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The Disappearing Phalanx: A Case Report of a Vascular Tumor of the Toe

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ABSTRACT

We report a unique case of an epithelioid hemangioma of the third middle phalanx in which the lesion replaced the phalanx, became symptomatic, and then required resection, bone grafting, and joint arthroplasty. To the best of our knowledge, this is the first report of an epithelioid hemangioma in the toe that was treated using this approach.

Hemangiomas are benign vascular tumors that arise from the blood vessels and can occur in any location and organ of the human body. The histologic classification encompasses a variety of types, such as capillary, cavernous, hobnail, spindle cell, and epithelioid, and also includes vascular malformations, either venous or arteriovenous (1,2). Capillary and cavernous hemangiomas are the most frequent types occurring in bone and are most common in the skull and vertebrae, although they are also observed less frequently in the long bones and ribs (3,4). Epithelioid hemangiomas occur infrequently in bone and can be misdiagnosed as low-grade malignant epithelioid hemangioendotheliomas or even high-grade malignant epithelioid angiosarcomas, especially from a limited tissue biopsy specimen, in part because of the lack of clear, objective criteria for the classification of this uncommon subset of vascular tumors. Rare case series have been reported (5,6), and most of the other cases were miniseries or single reports, some of which used the previous term histiocytoid hemangioma (7–16). In a series of 50 cases of epithelioid hemangioma of the bone, the most frequent location was the foot (18%), followed by the vertebrae (16%), tibia (14%), humerus (12%), and hand (8%) (5). Occurrence in the forefoot has been reported in the metatarsal bones, with only 1 case reported in the phalanx (5,6,17,18).

We report a case of epithelioid hemangioma of the left third toe middle phalanx with total involvement and extension into the adjacent soft tissues. The lesion became symptomatic and required resection, bone grafting, and joint arthroplasty. To the best of our knowledge, this is the first report of this type of treatment for a case of epithelioid hemangioma occurring in a toe.

Case Report

A 25-year-old female with no significant medical history presented with left third toe pain of insidious onset for 1 year without any trauma in April 2015. She also denied injury or trauma to any other musculoskeletal areas. She reported swelling, stiffness, and pain in the affected area during activity. On physical examination, the alignment of her foot and toes was within normal limits. Examination of the third toe showed soft tissue swelling over the middle phalanx and some decreased range of motion of the proximal interphalangeal joint and no restriction of the metatarsophalangeal joint.

Standard routine anteroposterior, lateral, and oblique radiographs of the left foot showed complete obliteration of the third middle phalanx with soft tissue swelling and a residual thin shell of cortical bone (Fig. 1). Previous radiographs showed that the lesion had expanded over time. No other osseous abnormalities were identified. These findings were consistent with a tumor with slightly aggressive growth features. Magnetic resonance imaging of the left forefoot was performed using coronal and axial inversion recovery, axial sagittal and coronal fast spin sequences, and axial multiple planar gradient recalled. The magnetic resonance imaging scan revealed an expansile

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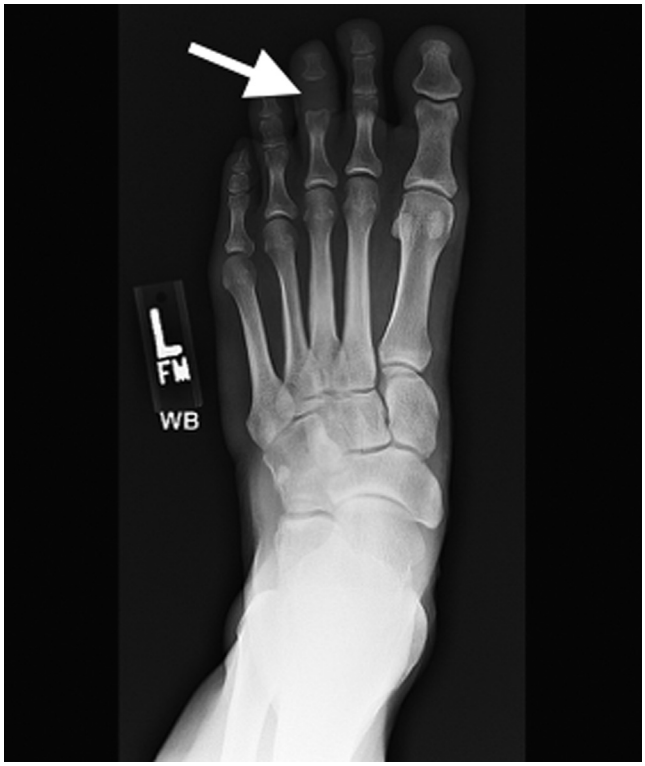

