

## Epithelioid Hemangioendothelioma of the Distal Radius: A Case Report

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Epithelioid hemangioendothelioma is a rare vascular tumor with cytologic behavior between angiosarcoma and hemangioma. We present the case of a 58-year-old male with primary epithelioid hemangioendothelioma of the distal radius measuring 6.2 x 5 cm with extension into the pronator quadratus and brachioradialis muscles. We discuss our approach to performing a limb-sparing resection combined with reconstruction to preserve upper extremity function. A review of the clinical, radiographic, and pathologic features of epithelioid hemangioendothelioma is also presented.

### Introduction

Hemangioendotheliomas represent a type of vascular tumor with cytologic behavior that is in between an angiosarcoma and hemangioma. There is variability in the predilection of these tumors to recur and metastasize. The histologic diagnosis of epithelioid hemangioendothelioma (EH) is difficult to differentiate from other vascular tumors, epithelioid tumors, or histiocytic tumors [1, 2].

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Abbreviations: EH, epithelioid hemangioendothelioma; CT, computed tomography; MRI, magnetic resonance imaging

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However, their aggressive and malignant behavior makes distinguishing this particular histologic subtype paramount.

Patients commonly present with nonspecific signs and symptoms, most commonly pain and swelling; few present with pathologic fractures [3]. The highest incidence of EH occurs in the second decade, with multicentric disease present in 55% of patients [4]. Previous series have reported that EH occurs at an earlier age than other vascular tumors of bone [4, 5]. Theoretically, these lesions can arise from any vascularized structure, but have a predilection for lung, liver, and soft tissues of the extremity.

Epithelioid hemangioendothelioma typically involves a single anatomic region or extremity, although multiple bones may be involved in 40% of cases [4]. When the upper extremity is involved, the humerus is the most common location. Radiographically, EH classically produces a lytic lesion on plain films. Cortical destruction and cortical expansion are infrequently seen [3]. Non-osseous soft tissue masses present in up to 40% of cases, although the majority of lesions are restricted to bone [6]. The aggressive radiographic characteristics of EH raises the possibility of a malignant lesion in the differential diagnosis.

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### Case Report

The patient is a 58-year-old right-hand dominant male, who was referred to our institution for evaluation of a right forearm mass. The patient described vague forearm pain and swelling that began three months prior. He otherwise denied numbness, weakness, or functional loss of the hand. The patient subsequently fell and fractured his distal radius at the site of the swelling. Plain radiographs revealed an associated lytic lesion. He was treated at an outside institution for the pathologic fracture. An incisional biopsy of the mass was inconclusive.

Neurovascular examination on presentation revealed a normal distal extremity and a well-healed biopsy incision. Past medical history was significant for diabetes and dialysis-dependent renal failure with subsequent kidney transplantation in 2001. His laboratory studies were otherwise unremarkable. Plain radiographs demonstrated an expansile lesion within the distal one-third of the radius (Figure 1).

Magnetic resonance imaging was obtained to delineate bone and soft tissue involvement. The lesion demonstrated a heterogeneously enhancing, aggressive-appearing lesion (Figure 2).

Ultrasound guided biopsy was performed. An 18-gauge



Figure 1. Preoperative oblique radiograph of the right distal forearm demonstrating an epithelioid hemangioendothelioma. A lytic, septated, expansile lesion is present without identifiable matrix. A pathologic fracture is visible along the radial border.

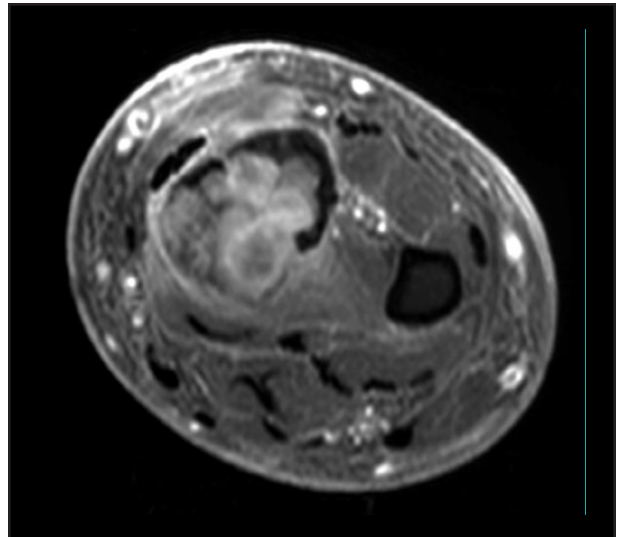


Figure 2. Sagittal (A, left) and axial (B, above) T1-weighted fat suppressed MR images with gadolinium enhancement demonstrate a heterogeneously enhancing, aggressive lesion that traverses the bone cortex and is encased by the pronator quadratus and brachioradialis muscles.

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