

Case report

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Epithelioid hemangioendothelioma of the brachial artery: A rare benign vascular tumor

Enes Ozsozgun; Esin Kurtulus Ozturk*; Saffet Ozturk

Department of Radiology, Ankara Etlik City Hospital, 06170, Turkey.

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***Corresponding Author:** Esin Kurtulus Ozturk, Department of Radiology, Ankara Etlik City Hospital, 06170, Turkey. Tel: +90.312 797 00 00, E-mail: e.kurtulus@hotmail.com

Introduction

Epithelioid hemangioendotheliomas are uncommon vascular tumors presenting with different biological behavior and clinical features. It is a pathology that should be diagnosed early because it is seen at earlier ages and can cause distant organ metastases. Due to its rarity, it is often not included in the differential diagnosis of soft tissue masses [1]. In this case, we aimed to present a malignant epithelioid hemangioendothelioma presenting with palpable swelling.

Case presentation

A 75-year-old male patient was admitted to the orthopedics clinic due to swelling on the medial side of his right arm. On ultrasonography, a 32x20x25mm ill-defined, lobulated soft tissue mass surrounding the brachial artery located medial to the right arm was observed. Significant peripheral vasculariza-

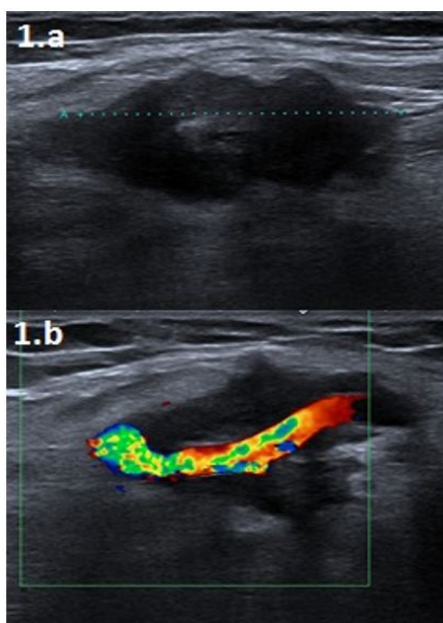


Figure 1: B-mod and colour Doppler ultrasound images (a and b) show an irregularly contoured soft tissue mass surrounding the brachial artery medially in the right arm midsection.



Figure 2: Axial upper extremity CT angiography image shows irregular course of the brachial artery within the mass (arrow).

tion was noted on Doppler examination (Figure 1). Irregular course of the brachial artery in the mass was observed on upper extremity Ct angi (Figure 2). The magnetic resonance imaging scan revealed a heterogeneous mass with irregular contours, surrounding the brachial artery and indistinguishable from the brachial vein (thrombosis), restricting diffusion, and showing peripheral irregular contrast enhancement was observed (Figure 3). The imaging findings suggested a vascular tumor but with difficulty in differentiating malignant or benign. After surgical excision of the lesion, the histopathology was reported as malignant epithelioid hemangioendothelioma originating from the brachial artery.

Discussion

Epithelioid hemangioendothelioma was first described in 1982 and was previously known as intravascular bronchioalveolar tumor. Histologically, it consists of pleomorphic spindle epithelioid cells with high mitotic activity. It contains various immunohistochemical endothelial markers (CD31, CD34, Fli-1 etc.). In its etiopathogenesis, Some molecules and chromo-

