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The chordoma arised from ilium: A rare case report

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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.
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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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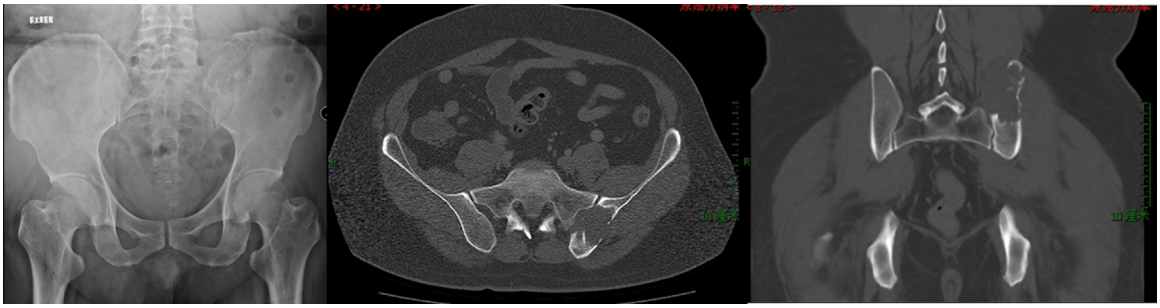

Fig.1. Preoperative X ray and CT showed osteolytic destruction of the left iliac wing. The density was uniform and the cortical bone was not continuous. The sacrum was not involved.

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

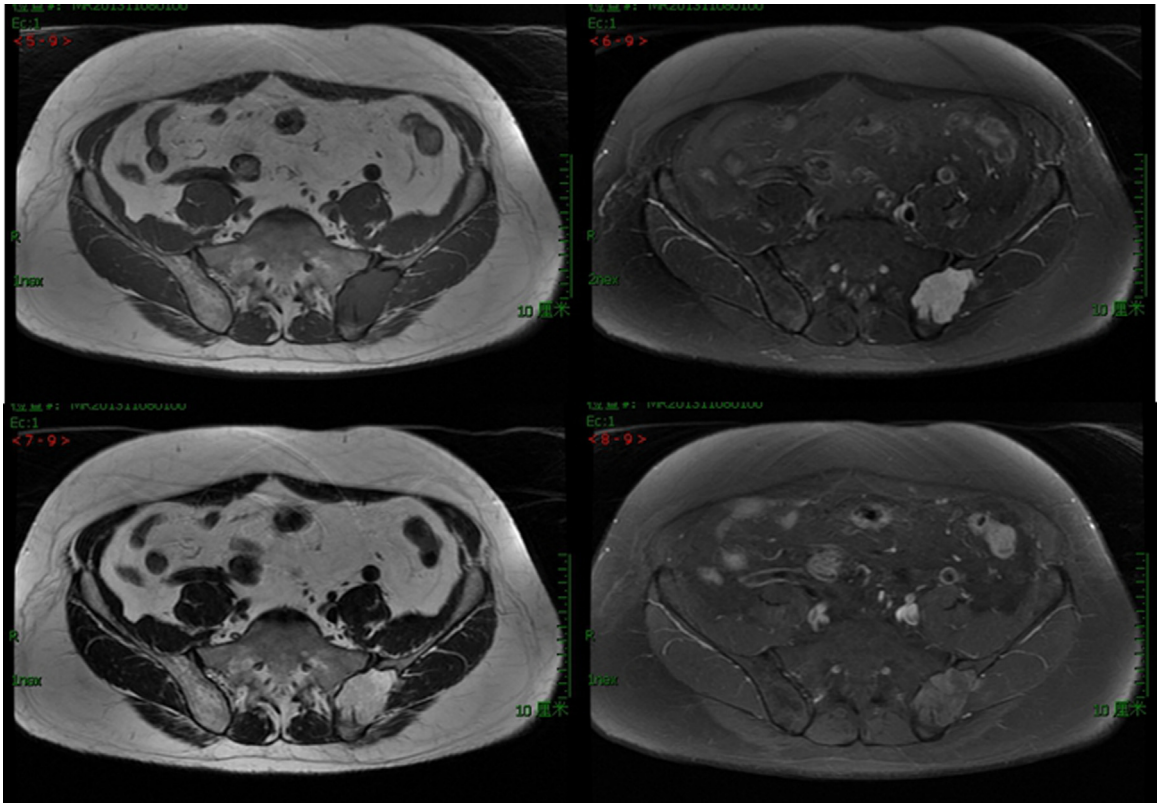

Fig.2. Preoperative MRI showed moderate signal on T1 and high signal on T2 with inhomogeneous enhancement.

