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CASE REPORT

MRI of hemangiopericytoma in the sacrum

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Abstract Osseous hemangiopericytoma is rare. We present a case of a 30-year-old woman with low-back pain with radiation to the left buttock for 1 month. Magnetic resonance imaging (MRI) showed a tumor mass with areas of serpentine signal void pattern in the sacrum suggestive of a vascular tumor. Neither calcifications nor layered blood serum were noted. Histological diagnosis was compatible with osseous hemangiopericytoma.

Keywords Sacrum · Hemangiopericytoma · Magnetic resonance imaging (MRI)

Introduction

Osseous hemangiopericytoma is rare, as the vast majority of these tumors arise in soft tissue. Osseous hemangiopericytoma accounts for less than 1% of vascular tumors [1] and most commonly is derived from the pelvis or femur, but any bone can be affected. Only a few cases of osseous hemangiopericytoma in the sacrum with magnetic resonance imaging (MRI) features have been reported. We present such a case with characteristic MRI findings and histological diagnosis.

Case report

A 30-year-old woman suffered from low-back pain with radiation to the left buttock for 1 month. She felt numbness over her left lower leg and walked with difficulty in the past few days. Physical examination revealed decreased left S1 muscle power and abnormal S1 dermatome sensation. Straight leg raising of the left leg was 40–50°, and the right leg was 60°. Radiography showed an osteolytic lesion in the left sacral ala (Figs. 1A,B). CT scan of the pelvis showed no matrix mineralization (Fig. 1C). MRI of the pelvis showed a large tumor with prominent areas demonstrating signal voids (Figs. 1D–G). The tumor involved the sacrum, the left sacroiliac joint, and the coccyx, with epidural extension. The urinary bladder was compressed, and the uterus was anteriorly displaced by the tumor. The rectum and sigmoid colon were displaced to the right aspect of the pelvic cavity. A hypervascular mesenchymal tumor was proposed before biopsy.

