Radiology Case Reports 16 (2021) 1428–1432

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Case report

Angioleiomyoma of the sacrum: a case report and literature review of similar sacral lesions ☆,☆☆,★

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ARTICLE INFO

Article history:
Received 13 December 2020
Revised 10 March 2021
Accepted 10 March 2021

Keywords:
Angioleiomyoma
Sacrum
Magnetic resonance imaging (MRI)
Computerized tomography (CT)

ABSTRACT

Angioleiomyoma is a solitary smooth muscle cell tumor that originates from the tunical media of arteries and veins. Its origin in the sacrum is rare, only one case has been recorded in the literature to date, but cases with bone destruction are even rarer.

We present a 49-year-old woman with lumbosacral pain, unsteady gait and right lower extremity pain for two years, accompanied by radiation pain. Through plain film, Computerized tomography (CT) and Magnetic resonance imaging (MRI) examination, we diagnosed this case as giant sacral schwannoma(GSS). After an operation, it was pathologically confirmed as angioleiomyoma.

The diagnosis of angioleiomyoma should not be based on imaging alone. It requires histopathological examination combined with immunostaining. It should be considered as one of the differential diagnosis of sacral tumors.

Introduction

Angioleiomyoma (ALM), also called vascular leiomyoma or angiomyoma, is a benign tumor that originates from the tunical media of the blood vessels. It is mostly composed of mature smooth muscle cells with significant vascular components. Examples of this tumor arising from the sacrum are exceedingly rare, only one case is recorded in the literature to date. Still, with obvious bone destruction of the sacrum, as far as we know, it is the first reported case thus far. Therefore, it should be considered in the differential diagnosis of the sacral tumors.

Case report

A 49-year-old woman who had lumbosacral pain, unsteady gait and right lower extremity pain for two years, accompanied by radiation pain, visited our outpatient department.

☆ Competing interest
☆☆ The authors declare that they have no conflicts of interest.
★ Patient consent: The study was done after agreement from the local ethics committee and with the patient's informed consent.

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https://doi.org/10.1016/j.radcr.2021.03.017

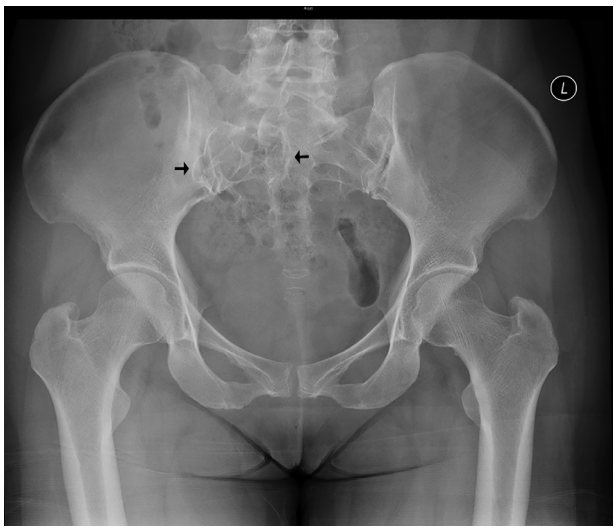

Fig. 1 – Radiographs of the pelvis. The frontal view shows bone destruction of the right sacral ala with peripheral sclerosis (arrows)

Percussion tenderness was elicited on the right side of the sacral region on physical examination. A unilateral positive straight leg raising test at 50° and numbness in the right lower extremity was noted. Sphincter pressure and rectal sensation were normal. Laboratory tests revealed no abnormalities.

