

Solitary fibrous tumor of the ilium

A case report

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Abstract

Rationale: Solitary fibrous tumors (SFTs) are rare spindle cell tumors that are most commonly found in the mediastinal pleura. Although there are increasingly more reports of extra-pleural SFTs, reports of SFTs in bone are very rare. To our knowledge, a SFT of the ilium has not yet been reported. With low specificity on computer tomography and magnetic resonance imaging, SFTs are easily misdiagnosed.

Patient concerns: A 33-year-old man visited our hospital due to repeated right ilium pain for 3 months. The pain was dull and bearable, with no hip joint dyskinesia. The relevant physical examinations are negative. The patient was healthy before and had a negative family history. Radiologically, a large mass with inhomogeneous attenuation and intensity and obvious heterogeneous enhancement was misdiagnosed as a giant cell tumor of ilium.

Diagnoses: The man was diagnosed as the solitary fibrous tumor of right ilium.

Interventions: The patient was performed an “incision biopsy of the right ilium” and “extended resection of tumor”.

Outcomes: The pathology and immunohistochemistry was confirmed as the solitary fibrous tumors. The patient was followed-up by computed tomography of pelvis in local hospital every 6 months, and there is no recurrence and any symptoms.

Lessons: We learned that the solitary fibrous tumor could locate in the ilium, and when we see imaging manifestations like this case, we should think it may be SFT.

Abbreviations: CT = computed tomography, ESFT = extra-pleural solitary fibrous tumor, GCT = giant cell tumor, HIPAA = Health Insurance Portability and Accountability Act, MRI = magnetic resonance imaging, SFT = solitary fibrous tumor, SMA = smooth muscle actin, WHO = World Health Organization.

Keywords: computed tomography, ilium, magnetic resonance imaging, solitary fibrous tumor, x-ray

1. Introduction

Solitary fibrous tumors (SFTs) are uncommon neoplasms that can be benign or malignant. They were first described by Klemperer and Rabin^[1], who observed a mesothelial tumor arising from the pleura. It was established that SFTs could also be found in numerous extra-pleural sites. In fact, extra-pleural SFTs (ESFTs) are universal neoplasms with pleural and extra-pleural distribution. ESFTs have been described within the head and neck region (e.g., petrous bone, skull,^[2] cervical spine,^[3] infratemporal fossa,^[4] nasal cavity, orbit, thyroid salivary gland, face, neck, parapharyngeal space, nasopharynx, larynx, and oral cavity),^[5]

and other organs (e.g., breast,^[6] bladder,^[7] thigh,^[8] the retroperitoneum, and the genitourinary tract).^[5]

Although reports of ESFTs have become more common in recent years, reports in the skeleton are rare. To our knowledge, SFTs of bone were only found in the petrous bone, skull, thigh, and spine. This is the first case of an ESFT in the ilium.

2. Case report

This Health Insurance Portability and Accountability Act (HIPAA) compliant study was approved by the institutional review board of the First Affiliated Hospital, Zhejiang University, China. The requirement for informed consent was waived because of the study's retrospective nature.

A 33-year-old man visited our hospital on December 2, 2014, due to repeated right ilium pain for 3 months (Table 1). The pain was dull and bearable, with no hip joint dyskinesia. The relevant physical examinations are negative. The patient was healthy before and had a negative family history. A pelvic x-ray showed an area of low density in the right iliac inhomogeneously. The cortical bone was discontinuous with an obscure boundary (Fig. 1). The computed tomography (CT) scan of the pelvis revealed that there was an 8.5 cm × 6.1 cm × 8.7 cm mass with slightly low attenuation, similar to muscle, in the right ilium. An osteophyte was seen in the mass and the cortical bone was discontinuous at the margin. The mass extended to the adjacent soft tissue with a well-defined margin while pushing on the surrounding tissue (Fig. 2). The magnetic resonance imaging (MRI) scan showed an abnormal 8.8 cm × 6.1 cm × 8.7 cm mass in the right ilium, which was slightly hypointense on T1-weighted

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Table 1**The important milestones of diagnoses and interventions.**

Date	Events
September, 2014	The beginning of pain
December 2, 2014	Visited hospital
December 9, 2014	Incision biopsy
December 18, 2014	Pathology
December 19, 2014	Extended resection of tumor
January 5, 2015	Immunohistochemistry
July, 2015	Follow-up
January, 2016	Follow-up
July, 2016	Follow-up
January, 2017	Follow-up
July, 2017	Follow-up