Fig. 1. Preoperative anteroposterior radiograph showing complete obliteration of the third middle phalanx *(arrow)*.

heterogeneously hyperintense mass replacing the near entirety of the third middle phalanx (Figs. 2 and 3). Areas of central dephasing were present within the lesion, consistent with calcification and the presence of a thin peripheral cortical shell of the middle phalanx. The differential diagnosis included enchondroma, vascular tumors, and giant cell lesions. Because the tumor was causing the patient considerable pain and the definitive diagnosis required histologic

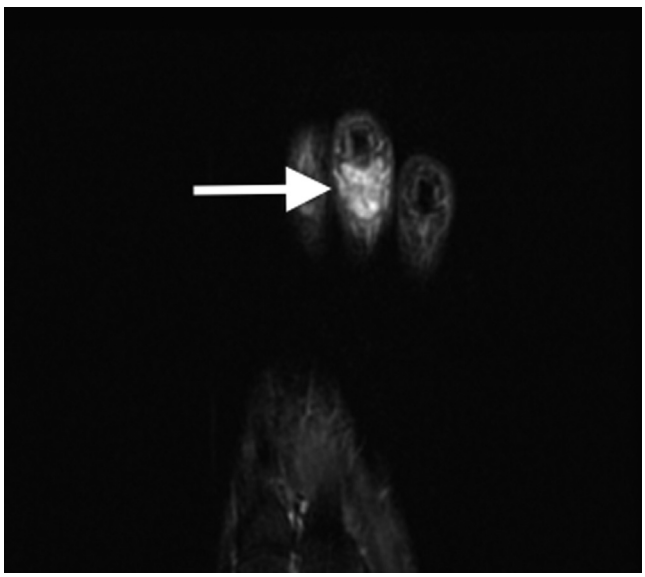

Fig. 2. Fat suppressed magnetic resonance image showing an expansile hyperintense mass that has replaced much of the third middle phalanx *(arrow)*.

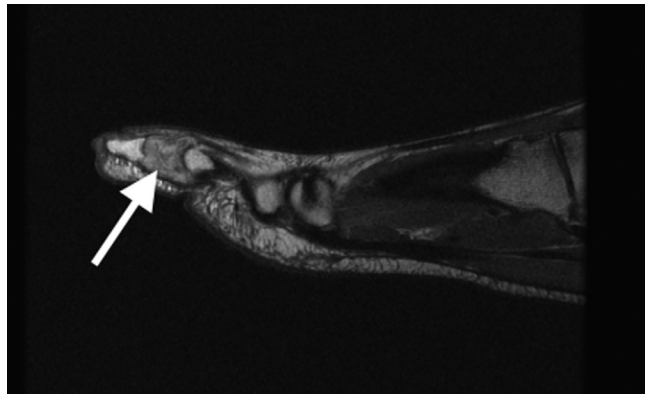

Fig. 3. Pro-time density magnetic resonance image showing an expansile hyperintense mass that has replaced much of the third middle phalanx *(arrow)*.

examination, resection of the lesion and third interphalangeal joint arthroplasty, with additional calcaneal bone grafting, was recommended. The patient underwent surgery in May 2015. She was placed in a supine position, and anesthesia was administered. A tourniquet was inflated to 250 mm Hg. At surgery, all visible tumor in the bone and its soft tissue extension were removed. On macroscopic examination, the tumor appeared confined to the middle phalanx without involvement of either the distal or proximal phalanx.

