Journal of Bone Oncology 4 (2015) 103–106

Contents lists available at ScienceDirect

Journal of Bone Oncology

journal homepage: www.elsevier.com/locate/jbo

The chordoma arised from ilium: A rare case report

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ARTICLE INFO

Article history:
Received 20 July 2015
Received in revised form
26 September 2015
Accepted 26 September 2015
Available online 1 October 2015

Keywords:
Chordoma
Ilium
Atypical site
Diagnosis
Treatment

ABSTRACT

Chordomas are malignant tumors that originate in embryonic notochordal remnants. The sacrum and skull are the most common sites; the mobile spine and other bones are extremely rare sites. We describe a 45-year-old man who presented with a lytic lesion in his left ilium. Imaging and pathology of a biopsy specimen suggested a malignant bone tumor; wide resection was accordingly performed. The morphology and immunohistochemistry of the operative specimen showed obvious characteristics of classic chordoma. To our knowledge, this is the first reported case of a chordoma originating in the ilium. Chordoma should therefore be considered in the differential diagnosis of lytic lesions in the ilium.

1. Background

Chordomas, rare malignant tumors that originate in embryonic notochordal remnants, account for 1–4% of primary malignant bone tumors [1]. Their origin from notochordal cells was first reported by Muller in 1858; the tumors were named by Ribbert in 1894. Their incidence is only 0.5–0.8/million and they account for 17.5% of primary malignant bone tumors in axial bones [1,2]. Most chordomas occur in the sacrococcygeal region. About 50–60% originate in the sacrum, and 15% in the skull and mobile spine [3,4]. Chordomas in other sites are extremely rare.

We here present the clinical, radiographic, and histologic findings as well as treatment and outcome in a patient with an iliac chordoma. To the best of our knowledge, a chordoma located in the ilium has never been reported.

2. Case report

A 45-year-old man presented to our institution with left low back pain with no obvious cause for two months. The pain occurred when climbing stairs or bending over, not at rest or during the night. Concurrent with its worsening, he had noticed an enlarging mass on his lower back.

On physical examination the patient appeared healthy, but in mild discomfort. No erythema, varicose or ruptured veins were found. The skin temperature over the lesion was normal. There was a tender, non-compressible, diffuse swelling over the posterior aspect of the left ilium that did not change with bending down, although this did increase his pain. His gait and range of hip joint motion appeared normal, as did muscle strength and sensation in his lower limbs.

Radiographs showed a lytic lesion in the left ilium adjacent to the sacroiliac joint (Fig. 1). Computed tomography (CT) (Fig. 2) showed a $5.5 \times 5.0 \times 3.4\ cm^3$ lytic lesion in the left ilium; the sacrum was not involved. The lesion was of uniform density with no bone or cartilage matrix formation. The cortex was discontinuous and the boundary well-defined. CT values were 39 HU before enhancement and 57 HU after enhancement. A chest CT revealed no lung lesions. Magnetic resonance imaging showed medium and high signals in T1- and T2-weighted images, respectively, with uneven enhancement. The maximum standardized uptake value on positron emission tomography was 2.9 and no other lesions were detected. Serum alkaline phosphatase, calcium and phosphate concentrations were normal.

A core needle biopsy showed a funicular, or cluster, structure of vacuolated tumor cells in a background of mucus. The cells resembled chordoma cells and appeared mildly atypical. No clearly differentiated cartilage or bone was found.

We made a diagnosis of malignant bone tumor based on the imaging and pathology findings, and made the decision to perform computer navigation-aided wide resection. We chose an iliac groin approach extending to the posterior inferior iliac spine. We protected the femoral nerve and lateral cutaneous nerve while exposing the sacrum and ilium. We cut the bone in accordance with the pre-operative plan and completely excised the tumor, leaving

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http://dx.doi.org/10.1016/j.jbo.2015.09.004

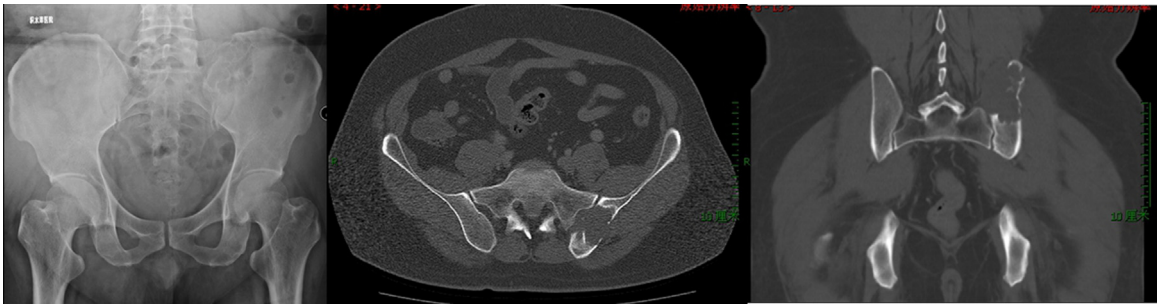

Fig.1. Preoperative X ray and CT showed osteolytic destruction of the left iliac wing. The density was uniform and the cortical bone was not continuous. The sacrum was not involved.

the pelvic ring intact. We used a large piece of autologous normal ilium to fill the defect.

