in the literature.[1,2] BFH of the spine traditionally causes nonspecific, poorly localized pain. The **Table** lists the reported cases of spinal BFH and their presenting symptoms, location, and treatment. BFH usually occurs in young adults, but the age range is 5 to 75 years.[2-4] Mean age of

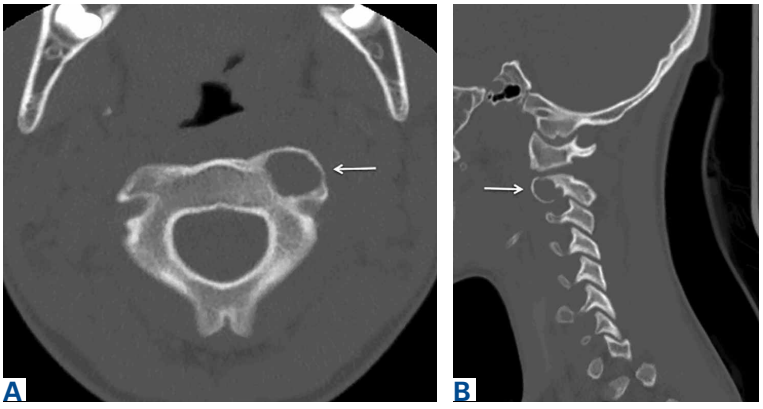

Figure 1. (A) Axial and (B) sagittal computed tomography shows discrete, expansile, radiolucent mass extending anteriorly from left C2 vertebral body.

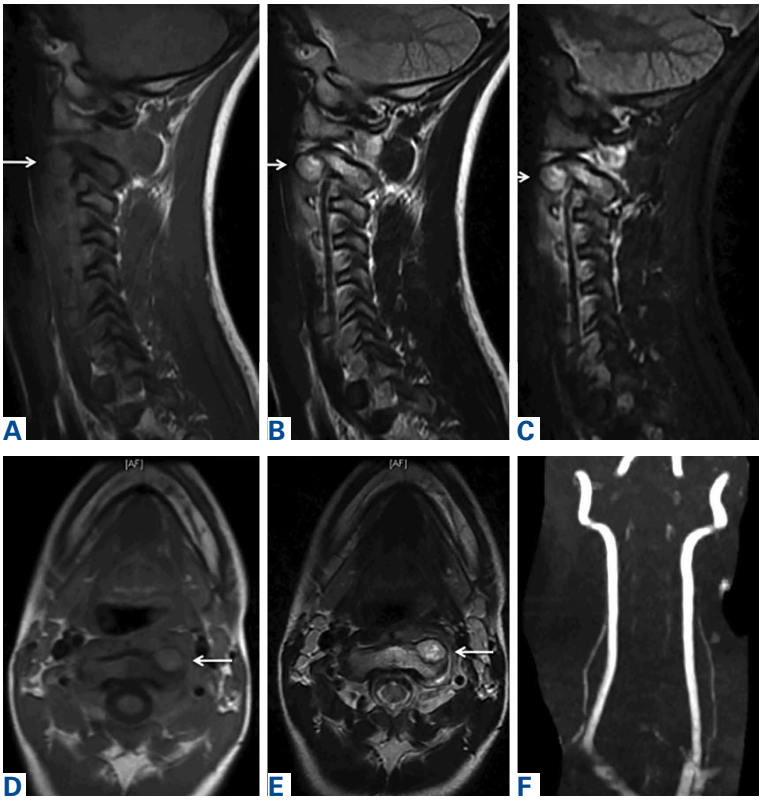

Figure 2. (A) Sagittal and (B) axial T1-weighted magnetic resonance imaging (MRI) shows discrete, expansile, homogenous lesion with low signal intensity. (C) Sagittal and (D) axial T2-weighted MRI shows discrete, expansile, homogenous lesion with primarily high signal intensity. (E) Sagittal short tau inversion recovery (STIR) MRI shows lesion with primarily low signal intensity. (F) Magnetic resonance angiography shows normal vertebral arteries.

the 12 patients with spinal BFH in the literature (including ours) is 25 years.[1] In addition, spinal BFH appears to have no predilection for sex.

