Benign fibrous histiocytoma of the thoracic spine

Case report and review of the literature

SARIN KURUVATH, M.R.C.S., DOMINIC G. O’DONOVAN, F.R.C.PATH.,
A. ROBERT ASPOAS, F.R.C.S.(SN), AND KAROLY M. DAVID, F.R.C.S.(SN)

Departments of Neurosurgery and Neuropathology, Essex Centre for Neurological Sciences,
Oldchurch Hospital, Romford, Essex, United Kingdom

Benign fibrous histiocytoma (BFH) is a rare skeletal tumor, accounting for approximately 1% of all surgically managed benign bone tumors.19 Approximately 87 cases have been reported in the literature.32 Location in the spine is rare, with nine reported cases of BFH involving the spine and only one in a thoracic vertebra. We present the second reported case of BFH of a thoracic vertebra, including its management, and review the literature concerning BFH of the spine.

KEY WORDS • benign fibrous histiocytoma • spinal tumor • thoracic spine • spinal stabilization

Benign fibrous histiocytoma is a rare skeletal tumor accounting for approximately 1% of all surgically managed benign bone tumors.19 Approximately 87 cases have been reported in the literature.32 Location in the spine is rare, with nine reported cases of BFH involving the spine and only one in a thoracic vertebra. We present the second reported case of BFH of a thoracic vertebra, including its management, and review the literature concerning BFH of the spine.

Case Report

Presentation and Examination. This 24-year-old man was referred by orthopedic surgeons in May 2002 with a presenting complaint of constant thoracic back pain that was worse at night and of 3 months’ duration. He had been involved in a snowboarding accident 1 month before the onset of pain and was admitted 2 months later. Two weeks before admission he experienced progressive weakness of the lower limbs and a feeling of incomplete voiding of urine.

Physical examination revealed that muscle tone was decreased in the lower limbs, muscle power was Medical Research Council Grade 4+/5 in all muscle groups, reflexes were increased, and ankle clonus and the Babinski sign was present bilaterally. Sensory level was T-8 bilaterally with saddle hypesthesia, but vibration sensation was preserved. A rectal examination did not reveal any abnormality. He had no neurological deficit in the upper limbs. The results of routine hematological and biochemical tests, including measurement of serum calcium, phosphorus, and alkaline phosphatase, were normal. The results of myeloma/plasmacytoma screening tests, including serum electrophoresis and urine test for Bence Jones proteins, were non-diagnostic. A chest radiograph showed no abnormality. Magnetic resonance imaging of the spine revealed a mass lesion involving the T-3 vertebra, causing severe cord compression (Fig. 1).

Operation. Percutaneous biopsy sampling of the lesion was not considered due to the acute presentation of progressive paraparesis, sensory deficits, and bladder dysfunction. Urgent spinal cord decompression was indicated to reverse the neurological signs and symptoms and to obtain tissue for histological diagnosis. The patient underwent a T-3 decompressive laminectomy with subtotal removal of the tumor. The theca and the left T-3 nerve root were adequately decompressed after the left T-3 pedicle was removed, although its lateral and anterior extension could not be excised completely through the posterior approach.

Histological Examination. Histological examination showed a neoplasm composed of predominant spindle cells arranged in a storiform pattern with a few mitoses, admixed with foamy macrophages (Fig. 2). Osteoid with scattered osteoblasts and osteoclasts was present. In addition, adjacent blood-filled cavities with septa of collagen were present. There were no features of malignancy. These appearances indicated a BFH of the bone with secondary aneurysmal bone cyst formation.

Postoperative Course. After surgery, the patient regained normal muscle power in his lower limbs and full control of the bladder within 2 days. He was discharged on the 5th postoperative day.

