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Case Report

Pseudomyogenic hemangioendothelioma—A case report and review of the literature

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ABSTRACT

Pseudomyogenic hemangioendothelioma (PMH) of bone is a very rare tumor and frequently presents at multiple locations. PMH is difficult to diagnose by imaging and histopathologic features. Various and partially discordant imaging findings have been reported in case reports and small case series. We report a case of a 63-year-old man with PMH isolated to the sacrum, presenting with chronic intermittent buttock pain that was incidentally identified on imaging for acute pancreatitis. We believe that learning about PMH of bone will help to include this disease in the differential diagnosis of lytic lesions of the sacrum. Becoming aware of the various and sometimes discordant imaging findings of this rare entity is important and helpful for radiologists, pathologists, and orthopedic surgeons.

Introduction

Pseudomyogenic hemangioendothelioma (PMH) is a rare vascular bone and soft tissue tumor, previously known as epithelioid sarcoma-like hemangioendothelioma [1], and pseudomyogenic (fibroma-like) variant of epithelioid sarcoma [2]. This is a neoplasm of intermediate malignant potential, which will rarely metastasize. The neoplastic cells mimic some histologic characteristics of skeletal myocytes [3]. The accurate preoperative diagnosis of this tumor is difficult given similarities with benign and malignant lesions and metastases [4,5].

PMH most commonly arises from soft tissue in the distal extremities, with a male predilection, and usually affects patients in the second to fourth decades of age. Primary soft tissue disease usually presents as multifocal tumors involving the mucosa, dermis, subcutaneous tissue, and skeletal muscles. About 25% of patients have concurrent osseous involvement [3]. Primary bone lesions have been rarely reported and if present, it usually presents as multifocal disease. To the best of our knowledge, PMH presenting as a solitary bone lesion is rare and no case report of PMH as a solitary lesion of the bone has been reported. In this paper, we describe the imaging and histologic features of a 63-year-old man with solitary primary PMH of the sacrum and review the reports of PMH localized to bone in the literature.

Case presentation

A 63-year-old man was referred to us with a sacral mass identified on a computed tomography (CT) of the abdomen and

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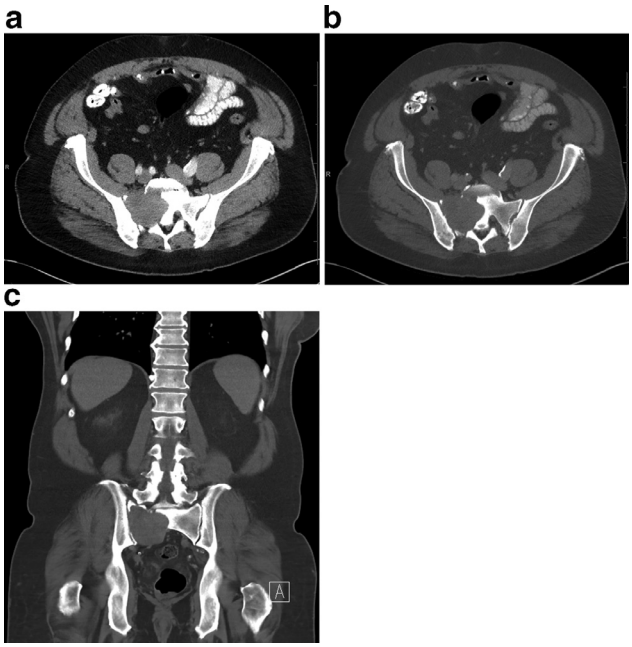

Fig. 1 – (a, b) Axial CT reformatted images with and without contrast administration and (c) coronal CT image showing the lytic expansile lesion involving the right sacral ala extending to the sacroiliac joint. Cortical breakthrough is appreciated without obvious aggressive soft tissue component.