Figure 1. Conventional radiograph of pelvis showed an irregular inhomogeneous lesion with low density in the right iliac, the bone shell of which was discontinuous (indicated by the white arrow), and the boundary was ill defined.

images and hyperintense on T2-weighted images, with some areas of much higher intensity within the mass. The mass showed obvious inhomogeneous enhancement with mottling and a hypointense band in the center. The adjacent tissue was compressed (Fig. 3). One week later, the patient was given an “Incision biopsy of the right ilium.” Histological analysis demonstrated spindle cell lesions accompanied by collagen fibers, the tumor may be solitary fibrous tumors (Fig. 4). On December 19, 2014, the patient was given an “Extended resection of tumor” (Table 1). The immunohistochemical

staining of the tumor cells was strongly and diffusely positive for CD34, BCL-2, and CD99; and negative for S-100, CD31, CKpan, EMA, P63, and SMA. The Ki-67 was 20% (Fig. 4, Table 1). The tumor was finally diagnosed as a SFT. The patient was followed-up by computed tomography of pelvis in local hospital every 6 months, and there is no recurrence and any symptoms (Table 1).

3. Discussion

SFTs are uncommon neoplasms that were originally reported as arising from mesothelial cells exclusively from the surface of the thoracic pleura.^[1] The most common hypothesis suggests that they originate from undifferentiated mesenchymal cells. Although SFTs most commonly occur in the pleura, in recent years, they have been found to occur anywhere in the body. The most common sites of ESFTs are the retroperitoneum, deep soft tissues of the proximal limbs, abdominal cavity, and the head and neck. There are only a few cases reported in the skeleton, such as the skull,^[2] cervical spine,^[3] and infratemporal fossa.^[4] This is the first report of an ESFT of the ilium. SFTs rarely occur in children^[9] of both sexes, and occur during the fourth to seventh decades of life with equal incidence by sex.^[10]

The imaging findings of SFTs are nonspecific. They are usually large (varying from 2 to 26 cm in diameter),^[11] solid with well-defined margins, polylobulated contour masses, and tend to displace adjacent structures. On CT, SFTs can show low or high attenuation relative to the muscle, depending on the collagen content. Calcifications may be present in 10% cases^[9] and bone erosion may also be present. Hemorrhage and central necrosis may be visible in larger lesions. In this case, the mass measured $8.8 \times 6.1 \times 8.7$ cm, with clear margins and had a polylobulated appearance. Bone erosion was also present, which is consistent with the imaging findings for SFTs in the literature. On MRI, SFTs are usually isointense or slightly hyper-intense on T1WI and variable on T2WI to skeletal muscle; SFTs are also hypervascular tumors that are vigorously enhancing. Enhanced patterns may be homogenous, heterogeneous, or patchy.^[12] However, the mass in this article was heterogeneous and we can see few necrosis as Garcia-Bennett et al^[13] reported. A vascular pedicle, although unambiguous, is a useful feature for distinguishing SFT. Heterogeneous signal intensity, which is often caused by necrosis and internal hemorrhage, is observed in malignant tumors.^[13]

SFTs are typically composed of bundles of spindle cells irregularly arranged with a background of collagen and a dilated “staghorn-like” vascular network. SFTs usually stain positive for CD34 (95%) and CD99 (70%). They may be positive for BCL2, EMA and smooth muscle actin (SMA) (~30%). A strong immunoreactivity for CD34 can not only diagnose but also allow

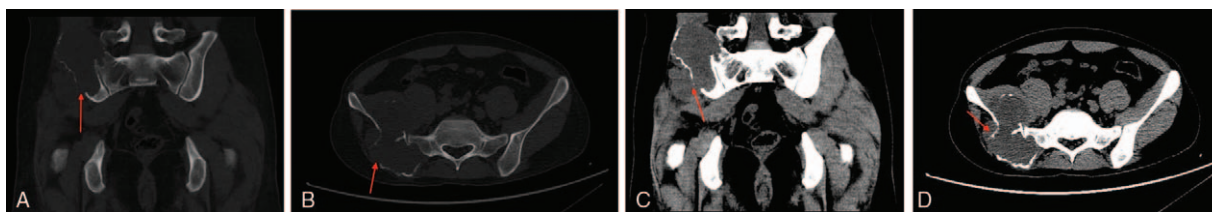


Figure 2. Conventional unenhanced pelvic CT (A. coronal image of bone window; B. axial image of bone window; C. coronal image of soft tissue window; D. axial image of soft tissue window) showed an irregular mass with low attenuation in the right ilium, which was about $85 \text{ mm} \times 61 \text{ mm}$. Bone ridge could be seen in the mass, and the bone shell was discontinuous (indicated by the white arrow). The mass extended into and pushed the surrounding structure, and margins of which was well defined. CT=computer tomography.