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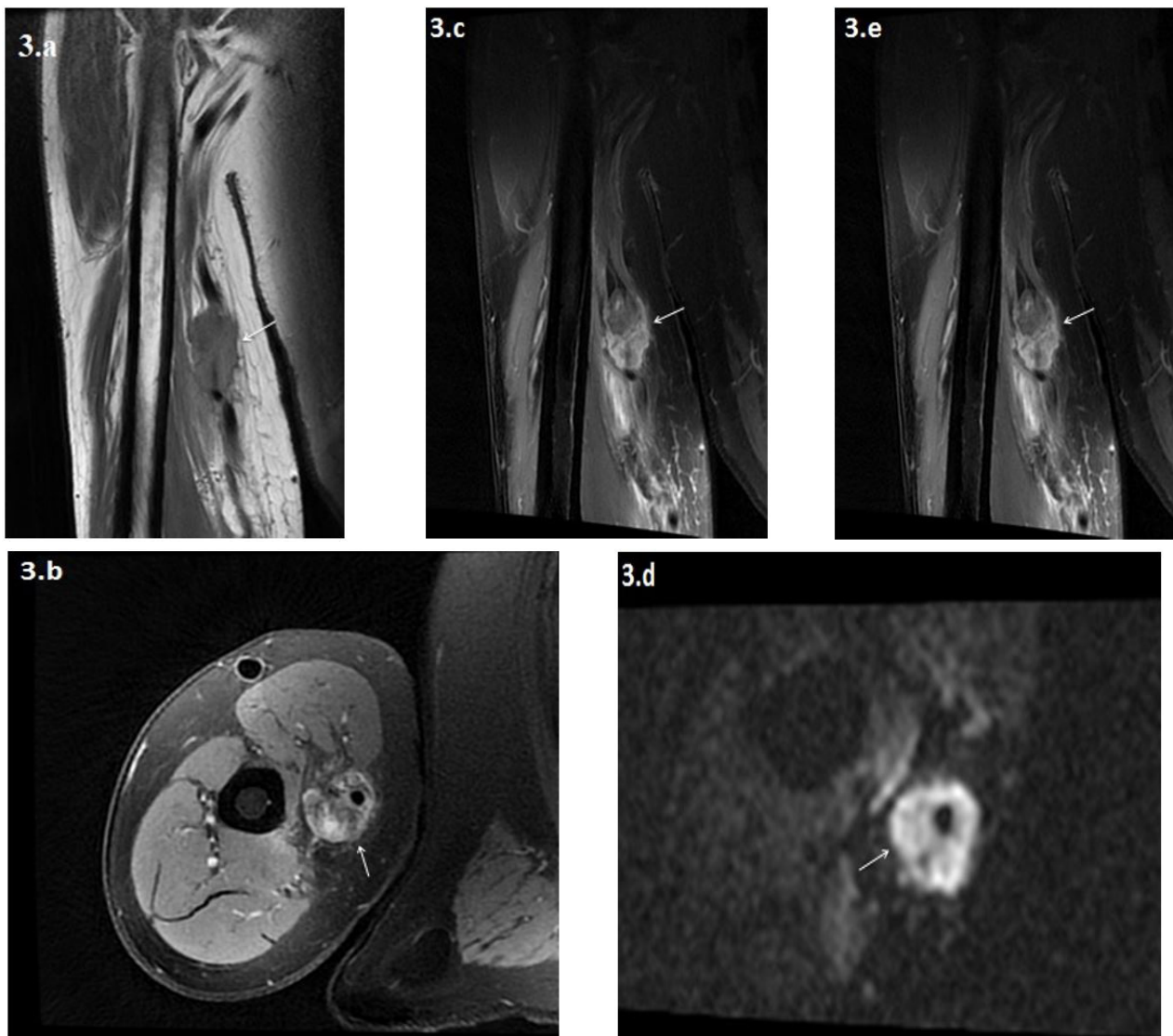


Figure 3: Axial T1W (a), fat-sat axial (b) and coronal (c) PD images show a heterogeneous mass lesion (arrow) with irregular contours surrounding the brachial artery, but where the brachial vein cannot be distinguished (thrombosed). Diffusion (d) and contrast-enhanced coronal T1W(e) images show that the lesion is diffusion-restricted and peripherally irregularly enhanced.