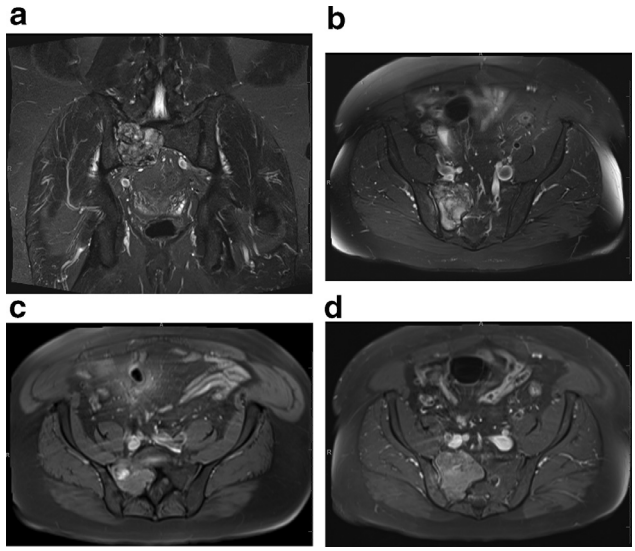

Fig. 2 – (a) Coronal short tau inversion recovery (STIR) image shows a heterogenous right sacral ala mass with mixed signal. (b) Axial T2WI shows the mass abutting the internal artery branches with serpiginous internal signal voids. (c) Axial pre–contrast T1W fat sat imaging shows area of increased signal suggestive of hemorrhage. (d) Mild heterogeneous enhancement on post–contrast T1W image.

pelvis, performed for epigastric pain related to acute pancreatitis. He had a past medical history of intermittent pain in the right upper buttock with radiation down the posterior aspect of the right leg over the last 3-4 years. He also reported discomfort while sitting for a long period of time, with occasional tingling in the tips of his toes. He had been seen by a chiropractor with no significant improvement. Overall, the pain was mild and well controlled with occasional pain medications. The patient denied any fevers, chills, weight loss, or any history of malignancy.

A CT scan of the pelvis was performed, which demonstrated a well-defined lytic lesion with soft tissue attenuation involving the right sacrum. Extension of tumor to the sacral foramina and sacroiliac articular surface with loss of cortical bone was noted (Fig. 1).

Magnetic resonance imaging (MRI) of the pelvis demonstrated an expansile, well-circumscribed mass within the right sacrum, extending to the sacroiliac joint. The mass was heterogeneous, with mixed signal on short tau inversion recovery images (Fig. 2a). On T2-weighted imaging, serpiginous signal voids with a branching pattern were appreciated. The mass was abutting the internal iliac artery branches (Fig. 2b). On pre–contrast T1-weighted imaging fat sat imaging, an area of increased signal was present (Fig. 2c). Mild heterogeneous enhancement after administration of IV gadolinium was appreciated (Fig. 2d). No abnormal radiotracer uptake by the sacral mass or adjacent osseous structures was appreciated on a whole- body bone scintigraphy (Fig. 3a). The mass was hypermetabolic on PET-CT (Fig. 3b and c). No distinct metastatic lesion was identified.

CT-guided biopsy was performed demonstrating a grade 1 angiosarcoma. Additional tissue was required for further testing. Thus, open core biopsy with intraoperative CT scan was performed. Histologic diagnosis was PMH.

Four months later, surgical treatment was performed including S1-S3 partial sacrectomy, tumor resection, and use of argon beam laser diathermy to create a broad area of zonal necrosis along the margin of tumor. Allograft cancellous bone chips were placed laterally along the defect in the sacral ala. Final histologic examination confirmed the diagnosis of PMH (Fig. 4).

Follow-up MRI, 9 months after resection, demonstrated postsurgical changes without evidence of tumor recurrence (Fig. 5). Two follow-up CT examinations also revealed no evidence of tumor recurrence 12 and 16 months after surgery (Fig. 6).

Discussion

Vascular tumors of bone include hemangioma, epithelioid hemangioma, PMH, epithelioid hemangioendothelioma, and angiosarcoma. Table 1 summarizes the distribution of vascular tumors of bone as reported by van Ijzendoorn and Bovee [6].

The term PMH was suggested for the first time in a study of 29 cases by Hornick and Fletcher. Of patients, 24% have concurrent osseous involvement [2]. In 2016, a series of 10 cases of PMH of bone was published by Inyang et al [3]. They reported a male predominance (9:1), and mean age of 36 (range 12-74 years). All cases had multicentric tumors and distinct regional distribution (45% restricted to the lower extremity, 25%

Table 1 – Distribution of vascular tumors of bone (data adapted from van IJzendoorn and Bovee [6]).

