Inaccurate Histological Documentation of Massive Desmoplastic Fibroblastoma With Scapular Invasion

To the Editor:

We read with interest the article “Massive desmoplastic fibroblastoma with scapular invasion” (http://www.orthosuper-site.com/view.aspx?rid=66666) in the August 2010 issue of ORTHOPEDICS. Desmoplastic fibroblastomas are rare fibrous soft tissue tumors that usually arise in subcutaneous tissue or skeletal muscle at a variety of anatomic sites. The authors describe an unusual case of desmoplastic fibroblastoma that presented with a massive 23-cm tumor. The tumor was also unique for its infiltration and destruction of the scapula. The aggressive clinical features prompted the original physicians to administer chemotherapy, but the tumor exhibited no response to systemic treatment. The patient eventually underwent limb-sparing surgery, which included en bloc resection, complete scapulectomy, and osteoarticular allograft replacement. The authors believe that the invasiveness of the tumor and its large size are distinctly unusual for desmoplastic fibroblastoma. Following surgical excision, the patient has remained continuously disease free for 5 years, which is in keeping with the intrinsically benign nature of the tumor.

This case demonstrates that desmoplastic fibroblastoma can occasionally reach an enormous size and may exhibit invasive characteristics, but this does not necessarily portend subsequent recurrence of disease. The microscopic features were notable for a hypocellular tumor composed of reactive-appearing stellate and spindle-shaped fibroblasts in a dense collagenous background. No mitotic figures were seen, and there were few vessels within the tumor. Tumor necrosis was negligible.

The combination of clinical, histopathological, and immunohistochemical features usually allow distinction of immunohistochemical from other cutaneous or soft tissue tumors. Adequate sampling is essential in the diagnostic differentiation. In the benign category, consideration must be given to nodular fasciitis, fibroma of tendon sheath, nuchal fibroma, sclerotic fibroma of skin (as seen in Cowden’s disease), calcifying fibrous pseudotumor, neurofibroma, and myxoma. For example, with skeletal muscle infiltration, a locally aggressive tumor such as fibromatosis might be entertained, and for large lesions one might contemplate the possibility of a low-grade fibromyxoid sarcoma (grade I fibromyxosarcoma). Histochemical studies, adequate sampling, and histological illustrations allow conclusive distinction between fibromatosis and grade I fibromyxosarcoma.

Merriman et al did not report the number of tissue blocks and the immunohistochemical features of the tumor; moreover, only 2 histological illustrations are present in the article. The magnification of Figure 4A does not permit observation of the cytological features of the lesion. Figure 4B is insufficient to appreciate the proliferative pattern of the enormous tumor measuring 23×13×11 cm. The immunophenotype of desmoplastic fibroblastoma is well known. The tumor cells are positive for vimentin and are variably positive for alpha-smooth muscle actin and occasionally for keratins AE1/AE3. They are negative for desmin, EMA, S100 protein, and CD34.

According to Miettinen and Fetsch,1 fibromatoses have greater infiltrative potential and are usually more cellular than desmoplastic fibroblastoma. They feature spindled cells with a distinct tendency to grow moderately to heavily collagenized fascicles, and they often contain a fairly uniform vascular pattern with mildly dilated vessels. Stellate-shaped fibroblasts may be seen in a fibromatosis. The tumor cells are positive for alpha-smooth muscle actin, factor XIIIa, CD34, and vimentin.2 Low-grade fibromyxoid sarcoma (grade I myxofibrosarcoma) has greater cellularity, more mitotic activity, and increased vascularity. Also, low-grade fibromyxoid sarcoma has a tendency to contain areas with a swirled growth pattern. Although cytologic atypia is not a pronounced feature, it is nonetheless usually evident. With immunohistochemical staining, the stromal cells are most consistently positive for vimentin,3-6 and there are sporadic reports of positivity for CD34,5 actin, and desmin.4 Tumor cells are typically negative for epithelial, neural, histiocytic, melanocytic, or vascular markers.3,5,6
We believe that the histological diagnosis of desmoplastic fibroblastoma is inaccurate and is unacceptable because fibromatosis and grade I myxofibrosarcoma cannot be excluded.

Teresa Pusiol, MD
Doriana Morichetti, MD
Maria Grazia Zorzi, MD
Trento, Italy

REFERENCES


Reply:

We appreciate the interest in our case report of a massive desmoplastic fibroblastoma. Some of the points raised in the letter highlight the limitations of the case report format. Unfortunately, we were only able to include 2 photomicrographs of this large tumor, so as to incorporate other illustrations of interest, such as imaging studies and patient and gross photographs. As to the question of sampling, 1 section per centimeter of tumor was taken as per the protocol of our institution. The tumor was more cellular than typical for desmoplastic fibroblastoma but was still composed of the stellate and spindle cells in a collagenous background that is usually seen in desmoplastic fibroblastoma.

Desmoid tumor was considered in the differential diagnosis; however, we felt this was not the best diagnosis, because desmoid tumors are even more cellular than the tumor reported. Furthermore, desmoid tumors tend to be arranged in bundles or fascicles and lack stellate cells. Low-grade fibromyxoid sarcoma has a whorled pattern and myxoid background that was absent in this tumor.

