Epithelioid Angiosarcoma of the Tibia: A Case Report and Literature Review

Qianqian Yao

The Second Affiliated Hospital of Shandong First Medical University

Xubo Ge

the Fourth People's Hospital of Taian

Yinghua Gao

The Second Affiliated Hospital of Shandong First Medical University

Changqin Li

The Second Affiliated Hospital of Shandong First Medical University

jian qin (✉ sdqinjian@126.com)

The Second Affiliated Hospital of Shandong First Medical University

Research Article

Keywords: Epithelioid angiosarcoma, Tibia, Computed tomography, Magnetic resonance imaging

Posted Date: September 13th, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-827733/v1>

License: This work is licensed under a Creative Commons Attribution 4.0 International License. [Read Full License](#)

Abstract

Background Epithelioid angiosarcoma (EA) is characterized by epithelioid-like neoplastic cells. In fact, there is little literature regarding EA of bone. Accordingly, this study presents an imaging analysis of a 66-year-old man who suffered from EA in his right tibia.

Case presentation: A 66-year-old man developed right ankle pain four months prior to initial evaluation at our institute, which was progressively worsening for six days. All laboratory data were within the reference ranges. Plain radiographs and computed tomography (CT) revealed osteolytic lesions with multiple separations in the distal tibia, measuring 7.9 cm × 4.6 cm × 4.4 cm in size. The lytic lesions were ill-circumscribed and lacked marginal sclerosis. Punctate irregular calcifications and low-density areas were observed within lesion's areas. The cortical bone was irregularly thinned and was discontinuous. On magnetic resonance imaging (MRI), the lesions were heterogeneous. Immunohistochemically, the tumor cells expressed the vascular markers CD31, CD34, and factor VIII and revealed 5% positivity for Ki67. Finally, the patient was diagnosed with EA of bone.

Conclusions: We reported a case of EA that occurred in the right tibia and summarized the imaging features of EA by reviewing the literature. Although pathological examination remains the gold standard for diagnosing EA, specific imaging features may assist in diagnosis.

Background

Epithelioid angiosarcoma (EA) is a rare variant of angiosarcoma that is characterized by epithelioid-like neoplastic cells. These tumors are usually poorly differentiated and biologically aggressive [1]. In small series, EA has been reported to occur in various parts such as thyroid, skin, adrenal gland, and deep soft tissue, whereas bone-originated EA is relatively rare [2, 3, 4]. EA has a male predilection and is more prevalent in older individuals [1, 5], a population with a higher rate of metastatic carcinoma. The current study presents the imaging analysis of a 66-year-old man who suffered from EA in his right tibia, and its imaging features were summarized by reviewing the literature. The patient provided written informed consent. To our knowledge, this is the first report to analyze and summarize the imaging features of EA.

Case Presentation

A 66-year-old man was admitted at our institute with right ankle pain four months before initial evaluation, which was progressively worsening for six days. At rest, symptoms were alleviated, while exercise aggravated them. On physical examination, his right ankle was markedly swollen and firm, he suffered from local pressing pain, and a surgical scar was visible on the inside aspect of his right tibia. All laboratory data were within the reference ranges. The patient had a prior medical history of right lower limb trauma and underwent surgical treatment in 2011 at the age of 57. He smoked and consumed alcohol for about 30 years. He denied that there was no family or personal history of malignancy or autoimmune disease.

In the distal tibia, plain radiographs revealed osteolytic lesions with multiple separations (Fig. 1). The lytic lesions were ill-circumscribed and lacked marginal sclerosis. Some were accompanied by thinning and ballooning of bone cortices. Besides, cement was observed in the proximal tibia following a trauma nine years ago. As a result, a CT (computed tomography) scan of the right ankle was performed and revealed osteolytic and expansive destruction of bone in the distal tibia (Fig. 2), measuring 7.9 cm × 4.6 cm × 4.4 cm in size. The normal trabecular bone structure disappeared and was replaced by heterogeneous density soft tissue mass. Punctate irregular calcifications and low-density areas were also observed within lesion areas. The cortical bone was irregularly thinned and was discontinuous. The mass extended into adjacent soft tissue through the discontinuous cortical bone, and punctate calcifications were also observed. On ankle MRI (magnetic resonance imaging), the lesions appeared to be isointense or slightly hyperintense to the muscles on T1-weighted images, and fat-suppressed T2 images, centrally scattered occurring patchy, with a slightly low signal (Fig. 3). Edema signal could be observed in the surrounding soft tissue.