Approximately 1 cm of bone void resulted from the tumor resection. Approximately 2 cm^3 of autologous calcaneal graft was harvested laterally just distal to the tuberosity and placed into the void area. We then prepared the distal aspect of the proximal phalanx and the proximal aspect of the distal phalanx with the goal of obtaining bony union. Two 3.5-in. Kirschner wires were used to maintain fixation (Fig. 4). The specimen consisted of a portion of articular bone

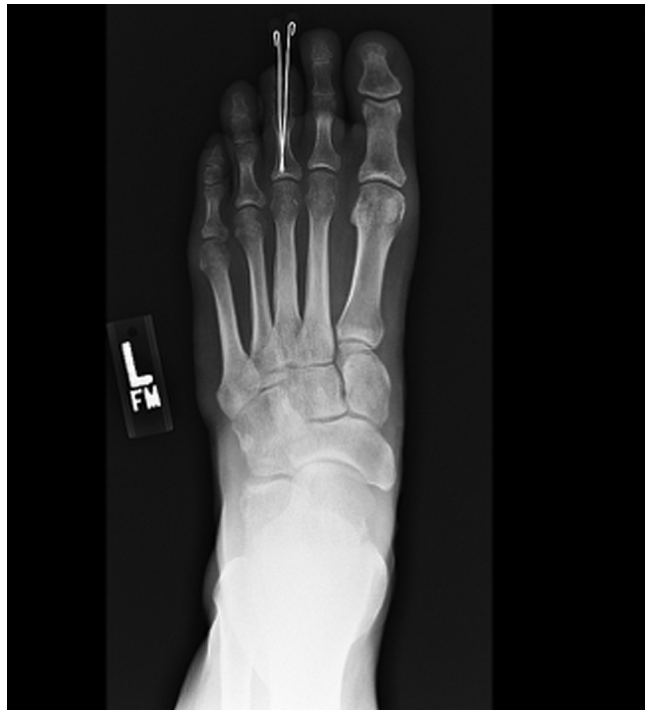

Fig. 4. Postoperative anteroposterior radiograph showing Kirschner wires used to maintain fixation of the bone graft.

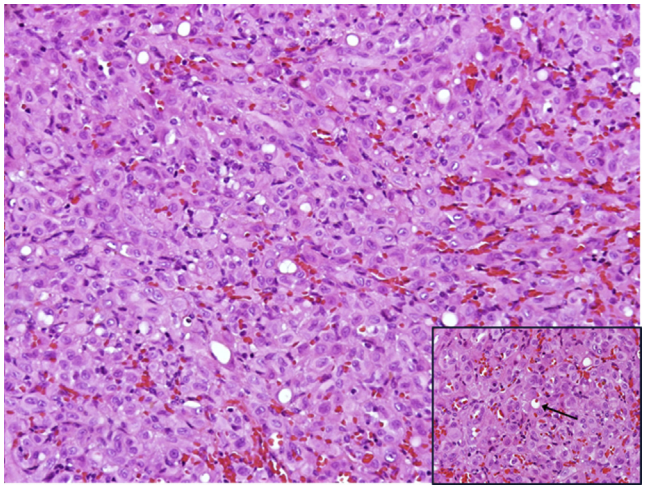

Fig. 5. Slide of epithelioid hemangioma with tightly packed epithelioid endothelial cells, polygonal eosinophilic cytoplasm, and round/slightly ovoid nucleus with small nucleolus and poorly anastomosing vascular channels (original magnification × 200; hematoxylin and eosin stain). *(Inset)* Intracytoplasmic lumen occasionally containing a red blood cell *(black arrow)* (original magnification × 400; hematoxylin and eosin stain).

from a small joint measuring approximately 7 mm in diameter and nodules of dark red soft tissue of variable size measuring approximately 1.5 cm in aggregate. On light microscopy examination, the tumor was composed of tightly packed epithelioid endothelial cells with frequent intracytoplasmic vacuoles occasionally containing a red blood cell and lining poorly developed anastomosing vascular spaces (Fig. 5). The endothelial cells had a distinct cytoplasmic border with abundant eosinophilic cytoplasm and round to ovoid nucleus with

