

Massive rare desmoplastic fibroma of the ilium and ischium in a young adult

A case report

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Abstract

Rationale: Desmoplastic fibroma (DF) is a rare, locally invasive but benign bone tumor. It represents one of the rarest bone diseases, with an incidence of only 0.11% of all primary bone tumors.

Patient concerns: Herein, a case of massive and unusual DF, with simultaneous involvement of ilium and ischium, is described. A 29-year-old man suffered minor pain in his right hip for 2 years. It worsened after sudden movements, which prevented him from walking normally. Physical examination showed a limitation when the right hip was flexed and a percussion pain on the hip region. A medical imaging examination showed that the right ilium and ischium had a massive bone lesion. The top of acetabular had very little bone left and a fracture was likely at any time. No prominent body weight loss was noted, because there was no extensive invasion to the adjacent soft tissue.

Diagnoses: DF of the Ilium and Ischium.

Interventions: The patient underwent a surgery involving curettage and grafting to maintain the stability of the pelvis.

Outcomes: The definitive pathological diagnosis was DF, without evidence of malignancy. The postoperative recovery course at 3-month follow-up was uneventful.

Lessons: To the authors' knowledge, such a massive DF involving both ilium and ischium has been rarely reported. Young patients require appropriate and timely treatment modalities.

Abbreviations: CT = computed tomography, DF = desmoplastic fibroma, MRI = magnetic resonance imaging.

Keywords: desmoplastic fibroma, ilium, ischium

1. Introduction

Desmoplastic fibroma (DF) of the bone is a rare, lytic, locally aggressive but nonmetastatic tumor that was first described by Jaffe^[1] in 1958. It belongs to the fibroblastic/myofibroblastic tumor group according to the 2013 World Health Organization Classification of Soft Tissue Tumors.^[2] It represents one of the rarest bone diseases, with an incidence of only 0.11%^[3] of all primary bone tumors. DF can theoretically affect any bone, but the majority of cases occur in the metaphysis of long bones (56%)

and the mandible (22%).^[4] To the best of our knowledge, this is the first case report of DF involving the ilium and ischium that was treated with curettage and grafting.

1.1. Ethical statement and consent

This study was approved by the Ethics Committee of Second Affiliated Hospital of Harbin Medical University. The clinical and imaging data were obtained with the patient's signature for use in the publication of scientific research papers.

2. Case presentation

A 29-year-old man felt occasional minor pain deep in the right hip region. The patient had first noted this pain approximately 2 years ago, but the claudication due to pain while walking was felt about 2 months ago.

The patient had no history of trauma. He once presented to the local physician. X-ray showed lesions in the right ilium and ischium (Fig. 1A), but a definite diagnosis was not made. At the current presentation, his pain was accompanied by a change in posture (from sitting or lying to standing) and did not radiate to other regions. On routine physical examination, his gait pattern was claudication. There was a limitation when the right hip was flexed and a percussion pain on the hip region. Neurovascular examination of the right lower extremity was within normal limits, with no lymphadenopathy. Laboratory findings were within normal limits. His medical history was unremarkable.

Editor: Niles Talele.

The authors have no conflicts of interest to disclose.

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Medicine (2017) 96:48(e8962)

Received: 5 July 2017 / Received in final form: 3 November 2017 / Accepted: 8 November 2017

<http://dx.doi.org/10.1097/MD.0000000000008962>



Figure 1. A, Oval on X-ray showed lesions in the right ilium and ischium. B, The area noted on CT showed an osteolytic lesion of the right iliac bone. C, The area noted on CT showed an osteolytic lesion of the ischial bone. CT = computed tomography, MRI = magnetic resonance imaging.

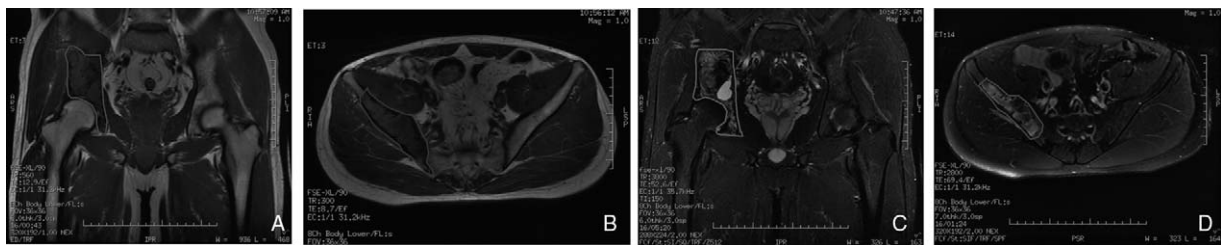


Figure 2. A, The mass showed in T1-weighted coronary MRI. B, The mass showed in T1-weighted transverse MRI. C, The mass showed in T2-weighted coronary MRI. D, The mass showed in T2-weighted transverse MRI. CT = computed tomography, MRI = magnetic resonance imaging.