Based on the findings from imaging studies and the patient's age, the differential diagnoses of osteolytic bone lesions combined with calcification included chondrosarcoma, angiosarcoma, and bone metastasis.

During operation, brown-red tumor tissue was observed in medullary cavity, which was sandy, soft, and partly calcified. For pain relief and histological diagnosis, curettage of the lesion in the distal tibia and artificial bone grafting were performed. Microscopically, the resected tumor cells exhibited nestlike epithelioid features between trabeculae with prominent nucleoli and abundant cytoplasm. Numerous red blood cells were also discovered between the epithelial nests. Immunohistochemically, the tumor cells expressed the vascular markers CD31, CD34, and factor VIII and revealed 5% positivity for Ki67 (Fig. 4). Finally, the patient was diagnosed with EA of the bone. The patient was fully recovered and was discharged.

Discussion And Conclusions

EA is relatively rare and only accounts for < 1% of primary bone

tumors [6]. Since EA did not exhibit characteristic performance in imaging, the definitive diagnosis is done using pathological examination [7]. The immunohistochemical examination relies on applying vascular markers for EA diagnosis, including CD31, CD34, and factor VIII [8, 9]. The most important vascular marker is CD31, which is considered to provide a relatively high index of sensitivity and specificity [10]. Factor VIII and CD34 markers are useful for diagnostic studies. Desphande et al. examined ten patients and discovered that 75% of them tested positive for factor VIII, and 40% tested positive for CD34 markers [1].

Although pathological examination remains the gold standard for diagnosing EA, specific imaging features may assist in diagnosis. To our knowledge, few reports have discussed EA imaging characteristics.

We searched the Pubmed and found 6 previously reported cases of primary EA of bone, the inclusion criteria were that they underwent CT and MRI at the same time. Including our case, clinical information and imaging features of the 7 cases are summarized in Table 1. There were four men and three women, who ranged in age from 31 to 76 years (mean, 59 years). The maximum diameter of lesions was greater than or equal to 5cm. More than half of the cases presented the disease in the lower limb(5/7), two were located in the femur. In the present case, the imaging examination mainly revealed osteolytic and expansive destruction of bone in the distal tibia, which were similar to that reported in the 6 cases. Besides, the cortical bone was thinning and discontinuous(7/7). The normal trabecular bone structure disappeared and was replaced by heterogeneous soft tissue. Phlebolithlike calcifications or hemorrhaging support this diagnosis of EA. However, only half of the cases(4/8) showed signs of calcifications and hemorrhaging at the same time. In CT and MRI scans, cortical breakthrough(6/7), no marginal sclerosis(5/7) and soft tissue extension(6/7) indicated the presence of an aggressive, high-grade bone tumor.

Malignant tumors of cartilage origin, such as chondrosarcoma, must be distinguished. While it is difficult to distinguish from metastases that cause osteolytic changes on imaging, it is easy to identify based on clinical symptoms and primary lesions.

This study aimed to help radiologists improve their EA understanding. Together, X-ray, CT, and MRI scans may effectively diagnose epithelioid angiosarcomas by eliminating a differential diagnosis of other osteolytic lesions masses of the bone.

Abbreviations

EA
Epithelioid Angiosarcoma
CT
Computed Tomography
MRI
Magnetic Resonance Imaging

Declarations

Ethics Approval and Consent to Participate

As it is a case report, ethics approval is unnecessary after consulting the Ethics Committee of The Second Affiliated Hospital of Shandong First Medical University.

Consent to publish

Written informed consent was obtained from the patient and his wife for publishing this case report and any accompanying images.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Competing interests

The authors declare no competing interests.

Funding

This study was funded by Academic Promotion Programme of Shandong First Medical University (No.2019QL017).