As an aside, Dr Harry Evans, our colleague who first described desmoplastic fibroblastoma and low-grade fibromyxoid sarcoma, believes that low-grade fibromyxoid sarcoma is distinct from myxofibrosarcoma (grade 1). This view is supported by the presence of a translocation involving the FUS gene in low-grade fibromyxoid sarcoma.

As Dr Pusiol et al describe in their letter, immunohistochemical markers can often be helpful in confirming the diagnosis, but in our particular case, they cannot definitely distinguish between the tumors that were being considered in the differential diagnosis.

Patrick P. Lin, MD
Michael T. Deavers, MD
David J. Merriman, MD
Bogdan A. Czerniak, MD
Houston, TX
doi: 10.3928/01477447-20110922-01

Identifying Multiple Reports From the Same Study

To the Editor:

We read with interest the article “A systematic review assessing the effectiveness of alendronate in reducing peri-prosthetic bone loss after cementless primary THA” (http://www.orthosupersite.com/view.aspx?rid=81547) in the April 2011 issue of ORTHOPEDICS. Two studies referenced in the article likely reported the same study (Tapaninen et al1 and Venesmaa et al2). We compared the 2 articles strictly following the instructions of the Cochrane Handbook for Systematic Reviews of Interventions (Table).3 The articles shared a group of authors who performed the same intervention in the same institution. The number of patients and the study period were also similar. The possibilities that 1 article reanalyzed another’s results of the same study with a longer follow-up might be considered.

Duplicate publication can introduce substantial biases if studies are inadvertently included more than once in a meta-analysis or systematic review.4 It can be difficult to detect duplicate publication when we are selecting studies for a systematic review or meta-analysis, and uncertainties may remain even after careful checking. In this case, corresponding with the authors of the reports for identification may be necessary. If this attempt is unsuccessful, the data from both studies might be conservatively considered as 1 trial for reporting purposes based on the similarities in the 2 articles.

Tiao Lin, MD
Shi-gui Yan, MD
Cong Wang, MD
Xun-zi Cai, MD
Hangzhou, China
REFERENCES


Reply:

We thank Dr Lin et al for their letter. We checked the 2 referenced reports carefully. Although the 2 articles are from 1 trial, the problem cannot be fully attributed to substantial biases. Venesmaa et al reported the short-term results of alendronate treatment in preventing periprosthetic bone loss after total hip arthroplasty, and Tapaninen et al reported the long-term results. We combined the short- and long-term results of these 2 trials, but we did not reanalyze the results as 1 trial; rather, we included the 2 papers separately, which was inappropriate. We should have pointed out that these 2 papers came from 1 trial and combined the results. However, the results and conclusions of this systematic review were not influenced by not doing this. We apologize for the inconvenience caused.

Shen Bin, MD
Chengdu, P.R. China
doi: 10.3928/01477447-20110922-02

54-year Follow-up of Lumbar Posterior Fusion With Tibial Graft

To the Editor:

We would like to briefly report on a recent follow-up of a lumbar posterior fusion procedure performed in 1954.

A 69-year-old woman presented for evaluation of chronic neck, shoulder, and arm pain of moderate intensity. The pain was exacerbated by an incident approximately 3 months prior in which the patient was roughly bumped into a bus seat; she did not seek immediate treatment afterward. The patient reported treating her discomfort with vitamins and some marijuana use prior to her visit.

Physical examination revealed an incidental surgical incision in the anterior abdominal area. The patient reported suffering from chronic back pain as a child for many years, most likely due to spondyloysis of the L5 vertebra that eventually caused spondylolisthesis. At age 16, she underwent a corrective L5/S1 posterior fusion with anterior approach and tibial graft. At the time of presentation, her graft was estimated to be 54 years old.
On graft condition assessment, anteroposterior and lateral radiographs (Figures A, B) revealed no instrumentation and an intact solid fusion of L5/S1, a finding supported by sagittal and L5-level axial magnetic resonance imaging (Figures C, D). Mild degenerative change was noted in the area with nothing particularly abnormal given her age. Cervical imaging revealed multilevel disk degeneration with no associated myelopathy, and the patient was advised to avoid fall risks, continue symptomatic treatment for her discomfort, and remain active.

Spondylolysis is a vertebral defect of the pars interarticularis that occurs in approximately 6% of the general population and is the most common cause of spondylolisthesis. Severe and refractory cases of spondylolisthesis are generally surgically treated, with fusion procedures considered as the gold standard. More than 300,000 lumbar spine fusion procedures were performed in the United States in 2004, and approximately 2.2 million bone graft procedures are performed annually worldwide. The modern orthopedist has a variety of graft materials, instrumentation, approaches, and techniques to consider when preparing for a spinal fusion procedure.

This case contributes to the literature by demonstrating the potential longevity of bone graft material in fusion surgery, and we believe it is the oldest intact posterior fusion without instrumentation described. Of particular note is the lack of lumbar symptomatology and significant lumbar degeneration. Given the growing body of evidence that fusion alters biomechanical stresses on adjacent segments, it is interesting that the graft did not progress to any major degree of adjacent segment disease over >50 years of this patient’s life.

Michael X. Liu, BA
David M. Cai, MS
Patrick J. Connolly, MD
Mark S. Eskander, MD
Worcester, MA

doi: 10.3928/01477447-20110922-03

REFERENCES