	Cranium	Flat bones	Vertebra	Long bones	Small bones of hand	Small bones of foot
Hemangioma	52%	9%	18%	12%		
Epithelioid hemangioma	2%	16%	16%	40%	18%	8%
Pseudomyogenic hemangioendothelioma		30%		30%	30%	10%
Epithelioid hemangioendothelioma		5%	14%	71%	10%	
Angiosarcoma		19%	15%	74%		

to the spine and pelvis, and 15% to the upper extremity). Only 1 patient presented with a single lesion involving only 1 bone (femoral head).

To the best of our knowledge, this is the first report of a solitary PMH of the sacrum. The patient's age was in the upper age range in comparison to reported cases of PMH tumors of bone. Histologically, a panel of immunohistochemical markers such as ERG, CD31, and CD34 are helpful to identify endothelial differentiation of vascular tumors [6]. However, the histologic diagnosis of PMH is challenging since vascular differentiation is essentially absent, and CD34 marker is consistently negative. In addition, some epithelial markers such as keratin AE1/AE3 immunoreactivity in PMH mimic epithelioid sarcomas [7]. CD31 is expressed in about 50% of PMH cases. The most important and characteristic immunohistochemical marker is nuclear staining for FOSB overexpression, resulting from the t(7;19)(q22;q13) SERPINE1-FOSB gene fusion . Fluorescence in situ hybridization (FISH) break-apart probe for FOSB gene rearrangement can be an excellent diagnostic marker as well. The upregulation of FOSB serves as the underlying molecular mechanism of tumorigenesis [6]. The immunohistochemical study of our case was positive for ERG, CD31, AE1/AE3, and FOSB and negative for TFE3, and CAMTA1, consistent with the diagnosis of PMH.

The reported histologic findings of PMH have been quite variable. Inyang et al [3] described the presence of spindled tumor cells arranged in fascicles, associated with scattered rhabdomyoblast-like cells with characteristic, intensely eosinophilic, eccentric, cytoplasm as common histologic findings. In addition, they described 3 histologic features unique to the PMH involving bones: (1) presence of epithelioid cells, (2) exuberant network of interanastomosing reactive woven bone lined by plump osteoblasts and surrounded by loose fibrovascular stroma (similar to osteoblastoma), and (3) focal to broad areas of hemorrhage harboring numerous osteoclast-like giant cells. These features resemble histologic features of giant cell tumor of bone (GCTB) and can present with similar imaging findings.

All but one of the reported cases of PMH of bone are multifocal. Pradhan et al [8] reported the clinicopathologic findings in 8 patients with PMH of bone, skin, and soft tissue; the only patient with unifocal bone disease was a 9-year-old patient with a 1.7-cm femoral head lesion. CT findings demonstrated a permeative osteolysis and marked osteopenia of the femoral head. The majority of the other reported bone lesions were located in the lower extremity such as the tibia, fibula, and foot [8]. In our case, the solitary lesion was centered in the sacrum.

PMH may not be identified with certain imaging studies and may show discordant findings across different modalities. This has been described in a case report by Krebs et al on an otherwise healthy 33-year-old man with multifocal PMH lesions in the femur and tibia. The fluorodeoxyglucose (FDG) and methylene diphosphonate (MDP) uptake and CT appearance of the lesions were not entirely concordant, with some of the lesions being lytic on CT and associated with FDG and MDP uptake, while other lesions were not identified on CT, and either MDP or FDG avid, with enhancement on MRI [9]. The most prominent imaging finding in our case was marked FDG uptake of the lesion and the lytic appearance. We reiterate the importance of using all the imaging modalities for evaluation of vascular tumors of bone. GCTB can have a similar imaging appearance and was on the differential diagnosis of our case. In a study by Muheremu et al, a wide range of FDG uptake by GCTB has been reported (SUVmax between 1.8 and 18.6) [10].

The largest series of 10 patients with primary PMH of bone was reported by Inyang et al. The lesions were well circumscribed, lobulated, and lytic. Some lesions had a sclerotic rim. On MRI, lesions were hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging. This was similar to our case. One patient had marked perilesional edema. Cortical breakthrough and soft tissue extension were present in 2 lesions. Variable PET-CT and bone scan findings were present in 3 and 4 patients, respectively [3].