Authors' Contributions

All authors read and approved the final manuscript. QQY reviewed the medical record and wrote the manuscript. XBG and YHG performed the histopathological diagnosis and immunohistochemistry. CQL performed the imaging diagnosis. JQ designed the case report and revised the manuscript for important intellectual content.

Acknowledgments

The authors would like to thank the patient's family for giving consent.

Authors' Information

^a Department of Radiology, The Second Affiliated Hospital of Shandong First Medical University, Taian, Shandong, 271000, China.

^b Department of Radiology, The Fourth People's Hospital of Taian, Taian, Shandong, 271000, China.

^c Department of Pathology, The Second Affiliated Hospital of Shandong First Medical University, Taian, Shandong, 271000, China.

References

1. Deshpande V, Rosenberg AE, O'Connell JX, et al. Epithelioid angiosarcoma of the bone: a series of 10 cases. *Am J Surg Pathol*. 2003;27(6):709–16.
2. Li Y, Zou X, Chang X, et al. Right femoral pathological fracture caused by primary bone epithelioid angiosarcoma: Case report. *Medicine (Baltimore)*. 2017;96:e6951.
3. Wang J, Zhao M, Huang J, et al. Primary epithelioid angiosarcoma of right hip joint: A case report and literature review. *Medicine*. 2018;97(15):e0307.
4. Lv L, Xu P, Shi Y, et al. Imaging features of soft tissue epithelioid angiosarcoma in the lower extremity: A case report. *Oncol Lett*. 2016;11(5):3457–60.
5. Fletcher CDM, Beham A, Bekir S, et al. Epithelioid angiosarcoma of deep soft tissue: a distinctive tumor readily mistaken for an epithelial neoplasm. *Am J Surg Pathol*. 1991;15(10):915–24.
6. Mendeszoon MJ, Mendeszoon ER Jr, Rasmussen S, et al. Epithelioid angiosarcoma of the talus. *J Foot Ankle Surg*. 2011;50(1):87–92.
7. Markaki S, Kokka H, Kyparidou E, et al. Primary vascular bone sarcomas. A clinicopathological and immunohistochemical study of two cases. *Arch Anat Cytol Pathol*. 1990;38(4):163–7.
8. Oc Y, Kilinc BE, Ertugrul R, et al. Epithelioid Angiosarcoma in Femur: A Case Presentation. *World J Oncol*. 2017;8(6):196–8.
9. Chen M, Zhang W, Qu J, et al. Epithelioid angiosarcoma of the ilium: a case report. *Int J Clin Exp Pathol*. 2014;7(12):9099–103.
10. Lang J, Chen L, Chen B, et al. Epithelioid angiosarcoma of the spine: A case report of a rare bone tumor. *Oncol Lett*. 2014;7(6):2170–217.
11. Sakamoto A, Takahashi Y, Oda Y, et al. Aggressive clinical course of epithelioid angiosarcoma in the femur: a case report. *World J Surg Oncol*. 2014;12:281.
12. Bürk J, Gerlach U, Baumann T, et al. Epithelioid angiosarcoma of the scapula. *In Vivo*. 2010;24(5):783–6.

Tables

Table 1

[illegible]

Figures

Figure 1

anteroposterior (A) and lateral (B). Radiographs showing the osteolytic lesion with multiple separations in the distal tibia.

Figure 2
 axial (A), coronal (B), sagittal (C). CT showing osteolytic and expansive destruction of bone in the distal tibia. Punctate irregular calcifications were observed within lesion's areas. The cortical bone was irregularly thinned and was discontinuous.

Figure 3
 MRI, sagittal fs PD (A), sagittal T1 FSE (B), coronal fs PD (C). On ankle MRI, the lesions showed isointense or slightly hyperintense to the muscles on T1-weighted images and fat-suppressed T2 images, centrally scattered occurring patchy and slightly low signal. Edema signal could be observed in the surrounding soft tissue.

Figure 4

Immunohistochemically, the tumor cells expressed the vascular markers CD31 (A) and CD34 (B). Microscopically (C), the resected tumor cells appeared to have nestlike epithelioid features between trabeculae with prominent nucleoli and abundant cytoplasm. Numerous red blood cells were also found between the epithelial nests. The magnification is 10×40.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- [CAREchecklistpdf..pdf](#)