Skeletal BFH presents as a discrete, well-defined, osteolytic lesion with sharp borders and potentially a sclerotic rim.[4-6] Cortical expansion and even cortical disruption with invasion into adjacent tissue have occurred in flat bones.[7] Histologically, BFHs contain spindle cells, multinucleated giant cells, and foam cells in storiform pattern.[6]

BFH shares many of its radiologic and histologic characteristics and clinical symptoms with other benign bone lesions (the tumors listed above). Therefore, accurate diagnosis of BFH requires appropriate correlation of clinical, radiographic, and histologic data.[2,3,8] Below is a comparison of BFH with related bone lesions.

Spinal BFH causes a nonspecific, poorly localized pain similar to that of EG, ABC, GCT, and OB.[3,9] NOF and fibrous dysplasia generally do not cause pain, unless these lesions are discovered secondary to a pathologic fracture.[8,10,11] Our patient had minor antecedent neck pain, which was brought to light by his football accident. ABC and OB are more locally aggressive than BFH and can cause neurologic symptoms by mass effect and spinal cord or nerve root compression.[1,8] In this case and in the 6 other cases of BFH of the cervical spine, there were no neurologic changes.[4,10]

Of the tumors mentioned, NOF and EG almost always occur in children. However, NOF usually occurs in the metaphyseal region of long bones, and EG is usually accompanied by systemic symptoms, such as lymphadenopathy, hepatomegaly, and increased inflammatory markers.[1,8] Fibrous dysplasia usually presents in childhood but does not become symptomatic until adulthood. GCTs and OB predominantly occur in adulthood.[12,13] Our patient's age and lack of other systemic symptoms supported the diagnosis of BFH.

Appearance on MRI is reported less with BFH than with other tumors, but heterogenous signal intensity similar to that of skeletal muscle on T1-weighted images and high signal intensity on T2-weighted images is typically reported.[8,14] NOF and fibrous dysplasia do not disrupt the bony cortex unless a pathologic fracture has occurred.[4] GCTs are more aggressive lytic lesions with more aggressive radiologic features. GCTs generally cause cortical expansion/attenuation, and lack a sclerotic rim. GCTs also have a heterogenous appearance on MRI and give a low to intermediate signal on both T1- and T2-weighted images.[12,15] The appearance of EG is similar to that of BFH as an osteolytic lesion with a sclerotic rim, though EGs typically break through the cortex and acquire a "punched-out" look.[1,8] ABC typically is described as an expansile osteolytic lesion with a "soap-bubble" appearance on radiographs; periosteal elevation and cortical attenuation can also be visualized. MRI shows the typical multilobular appearance of the lesion with fluid levels.[13]

OB appears as a radiolucent lesion, with or without calcifications, surrounded by a thin margin of reactive bone.[14,16] A distinguishing characteristic of OB was thought to be intense radioisotope uptake on bone scintigraphy, but recently a bony BFH demonstrated intense uptake.[17] OBs typically demonstrate nonspecific MRI results similar to those of BFH: low to intermediate signal on T1-weighted images and intermediate to high signal on T2-weighted images.[13] In our patient's case, the radiographic appearance and lack of specific radiographic findings consistent with the other tumors supported the diagnosis of BFH.

Histologically, BFHs contain spindle cells, multinucleated giant cells, and foam cells in a

