

Epithelioid hemangioma of bone: a report of two special cases and a literature review

Qin Zhou¹ · Liangji Lu¹ · YanBiao Fu² · KeWei Xiang¹ · Leiming Xu¹

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Abstract Intraosseous epithelioid hemangioma (EH) is a rare intermediate vascular neoplasm, characterized by locally aggressive and rarely metastasizing behavior. Occasionally, EH of bone can behave strangely and may simulate malignant neoplasm. Here, we report two cases of EH of bone. Of interest was the fact that the computed tomography and magnetic resonance images from one case showed an osteolytic lesion in the right scapula, with multiple swollen lymph nodes in the right supraclavicular and axillary areas. Another patient exhibited a local recurrence in the cervical vertebrae. The initial radiological diagnosis of both cases was metastatic tumor. EH should be included in the differential diagnosis of a radiographic osteolytic lesion with an aggressive appearance. Also, we reviewed the literature that reported EH of bone and summarized their radiological appearances. The cases of EH of bone that exhibited involvement of regional or draining lymph nodes were also summarized.

Keywords Epithelioid hemangioma · Bone · Scapula · Computed tomography · Magnetic resonance imaging

Introduction

Epithelioid hemangioma (EH) is a locally aggressive neoplasm composed of small vessels lined by epithelioid

endothelial cells. Recently, EH of bone was classified as an intermediate vascular neoplasm and separated from hemangioma in the World Health Organization classification of bone tumors (4th edition) [1, 2]. Intraosseous EH is rare and most frequently presents in the long tubular bones, followed by the flat bones and vertebrae [3]. In contrast to conventional hemangioma, EH is locally aggressive: recurrence occurs in 8–10 % of cases [1, 3], and metastasizing behavior is extremely rare. The rarity of this tumor has led to diagnostic difficulties, especially only with radiographic resources of the lesion. More radiographic materials of intraosseous EH need to be reported and reviewed for a better differential diagnosis in radiology.

Generally speaking, EH of bone occurs as a benign tumor, and the treatment and prognosis differ from other malignant hemangioma tumors. Correct diagnosis of EH of bone is important before performing treatment. Here, we report two special cases of EH of bone, one with draining lymph node enlargement and another with local recurrence. The findings of preoperative imaging studies, including radiographs, computed tomography and magnetic resonance imaging, are described along with a review of the literature describing similar cases.

Case report

Case 1

A 30-year-old woman had no history of trauma or fracture in the right shoulder. She presented with pain and swelling in the right shoulder region that had lasted for almost 1 year. A physical examination revealed an ovoid mass in the right scapula. Laboratory test values were within the normal ranges.

Radiographs revealed an expansive and osteolytic lesion, focal cortical thinning and destruction involving the middle and superior region of the scapula (Fig. 1a). The CT

✉ Leiming Xu
Leimixu@163.com

¹ Department of Radiology, Second Affiliated Hospital, Zhejiang University College of Medicine, Hangzhou 310009, China

² Department of Pathology, Second Affiliated Hospital, Zhejiang University College of Medicine, Hangzhou 310009, China

scan clearly demonstrated a mass with a size of 4.3×3.5 cm. The mass was osteolytic and extensive, contained coarse trabecular bone and was surrounded with a rim of osteosclerosis. Thinning and breaching of the cortex of the scapula were also seen (Fig. 1b). MR images disclosed a mass on the superior side of the right scapular. On T1-weighted images, the mass was heterogeneously iso- and hyperintense to muscle with multiple linear low signal intensities (Fig. 1c). Axial T2-weighted image shows the lesion to be of heterogeneously high signal intensities within a linear low signal area. (Fig. 1d). Gadolinium-enhanced T1-weighted imaging revealed relatively heterogeneous enhancement; linear non-enhancing low signal lesions were also observed (Fig. 1e). Multiple swollen lymph nodes were seen in the right supraclavicular region (arrow in Fig. 1e).

The initial radiological diagnosis was metastatic tumor in the bone. An open biopsy was performed, and a diagnosis of EH of the scapula was made. The surgery consisted of extirpation of the tumor; en bloc resection

