Epithelioid Hemangioma of the Distal Humerus With Pathologic Fracture

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abstract

Epithelioid hemangioma is a rare tumor that can have bone involvement. Its clinically and radiographically aggressive appearance mimics a malignant neoplasm. Although epithelioid hemangioma has been described as having an aggressive appearance on magnetic resonance imaging (MRI) and plain radiographs, this is the first reported case of pathologic fracture associated with this lesion to our knowledge. This article describes a case of epithelioid hemangioma involving the distal humerus, which initially presented with progressive pain and fracture of the lateral condyle. The aggressive appearance on plain radiographs and MRI suggested a malignant bone tumor. This preliminary diagnosis was confirmed due to the presence of local lymph node spread on positron emission tomography/computed tomography. After a core needle biopsy revealed nondiagnostic tissue, rather than performing a wide resection based on a presumptive malignant diagnosis, we followed the standard diagnostic algorithm and performed an open biopsy with temporary internal stabilization. The tissue sample was adequate and revealed a diagnosis of epithelioid hemangioma. Based on this finding, we were able to proceed with surgical management, including curettage of the lesion, placement of a bone graft, and internal fixation, rather than a wide resection with elbow joint replacement. This article emphasizes the need for careful adherence to the diagnostic algorithm for musculoskeletal tumors. In doing so, a definitive diagnosis was reached, and our patient was able to resume his occupation as a laborer without the restrictions that would have accompanied elbow arthroplasty.
Epithelioid hemangioma, previously termed histiocytoid hemangioma and angiolymphoid hyperplasia with eosinophilia, is a rare tumor that can have bone involvement. The radiographic appearance of epithelioid hemangioma ranges from a lytic lesion confined to the bone with sclerotic margins to a mixed lytic and sclerotic lesion with intraslesional bone formation.\(^1,2\) Cortical bone thinning has also been reported, as well as expansion of the cortex on imaging, which are aggressive radiologic features often seen with malignant lesions.\(^3,4\)

Few case reports evaluating this benign tumor have been published in the literature. This tumor can present as cutaneous skin lesions accompanying bone tumors.\(^5\) One recent report of multifocal disease involving the tibia and talus was presented with evidence of reactive bone formation.\(^9\) After development of subsequent lesions, new biopsies were obtained, and the diagnosis was changed to epithelioid hemangioma.

A recent report of multifocal disease involving the tibia and talus was presented with evidence of reactive bone formation.\(^9\) Previously, multifocal disease with similar findings had also been documented in a child with tibia and femur involvement.\(^10\) Contiguous bone involvement has also been reported in the literature, with a single lesion involving the tenth rib and ninth and tenth vertebral bodies.\(^11\) One other case of involvement of the distal humerus has been reported.\(^12\) Although this report indicated aggressive radiologic features, no pathologic fracture or lymph node spread was present as noted in our report. A small series documenting pathology, appearance, and radiographic follow-up of various treatment modalities has been reported, but no functional data were presented.\(^13\) This report also noted that local lymph node involvement could be associated with this lesion but did not require treatment.

**Case Report**

A 61-year-old male laborer presented to the orthopedic oncology clinic with a 2-month history of pain and swelling about the right elbow. He was initially seen by his primary care physician and was referred to an orthopedic surgeon. Radiographs of the elbow showed a primarily lytic lesion of the distal lateral humerus. This rapidly evolved into a painful, stiff elbow, with soft tissue swelling and a mass. He was referred to an oncologist and an orthopedic oncology specialist for further workup. At evaluation, the patient was taking narcotics for severe pain, with little relief. The patient was self-splinting and had limited use of his right upper extremity due to pain and swelling. He had no constitutional symptoms. His medical and surgical history were negative for prior malignancy.

On physical examination, significant edema of the right upper extremity from the mid-arm to the mid-forearm was present. The patient had tenderness to palpation of the elbow over the lateral aspect. Range of motion (ROM) was limited from 30° to 60° and was painful through this small arc of motion. He had decreased strength, with wrist extension and grip at 4/5. Radiographs showed a lytic lesion of the distal humerus with posterior and lateral cortical loss (Figure 1). Magnetic resonance imaging (MRI) (Figure 2) showed a mass involving the lateral aspect of the distal humerus extending to the articular surface. The lateral and posterior cortices had significant thinning, with breakthrough of the posterior cortex. Extensive soft tissue edema was also present. A bone scan showed increased uptake around the distal humerus. Core needle biopsy was performed by interventional radiology 1 week after the clinic visit. Positron emission tomography/computed tomography (CT) showed the lesion involving the distal humerus, as well as 2 local lymph nodes with high standard uptake value and increased signal in an ipsilateral axillary lymph node.