The role of imaging in differentiating low- from high-grade malignant vascular tumors of bone is limited. However, Xu et al [11] have reported some imaging features that are helpful in differentiation. They reported that low-grade malignant vascular tumors of bone tend to have multifocal, well-defined lesions with residual bone, peripheral sclerosis, and slightly heterogeneous enhancement, whereas high-grade lesions are more likely to be expansile, ill defined, with necrosis and cystic areas, often hemorrhagic, and demonstrate obvious heterogeneous enhancement. Only 1 of the18 cases in that series was diagnosed as PMH, previously called epithelioid sarcoma-like hemangioendothelioma, and that case demonstrated a lytic, expansile, relatively well-defined metacarpal lesion with cortical destruction. MRI demonstrated only mild enhancement.

In conclusion, we present a rare case of PMH isolated to the sacrum in an older patient, incidentally identified on a CT scan in a patient presenting with epigastric pain, and concern for malignancy given the worrisome and discordant imaging features of the sacral lesion. The imaging and histologic findings of PMH of bone have similarities to that of GCTB.

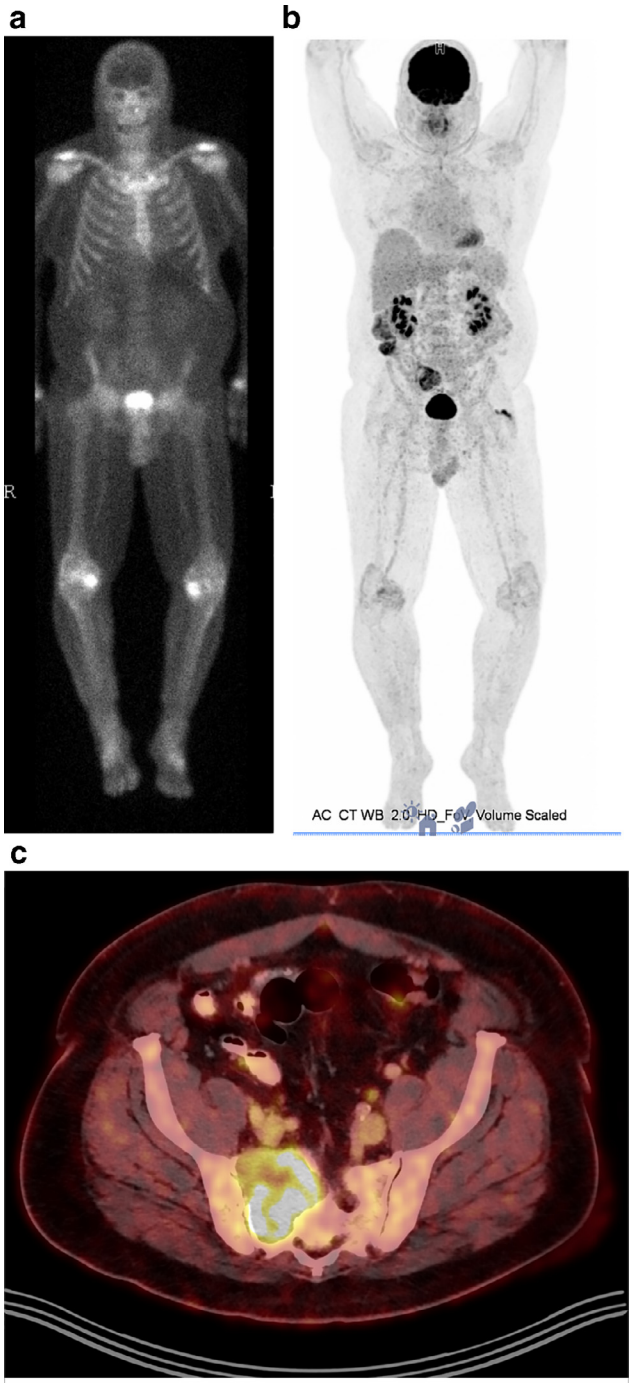

Fig. 3 – (a) Whole-body bone scintigraphy showing no suspicious radiotracer uptake to suggest osteoblastic activity. (b) Maximal intensity projection (MIP) of whole-body PET scan showing intense FDG uptake by the right sacral alar mass and no evidence of metastasis elsewhere. (c) Axial PET-CT fusion showing intensely hypermetabolic sacral mass with SUVmax 10.5.