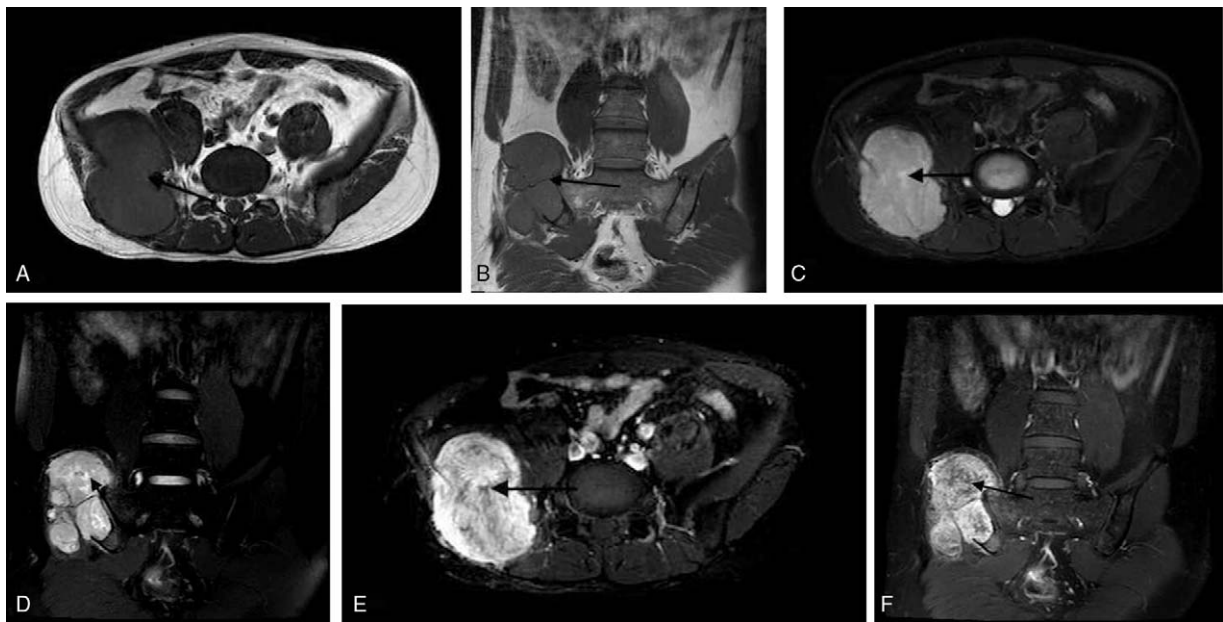


Figure 3. MR images of pelvis showed an iliac mass with abnormal signal intensity, which was about 88 mm×61 mm in size. Axial T1WI (A) and coronal T1WI (B) showed a slightly hyperintense mass compared to the muscle. And the adjacent structure was compressed. Axial T2WI with fat saturation (C) and coronal T2WI with fat saturation (D) showed a hyperintense mass, and in the mass there were mottling and band much higher hyperintense area (indicated by the white arrow). Contrast enhanced axial T1WI with fat saturation (E) and coronal T1WI with fat saturation (F) showed obviously inhomogeneous enhancement, with some mottling and band hypointense area in the lesion. MR=magnetic resonance.

the distinction of SFT from most of the other spindle cell neoplasms. An immunohistochemical profile showing strong expression of CD34 and bcl-2, with a weaker but still positive reaction for CD99, is diagnostic for an SFT. In this case, the tumor showed the same immunohistochemical profile as described in several previous cases.^[14] The proliferation marker Ki-67 can be used to stratify lesions according to their clinical outcome. Recently, a cut off level of a proliferation rate of 12% (Ki-67) was suggested to distinguish benign and malignant lesions.^[15] In our report the proliferation rate of Ki-67 was 20%, so the lesion may be malignant.

