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Epithelioid hemangioma occurring in the radial styloid of a 17-year-old boy—an unusual presentation of an uncommon neoplasm

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ABSTRACT

Presented is a case of epithelioid hemangioma (EH) of bone occurring in the radial styloid of a 17-year-old boy. EH is a benign vascular tumor whose name and classification have changed over the years, adding potential confusion to an already existing diagnostic challenge. Overlapping imaging and histopathologic features with malignant vascular neoplasms and occasional aggressive clinical features have resulted in misdiagnoses and inappropriate treatment. The goal of this case report is to raise awareness of EH and related vascular neoplasms.

1. Introduction

Epithelioid hemangioma (EH) is an uncommon benign vascular tumor that occurs most commonly in the skin and subcutis and is only rarely described in bone [1]. The vast majority of osseous EH are solitary and occur in the diaphysis or metaphysis of long tubular bones with a predilection for the lower extremity; only 3% of EH occur in the long bones of the upper extremity [1]. Lesions are typically lytic and well circumscribed but a surrounding zone of sclerosis and cortical expansion may be present. Cortical destruction and thick periosteal new bone formation have been observed as well but are rare. Males and females are affected with equal frequency. The peak incidence occurs in the adult years (third through sixth decades) with only a few cases reported in children or adolescents. The mean age of presentation is 34 years [1–4].

In this paper, we present a case of EH occurring in the radial styloid of a 17-year-old boy. Both the location of the tumor and the age of the patient are unusual. While prospectively evaluating this patient and all of his imaging studies, a vascular neoplasm such as EH was not considered. Chondroblastoma was the leading diagnostic consideration and other entities such as giant cell tumor, Brodie's abscess, and eosinophilic granuloma (Langerhans cell histiocytosis) were included in the differential. The goal of this paper is to raise awareness of EH and other related vascular neoplasms and to illustrate the imaging and histologic features of the condition. The correct identification of EH is important as its treatment and prognosis differ from those of other low- and high-grade vascular neoplasms.

2. Case report

A 17-year-old male baseball pitcher presented with several weeks of increasing discomfort in and around his distal right radius. A focused clinical exam of his right wrist revealed tenderness to palpation over the radial styloid without skin changes, edema, or swelling. Radiographs of the right wrist demonstrated the presence of a well-circumscribed oval lytic lesion in the radial styloid surrounded by a faint rim of sclerosis. The lesion extended to the subarticular surface of the distal radius (Fig. 1). There was no appreciable intralesional matrix calcification. There was no cortical destruction or expansion, pathologic fracture, periosteal new bone formation, or radiographic evidence of associated soft tissue mass. The joint spaces were normal.

Magnetic resonance imaging (MRI) was subsequently performed (Fig. 2). The lesion measured 17 mm×4 mm×15 mm in size. On the T1-weighted images, the tumor was predominantly isointense to muscle with a few small, scattered central hyperintense foci. On the T2-weighted images, the lesion was predominantly hyperintense with a few central foci of hypointense signal. The Gadolinium-enhanced fat-saturated T1-weighted images demonstrated homogeneous enhancement of the lesion. Surrounding T2 hyperintense signal and abnormal enhancement attributable to marrow edema and inflammation were present as well. The cortex was intact. Given the patient's age, the imaging characteristics of the lesion on radiographs and MRI, and the clinical presentation, chondroblastoma was the

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favored diagnosis. Giant cell tumor, Brodie's abscess, and eosinophilic granuloma were included in the differential as well.

The patient was taken to the operating room and intralesional curettage was performed. Gross examination of the excised specimen did not reveal the chondroid matrix expected in the setting of chondroblastoma. Instead, the intraoperative frozen analysis revealed vascular features. This was inconsistent with either chondroblastoma or the other entities on our list of differential diagnoses. Since the final pathology was still uncertain at the time of surgery, the remaining defect was packed with antibiotic laden cement and hardware was not inserted.

Hematoxylin and eosin (H&E) and immunohistochemical stains led to the final histopathologic diagnosis of EH. Fig. 3a is an H&E stain demonstrating well-formed vascular channels with hypertrophic enlarged epithelial cells that protrude slightly into the vascular lumina. Note the lack of any significant cytologic atypia. Fig. 3b shows other typical histologic features of EH: cords and strands of plump endothelial cells embedded within a loose connective tissue stroma. A characteristic intracytoplasmic vacuole containing an erythrocyte is present. Immunohistochemical stains (data not shown) demonstrated that the tumor cells were positive for the endothelial markers CD31 and FLI1, confirming the vascular nature of the lesion.

At last check-up 5 months after the surgery, the patient was doing well. He had returned to pitching and reported no significant pain or swelling and no limitations. Radiographs obtained at that time show cement within the curettage defect with no evidence of residual or recurrent disease and no new radiographic abnormality.

3. Discussion

Most vascular tumors, benign and malignant alike, are readily diagnosed as such based on the histological features of vascular spaces formation and the expression of endothelial markers. However, the classification of vascular bone tumors remains challenging due to significant morphologic overlap between benign and malignant entities. Atypical histologic features such as solid growth pattern, epithelioid change, and spindle cell morphology complicate their diagnosis [2].