Abbreviations used in this paper: BFH = benign fibrous histiocytoma; MR = magnetic resonance; VB = vertebral body.
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Second Examination and Operation. Follow-up MR imaging 6 months after surgery demonstrated growth of residual tumor with further extension into soft tissue. Because of the benign histological findings and expanding character of the bone lesion, complete excision of the tumor was performed via a left-sided costotransversectomy and bilateral transpedicular approach and T-3 vertebrectomy. The left T-3 nerve root was sacrificed. An expandable titanium cage packed with freeze-dried irradiated coarsely ground human bone allograft (National Blood Service Tissue Services, Wakefield, Yorkshire, United Kingdom) was placed between the T-2 and T-4 vertebral bodies and stabilized with a T-1, T-2, T-4, and T-5 pedicle screw/rod system.

Second Histological Examination. Histological examination confirmed benign fibrous histiocytoma of bone, similar to the previous biopsy finding.

Second Postoperative Course. The patient made a good postoperative recovery. Repeated radiographs confirmed good positioning of the cage and pedicle screws (Fig. 3). He was discharged 10 days later after intensive physiotherapy. On follow-up examination, the patient indicated he was able to walk 3 miles every day and had resumed playing golf. The only neurological sequelae were bilateral hypesthesia of the T-3 dermatome and a left-sided present Babinski sign. The decreased sensation on the right side was probably due to injury during manipulation of the right T-3 nerve root. Repeated MR imaging 1 and 2 years after surgery demonstrated no recurrence of the tumor (Fig. 4).

Discussion

Benign fibrous histiocytoma is a well recognized but very rare bone tumor. It accounts for approximately 1% of all surgically treated benign bone tumors.19 The age of patients with BFH has been reported to range from 5 to 75 years, but it has most frequently been seen in young adults. No sex predilection has been reported.13,32 Dahlin8 and Unni and Dahlin31 have found only 10 cases of BFH in a total of 11,087 skeletal tumors. All patients were adults between the ages of 23 and 60 years, with a male to female ratio of 4:3. Grohs, et al.,19 in their 15-year study of primary bone tumors, have reported 10 cases of BFH in a total of 11,087 skeletal tumors. All patients were adults between the ages of 23 and 60 years, with a male to female ratio of 4:3. Grohs, et al.,19 in their 15-year study of primary bone tumors, have reported 10 cases of BFH among a total of 1037 patients, with a median age of 28 years (range 24–52 years) in the patients with BFH. The male to female ratio

Fig. 1. Left: Unenhanced sagittal T2-weighted MR image revealing a mass lesion involving posterior elements of the T-3 vertebra, causing severe cord compression. Increased sagittal density is also seen in the VB. Right: Axial T1-weighted MR image revealing an expansile mass lesion in the left half of the VB, pedicle, lamina, spinous and transverse processes, and head of adjoining rib.

Fig. 2. Photomicrographs showing BFH. Left: A portion of bone with trabeculae and a sharply demarcated spindle cell neoplasm. Center: Spindle cells and foamy cells showing no atypia are present; mitoses are inconspicuous, with scanty osteoclasts. Right: Tumor with two cavities containing red blood cells separated by a septum, indicating secondary bone cyst formation. H & E, original magnifications × 2 (left), × 20 (center), and × 4 (right).
was 4:6. Benign fibrous histiocytoma usually occurs in the dermis, superficial subcutaneous tissue, and deep soft tissues and more rarely in parenchymal organs. Occurrence in bone is frequent in the ilium, femur, pelvis, humerus, tibia, fibula, and ribs and is less frequent in vertebrae and in the sacrum, skull, and mandible. Benign fibrous histiocytoma is usually epiphysial or metaepiphysial in location.

Local pain is the cardinal symptom in patients with BFH. Duration of pain ranges from weeks to several years. Pathological fracture very rarely occurs. As in our case, investigations are usually initiated because of pain. Benign fibrous histiocytoma is commonly visualized on plain radiographs as an osteolytic lesion with sharply defined borders. Magnetic resonance imaging of BFH is nonspecific. Combined consideration of clinical symptoms, tumor location, and radiographic and histological characteristics is used to distinguish BFH from other benign lesions such as nonossifying fibroma, fibrous dysplasia, aneurysmal bone cyst, osteoblastoma, Langerhans cell histiocytosis, enchondroma, desmoplastic fibroma, chordomyoid fibroma, and giant cell tumor.

