Tufted Angioma of the Index Finger Middle Phalanx

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ABSTRACT

A tufted angioma is a benign vascular tumor of the skin and subcutaneous tissue that classically presents as a violaceous nodule on the trunk or extremities in early childhood. Tufted angiomas of the finger are uncommon, and intraosseous involvement of these tumors is exceedingly rare. When present in the bone, these lesions may be difficult to distinguish from the more common pediatric condition of osteomyelitis or osteoid osteoma. We present the clinical, radiographic, and histopathologic findings for a unique case of a tufted angioma with intraosseous involvement in the middle phalanx treated by surgical excision and curettage with preservation of function.

Tufted angioma is a rare type of benign vascular tumor of the skin and subcutaneous tissue, thought to be on the same clinical spectrum as the more locally aggressive vascular tumor, kaposiform hemangioendothelioma (KHE). A tufted angioma typically presents in childhood with 50% developing in the first year of life and up to 70% before the age of 5 years. The most common locations are the trunk and extremities, and there is no sex predilection.

Clinically, tufted angiomas usually present as isolated violaceous tender nodules, although extensive and multifocal lesions have been reported. Definitive diagnosis is made with biopsy demonstrating hypercellular tufts of capillaries in a cannonball distribution. Approximately 10% of tufted angiomas are complicated by the Kasabach-Merritt phenomenon (KMP), characterized by thrombocytopenia and coagulopathy. There does not seem to be a risk of malignant transformation.

Spontaneous regression occurs in over 50% of congenital tufted angiomas, favoring an active surveillance approach in patients with minimal symptoms. For symptomatic patients with small lesions, surgical excision is a definitive therapy, although there is a recurrence risk with incomplete resection. Previous case reports have demonstrated successful surgical excision of soft-tissue tufted angiomas in the finger without osseous involvement.

Case Report

A 6-year-old boy presented with 6 weeks of insidious onset pain, mild swelling, and stiffness of the right index finger, partially responsive to ibuprofen and ice. He denied preceding trauma or systemic symptoms. On examination, he had mild edema and erythema over the middle phalanx without bruising,
angulation, or nail bed changes. The middle phalanx was tender to palpation, and the digit had slightly limited end-range flexion.

Radiographs demonstrated a rounded lucent lesion with sclerotic margins in the middle phalanx abutting the cortex (Figure 1). The initial leading diagnosis was osteoid osteoma. Magnetic resonance imaging (MRI) with and without contrast showed an eccentric intramedullary lesion along the radial aspect of the proximal metadiaphyseal region of the middle phalanx. The lesion was hyperintense on T2-weighted images and demonstrated solid enhancement on postcontrast sequences (Figure 2). The MRI revealed an extraosseous soft-tissue component that appeared to cross the cortex into the intraosseous portion (Figure 2, B and C).

Based on findings of cortical disruption and contrast enhancement, the differential diagnosis was expanded to include vascular tumors and chronic infection. The patient’s complete blood count and inflammatory markers were normal. The blood culture did not identify a pathologic organism.

Surgical excision with curettage was done for diagnostic and therapeutic purposes. A 3-mm circular defect was noted along the radial and volar aspects of the cortical bone with protrusion of yellow fibrinous tissue. The tissue was removed and sent for histopathologic examination and microbiology testing. Curettage and irrigation of the cavity were done before wound closure. Bacterial and fungal tissue cultures showed no growth. Histopathologic examination revealed lobules of endothelial cells with CD31-positive, focal Prox-1–positive, and GLUT-1–negative staining (Figure 3). These findings confirmed the diagnosis of tufted angioma.

The patient wore a protective splint for 2 weeks postoperatively and then progressed through a 1-month range-of-motion and strengthening home exercise program. The patient returned to all activities 6 weeks postoperatively. His pain had resolved by his 6-month postoperative visit. Examination at 12 months revealed minimal swelling over the middle phalanx with full range of motion, sensation, and strength. Repeat radiographs demonstrated interval healing (Figure 4).

**Discussion**

Tufted angiomas of the finger are extremely rare with only three previously published case reports. Each of these cases described solitary, progressive, violaceous nodules without osseous involvement or KMP. Histopathology revealed overlapping features of tufted angioma and KHE in one case and confirmed the diagnosis of tufted angioma in the other two. Compromised finger function and associated pain prompted surgical excision. One case was complicated by rapid recurrence requiring repeat excision using a free vascularized groin flap.

Our case highlights the importance of considering tufted angioma on the differential for bone tumors, particularly in pediatric patients. Only three previous case reports have described intraosseous involvement of tufted angiomas. The first case reported on a 73-year-old patient with eye pain, swelling, and compromised vision secondary to a large tufted angioma with orbital and cranial bone involvement. The second case reported on a tufted angioma in the maxillary bone of a 10-year-old child. The third case described a 54-year-old patient with right knee pain found to have a superficial tufted angioma and an adjacent bone lesion with histologic findings suggesting an intermediate between tufted angioma and KHE.

Three clinical patterns have been proposed for pediatric tufted angiomas: tufted angiomas without complications, with chronic coagulopathy without thrombocytopenia, or with KMP. Chronic coagulopathy without thrombocytopenia is the least common. KMP is characterized by thrombocytopenia, low fibrinogen, and elevated D-dimer levels. It is important to assess for KMP in patients with tufted angioma because of the implications for prognosis.

![Figure 1](image1.png)

A and B AP and oblique radiographs showing the index finger at preoperative evaluation. In the middle phalanx, there is a nonaggressive-appearing lucent lesion with a sclerotic rim abutting the volar/radial cortex.
Figure 2

A–C, (A) Coronal short tau inversion recovery image of the index finger showing a T2 hyperintense lesion (arrowhead) in the middle phalanx abutting the radial-sided cortex. Axial (B) and sagittal (C) T1 postcontrast fat-saturated images demonstrating a solid hyperenhancing lesion with extraosseous soft-tissue (arrow) and intraosseous (arrowhead) components.

Figure 3

A–D, (A) ×10: Image showing that within the subcutaneous tissue, there are lobules of closely packed capillaries in a cannonball distribution. B, ×40: Image showing that the capillaries form slit-like spaces and are lined by endothelial cells without cytologic atypia. C, ×40: Image of CD31 showing membranous positivity within the vessels supporting the vascular origin. D, ×40: Image showing that Prox-1 staining highlights scattered endothelial cells (lymphatics).
Figure 4

A and B AP and oblique radiographs showing the index finger at 12-month postoperative evaluation. The middle phalangeal lesion has become more sclerotic, and there is cortical thickening on the volar/medial aspect, compatible with healing.

KHEs have a higher rate of associated KMP and will not regress if left untreated.5

Treatment of tufted angiomas with medical and procedural interventions has variable efficacy, so surgical excision remains the treatment of choice for small, solitary, symptomatic lesions.3 Given the rates of spontaneous regression, treatment with active surveillance is reasonable for uncomplicated tufted angiomas with minimal symptoms.9 Other nonprocedural treatments of uncomplicated tufted angiomas include low-dose aspirin, topical immunomodulators, systemic corticosteroids, and vincristine monotherapy.2,19-21 Additional possible procedural interventions include pulsed dye laser, intralesional steroid injections, and embolization.7,9,21 For those patients with KMP, some studies suggest treatment with varying combinations of systemic corticosteroids, vincristine, aspirin, and ticlopidine.2,15,21 Our case adds to the literature suggesting that surgical excision of a finger tufted angioma with osseous involvement can be achieved with resolution of pain and maintenance of finger function.

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