Computed tomography (CT) scan demonstrated an osteolytic lesion of the right iliac bone and ischial bone with cortical breakthrough, marginal sclerosis, slight pseudotrabeulation, and bone expansion (Fig. 1B and C). T1-weighted magnetic resonance imaging (MRI) revealed an irregular mass with low signal intensity (Fig. 2A and B). T2-weighted MRI with fat suppression revealed high signal intensity (Fig. 2C and D). Gross inspection showed a 10×10×5 cm³ tumor in the right ischium and ilium.

The surgical treatment included curettage and grafting. We performed the Smith-Peterson approach (Direct anterior approach) combined with Moore (Posterior approach), which could sufficiently expose the focus so that the pathological tissue could be completely removed. The tumor was removed in

several parts. The tissue was beige colored, with hard and soft areas (Fig. 3A). An intraoperative frozen section analysis showed no signs of malignancy. The lesion was filled with allograft spongy bone. After proper hemostasis, the wound was closed with 2 drainage tubes. The wound completely healed in 2 weeks.

Microscopically, the tumor was composed of scattered spindle cells in an abundant collagenous stroma (Fig. 3B). Neither cellular atypia nor mitotic figures were observed.

Immunohistochemically, β-catenin was positive (Fig. 3C); while SMA, Desmin, S-100 and CK were all negative; the proliferation marker Ki-67 was <1%. The postoperative course was uneventful, and the patient is doing well without local recurrence 3 months after the surgery.

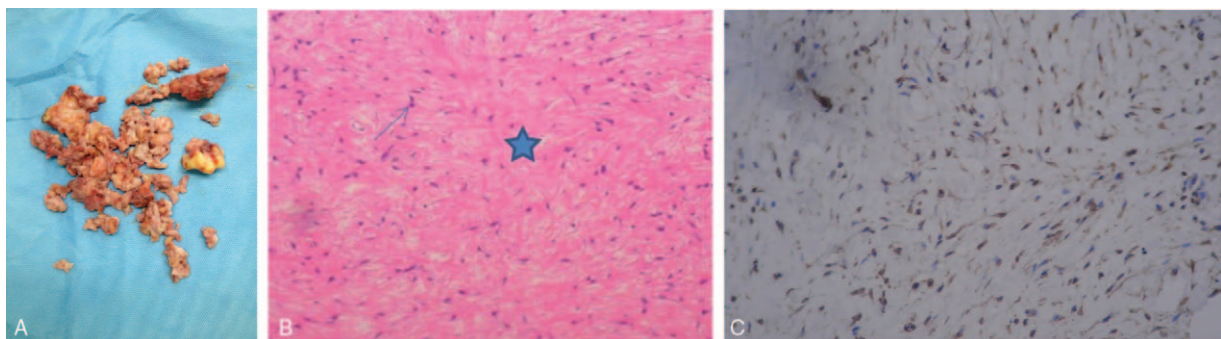


Figure 3. A, The tissue was beige in color with hard and soft areas of consistency. B, The tumor was composed of scattered spindle cells in an abundant collagenous stroma. C, Immunohistochemistry staining results turn out that β-Catenin is positive.