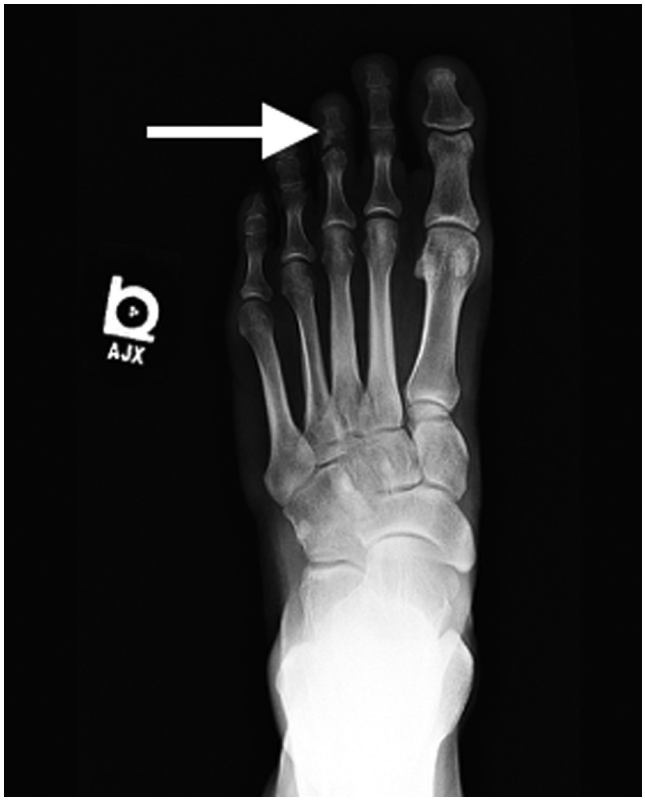

Fig. 7. Postoperative anteroposterior radiograph showing bone filling in the area with the defect *(arrow)*.

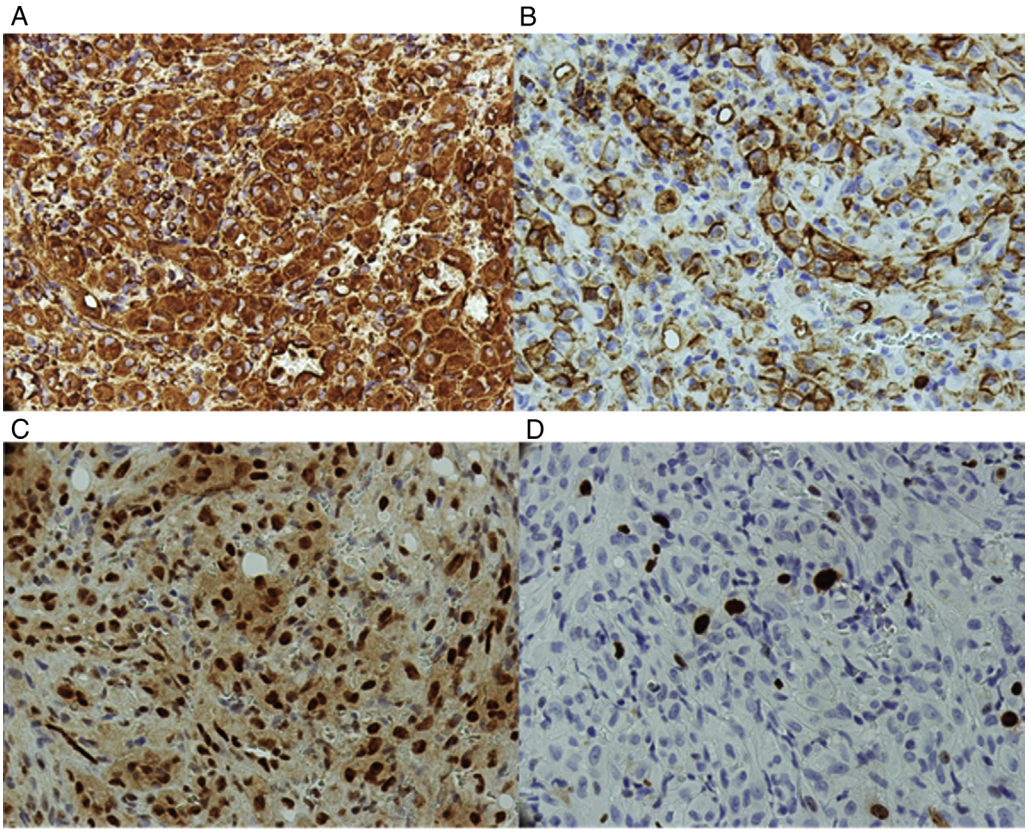

Fig. 6. Epithelioid hemangioma immunohistochemistry (original magnification × 400 for all). (*A*) Vimentin was diffusely positive with a cytoplasmic pattern; (*B*) CD31 was diffusely positive with a membranous pattern; (*C*) FLI-1 was diffusely positive with a nuclear pattern; and (*D*) Ki-67 showed <5% of cells positive with a nuclear pattern.