The symptoms of SFTs depend on their location, and size, however, the clinical behavior of SFTs is unpredictable. In this case, the pain was a unique symptom. SFTs present as slow-growing, painless masses^[9] and are frequently found incidentally based on radiological findings or symptoms related to their location.^[16] Two paraneoplastic syndromes have been reported: non islet cell tumor hypoglycemia and hypertrophic osteoarthropathy, neither of which was present in our patient. The World Health Organization (WHO) classifies SFTs as intermediate risk for aggressive behavior.

The behavior is usually benign but can be difficult to predict.^[17] The percentage of SFTs that are found to be malignant varies in the literature, with most studies reporting ranges from 5 to 26% of cases,^[9] though even benign SFTs have indeterminate malignant potential. Malignancy potential increases with the presence of necrosis, four or more mitotic figures per 10 high power fields, increased cellularity, and cellular polymorphism.^[9] Increased tumor size (>10cm), the presence of hemorrhage, infiltrative margins, and the presence of anaplastic foci also point to malignancy. Local recurrence after resection can also occur, with reported rates of around 6% in benign disease and around 23% in malignant disease.^[9] Malignant SFTs are diagnosed with atypical histologic features such as nuclear atypia, hypercellularity, a large necrotic portion, and a high mitotic activity (C4 mitoses per 10 high-power fields).^[18]

The preoperative diagnosis for this case is giant cell tumor (GCT). GCTs mainly affect young adults between the ages of 20 and 35 years. The most frequent locations are the distal femur and proximal tibia, however, some reports say that they can also occur in ilium.^[19] The imaging findings of the GCT are the following: (1)

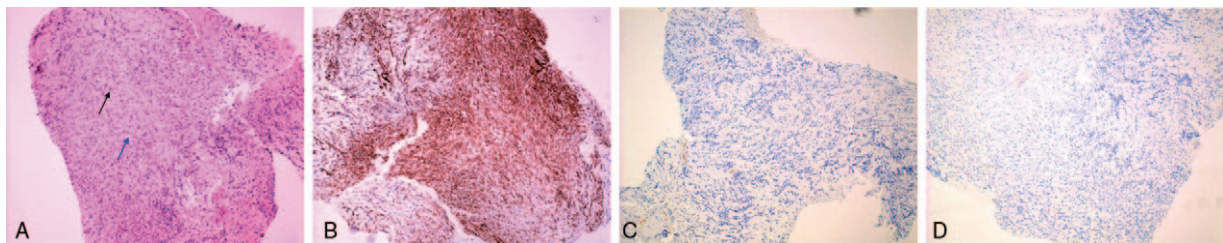


Figure 4. Histopathology of resected specimen showed spindle cell lesions (indicated by the black arrow), accompanied by collagen fibers (indicated by the white arrow) (A. H&E stain, ×100 magnification). Immunohistochemical stain of CD34 (B. CD34 stain, ×100 magnification) showed apparently positive component within the tumor, and immunohistochemical stain of SMA (C. SMA stain, ×100 magnification) and S100 (D. S100 stain, ×100 magnification) were both negative within the tumor. SMA = smooth muscle actin.

the lesion grows expansively and has shell formation; (2) the lesion has sclerotic margins; (3) the “soap bubble sign,” which is a characteristic sign of a GCT; (4) the cortex is discontinuous, a soft tissue mass is present, and there is no periosteal reaction. This case report contained several imaging findings consistent with GCT, with the exception of sclerotic margins. It can be seen that SFT and GCT of the ilium are difficult to distinguish.

The main treatment of SFT is surgical, and postradiotherapy may be useful and SFTs are regarded as chemo resistant.^[9] It has been reported in SFTs that about 6% of patients with histologically benign tumors have the possibility of recurrence. Long-term clinical follow-up is recommended for all patients with a SFT. The potential adverse biological behavior of this tumor may lead to repeated recurrences or malignant transformation.

4. Conclusion

To our knowledge, this is the first case of a SFT in the ilium. The radiological findings for an SFT were difficult to differentiate from other tumors such as GCTs. Positive immunohistochemical findings such as strong immunoreactivity to CD34 can facilitate diagnosis and can prompt a consideration of a SFT.

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