Desmoplastic fibroma of the spine: a series of 12 cases and outcomes

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Abstract

BACKGROUND CONTEXT: Desmoplastic fibroma (DF) is a benign, yet locally aggressive, tumor of the connective tissue. Desmoplastic fibroma in the spine is extremely rare, and only a few cases have been reported. Although surgical resection of DF arising in the spine is commonly regarded as a recommended treatment, it is difficult to achieve satisfactory results.

PURPOSE: This study reviews the clinical patterns and follow-up data of patients with DF in the spine who underwent surgical treatment. We attempted to correlate surgical treatment and outcomes over time.

STUDY DESIGN: A retrospective clinical study of the surgical managements, including subtotal resection, total spondylectomy, and en bloc resection, for DF in the spine. Desmoplastic fibroma of the spine treatment occurred from 2004 to 2009 at the Department of Bone Tumor Surgery, AA Hospital.

PATIENT SAMPLE: Twelve consecutive cases of DF of the spine underwent surgical treatment at our center between 2004 and 2009.

OUTCOME MEASURES: Neurologic outcomes were evaluated using Frankel score system and recurrence and metastasis were evaluated by computed tomography or magnetic resonance imaging of the surgical segments involved. Imaging was performed 3, 6, and 12 months after surgery, every 6 months for the next 2 years, and then annually for life.

METHODS: Overall, two different surgery protocols were applied. One protocol involved subtotal resection followed by radiotherapy (n=4), whereas the other involved total tumor resection (n=8). Postoperative radiotherapy was administered in six cases. Clinical data and surgery efficacy were analyzed via chart review.

RESULTS: Eleven patients were disease-free during their follow-up period, whereas one patient experienced recurrence without metastasis. Radicular pain nearly disappeared, and patients suffering from spinal cord compression recovered well. Local recurrence was detected in one-fourth (25%) of the cases that underwent subtotal resection and was not detected in any of the cases involving total spondylectomy.

CONCLUSIONS: Local recurrence of DF is not uncommon after insufficient removal. Therefore, total excision, while also preserving neural function, is recommended. In our study, patients who underwent a total spondylectomy had significantly lower local recurrence rates for DF in the spine. Radiotherapy may be an acceptable alternative therapy, whereas en bloc resection has the potential to result in significant functional impairment. © 2014 Elsevier Inc. All rights reserved.

Keywords: Desmoplastic fibroma; Spine; Case series; Outcomes; En bloc resection; Total spondylectomy; Radiotherapy

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Introduction

Desmoplastic fibroma (DF) is a rare benign neoplasm of the bone that currently accounts for 0.06% of all bone tumors and 0.3% of benign bone tumors [1–4]. In 2002, the World Health Organization defined DF as a rare, benign bone tumor composed of spindle cells with minimal cytologic atypia and abundant collagen production [5]. Until Jaffe [6] differentiated it from other bone fibromas and established DF as a specific term in 1958, DFs had previously been referred to as desmoid tumors, ligament fibromas, or aggressive fibromas.

Although it can present in any bone, DF is often localized to the mandible or the metaphysis of long bones. Specifically, DF usually arises in the mandible, femur, humerus, or pelvis [1–3] and rarely involves the calvaria or spine. Most spinal DF tumors have been located in the thoracic or lumbar level [7–12]. Epidural DFs are particularly rare, and there are no published cases of an epidural spinal cord tumor.

Although DF is a slow-growing benign tumor, it exhibits locally aggressive and infiltrative growth. For example, in a study by Böhm et al. [2], soft-tissue infiltration was detected in 48% of the patients. To our knowledge, there are no reports of metastases associated with bone DF in the literature. Because of the infiltration exhibited by DF in many cases, complete excision of the tumor leads to high rates of local recurrence [2]. Radical excision is considered the best treatment option, however complete resection is difficult to achieve in the spine. In this series, a retrospective review of 12 cases with DF in the spine that were treated with surgery at our center was performed, and to our knowledge, represents the largest cohort reported to date.