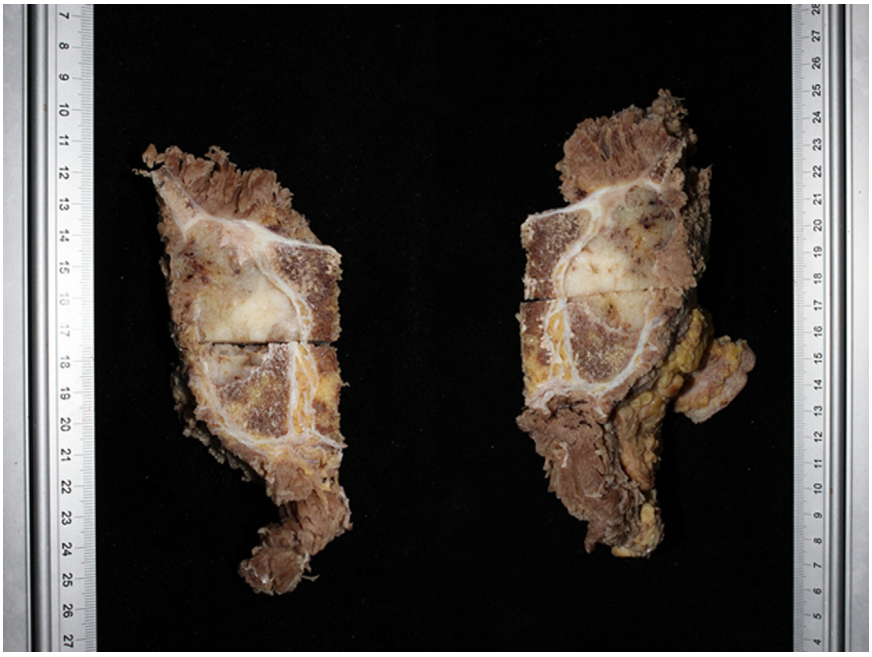

Fig.3. Assessment of postperative specimen showed tumor located in ilium without involvement of the sacrum.

the sacrum, base of the cranium, and mobile spine. We have not found any previous reports of iliac chordomas. In theory, chordomas can only occur in sites that were occupied by the embryonic notochord. These sites include the whole length of the spine from the skull base to the sacrum, most commonly at one end or the other. Imaging studies of this rare tumor showed the iliac lesion was adjacent to the sacrum; however, it appeared to have originated from the ilium without any involvement of the sacrum. An biopsy revealed chordoma, which was completely unexpected. The pathologists therefore carefully reviewed the pathology of the operative specimen and again concluded the diagnosis was chordoma. This rare case changed our understanding of both chordomas and primary iliac tumors. We could find only case reports of a chordoma occurring in non-axial bone [3].

The most common primary malignant tumor of the ilium is chondrosarcoma; osteosarcoma, Ewing sarcoma, and malignant fibrous histiocytoma are also common, whereas other primary malignant bone tumors are rare [4]. Common benign tumors of the ilium include giant cell tumors, osteochondromas, simple bone cysts, and fibrous dysplasia; chondroblastomas, Langerhans cell disorders, and bone hemangiomas can also occur [4].

Our patient was a 45-year-old man, which is consistent with the reported age range and sex for subjects with chordomas. These lesions should be differentiated from other benign and malignant tumors. Chondrosarcomas are the most frequent primary malignant bone tumor of the ilium, usually occurring in persons aged