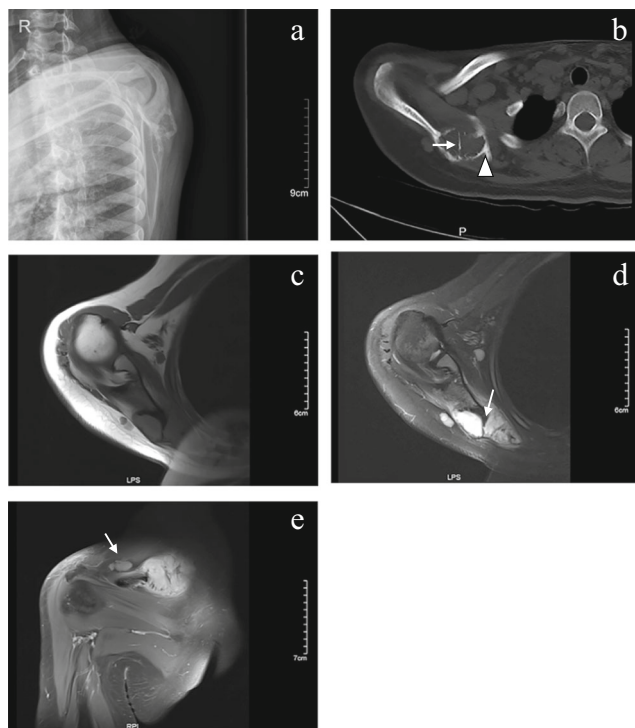


Fig. 1 a–e Case 1. A 30-year-old woman with EH of the scapula. **a** Lateral radiograph shows a lytic lesion on the right scapular region. **b** Axial CT image demonstrates an expansive and lobular lytic lesion on the right scapular region with coarse trabecular bone (arrow). Thinning and breaching of the cortex of the scapula was observed, surrounded by a rim of reactive sclerosis (arrowhead). **c** Axial T1-weighted MR image reveals an intraosseous lesion of heterogeneous iso- and hyperintensity to muscle. **d** Axial T2-weighted image shows the lesion to be of mixed high signal intensities within a linear low signal area (arrow). **e** Coronal fat-suppressed contrast-enhanced T1-weighted MR image demonstrates obvious enhancement within the lesion as well as swollen lymph nodes in the right supraclavicular region (arrow)

of the tumor was completed. Microscopy showed well-formed blood vessels lined by prominent epithelioid endothelial cells (arrow in Fig. 4a). The epithelioid endothelial cells protrude into the lumens in a tombstone-like fashion (arrow in Fig. 4b). This patient refused the lymph node biopsy. Therefore, the pathology feature of these swollen lymph nodes in this case was not known. At 6 months after surgery, follow-up showed that the patient was doing well with no significant pain or swelling in the right scapula area, and no limitations were reported.

Case 2

A 43-year-old man who had been diagnosed as having EH in the cervical vertebrae 4 years ago was treated using the curettage and posterior internal fixation procedures. After treatment, this patient had a good outcome. For the last year, he had experienced a progressively worsening pain in the neck. His body, arms and legs could not cope well with the activities of daily life. In addition, somatosensory abnormalities occurred.

Preoperative CT scan demonstrated a lytic lesion in the vertebral body (C2, C3) and the involvement of the left pedicle (Fig. 2a). The lesion was osteolytic and extensive, surrounded with a rim of osteosclerosis. Breaching of the

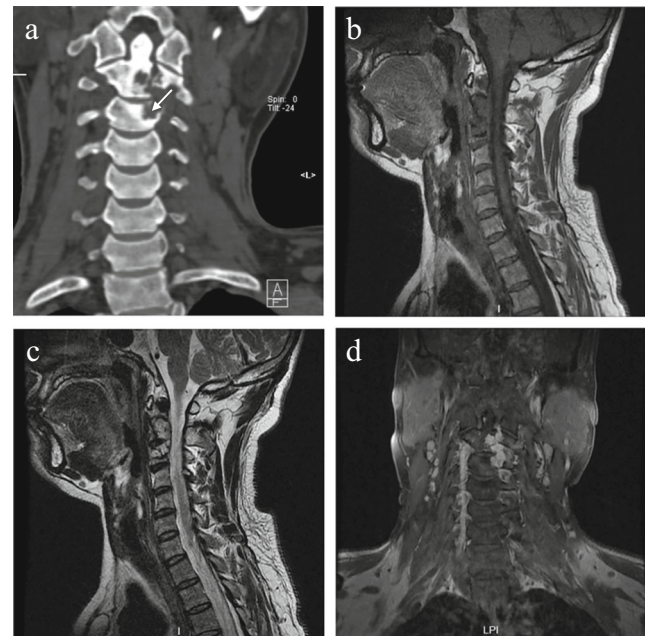


Fig. 2 a–d Case 2. A 43-year-old man with EH of cervical vertebrae. **a** Coronal CT image demonstrates a lytic lesion in the vertebral body and left pedicle of vertebrae C2–C3, surrounded with a rim of osteosclerosis (arrow). **b** Sagittal T1-weighted image disclose that the mass had heterogeneous isointensity relative to the muscle. **c** Sagittal T2-weighted image shows the lesion with hyperintensity. **d** Coronal fat-suppressed contrast-enhanced T1-weighted image reveals strong enhancement