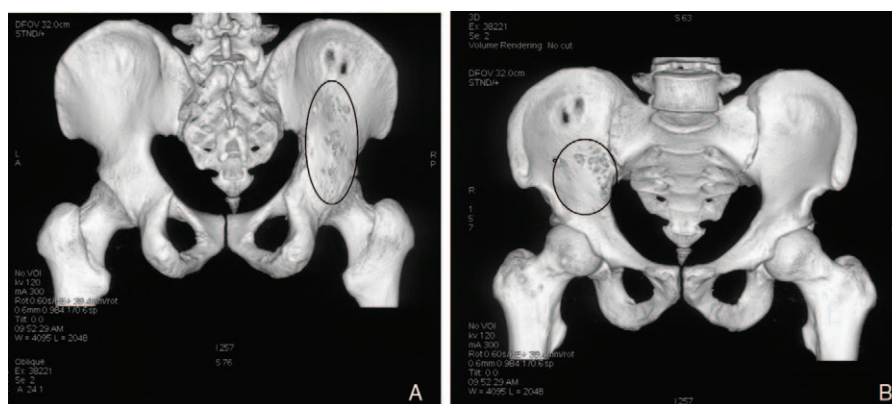


Figure 4. A, 3D-CT for posterior aspect of pelvis. B, 3D-CT for anterior aspect of pelvis. 3D = three dimensional, CT = computed tomography.

3. Discussion

Desmoplastic fibroma (DF) is a benign bone tumor that is locally invasive. It usually occurs in young patients within the first 3 decades of life. The pathogenesis of DF remains unclear. By cytogenetic analysis, some cases of DF revealed a t(2;11)(q31;q12) reciprocal translocation^[5], which would result in fusion genes and encode novel fusion proteins or a normal but ectopically expressed protein. The translocation may partially serve as a useful diagnostic adjunct to distinguish DF from desmoid tumor (which closely resembles DF, but tends to be more cellular), but was not performed for our patient. The preoperative CT scans showed cortical breakthrough in several areas, indicating bone instability. In addition, a three-dimensional CT scan was helpful to determine the stability of the pelvis. No prominent fracture was demonstrated (Fig. 4A and B). While CT best illustrates the extent of bone destruction, MRI better visualizes the medullary as well as soft tissue extension of the tumor, and is the “gold standard” to define the local extent of the tumor before surgery.^[6–8] In the current case, the MRI showed no isolated extraosseous tumor growth or intraosseous lesions, with strong evidence of extension in adjacent soft tissue. Unlike DF, desmoid tumors, also called aggressive fibromatosis, can be intra-abdominally or extra-abdominally localized. However, it is difficult to distinguish DF from many other tumors by imaging studies alone. Further diagnostic tests are required for differential diagnosis. The histological examination of the tissue may exclude some types. Fibrosarcoma presents with more atypical cells and less collagen fibers. Nonossifying fibroma presents with fibers and system tissue cells, such as monocytes, foam cells, and multinucleated giant cells. Immunohistochemistry on the patient’s tumor tissue revealed high expression of β -catenin. Hauben et al^[9] showed that all 13 cases of DF studied expressed β -catenin, which is in accordance with the current case. Unlike DF, fibromatosis and grade-I myxofibrosarcoma cells are positive for vimentin. High β -catenin and vimentin typically indicate mesenchymal lineage, but are also seen during epithelial mesenchymal transition in some aggressive tumors. β -catenin participates in the canonical Wnt signaling pathway, which influences several physiological processes, such as embryonic development and energy metabolism, and leads to tumor growth, progression and metastasis. Higher β -catenin protein level could mediate activation of transcription and expression of related genes, which would be potential drivers of cancer.

This is an unusual case because of the low incidence of DF and its simultaneous involvement of 2 regions as well as for the individualized treatment, which involved excision, curettage, grafting, and radiotherapy. Sanfilippo et al and Nag et al reported radiotherapy for DF in the ilium and in the distal end of the femur, respectively.^[10,11] Both the patients from the above-mentioned studies are now free of pain. Goyal et al^[12] reported a large DF in the right ulna of a 15-year-old male. The tumor was excised with a wide margin, and the bony defect was reconstructed with nonvascular autologous fibular graft. However, in this case, the critical issue was that the massive tumor had nearly damaged the frame of the pelvis. Gebhardt et al^[13] performed marginal resection with hemipelvic allograft to treat a patient whose pelvis suffered extensive invasion. The current case had comparatively smaller lesions but required immediate operation since a fracture could occur at any time. Fortunately, the patient sought treatment in a timely manner; curettage and allogenic grafting were thus sufficient and feasible, leaving the adjacent soft tissue intact. Given that DF has a high recurrence rate after curettage and grafting, a close postoperative observation is essential.

4. Conclusion

Massive DF involving 2 locations is rare. Young patients should be provided appropriate modalities, such as excision, curettage, grafting, radiotherapy, and follow-up, in a timely fashion.

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