Patients and methods

Epidemiology

A total of 12 patients with DF in the spine were analyzed in this study (seven men and five women). At diagnosis, the patients’ ages ranged from 15 to 77 years (mean, 37.5 years), with 8 of 12 (66.7%) patients’ ages ranging from 15 to 40 years. Lesions were detected in the cervical spine (n=5), thoracic spine (n=3), lumbar spine (n=3), and the sacrum (n=1). Moreover, tumors involved one vertebral level in three cases, two vertebral levels in five cases, three vertebral levels in three cases, and six vertebral levels in one case (Table). For most cases, chronic endurable pain in the spine was the most consistent complaint, and the duration of preoperative atypical symptoms ranged from 3 to 96 months (mean, 18.9 months). Additional patient characteristics included three patients had a previous spinal injury; four patients presented with chronic pain; two patients presented with a palpable mass; and seven patients had varying degrees of cord compression at diagnosis. For all 12 patients, neurologic status was classified according to the Frankel scoring system.

Radiologic studies

Radiographic features of the tumors analyzed included geographic bone destruction, a narrow zone of transition, pseudotrabeculation, and bone expansion. In addition, tumors were not observed in the intervertebral spaces in the radiographs obtained. Although no pathologic fractures or dislocations were found, some patients exhibited characteristics of marginal sclerosis and cortical breakthrough. Computed tomography (CT) images also showed well-defined, expansile lytic lesions with a soap-bubble appearance and thinning of the bone cortex with little or no sclerotic reactions. T2-weighted magnetic resonance imaging (MRI) also showed low or intermediate signal intensity foci because of collagen, inhomogeneous areas of contrast enhancement, and higher signal areas associated with cellular areas.

Staging

Tumor extension was described according to the Weinstein-Boriani-Biagini (WBB) classification criteria (except for one case in which the Enneking grading system was used) based on CT and MRI. Extraosseous paravertebral (layer A) involvement was found in all cases, whereas lesions only involving the anterior column (zones 4–9) were identified in three cases. In contrast, 6 of 12 (50%) cases, including the sacrum DF case, involved tumor extension into both the anterior and posterior columns. Moreover, in only 3 of 12 patients (25%) the lesion was located in the posterior element (eg, sectors 10–12 and 1–3). Tumor involvement in the epidural space (layer D) was observed in all cases.

Treatment history

Of the 12 cases analyzed, only one patient (Case #3) had already been subjected to incomplete tumor resection.

EVIDENCE & METHODS

Context

DF is a benign locally aggressive tumor that rarely involves the spine. The authors present their experience.

Contribution

In this case series of 12 patients, total excision and subtotal resection with radiotherapy were found to be treatments associated with good outcomes. Only one recurrence was noted.

Implications

The findings suggest that total excision, if safe, is the best approach. However, if the risks associated with this aggressive surgery are too great, then subtotal resection with radiotherapy is reasonable, although the answer is best assessed by whether recurrences locally cause significant functional problems.

—The Editors
without adjunct therapy at another institution. The patient was admitted to our center because of tumor recurrence. The remaining 11 patients in this series were regarded as “intact” cases because they did not receive surgical intervention, radiation therapy, or any other treatment before the surgical treatment applied at our institution.

**Treatment**

Four patients received needle biopsies that led to the diagnosis of DF before admission to our center. Although we had communicated the importance of biopsies and suggested that they be performed to the other eight patients, they chose to have an intraoperative fast pathology examination only and not be subjected to a biopsy because of fears of possible nerve damage. Intraoperative fast pathologic examination was performed in all 12 cases. Depending on the location and extension of the tumor detected in each case, the WBB system was applied to select a treatment protocol. In four cases, curettage was performed, whereas total spondylectomy was performed in the remaining eight cases (using an anterior, posterior, or combined approach) (Table). En bloc removal was performed in one case where the tumor only involved the anterior element of L4.