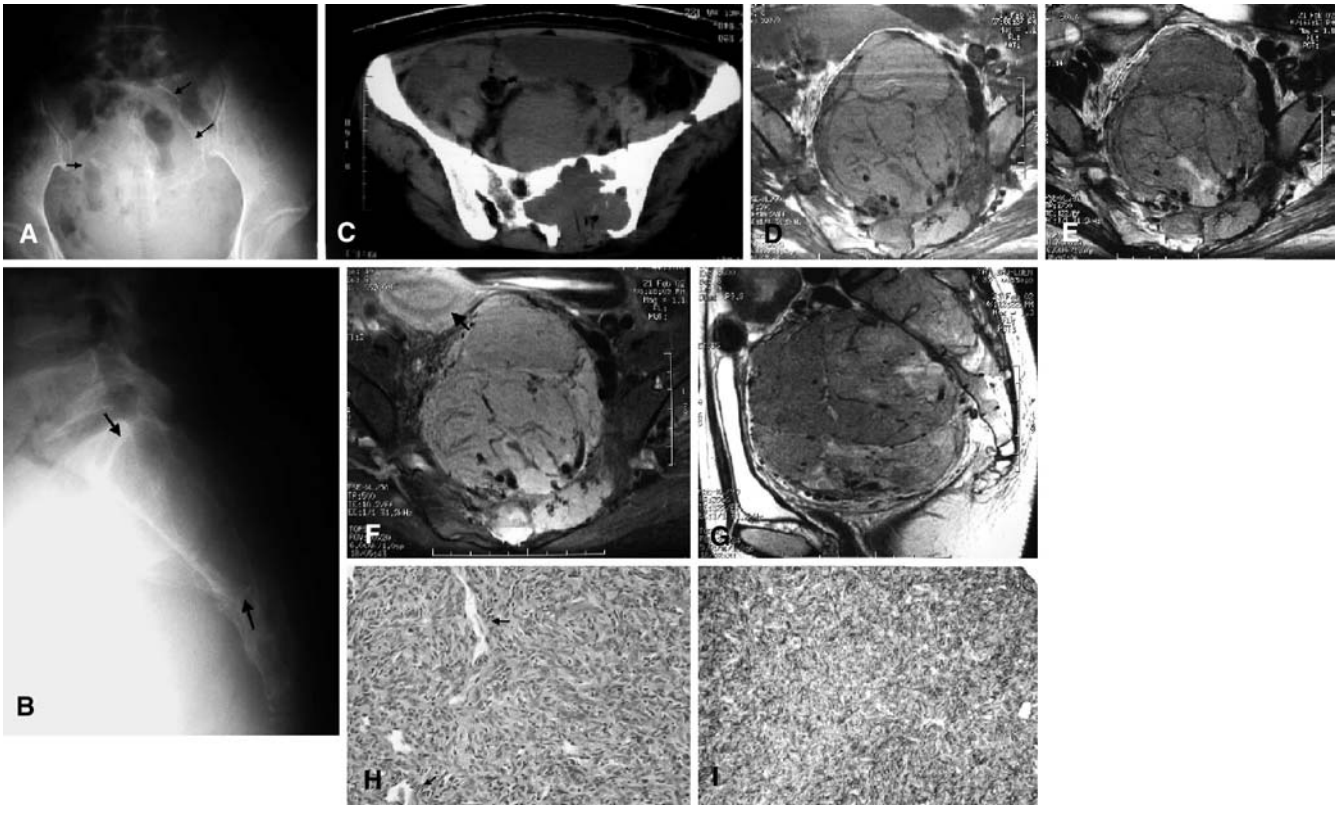

Fig. 1 **A** Anteroposterior and **B** lateral radiographs of the sacrum show a destructive osseous lesion (*arrows*) in the sacrum. **C** CT scan of pelvis after biopsy shows no calcifications within the osteolytic bony lesion. The tumor has air due to being post-biopsy. **D** Axial T1-weighted (TR/TE=500/10.2 ms), and **E** axial T2-weighted (TR/TE=3700/122 ms) MR images show a well-demarcated soft-tissue mass in the sacrum with anterior extension to the pelvic cavity. Note signal-void tubular vascular structures within the tumor. **F** Gadolinium-enhanced T1-weighted (TR/TE=500/10.2 ms) image with fat saturation shows marked enhancement of the tumor. The curvilinear structures are arranged radially as a "spoke-wheel" pattern within the tumor. The vascular channels do not enhance because of high flow, and this also explains the low signal intensity on T2-weighted image (see Fig. 1**E**). The uterus (*arrow*) is displaced ventrally to the right side of the pelvic cavity. **G** Midsagittal, T2-weighted (TR/TE=3986/122 ms) image shows hypervascular tumor mass in the sacrum, corresponding to the destructive osseous lesion, as seen on radiography. The tumor mainly involves the left-sided sacrum (as shown on CT scan) with anterior extension to the pelvic cavity. **H** The tumor is composed of hypercellular spindle cells arranged in short fascicles. Small-caliber, irregular-shaped, thin-walled blood vessels (*arrows*) are seen in the tumor (H&E ×200). **I** The tumor cells are strongly diffusely positive for CD34 on immunohistochemical study (Immunostain ×200)

CT-guided biopsy was performed 5 days later. Histologic examination revealed a hypercellular spindle-cell tumor arranged in short fascicles with focal storiform pattern (Fig. 1H). The tumor cells revealed mild-to-moderate nuclear pleomorphism. There were frequent small-caliber, irregular-shaped, thin-walled blood vessels in the tumor. No mitotic activity was found in this specimen. Neither necrosis nor hemorrhage was seen. The tumor cells were strongly diffusely positive for CD34 and vimentin (Fig. 1I) and negative for cytokeratin, EMA, NSE, S-100, CD117, actin and desmin immunohistochemically. Histological diagnosis was compatible with hemangiopericytoma. The patient received pain control medication because the tumor was not resectable.

After 24 months of follow-up, the patient was bothered by exacerbation of low-back and left-buttock pain. Difficulty in urination was noted. The sphincter function was preserved. Follow-up MRI showed progressive enlargement of the pelvic mass. No surgery was performed. The patient is still alive.