The histogenesis of BFH is a matter of discussion. Arguments for both fibroblastic and histiocytic origins have been proposed. Histologically BFH is indistinguishable from nonossifying fibroma and metaphysical fibrous defect, which are exclusively located in the metaphysis of long bones, but it is distinguished with typical radiographic findings and its occurrence in children and adolescents. Both present with fibroblastic components and focal histiocytic differentiation. Grossly BFH is usually partly composed of cysts containing yellow or hemorrhagic fluid. The solid component varies from firm to soft and from gray to ruddy brown. These components are well demarcated but unencapsulated. The fibroblasts form fascicles or a storiform pattern. The matrix comprises collagen including reticulin, foamy histiocytes, multinucleate giant cells (sometimes of the classic Touton type), and collections of lymphocytes and scanty plasma cells. Immunohistochemically the most widespread reactivity is found for vimentin and CD68 (granular cytoplasmic staining), S100 protein, lysozyme, α1-antitrypsin, and antichymotrypsin, indicating histiocytic differentiation. Although occasionally BFH shows indistinct borders or features of atypia and mitoses, BFH must be distinguished from giant cell tumors and malignant conditions such as malignant fibrous histiocytoma. Soft-tissue extension was an unusual finding in our case.

The treatment of benign spinal tumors is individualized based on location, extent, and clinical behavior. Some benign lesions are observed without requiring intervention, whereas others require complete en bloc excision followed by complex reconstruction. Some of these lesions also possess the potential for malignant transformation.
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Followed by metastasis.26 Treatment of primary bone tumors of the spine should include the following: 1) diagnosis using appropriate biopsy procedures (in nonacute presentations); 2) staging and decision about the surgical margin using the Enneking oncologic surgical staging system11 with the goal of defining the biological behavior of the lesion;1,4,21 and 3) surgical planning aided by the Weinstein-Boriani-Biagini classification system, which involves a pretreatment three-dimensional radiographic study of the lesion. This system divides the vertebrae in the transverse plane into 12 sectors in a clock-face arrangement from right to left and into five tissue layers, allowing distinction between extensions in various tissue planes.1,4,5,21 Use of these systems maximizes diagnostic and treatment efforts and clinical success and allows reliable communication about the results among treatment centers.1

The operative procedures typically used for the treatment of benign lesions include intralesional excision, marginal en bloc excision, and wide en bloc excision. The vast majority of spinal tumors are located in the posterior elements and hence can be approached through a posterior route. Tumors in which a significant proportion of the anterior elements are involved are approached via an anterior route, posterior route (transpedicular approach, costotransversectomy, lateral extracavitary approach), or a combined anterior–posterior route. For resection of lesions involving the VBs of the upper thoracic vertebrae (T1–4), the anterior approach often necessitates a sternotomy via a manubrial window. Due to the presence of vital structures at the thoracic inlet, this route is technically demanding and associated with higher risk of injury to the aorta, thoracic duct, and recurrent laryngeal nerve.2,24,33 For these reasons, in our case we elected to resect the tumor via the posterior transpedicular approach supplemented by costotransversectomy. This approach has been well described in the literature.2,6,14,25 After excision of benign vertebral tumor, VB replacements composed of artificial materials and implants such as ceramics, glass, polymethylmethacrylate, carbon fiber, and titanium mesh cage may be used with bone grafting to provide anterior column support and correct sagittal plane deformity. Implants with posterior pedicle screw fixation offer immediate segmental stability.

Surgery with complete excision is the treatment of choice for BFH. The prognosis after resection or curettage is usually good, and recurrences are observed only in rare cases.Clark, et al.,7 reported local recurrences in three of eight cases, all of which involved BFH in the long bones of the leg treated with curettage and grafting. There is no reliable report in the literature of malignant change of BFH. Unni and Dahlin,31 in their study of 10 cases, described one patient with pulmonary metastasis 2 years after a local recurrence in the distal femur in which amputation was required. There is no evidence in the literature to support the role of radiotherapy in the treatment of BFH. However, radiotherapy has been advocated in the treatment of selected other benign spinal tumors, such as aggressive osteoblastoma, aneurysmal bone cysts, and giant cell tumors.18 The use of radiosurgery has been reported for benign spinal tumors such as neurofibroma, paraganglioma, schwannoma, meningioma, chordoma, and hemangioma.17 Spinal stereotactic radiosurgery may have a role for recurrent lesions that are not amenable to complete excision or open