cortex of the vertebral body was also seen. MR images disclosed that the mass had heterogeneous isointensity relative to the muscle on T1-weighted images, with hyperintensity on T2-weighted images (Fig. 2b and c). Gadolinium-enhanced T1-weighted imaging revealed strong heterogeneous enhancement (Fig. 2d). The initial radiological diagnosis was malignant tumor, but the histology of the tissue from the vertebrate curettage treatment indicated epithelioid hemangioma with chronic inflammatory infiltration. CT and MR images at a follow-up time of 4 years after curettage treatment showed tumor recurrence and enlargement (Fig. 3a–c); the spinal cord was obviously compressed (Fig. 3b and c). Surgery was not recommended because of the tumor's size and location. This patient received radiotherapy treatment; 4 months later, the MR images revealed that the tumor was still growing (Fig. 3d).

Needle biopsy was performed, and pathological examination revealed the recurrence of EH (Fig. 4c). Microscopy showed epithelioid hemangioma with prominent inflammatory infiltrate containing numerous eosinophils (arrow in Fig. 4c).



Fig. 3 a–d Case 2. Radiological images at a follow-up time of 4 years after curettage treatment. CT and MR images show tumor recurrence and enlargement. **a** Coronal CT image shows the nail with high density on the axis (black arrow). The vertebral body and left pedicle of vertebrae C2–C3 were damaged more seriously; a rim of reactive sclerosis can also be observed (arrow). **b** Sagittal T2-weighted MR image shows the spinal cord was obviously compressed (arrow). **c** Axial fat-suppressed contrast-enhanced T1-weighted image reveals the lesion with strong heterogeneous enhancement, and the spinal cord was compressed seriously (arrow). **d** Sagittal T2-weighted image shows that the lesion was enlarged with more severe spinal cord compression at a follow-up time of 4 months after radiotherapy

Discussion

No specific radiographic appearance has been reported to date concerning EH [4]. A lucent lesion usually presents on conventional X-rays and computed tomography scans with well-defined margins; however, they have a different appearance on MR images. In several cases, especially those involving small tubular bones, the expansion of the destroyed bone extended to the adjacent soft tissue [5, 6]. In our cases, the most confusing radiographic features for case 1 were focal cortical destruction as well as multiple swollen lymph nodes in the right supraclavicular and axillary areas and for case 2 were the recurrences. All of these factors resulted in the incorrect diagnosis of a metastatic tumor. Through retrospective analysis we found that when we observed radiographic features

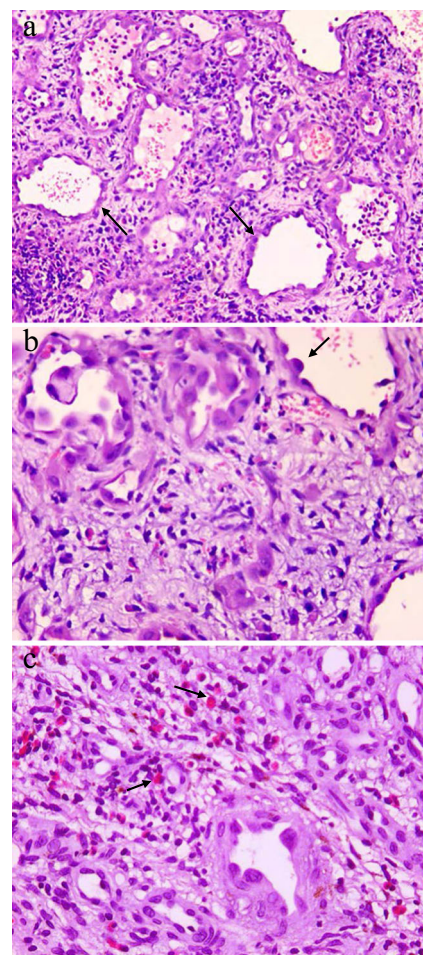


Fig. 4 a–c Microscopic pathological findings of epithelioid hemangioma. **a** Specimen from case 1 shows epithelioid hemangioma containing well-formed blood vessels lined by prominent epithelioid endothelial cells (arrow) [hematoxylin eosin (H&E) stain, $\times 200$]. **b** Specimen from case 1 clearly demonstrates the epithelioid endothelial cells protrude into the lumens in a tombstone- or spike-like fashion (arrow) (H&E stain, $\times 400$). **c** Specimen from case 2 demonstrates epithelioid hemangioma with prominent inflammatory infiltrate containing numerous eosinophils (arrow) (H&E stain, $\times 400$)

such as lytic expansile lesions surrounded by a rim of osteosclerosis, creating a sunburst soap-bubble appearance, containing coarse trabecular bone, high signal intensity on T2-weighted images and obvious degrees of enhancement on contrast T1-weighted images, a vascular tumor should be considered [7]. However, it is difficult to distinguish EH from other intraosseous vascular tumors including epithelioid hemangioendothelioma, endothelioma, epithelioid angiosarcoma as well as atypical intraosseous hemangioma. The final diagnosis depended on the findings at histopathological examination.