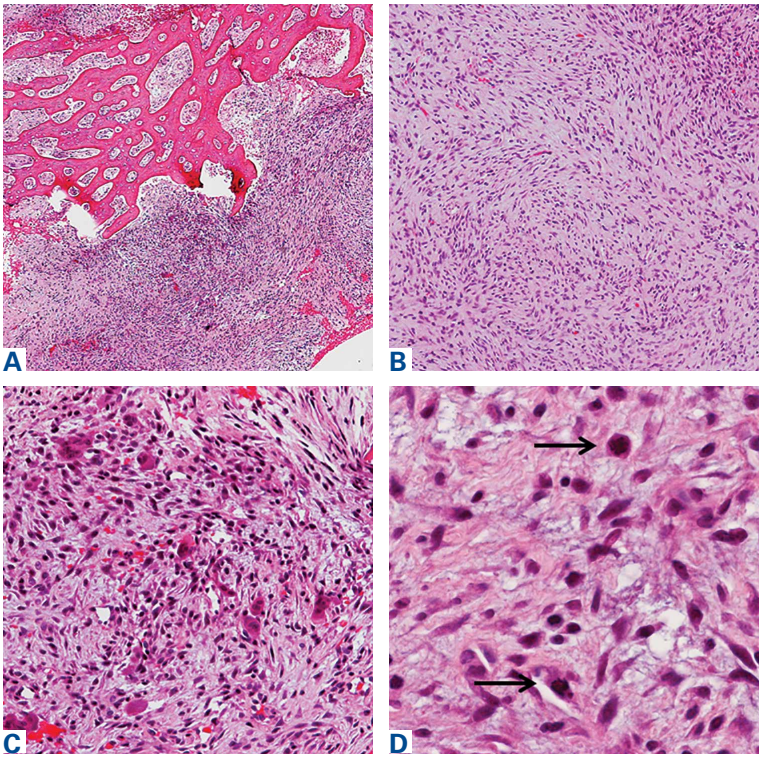

Figure 3. (A) Benign fibrous histiocytoma involving bone (hematoxylin-eosin, original magnification x4). (B) Bland spindle cells in storiform pattern (hematoxylin-eosin, original magnification x10). (C) Scattered multinucleated giant cells (hematoxylin-eosin, original magnification x20). (D) Mitotic figures (arrows) (hematoxylin-eosin, original magnification x60).

storiform pattern[6] which was demonstrated in our patient's case. In addition, significant nuclear pleomorphism, mitotic activity, and necrosis were absent—a difference between BFH and malignant fibrous histiocytoma.[4,15] The microscopic characteristics of BFH readily differentiate it from OB, ABC, EG, and GCT, but not from NOF on microscopic appearance alone. Clinical and radiographic findings must be consistent, as mentioned.[7,18]

Complete surgical excision is the reported treatment for BFH. Prognosis after resection or curettage is usually good, and recurrences have been rare.[1,2] Depending on the intraspinous location of BFH, stabilization after resection or curettage may be necessary to prevent residual instability. Three of the 11 reported cases of spinal BFH required stabilization by anterior fusion or posterior pedicle screw fixation after resection.[1,2] The other 8 cases underwent excision alone or excision and grafting. All 11 patients were disease-free at a mean follow-up of 3.5 years.[1] In nonspinal BFH, however, both local recurrence and lung metastasis have been reported.[2,5,9,19]

Clarke and colleagues[9] reported local recurrences in 3 of 8 cases. These recurrences involved BFH in long bones of the leg, which had been treated with curettage and grafting. There has been no reliable report of a malignant change in BFH.[2,9] The only case of lung metastasis, reported by Unni and Dahlin[6] in their study of 10 cases, occurred 2 years after local recurrence in the distal femur.

Our patient was doing well at most recent follow-up, 6 months after surgery. He had no pain and had returned to normal activities. Although there are no reported cases of spinal BFH recurrence, we will follow this patient with imaging on an annual basis. His case is of particular interest to orthopedic surgeons because they encounter benign bone lesions every day, and many of these lesions are in difficult anatomical locations. Knowing the characteristics, differential diagnoses, and appropriate diagnostic workups for benign bone lesions is important for optimal and timely patient care.

Table. **Reported Cases of Benign Fibrous Histiocytoma of Spine**

Study	Year	Age, y	Sex	Symptom(s)	Location	Treatment	Follow-Up,[a] y
Destouet et al[19]	1980	24	M	Neck pain	C2 posterior elements	Curettage and graft	3.25
Roessner et al[18]	1981	41	M	Neck pain, stiffness	C3, C4	Resection, anterior fusion/graft with C2–C5 laminectomy	0.25
Mirra et al[15]	1989	18	M	Neck pain	C2 posterior elements	Excision	5
		24	M	Neck pain	C2 spinous process	Excision	Not reported
		28	M	Neck pain	C6 posterior elements	Excision	4.9
Hoeffel et al[20]	1992	13	M	Scoliosis, stiffness	T12	Excision and graft—combined approach	2
Peicha et al[10]	1999	44	F	Minor trauma, neck pain	C2, odontoid fracture	Excision and graft—anterior approach	5
Grohs et al[5]	2002	33	F	Abdominal, back, and leg pain	L3 posterior elements	Excision and graft	5.4
van Giffen et al[8]	2003	6	M	Neck pain, stiffness	C1 posterior arch	Excision and hemilaminectomy	1
Kuruvath et al[2]	2006	24	M	Thoracic back pain	T3 posterior elements	T3 laminectomy/vertebrectomy, posterior pedicle screw fixation	2.5
Demiralp et al[1]	2009	21	M	Low back and leg pain	L2 posterior elements	Curettage, grafting, posterior pedicle screw fixation	6
Present case	2016	14	M	Minor trauma, neck pain	C2 vertebral body	Excision/curettage	0.5

[a]Patients were found to be disease-free at follow-up (finding for 1 patient was not reported).

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