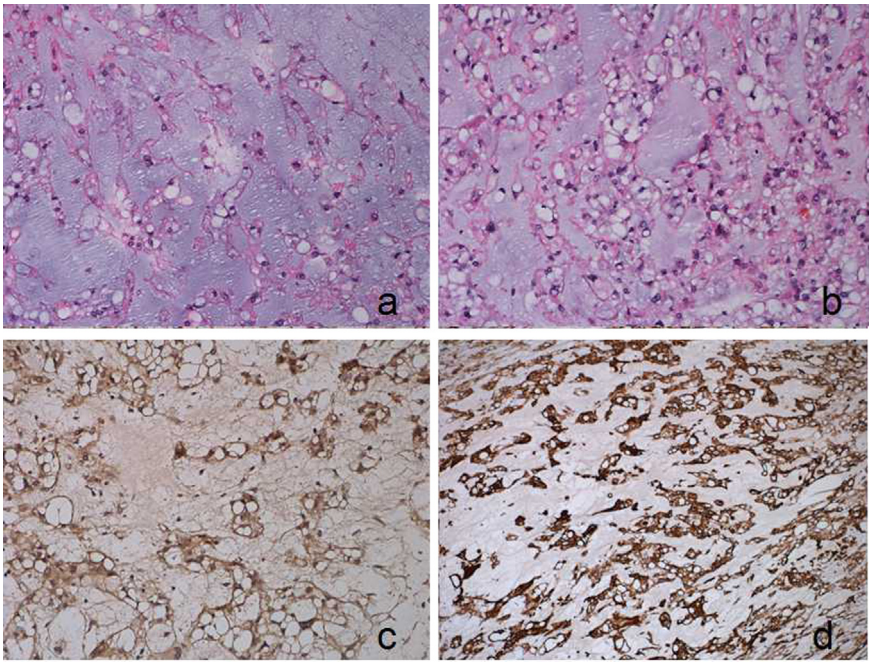

Fig.4. The postoperative pathology showed funicular or cluster structure atypia vacuolated tumor cells in background of mucus (a). Droplet like tumor cells with round nuclei and mild atypia. Multiple vacuoles were showed in cytoplasm (b) (HE staining X 200). The tumor cells were positive for Brachyury (c) and CK8 (d) (EnVision X 200).

more than 40 years [7]. Imaging usually shows osteolytic destruction with well-demarcated boundaries, as was true in our case. However, a calcified matrix and obvious soft tissue mass are characteristically found in subjects with chondrosarcomas. Osteosarcomas occur mainly in adolescents, but have been reported in older patients with pelvic lesions [4]. These high grade tumors usually cause severe local symptoms and have aggressive characteristics. About 5% of giant cell tumors occur in the pelvis [8], and affected patients are characteristically younger than our case. Imaging shows osteolytic destruction with no bone or cartilage matrix formation. These lesions usually have obvious cortical expansive growth with soap bubble-like changes. A liquid plane can be found on MRI examination. Thus, the findings in our case are inconsistent with any other common malignant tumor.

In our case, radiographs showed osteolytic destruction of uniform density. No bone or cartilage matrix formation was evident, these findings being similar to the classic manifestations of chordoma; however, there were some differences. The MRI signals were fairly uniform and there was no obvious soft tissue mass, whereas sacral chordoma often appear as large, lobulated, heterogeneous, soft tissue masses; possibly because sacral chordomas tend to have occult symptoms and diagnosis is usually delayed. However, our case was diagnosed only two month after onset of symptoms.

The standard therapy for chordoma is excision. Local recurrence, but not metastasis, is a major determinant of prognosis. Because local recurrence is related to initial surgical margin and intra-operative tumor cell contamination, achieving a safe margin is very important. Boriani [6] reported 153 cases of chordoma patients who underwent surgery. The recurrence and survival rates were 26.2% and 86.2%, respectively, for patients with safe margins; they were 78.4% and 58%, respectively, for those receiving intralesional resection. Others have reported recurrence rates of 43–85% [9–12]. Wide resection can significantly reduce the recurrence rate; however, it is often very difficult to achieve. Fuchs [13] reported 31 cases with wide resection, only one of whom relapsed. However, 22 of 31 patients in whom wide margins were not achieved relapsed. Yonemoto [14] reported a recurrence rate after intracapsular resection of 60%; no patients in whom wide resection was achieved relapsed. In our patient, wide excision of the iliac lesion was achieved and the patient made a good functional recovery and has had no recurrence.

Boriani [6] reported that patients who have undergone intralesional resection and have residual tumors can benefit from postoperative radiotherapy. Many recent studies [12,15,16] have reported that postoperative radiotherapy can relieve pain and prolong disease-free survival. Reported 5-year and 10-year survival rates are 45–87% and 28–71%, respectively. Some factors such as being male, younger age, smaller tumor, and early treatment may be associated with a good prognosis [1,6,9–14,17,18].

A possible explanation for the occurrence of our patient's chordoma in the ilium near the sacrum is ectopic growth of notochord tissue. Our patient was fortunate to receive an early, accurate diagnosis and effective treatment.

Conflict of interest statement

Each author certifies that he or she has no commercial associations (eg, consultancies, stock ownership, equity interest, patent/licensing arrangements, etc) that might pose a conflict of interest in connection with the submitted article.

Ethical review committee statement

Each author certifies that his or her institution approved the reporting of this case report and that all investigations were conducted in conformity with ethical principles of research.

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None.

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