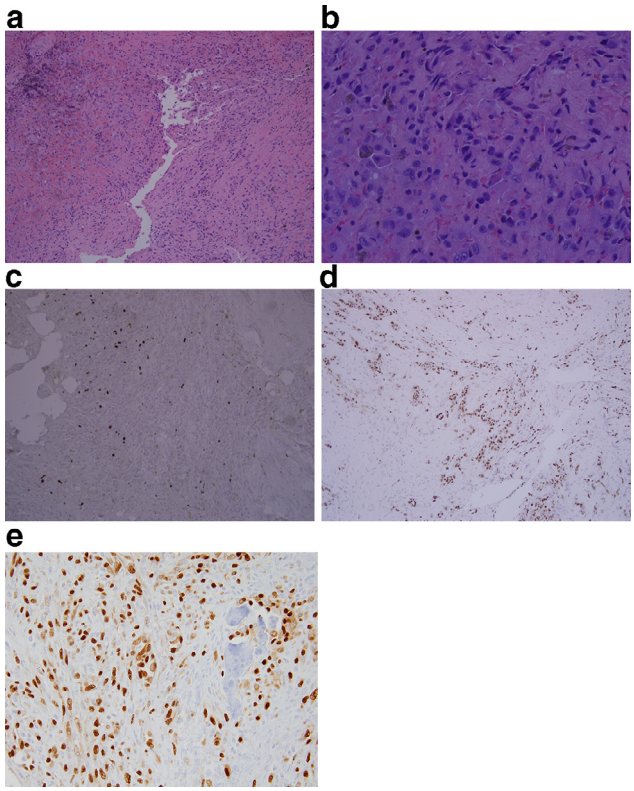

Fig. 4 – On hematoxylin and eosin staining [(a) magnification 100×; (b) magnification 400×], the tumor is composed of round/plump cells with bright eosinophilic cytoplasm with round vesicular nuclei and few cells with prominent nucleoli. A subset of these plump, eosinophilic cells has eccentric nuclei, morphologically resembling rhabdomyoblasts. Mitotic figures were inconspicuous in keeping with low Ki-67 proliferation index of 1%-5% (c). No areas of necrosis are seen. Immunohistochemistry staining demonstrates that lesional cells are diffusely positive for ERG (d), and FOSB (e) and negative for S100, TFE3, and CAMTA1 (data not shown).

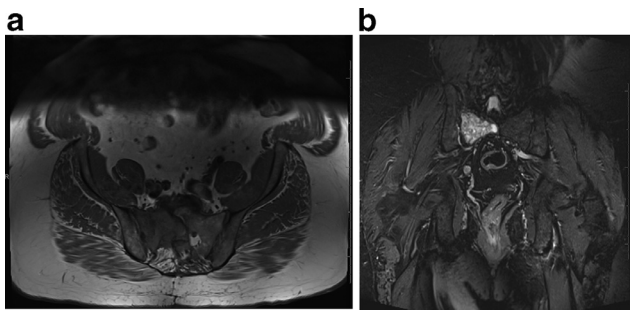

Fig. 5 – (a) Axial T1W image and (b) coronal short tau inversion recovery (STIR) image showing postsurgical changes without evidence of tumor recurrence 9 months after tumor resection and bone chips allograft placement.