Conventional radiographs of the pelvis revealed bone destruction of the right sacral ala with sclerotic margins (Fig. 1). Computerized tomography (CT) of the pelvis revealed a lobulated neoplasm with both intraspinal and extraspinal components along the path of the sacral spinal nerves extending to the presacral area. It had caused widening of the right sacral foramina (S1-2) and bone destruction of the right sacral ala and the iliac bone (Fig. 2), with peripheral sclerosis. After careful observation of the CT scan of the lumbar intervertebral disc performed two years ago, it was found that there was a mass in the same area, which was suspected to be a slow-growing tumor. Besides, MRI scans of the pelvis revealed a mass, which showed isointense signal intensity on T1-weighted (T1W) images (Fig. 3A) and inhomogeneous high signal intensity on T2-weighted (T2W) images (Fig. 3B)and short tau inversion recovery (STIR) images (Fig. 3C) compared with the surrounding muscles. Gadolinium-enhanced fat-suppressed T1-weighted images (Fig. 3D) showed slightly inhomogeneous enhancement. These performances are consistent with previous reports. Yoo et al. [1]reported eight patients with angioleiomyoma and all of them had a well-confined fibrous pseudocapsule. Still, in our case, we can not see it very clearly on MRI images. During the operation, a mass with a complete capsule was observed and bone destruction was found. There were no postoperative complications.

Macroscopically, the excised specimens revealed a 9.0 × 7.0 × 4.0cm gray-reddish, solid mass with a complete fibrous pseudocapsule. Histological examination showed smooth muscle bundles and many slit-like vascular channels (Fig. 4A). And the results of immunohistochemistry were as follows: SMA (Positive), desmin (Positive), S-100 (Negative), CD34 (Negative), CD117 (Negative), Ki-67 (< 5% Positive). These examination results confirmed the diagnosis of angioleiomyoma.

Discussion

Angioleiomyoma of the sacrum with bone destruction is relatively infrequent. The only case was published by Sang Min Lee et al. [2]. They showed that a giant mass was detected in the foramen of the sacrum, accompanied by a branching pattern of growth and bone erosion of adjacent bones, but there was no significant bone destruction. It showed an isointense to hypointense signal intensity on T2-weighted (T2W) images compared to muscle. In comparison, our case showed inhomogeneous high signal intensity on T2W images and short tau inversion recovery (STIR) images. Both lesions showed an isointense signal intensity on T1-weighted (T1W) images and obvious enhancement on fat-suppressed T1-weighted images compared to muscle. However, obvious cortical destruction of the sacrum was observed only in this case. We present it here to improve the identification and further understanding of clinical, pathological, radiological characteristics of this tumor.

Angioleiomyoma, a rare tumor consisting of mature smooth muscle cells and prominent vascular components, is classified by the World Health Organization (WHO) in 2013 as benign pericytic (perivascular) tumors [3]. It usually happens in the subcutaneous tissue of the limbs, presenting as painful, small, isolated solid mass [2]. Incidence peaks in people aged 40 to 60 years, mainly in women. Hachisuga et al. [4]reported that women were 1-1.7 times more frequently affected than men by reviewing 562 cases. Because intraosseous angioleiomyomas are very rare, few cases have been reported at present. There is no exact incidence and differences between men and women. Clinical manifestations and physical examination of angioleiomyoma are nonspecific. Most clinical symptoms are relevant to the location of the disease and local space-occupying compression. Pain is considered to be the most significant clinical feature. In Hachisuga's study [4], pain/tenderness was present in 58% of the cases of angioleiomyoma. In our study, the patient felt pain in the lumbosacral and right lower extremity, accompanied by radiation pain. Although suspected etiologies such as trauma, infection and hormone have been put forward, the definite etiology of angioleiomyoma (ALM) is not exact yet [2].

According to the classification of angioleiomyomas in Morimoto's theory [4], the tumor was classified into three histologic types: capillary or solid, cavernous, and venous. The solid subtype is marked by small in size and slit-like vascular channels encompassed by smooth muscle bundles. The cavernous tumors consist of fewer smooth muscle bundles and dilated vascular channels. Lastly, venous angioleiomyomas are made up of non-compact smooth muscle bundles and thick-walled vascular channels. Changes in the proportion of different components may lead to various signal features on magnetic resonance imaging (MRI). JaeWoongHwan et al. [5] compared the magnetic resonance (MR) images of angioleiomyoma of the extremity with histopathologic find-