distinct nucleolus and dispersed chromatin. Occasional small foci of woven bone were interspersed throughout the tumor, with scattered lymphocytes and focal clustering. The articular bone from the specimen showed focal erosion of the cartilage and doubling of the tide mark without tumor involvement. Cellular atypia was not present nor were atypical mitotic figures. Immunohistochemical studies were performed for characterization of the vascular tumor and showed diffuse, positive staining of tumor cells for vimentin with a cytoplasmic pattern, CD31 with a membranous pattern, and FLI-1 with a nuclear pattern (Fig. 6). The capillary walls were positive for CD34. The Ki-67 proliferative index showed positivity of <5% of the neoplastic cells (Fig. 6). The morphologic findings and the immunohistochemical profile with diffuse positivity for the endothelial cell markers CD31 and FLI-1 were diagnostic of epithelioid hemangioma.

The patient was placed in a non-weightbearing splint for the first 2 weeks after surgery. She was then transitioned into a controlled ankle motion boot for 4 weeks, with continued non-weightbearing. She was transitioned into a Darco-type shoe at 6 weeks postoperatively and was allowed to begin partial weightbearing at this point. The Kirschner wires were removed at 10 weeks postoperatively. At the 6-month postoperative follow-up examination, radiographs showed no evidence of local recurrence or distant involvement of the tumor (Fig. 7). The patient reported that her pain had completely resolved and she had been able to return to athletic activities such as running. Partial bony fusion had occurred across the proximal interphalangeal and distal interphalangeal joints (Fig. 7). At 1 year postoperatively, she was clinically asymptomatic, and the alignment was stable without flexion deformity of the phalanges.

Discussion

The spectrum of vascular tumors of bone ranges from hemangioma to high-grade angiosarcoma. Epithelioid hemangioma is a subtype of benign vascular tumors that usually occurs in the skin, blood vessels, and soft tissue but is rarely observed in bone. In a series of 50 cases, the most frequent sites were the long bones of the lower and upper extremities followed by the short tubular bones (5). The terminology and classification of epithelioid vascular tumors of bone has been controversial regarding the definition of specific entities and the consideration of a continuous spectrum of the biology of these tumors from low-grade to high-grade malignancies (5,6). Owing to the various morphologic patterns and possible multifocal growth of epithelioid hemangiomas, they can be misdiagnosed as malignant tumors, such as epithelioid hemangioendothelioma and even epithelioid angiosarcoma (7,8,10).

Radiographically, epithelioid hemangiomas have been reported as well-defined lytic lesions predominantly involving the diaphysis and less frequently the metaphysis of the affected bone, such as other benign vascular tumors. They have also been confused with other entities, such as enchondromas or giant cell lesions (5,15). These tumors appear osteolytic and are often associated with peripheral bone sclerosis and expansion with thinning of the cortex, although their vascular nature cannot be determined with certainty and requires additional studies such as magnetic resonance imaging with or without contrast. The definitive diagnosis, however, can be established only by histologic examination. Recently, *FOS* gene rearrangements have been detected in cases of epithelioid hemangioma, providing an additional tool for the differential diagnosis in difficult cases or in cases with a limited tissue biopsy specimen (19).

The recommended treatment is surgical, including complete excision of the lesion with clear margins when feasible. The tumor has an indolent course with the potential for local recurrence in about 10% of the cases and rare involvement of the regional lymph nodes (20). It is important to differentiate epithelioid hemangioma from epithelioid hemangioendothelioma and angiosarcoma, because the treatment of the malignant tumors must be more aggressive, with wide excision and eventual radiotherapy and/or chemotherapy according to the histologic stage and grade of the tumor. In our patient, we were able to successfully perform a digit-sparing procedure without tumor recurrence, although with a short-term follow-up period. If a simple resection had been performed, it would have most likely resulted in floppy toe, leading to a shorter toe with poor cosmesis. Using autologous bone grafting, we maintained the alignment, stability, and original length of the toe.

In conclusion, and to the best of our knowledge, this is the first case report of treatment with bone grafting of an epithelioid hemangioma involving the phalanx of a toe. The distinction between epithelioid hemangioma and epithelioid hemangioendothelioma or angiosarcoma is important, because the latter diagnosis requires more extensive surgery and possible amputation and has a less favorable prognosis.

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