**Reconstruction**

For lesions in C3 and below, the vertebral body was replaced by a purely autologous iliac crest or combined with an anterior titanium plate. In 11 cases, a screw-rod system was used in combination with autologous or artificial bone grafts to reconstruct the stability of the spine. However, one patient did undergo a total resection of tumor without fixation. Clinical summaries of each patient are provided in the Table and example images of reconstructed spines are shown in Figs. 1 and 2.

**Pathology**

A histology-based diagnosis was obtained in all cases. In addition, for most cases, margin tissues (eg, gross normal surrounding tissue) were submitted for histologic study to determine if further treatments were needed. A pathologic specialist confirmed the diagnoses (Fig. 3).

**Radiotherapy**

Patients were referred to the radiotherapy center if the tumor margin analysis revealed residual disease or if the disease-free margins were uncertain. Radiotherapy was advised as an adjunct therapy to address surgical contamination associated with a procedure involving piecemeal curettage in cases prescribed total spondylectomy. There were six patients who did not receive postoperative radiotherapy, including five patients ranging in age from 15 to 27 years and another patient who underwent en bloc resection. In contrast, the other six patients received local conventional radiotherapy 4 to 6 weeks after surgery, and the total dose administered over 20 applications ranged from 30 to 50 Gy.

**Follow-up evaluations**

The mean follow-up period was 48.8 months (range, 24–91 months). All 12 cases included radiograph, CT or MRI of the surgical segments involved, and adjacent vertebrae. Imaging was performed 3, 6, and 12 months after surgery, every 6 months for the next 2 years, and then annually for life. Follow-up data were also obtained from office visits and telephone interviews. Our criteria for spinal fusion and

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### Table

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<th>Case no.</th>
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<th>LC</th>
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<th>F-S pre</th>
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<th>F-S post</th>
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<th>LR (mo)</th>
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DF, desmoplastic fibroma; M, male; F, female; LC, location; OCF, occipitocervical fixation; AP, anterior plating; TM, titanium mesh; TR, tumor resection; PF, posterior fixation; F-S, Frankel score; Subtotal, subtotal resection; Total, total spondylectomy; pre, preoperation; post, postoperation; AT, adjunctive therapy; RT, radiotherapy; LR, local recurrence; FU, follow-up; NED, no evidence of disease; AWD, alive with disease; τ, mean.

* Weinstein-Boriani-Biagini staging was used in all cases, except Case 5 for which Enneking staging was used.  
† Only case with an operation treatment history.
stability were the following: (1) a clear through trabeculae between the host bone and vertebral bodies; (2) no bright belt around the host bone; and (3) no obvious instability of the fused segments apparent in dynamic radiographs. In addition, neural function was reevaluated 3 months after surgery according to the Frankel score system (Table).

Results

Intraoperative fast pathology examination yielded results consistent with a final diagnosis of DF in 9 of 12 cases. All the tumors were subjected to pathology studies after surgical resection, and all were diagnosed as DF at that time. Intraoperative blood loss ranged from 400 to 3000 mL (mean, ~1,100 mL). The rate of fusion for bone grafts was 100%, and there were no observed incidences of spine instability. None of the patients died of surgical complications shortly after surgery. In one case, a lesion present in an anterior element of L4 was treated with en bloc resection without adjunct therapy, and no relapse was detected during the follow-up.

Neurologic status

For seven patients diagnosed with spinal cord compression before surgery, their pain was mostly absent by their 3-month follow-up visit and limb numbness had improved.
to varying degrees. These patients also experienced a decrease in Frankel scores of 1 to 2 grades. Moreover, the neurologic status of all 12 cases did not worsen in the early postsurgical period.

Local control and survival

The patients in this series underwent observation periods ranging from 24 to 91 months (mean, 48.75 months). Local recurrence was only detected in 1 of 12 (8.3%) patients. This patient had undergone curettage and then experienced a relapse 24 months after surgery. Total resection was performed in eight patients, and they did not experience recurrence, metastasis events, or death (Table). For six patients, radiotherapy was administered after surgery, and none of these patients experienced radiation myelopathy or sarcomatous transformation.