Pathological examination of the operative specimen showed that a wide resection margin had been achieved (Fig. 3). We again observed a funicular structure of vacuolated tumor cells in a background of mucus. The cells had a mildly atypical appearance. Immunohistochemical staining showed the following: cytokeratin (CK) (+++), CK7 (focal +), CK8 (+++), CK18 (focal +), CK19 (+++), epithelial membrane antigen (focal weak +), S-100 (scattered cells +), D2-40 (−), brachyury (+), and vimentin (+). Therefore, the microstructure, morphology and immunohistochemistry results were consistent with a classical chordoma diagnosis (Fig. 4).

No tumor recurrence was detected during 9 months of follow-up, during which time the patient resumed normal daily life activities and achieved a 90% Musculoskeletal Tumor Society score.

3. Discussion

The most common site of chordomas is the sacrum, but they can also occur in the base of the cranium and the mobile spine [5,6]. Most patients with chordomas are in their fifth to seventh decades. The ratio of male to female patients is about 2–3:1. Because affected patients often present with a long history of sacral or flank pain, diagnosis and treatment are often delayed. When these tumors enlarge, they can invade pelvic organs or nerve roots, causing organ and lower limb dysfunction.

Radiographs usually show cystic and expansible osteolytic lesions with bone shell discontinuity. CT scans can clearly show the extent of the soft tissue mass and associated destruction. These tumors often show low to moderate signals and high signals in MRI T1- and T2-weighted imaging, respectively, and can appear lobulated. Pathological examination characteristically shows a gray-brown or white, translucent, jelly-like lesion with uneven texture and a pseudo capsule. The tumor cells usually form masses or cords. Chordomas are classified into three types according to cell morphology: common, chondroid, and poorly differentiated. The common type is low grade with droplet-like vacuoles in the tumor cells.

It is extremely rare for chordomas to occur in sites other than

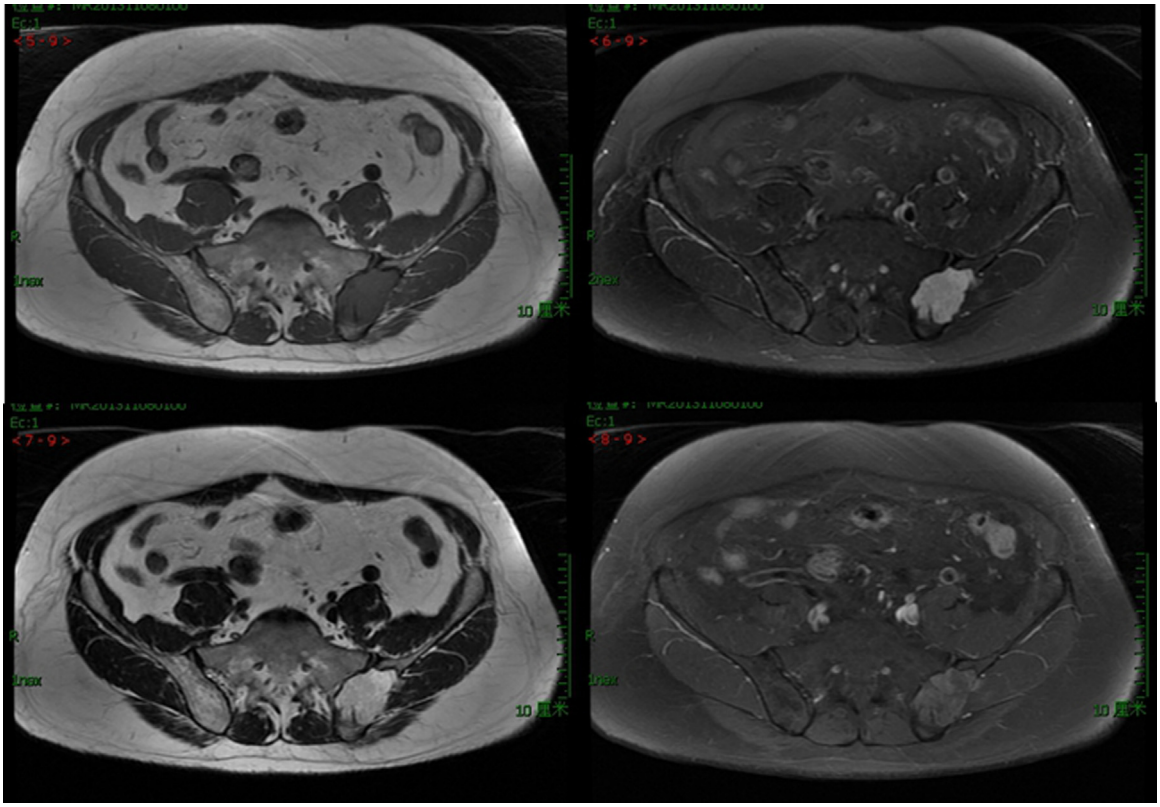

Fig.2. Preoperative MRI showed moderate signal on T1 and high signal on T2 with inhomogeneous enhancement.