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Clinical Study

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.

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

FDA device/drug status: Not applicable.

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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, incomplete 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.

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

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