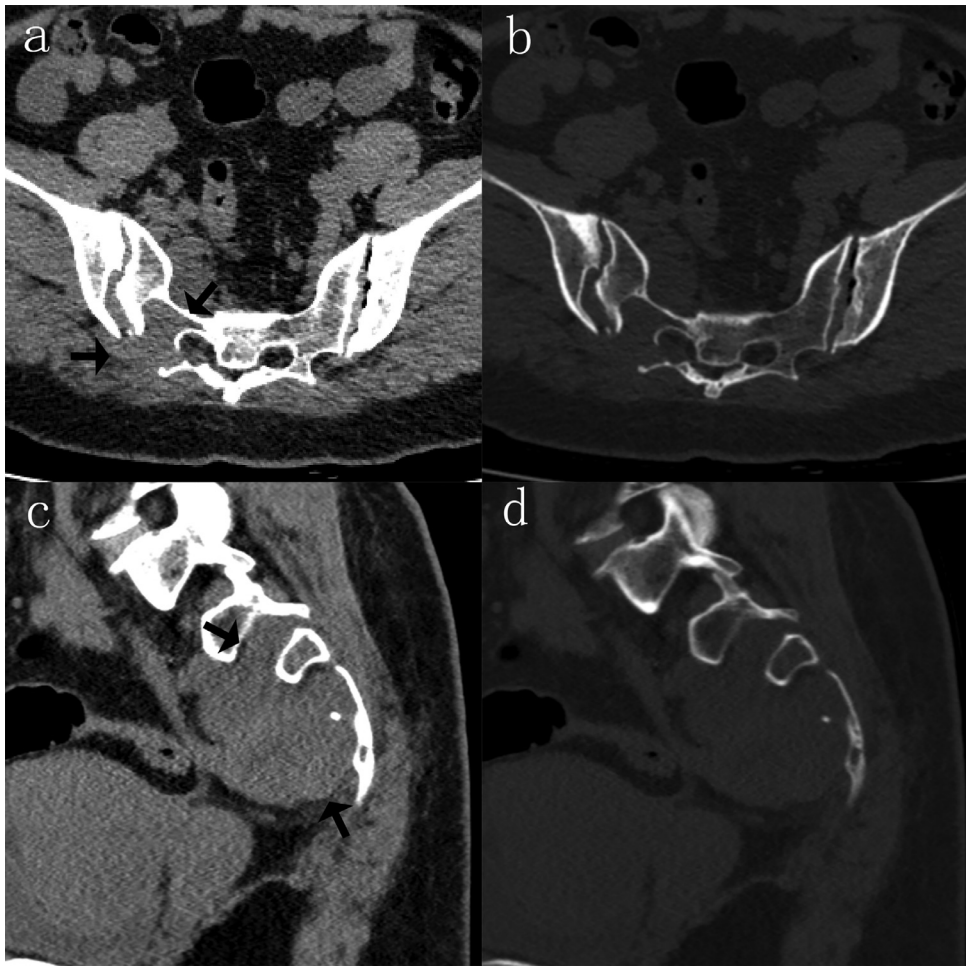

Fig. 2 – Computerized tomography (CT) of the pelvis reveals a lobulated soft tissue mass with both intraspinal and extraspinal components along the path of the sacral spinal nerves extending to the anterior sacral region. It has caused widening of the right sacral foramina (S1-2) and bone destruction of the right sacral ala and the iliac bone, with peripheral sclerosis (arrows)

ings. They found that hyperintense regions are equivalent to the smooth muscle bundle cells and isointense areas are related to the fibrous tissue or endovascular thrombosis on T2-weighted (T2W) images. Although MRI is helpful in the differential diagnosis of angioleiomyoma, it can not distinguish different histological subtypes [6]. MRI surpasses CT in the evaluation of parenchyma, bone marrow, nerve and vascular structures. Still, CT is better in assessing the characteristics of bone tumor, like micro bone destruction, periosteal reaction and differentiation of matrix mineralization [7].

Angioleiomyoma involving skeleton is extremely rare. Through the analysis of previous cases [8,9], it can be classified into two categories in imaging: the first is the primary angioleiomyoma of bone, it is considered to originate from smooth muscle cells in the walls of bone vasculature in previous studies [9]. The second is the angioleiomyoma that originating from the structures closely abutting the bone and causing scalloping of the cortex, even penetrating within the bone, like ours.