These two cases of EH of bone have an abnormal appearance, which makes them special. In case 1 in our study, multiple swollen lymph nodes were seen beside the lesion, located in the right supraclavicular and axillary areas. For those swollen lymph nodes, two possibilities may exist. One is the reactive hyperplasia (there may be an infected lesion). Another may be metastasis of the lesion in the right scapula, or, as some researchers pointed out, it is just the phenomenon of multifocality of EH [3]. Though lymph node enlargement in this case was not pathologically confirmed as metastasis, this phenomenon had misled the radiologist to diagnose this patient as having malignant tumor of bone. After reviewing the literature about EH of bone, we found three cases of EH of bone that exhibited involvement of a regional or draining lymph node (Table 1). Two of them were diagnosed as EH [3, 8], and the third was diagnosed as epithelioid hemangioendothelioma in the original case report [9]; however, Nielsen et al. in their study mentioned this case and considered that it “showed the features of EH” [3]. Two of the three patients underwent treatment with excision of the involved lymph node, and the pathological examination indicated EH in both cases [8, 9]. In Nielsen et al.’s case [3], the 42-year-old woman had EH in the metacarpal with axillary lymphadenopathy, and a biopsy was performed. Histological examination showed a single vessel lined by epithelioid endothelial cells in the lymph nodes.

Although EH is most commonly a solitary lesion, approximately 18–25 % of EH cases demonstrate multifocality [3, 10]. Very rarely, EH displays local nodal involvement and radiological examination reveals EH of bone with enlarged local draining lymph nodes, which might suggest metastasis. It is difficult to determine whether the involved local draining lymph nodes are the multifocal focus of EH or metastasis [3]. However, regardless of whether these cases are multifocality or metastasis, the patients presented in Table 1 have been followed up for a relatively long period and remain healthy.

Treatment of EH varies widely, ranging from biopsy to segmental resection. Most patients can be effectively treated with intralesional curettage. Regardless of the treatment EH patients receive, the prognosis is good [3]. In some cases, the EH in bone can result in a bad outcome [11], even though the tumor itself is benign, just like our case 2. The take-home

Table 1 Summary of clinical findings including the previously reported cases of EH of bone with local lymph node involvement

Case reported by	Age/gender	EH location/ size	Involved lymph nodes	Histological examination of involved lymph nodes	Treatment	Outcome
Floris et al. [8]	15/man	Second toe in the left foot	Left inguino-femoral region	Epithelioid hemangioma	Second ray resection of the left foot	36 months/FU, NED
Nielsen et al. [3]	42/woman	Second metacarpal	Left axillary lymph node	Reactive follicular hyperplasia with interfollicular eosinophilia and numerous plasma cells	Curettage/excision of lymph node	38 months/FU, NED
Chiriac et al. ^a [9]	33/woman	Local recurrence	Left axillary lymph node	Single vessel lined by epithelioid endothelial cells	Ray resection of the involved finger and axillary lymph node biopsy, radiation therapy	
		Head of the right tenth rib	Intercostal lymph nodes	Epithelioid hemangioendothelioma ^a	En bloc resection of the posterior portions of 9th and 10th ribs	32 months/FU, NED

^a This case was diagnosed as epithelioid hemangioendothelioma in the original paper, but in the review of Nielsen et al., the authors thought this case should be diagnosed as EH
FU follow-up, NED no evidence of disease

message from this patient is that while surgery is the chosen treatment for EH of the bone, particularly in the vertebrae, surgeons should be aware that local recurrence of EH (approximately 11 % [3]) might occur, and the surgical methods should be assessed carefully [12, 13].

It can be concluded that EH is an intermediate vascular neoplasm with distinctive vascular formative features that occasionally shows local-regional aggressive growth and rarely shows metastasizing behavior. EH involving the bone can present with a wide variety of radiological features. It can present as an expansive osteolytic lesion with cortical destruction, adjacent soft tissue tumor extension and local nodal involvement, mimicking a malignant bone tumor; this results in a challenge in differentiating EH from malignant neoplasms during diagnosis. Distinguishing EH of the bone from other malignant vascular tumors of the bone is important; their management and clinical outcome have major differences.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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