TABLE 1
Summary of all published case reports of BFH involving the spine

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Presenting Symptom</th>
<th>Lesion Location</th>
<th>Treatment</th>
<th>Follow Up (yrs)</th>
</tr>
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<tbody>
<tr>
<td>Destouet, et al., 1980</td>
<td>24, M</td>
<td>neck pain</td>
<td>C-2 lamina, pedicle, spinous process</td>
<td>excision (curettage) &amp; graft</td>
<td>3.25, disease free</td>
</tr>
<tr>
<td>Roessner, et al., 1981</td>
<td>41, M</td>
<td>neck pain &amp; stiffness</td>
<td>C-3 &amp; C-4</td>
<td>complete resection, anterior fusion w/ iliac graft &amp; C2–5 laminectomy</td>
<td>0.25, disease free</td>
</tr>
<tr>
<td>Mirra, et al., 1989</td>
<td>18, M</td>
<td>neck pain</td>
<td>C-2 spinous process, posterior arch</td>
<td>excision</td>
<td>5, disease free</td>
</tr>
<tr>
<td></td>
<td>24, M</td>
<td>neck pain</td>
<td>C-2 spinous process, posterior arch</td>
<td>excision</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>28, M</td>
<td>neck pain</td>
<td>C-6 spinous process, posterior arch</td>
<td>excision</td>
<td>4.9, disease free</td>
</tr>
<tr>
<td>Hoeffel, et al., 1992</td>
<td>13, M</td>
<td>scoliosis, spine stiffness</td>
<td>T-12</td>
<td>combined approach: VB excision &amp; tibial graft; excision of pedicle, lamina, articular process, &amp; graft; anterior approach</td>
<td>2, disease free</td>
</tr>
<tr>
<td>Peicha, et al., 1999</td>
<td>44, F</td>
<td>minor trauma, neck pain</td>
<td>C-2 VB, odontoid fracture</td>
<td>anterior approach: excision &amp; graft</td>
<td>5, disease free</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>excision w/ wide margins &amp; graft</td>
<td>(? recurrence)</td>
</tr>
<tr>
<td>Grohs, et al., 2002</td>
<td>33, F</td>
<td>abdominal, back, &amp; leg pain</td>
<td>L-3 posterior elements</td>
<td>excision (hemilaminectomy)</td>
<td>5.4, disease free</td>
</tr>
<tr>
<td>van Giffen, et al., 2003</td>
<td>6, M</td>
<td>neck pain, restricted rotation</td>
<td>C-1 posterior arch</td>
<td>2-stage op: T-3 decompressive laminectomy &amp; T-3 vertebrectomy &amp; expandable cage &amp; T-5 pedicle screw fixation</td>
<td>1, disease free</td>
</tr>
<tr>
<td>present case</td>
<td>24, M</td>
<td>thoracic back pain</td>
<td>T-3</td>
<td>—</td>
<td>2.5, disease free</td>
</tr>
</tbody>
</table>
surgery and in the treatment of medically inoperable patients. Table 1 lists reported cases of BFH involving the spine, including our clinical presentation, and their management and follow up. Careful clinical and radiological follow up is indicated in the management of BFH.

Conclusions

Spinal BFH is a very rare bone tumor. It can be distinguished from other benign lesions by characteristic clinical and radiographic features and through histological evaluation. Complete excision is the treatment of choice, but the management of spinal BFH should be individualized and carefully planned with reference to classification systems based on tumor location and extent of involvement. Treatment incorporates complex spinal surgery with principles of excision of tumor, spinal cord decompression, and spinal column reconstruction and stabilization. Treated patients have had excellent clinical outcomes, and there have been no reported cases of recurrence after complete resection of the tumor.

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References