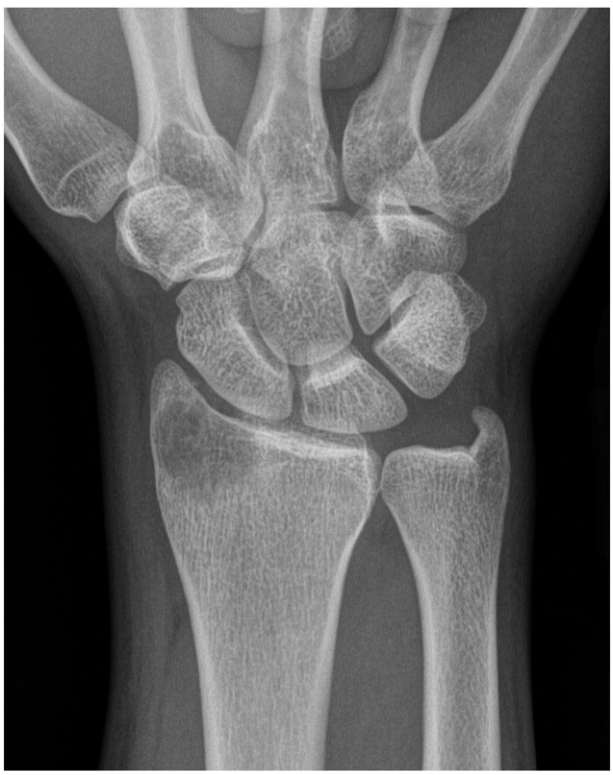

Fig. 1. PA radiograph of the right wrist demonstrates a well-circumscribed oval lytic lesion in the radial styloid surrounded by a faint rim of sclerosis. The lesion extends to the subarticular surface of the distal radius. There is no appreciable intralesional matrix calcification. There is no cortical expansion or destruction, pathologic fracture, periosteal new bone formation, or radiographic evidence of associated soft tissue mass. The joint spaces are normal.

EH is a well-recognized distinct clinicopathologic entity, which was previously designated as angiolymphoid hyperplasia with eosinophilia and histiocytoid hemangioma. The World Health Organization classifies EH as a benign neoplasm [3]. However, even to this day, a minority of investigators maintain that EH is not a unique histopathologic entity but, rather, a misdiagnosed hemangioendothelioma—a tumor with malignant potential [2,3]. The continuing debate over this distinction impacts the assessment of patient outcomes after treatment for vascular bone tumors. Old literature reporting higher than expected success rates in the treatment of hemangioendothelioma is most likely flawed due to unintentional inclusion of EH patients in the study population [5].

EH affects males and females equally. Patients are typically in their third through sixth decades of life [1]. There are rare cases, however, of EH occurring in children as young as 7 years of age [4]. EH typically presents as a painful mass and most commonly affects the diaphysis or metaphysis of long tubular bones. Rarely, the tumor occurs in the epiphysis of a long bone [4]. One study of 50 EH cases by the Nielsen group demonstrated that 40% of cases involved long tubular bones; 18%, distal lower extremities; 18%, flat bones; and 16%, vertebrae [3]. Another compilation of EH cases by the Wegner group demonstrated a skeletal distribution of 24% in the distal femur, 13% in the proximal tibia, and only 3% in the upper extremity long bones [1].

While EH is most commonly a solitary lesion, approximately 25% of EH cases demonstrate multifocality. This is thought to be due to multicentric primary disease rather than metastatic involvement. By contrast, roughly 50% of epithelioid hemangioendothelioma cases display multifocality [3,4]. Very rarely, EH displays local nodal involvement—a phenomenon known as "benign metastases" that is also observed with giant cell tumors [2].

EH is typically a well-circumscribed lytic lesion with a surrounding rim of reactive sclerosis. Bony expansion may be present. Rarely, EH can cause focal cortical destruction with adjacent soft tissue tumor extension. This latter presentation is more common when the lesion occurs in small tubular bones. When cortical destruction is present, thick reactive periosteal new bone formation is usually present [1,3]. A sclerotic appearance has been shown as well, occurring in the spine [6]. By contrast, epithelioid hemangioendothelioma is a poorly demarcated lesion with more infiltrative margins. While periosteal reaction is still infrequently seen in the latter, a small associated soft tissue component is present more commonly, occurring in approximately 40% of cases [2].

Histologically, the main differential diagnosis of EH includes epithelioid hemangioendothelioma and epithelioid angiosarcoma [2]. EH can be differentiated from epithelioid angiosarcoma rather easily based on the absence of cellular atypia or increased mitotic activity. The differentiation between EH and epithelioid hemangioendothelioma is more challenging. The following histopathologic features favor EH: the presence of multiple well-formed vessels lined by epithelioid cells that protrude into the vascular lumina in a "tombstone" fashion; mitoses are infrequent and, when present, appear normal; absence of cellular atypia; abundant, deeply eosinophilic cytoplasm that occasionally contains vacuoles with intact or fragmented erythrocytes; loose connective tissue stroma that may contain a prominent inflammatory infiltrate rich in eosinophils; lack of hyalinized, myxoid, or chondroid matrix [1,3]. On the other hand, epithelioid hemangioendothelioma is comprised of cells that show a greater degree of atypia and are arranged in cords. Importantly, unlike EH, epithelioid hemangioendothelioma cells are enmeshed in hyalinized or chondroid matrix [3]. A t(1;3), chromosomal translocation has recently been shown to be a specific genetic marker of hemangioendothelioma and can assist in the