Evolution of the recurrent case

One patient (Case 5) was found to have recurrence on an MRI examination conducted 2 years after a surgical tumor resection (Fig. 1). The patient had no obvious pain and no spine instability was apparent in the patient’s radiographs or CT scans. We advised him to undergo a second tumor resection but he refused. At the patient’s most recent follow-up (45 months), he was asymptomatic despite the presence of the tumor.

Discussion

In the present series, there were five cases of DF in the cervical spine, three in the thoracic spine, three in the lumbar spine, and one in the sacrum. Our cohort included an

Fig. 2. Images of the spine of a 15-year-old girl (Case #4). (A and B) The magnetic resonance image (MRI) of the tumor showed the destruction of vertebral bodies and the left accessories of C6 and C7. (C) Preoperative computed tomography (CT) also showed the tumor was located in the vertebral bodies and left accessories of C6 and C7. (D) Postoperative lateral radiograph showed the reconstruction performed from C5 to T1. (E) MRI performed 3 years after surgery and no signs of recurrence were observed.

Fig. 3. Histopathology of a DF tumor (Case #4). The tumor was characterized by interlacing fascicles with low fibroblastic cellularity. No mitotic activity was evident (hematoxylin and eosin staining; 120×).
unusually broad age demographic (range, 15–77 years; mean, 37.5 years). Typically, DF develops during the second and third decades of life, with nearly 75% of the cases occurring before the age of 30 [7,13].

Gender predilection and etiology of DF remain elusive, however, the most common clinical characteristics of DF include pain or swelling that is experienced over a long period, resulting in functional disability. In contrast, pathologic fractures are a rare symptom at the initial presentation of DF [2,14,15]. For the patients in this study, males and females were similarly represented and our cases were heterogeneous in terms of history and etiology. The duration of the preoperative atypical symptoms was very broad (3–96 months; mean, 18.9 months), and the symptom presentation was variable: three patients had a history of a spine injury, four presented with radicular pain, two patients presented with a palpable mass, and five patients exhibited some degree of cord compression.

Of the 12 patients in our case series, nine (75%) had multilevel involvement and none had intervertebral space involvement. We have reviewed 16 cases with spinal DF from 1963 to 2011 and found that none of these previous reports included any cases with either intervertebral space involvement or multiple interval lesions [8–11,15–22]. We could not find information on the mechanism by which multiple levels become involved without intervertebral space involvement in the literature, hence we analyzed all the cases in the studies reviewed to develop some clarification of the spread patterns. We found that tumors infringing on the posterior element were able to invade an adjacent segment without encountering an intervertebral disc [18,20], whereas tumors involving a vertebral body could contact an adjacent vertebral body through intraspinal growth and across the intervertebral disc [19,21].

The radiographic appearance of DF is nonspecific and somewhat similar to that of other lytic bone lesions. As a result, a wide range of radiologic differential diagnoses have been reported in literature [2], including nonossifying fibromas, giant cell tumors, unicameral bone cysts, aneurysmal bone cysts, fibrous dysplasia, fibrosarcoma, osteosarcoma, and metastases. In the present series, thinning of the overlying bony cortex in the absence of a periosteal reaction was common, and in most cases, a significant T2 shortening was observed through MRI.

There is no doubt that a preoperative biopsy can facilitate surgery protocol formulation. Thus, we find it regrettable when patients forego it based on fear of possible nerve damage. The recommended treatment for DF is tumor excision and restoration of spinal stability. In the present case series, surgical treatment resulted in a significant decrease in pain 3 months after surgery and limb numbness was relieved to varying degrees in 7 of 12 patients who were diagnosed with spine instability and spinal cord compression before surgery. Moreover, for these seven patients the Frankel scores decreased by 1 to 2 grades after surgery.