Considering the morphology and atypical localization of the lesion, the following differential diagnoses were proposed, but not limited to: giant sacral schwannoma (GSS), sacral chordoma (SC), sacral giant cell tumor (SGCT).

In the beginning, we diagnosed this case as GSS and we found that GSS and our case share many clinical and imaging similarities. Firstly, schwannomas located in the sacrum are relatively rare, accounting for 1%-5% of all spinal schwannomas [10]. Due to few reports of angioleiomyoma, the incidence rate of sacrum has not been reported so far. Secondly, they have no clinical symptoms, or just mild, until they reach a certain scale. In our case, the patient suffered from lumbosacral pain, unsteady gait and right lower extremity pain. Thirdly, on MR images, GSS show isointense to surrounding muscles on T1W images and hyperintense on T2W images. And the giant invasive (Type V) schwannomas, according to Sridhar's classification system [11], can grow in all directions and erode the vertebral body. The above findings are similar to our case. Ming-Jue Si et al. [12]reviewed 8 cases of GSS and found that seven of the eight (87.5%) showed large and centrally located cystic area instead of multiple small cysts. This feature may be helpful for the differential diagnosis of this tumor.

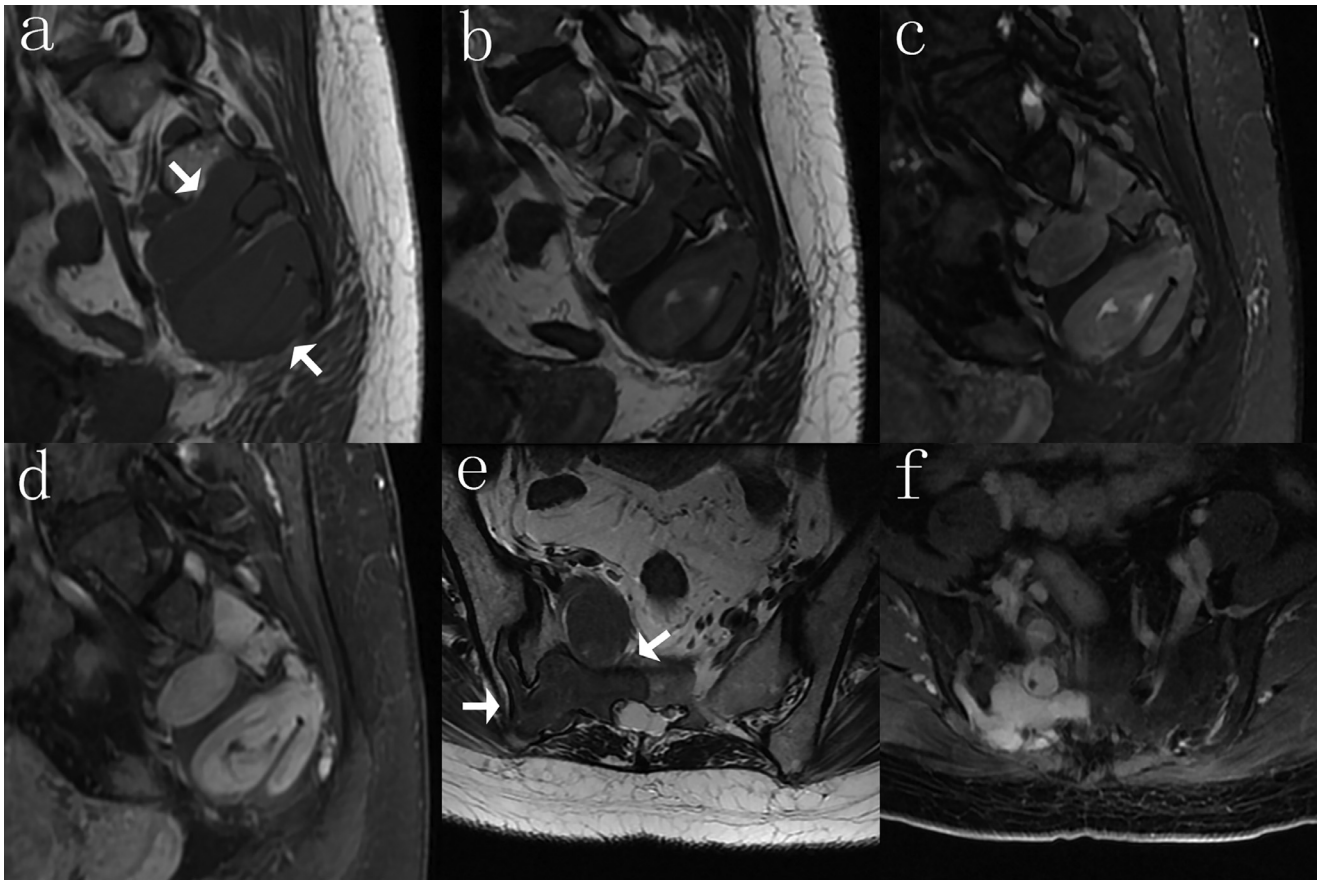

Fig. 3 – Magnetic resonance imaging (MRI) scans of pelvis demonstrating a soft tissue mass, which shows isointense signal intensity on T1W images (Fig. 3a) and inhomogeneous high signal intensity on T2W images (Fig. 3b.e) and STIR images (Fig. 3c) compared with the surrounding muscles. Gadolinium-enhanced fat-suppressed T1-weighted images (Fig. 3d.f) shows slightly inhomogeneous enhancement

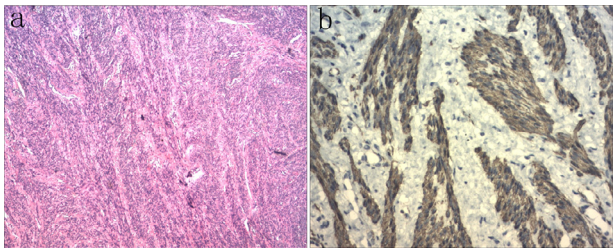

Fig. 4 – Histological appearance of angioleiomyoma. (a) photomicrograph demonstrating smooth muscle bundles with slit-like blood vessels (hematoxylin and eosin; original magnification × 20). (b) Immunohistochemical staining of smooth muscle actin expresses diffuse and strong positive reaction of the tumor cells (magnification × 100)