Discussion

Hemangiopericytoma was first introduced by Stout and Murray in 1942 [2] and was one of the first peripheral tumors in soft tissue to be described. It is derived from the pericytes of Zimmerman, which are pericapillary mesenchymal cells with contractile ability [3]. Hemangiopericytoma can arise in any part of the body owing to its vascular origin. The tumor frequently occurs in soft tissue, but the cranial region also has been reported [4]. Primary hemangiopericytoma of bone is very rare, accounting for 0.1% of malignant primary bone tumors and 11% of malignant vascular bone tumors [1]. The ages of patients range from 12 to 90 years, predominantly in the fourth and fifth decade, with a male-to-female ratio of 1.8:1 [5]. The most common symptoms at presentation are pain and a palpable mass.

Radiographic features are nonspecific [5], including osteolytic bone (100%) and cortical destruction with soft-tissue extension (72%). The margin can be well defined (40%) or ill defined (60%). A sclerotic rim has been described in some cases (16%). Periosteal new bone formation is not uncommon (32%), and pathologic fractures are rare (8%). In our case, the tumor mass exhibited a large osteolytic lesion with a lobulated margin and cortical destruction but without a sclerotic rim.

CT scan is superior to radiographs in evaluating bone destruction, but MRI is more accurate in detecting extraosseous extent of the tumor [6]. On MRI, hemangiopericytoma typically shows intermediate signal intensity on T1-weighted images and hyperintense serpentine channels on gadolinium-enhanced images [4]. T2-weighted or short-tau inversion recovery (STIR) images usually show hyperintensity of the tumor, relating to the fluid in the exceedingly vascularized tumor matrix. However, intermediate signal intensity or hypointensity of the tumor on T2-weighted images also have been described [6, 7]. Kehagias et al. [8] reported a case of pelvic hemangiopericytoma with prominent vascular channels in the periphery of the tumor. Mahnken [4] reported that three of his four cases with documented hemangiopericytoma showed a signal void pattern on T1-weighted images and T2-weighted hyperintense with marked contrast enhancement. These findings were highly suggestive of slow-flow blood vessels. However, Juan et al. [6] reported a case of primary hemangiopericytoma in the tibia of having several radially arranged signal-void vessels in the tumor on both T1- and T2-weighted images. These findings were suggestive of high-flow vessels, which was also noted in our case.

The prominent serpentine vascular channels within the tumor, although not specific, should suggest the diagnosis of hemangiopericytoma. Only limited types of tumors demonstrate high-flow vessels, including hemangioendothelioma, angiosarcoma, alveolar soft-part sarcoma, synovial sarcoma, and rhabdomyosarcoma [4]. Sarcomas occur most often with hemorrhage and necrosis within the mass, and therefore, sarcomas were not considered in our case. Tumors with high-flow vessels have clinical impact in that percutaneous or open biopsy can lead to extensive bleeding and exsanguinations. On MRI, the flow-void signal can play an important role in management planning.

Hemangiopericytoma in this location should be differentiated from chordoma, chondrosarcoma, and giant cell tumor. Chordoma typically arises in the midline and involves the S4 or S5 vertebrae. The lesion is purely osteolytic and relatively well defined, usually being oval or slightly lobulated in appearance. However, chordoma is characterized by slow growth, and it lacks the serpentine signal void feature in the tumor matrix. Chondrosarcoma usually exhibits lucent and destructive areas containing chondroid matrix calcification. Giant cell tumor is an eccentric, large, lytic lesion without surrounding sclerosis.

On histology, the tumor cells typically cluster around numerous capillaries and usually contain round-to-oval nuclei, a feature generally lacking in anaplasia [5]. Anaplastic hemangiopericytoma is characterized by the presence of necrosis and/or more than five mitoses per ten 400× microscopic fields and at least two of the following microscopic features: hemorrhage, moderate-to-high nuclear atypia, and moderate-to-high cellularity [9]. In our case, the tumor exhibited high cellularity and numerous capillaries. There was no necrosis, hemorrhage, mitotic figure, or high nuclear atypia in the tumor.

Wide surgical excision is the treatment of choice for hemangiopericytoma. Preoperative embolization can facilitate tumor resection in diminishing blood loss during surgery. Postoperative radiotherapy may improve survival. The role of chemotherapy is still unclear.

In summary, we presented a case of hemangiopericytoma in the sacrum with characteristic MRI findings. Hemangiopericytoma in the sacrum should be included in the differential diagnosis in patients with a tumor mass having a serpentine signal void pattern but without calcifications or layered blood serum on MRI images.

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