Because of the local infiltration associated with DF, local recurrence rates can be high if surgical resection is incomplete [3]. Correspondingly, local recurrence for cases of DF after intralesional curettage ranges from 20% to 72% [14,23]. Particularly in the spine, aggressive surgical treatment is needed to achieve local tumor control of DF. However, anatomical constraints associated with the spine limit the extent of radical excision that can be performed. This is particularly relevant when a tumor is detected in its later stages. From an anatomical point of view, the vertebral body, vertebral arch, transverse process, articular process, vertebral plate, and spinous process comprise a relative compartment. Therefore, a total spondylectomy is consistent with the recommended complete excision of a DF tumor. In the present series, curettage or total spondylectomy were performed depending on the location and extent of the tumor evaluated according to the WBB system. Although only a limited number of cases were analyzed, it was evident that patients who underwent total spondylectomy had a lower local recurrence rate and a delayed onset of recurrence.

With surgical and internal fixation techniques rapidly improving, the use of total spondylectomies for the treatment of nonmalignant, yet aggressive, tumors has increased, as well as for malignant tumors of the spine. In our retrospective review of these cases, it appears that limited application of this technique led to a high rate of tumor recurrence. For example, for a patient who was treated at other hospitals for a single vertebral lesion, MRI detected recurrence of the lesion in the original segment and in the lower adjacent vertebrae and surrounding soft tissue. As a result, surgical resection and reconstruction of the spine was more difficult. Based on our experience with surgical treatments for DF in the spine, we have found that the initial surgical intervention represents the best opportunity to successfully treat the disease. Correspondingly, treatment of recurrences, even very aggressive treatments, has been found to be unsuccessful in a long term. Therefore, DF in the spine should be locally controlled by excising as much of the tumor as possible, while still preserving the nerve function.

In recent years, the en bloc technique for tumor excision in the spine has been reported [24–28]. En bloc has been shown to be an improved method for the surgical intervention of tumors of the spine. Unfortunately, however, surgical revision and the presence of a soft-tissue mass constitute significant surgical challenges and make en bloc excision much more difficult. For the total spondylectomies performed in the present series, extracapsular piecemeal excision was performed using a posterior approach, or a combined anterior and posterior approach, to remove not only the whole tumor mass, but also as much of the surrounding tissue as possible (Figs. 1 and 2). Radiotherapy was then recommended as an adjunct therapy for these patients, and a satisfactory rate of long-term local control was achieved.
There is some dispute regarding the application of radiotherapy for the treatment of spinal DF because of the possibility of uncertain effects, radiation myelopathy, or radiation-induced development of sarcomas. While some researchers hypothesize that radiation therapy is of no benefit for this type of tumor, others hypothesize that radiation can control symptoms and inhibit DF growth [29]. In a study by Nag et al. [29], DF in the femur of a 28-year-old woman was treated with radiotherapy alone and a substantial decrease in analgesic use and an absence of disease progression were reported at a 3-year follow-up evaluation. In cases involving total spondylectomy, the potential for contamination, or if the margin is intralesional, leads to a recommendation for administering adjunct therapy. In the present series, 6 of 12 patients (including those older than 25 years with children who underwent total spondylectomy) received radiotherapy after surgery and none presented with radiation myelopathy or malignant transformation. However, because of the small number of total spondylectomy cases included in this study, statistical conclusions regarding local recurrence rates associated with postoperative radiotherapy could not be established. However, we hypothesize that radiotherapy should be regarded as a beneficial supplement after the removal of a gross mass.

Conclusion

Desmoplastic fibroma is a benign, yet locally aggressive, tumor of the connective tissue. Histologic confirmation of the diagnosis is required, and complete excision of the tumor is the optimal treatment. Although DF may be locally aggressive, complete excision can be curative, and subsequent reconstruction to achieve spinal stability should be performed. Subtotal excision with postoperative radiotherapy can be an acceptable alternative therapy when en bloc resection is not feasible. Nevertheless, we still recommend en bloc excision as the preferred curative treatment whenever it is technically possible. If local recurrence does occur, it can be very difficult to treat. However, because of the limited number of cases included in this retrospective analysis, a larger cohort and further studies are still needed to provide additional insight into this challenging disease.

References