TYPE OF CONTRIBUTION	DESCRIPTION	CONTRIBUTORS
CONCEPTION	Constructing an idea or hypothesis for research and/or manuscript	EKO
DESIGN	Planning methodology to reach the conclusion	EKO, EO
SUPERVISION	Organising and supervising the course of the project or the article and taking the responsibility	EKO
FUNDINGS	Providing personnel, environmental and financial support and tools and instruments that are vital for the project	SO
MATERIALS	Biological materials, reagents and referred patients	EKO
DATA COLLECTION AND/OR PROCESSING	Taking responsibility in execution of the experiments, patient follow-up, data management and reporting	EO, SO
ANALYSIS AND/OR INTERPRETATION	Taking responsibility in logical interpretation and presentation of the results	EKO, EO
LITERATURE REVIEW	Taking responsibility in this necessary function	EKO, EO
WRITER	Taking responsibility in the construction of the whole or body of the manuscript	EO
CRITICAL REVIEW	Reviewing the article before submission not only for spelling and grammar but also for its intellectual content	SO, EKO

somal mutations are thought to cause the stimulation of angiogenesis and endothelial cell proliferation [1].

Epithelioid hemangioendotheliomas are rare tumors of vascular origin that usually occur in young adults. In the literature, there are disagreements about the incidence of the most frequently involved organ and tumor. Epelboym et al. stated that the most frequently involved organs were liver, lung and bone, respectively, while in the study of Requena and Kutzner, more than half of the cases were related to vascular structures [2,3]. Epithelioid hemangioendotheliomas are mostly seen in the vascular structures of the venous system (most commonly the femoral vein) [4].

More rarely, it presents as a painful mass lesion originating from arterial vascular structures. It can be confused with aneurysms and arteriovenous malformations as it wraps around the vascular structures in the form of a sheath [5]. Depending on the tumor's occlusion of the vascular structure, findings such as edema and thrombophlebitis may be observed. Distant organ metastases are most common to the lung [4,6]. Fewer cases were reported in the upper extremity arteries than in the lower extremity arteries. Cases of epithelioid hemangioendothelioma seen in the superficial palmar arch, ulnar artery and radial artery have been reported in the literature [7-9].

On ultrasound, it is observed as a hypoechoic mass surrounding the vascular structure to a certain extent, adjacent to the vascular structure. In Doppler ultrasound examination, it is in the form of a mass with vascular blood supply that proves angiogenesis. Venous thrombus can be observed in masses originating from the venous system. Similar to US findings, it is observed as a non-specific soft tissue mass in CT examination. CT angiography can be used to guide surgical excision, evaluate the intraluminal extension of the mass and determine its borders. On MRI, it shows low-to-moderate contrast on T1-weighted images, hyperintense on T2-weighted images, and intense homogeneous enhancement on contrast-enhanced images, also with restriction on diffusion-weighted images. Compared to other imaging methods, MRI is more valuable in evaluating the invasion of the mass into the vascular structure and its relationship with the surrounding structures. On angiography, it is observed as a well-circumscribed dense mass with early venous drainage [1,2]. PET CT can be used to evaluate distant organ metastases. Since there are no typical imaging findings, the diagnosis is always made by pathology. Main treatment is surgery and can be combined with radiotherapy. In surgical treatment, resection of the affected vascular structure and appropriate graft application are performed. Chemotherapy can also be used in metastatic cases. Recently antiangiogenic drugs and monoclonal antibodies have been tested for usability [1,10]. The aim of the treatments is to prevent local recurrences and increase survival. Since it is a rare pathology and there are few case examples in the literature, there is no consensus on its clinical course, imaging findings and treatment.

Conclusion

Epithelioid hemangioendotheliomas are intensely enhanced

vascular lesions with low or 'borderline' malignant features between benign hemangioma and malignant hemangiosarcoma, and their clinical and radiological findings are nonspecific. It should be kept in mind in the differential diagnosis of soft tissue masses especially extending into the vascular structure and with intense homogeneous contrast enhancement. Imaging modalities especially MRI play a crucial role in identifying vascular tumors and directing the therapy. More and more patients are undergoing MRI for soft tissue mass, thus necessitating the interpreting physicians and radiologists to be familiar with MRI findings differentiating from malignancy.

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