The patient continued to have worsening pain not controlled by narcotics and was admitted through the emergency department. Radiographs taken at this visit showed a nondisplaced pathologic fracture involving the capitellum (Figure 3). During his hospitalization, an open biopsy was performed with adequate specimens for diagnosis noted on the frozen section. Provisional internal fixation with a short locked plate was placed so it could be resected with the distal humerus if needed.

The final pathology diagnosis was a benign epithelioid hemangioma. The photomicrograph is shown in Figure 4. The appearance has been described as large polyhedral endothelial cells in sheets or solid cords or lining numerous well-defined vascular spaces. The cells are oval in
shape with vesicular nuclei and variably sized nucleoli.\textsuperscript{14,15} Findings of a lobular growth pattern, cellular lobules that were separated by loose connective tissue containing well-formed vessels, and a lack of cytologic atypia supported the diagnosis.

Given the benign nature of this tumor, the patient underwent curettage, bone graft placement, and plate and screw fixation of the distal humerus. After the procedure, the patient’s pain significantly improved, and he was able to transition off narcotic pain medications. He had a gradual improvement of elbow ROM and strength in the immediate postoperative period and has been able to return to work as a gunsmith.

**DISCUSSION**

Few reports of epithelioid hemangioma of bone have been published in the literature. Epithelioid hemangioma have been described with aggressive radiologic features such as bone erosion, lytic appearance, and changes in bone morphology, which suggests a diagnosis of a malignant lesion but has not been previously described as being associated with a pathologic fracture. A few cases have been reported with lymph node involvement, as in our case.\textsuperscript{13,16}

As in previous reports, the initial aggressive radiographic presentation of our patient was thought to be that of a malignant tumor involving bone. Because a core needle biopsy obtained insufficient tissue for diagnosis, we opted to proceed with an open biopsy and temporary internal stabilization of the pathologic fracture with a plate and screws for pain control. However, given the malignant appearance, consideration had been given to more aggressive management, including resection of the distal humerus with joint reconstruction. A benign diagnosis was obtained, which would have made wide resection of the distal humerus with elbow arthroplasty an inappropriately aggressive treatment. This case demonstrates the need to adhere to the basic tenets of diagnosis, staging, and grading of bone tumors prior to definitive treatment.

General diagnostic strategy for the evaluation of a bone tumor begins with clinical and radiographic evaluation. If the lesion is felt to represent a stable, benign process, then it may be observed with repeat radiographic studies at a set time period for further confirmation. However, if the bone tumor appears to be a progressive benign lesion or a primary malignancy of bone, MRI or CT scan of the involved bone and bone scan may be performed for further evaluation. If primary malignancy is suspected, the workup includes evaluation of the lungs with chest radiographs or a CT scan of the chest looking for metastatic disease. Once the radiographic workup is complete, the diagnosis is generally confirmed or made with a biopsy of the lesion.

Similar studies, including bone scans and chest radiographs, are used for the diagnostic evaluation of suspected metastatic lesions of bone. However, in these cases, MRIs and CT scans are not always necessary. Also, evaluation for a primary tumor necessitates a CT scan of the chest, abdomen, and pelvis. Laboratory studies such as protein electrophoresis are also included in the diagnostic workup of metastatic bone lesions.\textsuperscript{17}

Due to the relative rarity of these lesions, no specific treatments have been scientifically compared. In the largest report of 50 cases, in which 36 patients were available for follow-up, it was recommended that these bone lesions be treated with curettage or marginal en bloc excision.\textsuperscript{13} This study reported good results, with 4 patients having recurrence but no metastatic disease or mortality due to the tumor. A report of spontaneous remission of the tumor without treatment in a pregnant patient was reported.\textsuperscript{18} Although we do not have long-term follow-up data for our patient, we treated him in similar fashion to that described in the literature, and his short-term outcome is encouraging.

Although epithelioid hemangioma can be confused with malignant tumors such
as angiosarcoma partly due to its aggressive appearance on MRI and plain radiographs, this is the first reported case of pathologic fracture associated with epithelioid hemangioma to our knowledge. The rarity of benign and malignant vascular tumors of bone combined with variability in presentation, as outlined in our case, make diagnosis challenging. However, by maintaining sound oncologic methodology, the proper diagnosis can be obtained and will guide appropriate treatment.

REFERENCES