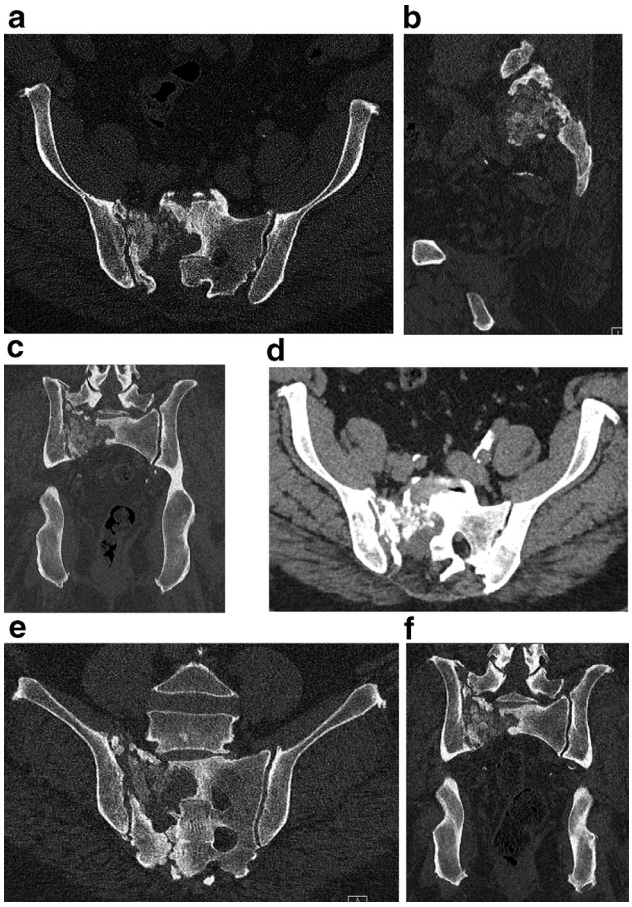

Fig. 6 – (a-c) Axial, sagittal, and coronal reformatted CT imaging showing postop changes with no evidence of recurrence 12 months after surgery. (d-f) Axial, oblique (sacral view), and coronal reformatted CT images showing stable postop changes 16 months after surgery.

We believe that familiarity of this case will allow PMH of bone to be included in the differential diagnosis of lytic lesions of the sacrum, with knowledge of the various and sometimes discrepant imaging findings of this rare entity. This case will be important and helpful for radiologists, pathologists, and orthopedic surgeons in making the correct diagnosis that will influence treatment decision-making.

Declaration of competing interest

None.

REFERENCES

[1] Billings SD, Folpe AL, Weiss SW. Epithelioid sarcoma-like hemangioendothelioma. Am J Surg Pathol 2003;27(1):48–57.
[2] Hornick JL, Fletcher CD. Pseudomyogenic hemangioendothelioma: a distinctive, often multicentric tumor with indolent behavior. Am J Surg Pathol 2011;35(2):190–201.
[3] Inyang A, Mertens F, Puls F, Sumathi V, Inwards C, Folpe A, et al. Primary pseudomyogenic hemangioendothelioma of bone. Am J Surg Pathol 2016;40(5):587–98.
[4] Wenger DE, Wold LE. Malignant vascular lesions of bone: radiologic and pathologic features. Skeletal Radiol 2000;29(11):619–31.
[5] Wenger DE, Wold LE. Benign vascular lesions of bone: radiologic and pathologic features. Skeletal Radiol 2000;29(2):63–74.
[6] van Ijzendoorn DGP, Bovee J. Vascular tumors of bone: the evolvement of a classification based on molecular developments. Surg Pathol Clin 2017;10(3):621–35.
[7] Ko JS, Billings SD. Diagnostically challenging epithelioid vascular tumors. Surg Pathol Clin 2015;8(3):331–51.
[8] Pradhan D, Schoedel K, McGough RL, Ranganathan S, Rao UNM. Pseudomyogenic hemangioendothelioma of skin, bone and soft tissue-a clinicopathological, immunohistochemical, and fluorescence in situ hybridization study. Hum Pathol 2018;71:126–34.
[9] Krebs S, Monti S, Lewis N, Luciano A, Rastrelli L, Mannelli L. Lower-extremity pseudomyogenic hemangioendothelioma on bone scintigraphy and PET/CT. Clin Nucl Med 2017;42(5):383–5.
[10] Muheremu A, Ma Y, Huang Z, Shan H, Li Y, Niu X. Diagnosing giant cell tumor of the bone using positron emission tomography/computed tomography: a retrospective study of 20 patients from a single center. Oncol Lett 2017;14(2):1985–8.
[11] Xu JX, Yang L, Chen Y, Peeroo MW, Zhu XL, Fu YB, et al. Differential CT and MR imaging diagnosis between low- and high-grade malignant vascular tumors of bone. Springerplus 2016;5(1):1772.