Sacral chordoma is the commonest primary malignant spinal tumor (20%-34%), 50% of chordomas occurred in the sacrum, the most common age of chordomas was 30-60 years old and the male to female ratio was 2-3:1 [13].SC usually occurs in the midline or paramedian position of the third, fourth or fifth sacral vertebrae. It is often considered to be a lesion with osteolytic destruction and extraosseous expansion. Internal calcifications and fibrous pseudocapsule are common on plain films and CT. On MRI, chordomas show a low-to-moderate signal intensity on T1W images and high signal intensity on T2W images. The septations of the tumor are hypointense on T2W images and the hemorrhage or proteinaceous material within the tumor is hyperintense on T1W images. The enhancement of extraosseous components ranges from mild to moderate [13,14].

Giant cell tumor (GCT) is the most common benign tumor of the sacrum (71%) [15]. And it is also the second, only to chordoma, the most common primary sacral tumor, accounting for 13% of sacral tumors. It occurs most commonly between 20 and 45 years old [16], affecting females more frequently than males [13]. Radiographically, sacral GCT is an expansile lytic lesion. It often occurs in an eccentric position of the sacrum, and it can extend to either side of the central line, even pass through the sacroiliac joint. Computerized tomography (CT) shows that there is usually a thin cortical rim of the GCT. On T2W images, 63% -96% of the cases show heterogeneous low to moderate signal intensity, making the differential diagnosis of GCT more clear [17].

In conclusion, it is very difficult to diagnose sacral angioleiomyoma on imaging. Its diagnosis requires histopathological examination combined with immunostaining. Therefore, it should be considered as one of the differential diagnoses of sacral tumors.

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