

Vascular Tumors of the Hand: A Report of 3 Cases

Mark A. Pollard, MD

Tumors of vascular origin are relatively rare. These tumors may be a manifestation of a systemic syndrome, or they may occur as the only clinical abnormality. Vascular tumors can be classified as congenital or acquired and as benign or malignant. Benign congenital vascular tumors include hemangioma, lymphangioma, and congenital arteriovenous fistula. Acquired benign vascular tumors include arteriovenous fistula, pyogenic granuloma, glomus tumor, and true and false aneurysms. Malignant vascular tumors include hemangiosarcoma, hemangioendothelioma, and Kaposi's sarcoma.

Approximately one fourth of all vascular tumors occur in the extremities, with a large proportion occurring in the hand and forearm.¹ Vascular tumors of the hand account for approximately 7% of all tumors of the hand, and malignant vascular tumors account for fewer than 1% of all vascular hand and forearm tumors.^{1,2} All of these tumors share a common origin from cells comprising the vascular system. Possible cells of origin include endothelial cells, pericytes, and myocytes.³ Classification of these tumors is based on the degree of cell differentiation, the type of cells primarily involved, and the architecture of the tumors.

In order to provide proper care for patients with vascular lesions, physicians must be able to accurately identify the tumors. Ambiguous nomenclature attributed to vascular neoplasms sometimes makes diagnosis confusing.^{1,4,5} For example, the terms hemangiosarcoma, angiosarcoma, hemangioendothelioma, and hemangioendothelial sarcoma have all been used either as synonyms or as terms denoting separate pathologic entities.² This article uses a case presentation format to describe the etiology, clinical features, and management of 3 types of vascular tumors of the hand: multiple hemangiomas associated with Maffucci's syndrome, recurrent hemangioma after traumatic injury, and aggressive hemangioendothelioma.

MULTIPLE HEMANGIOMAS OF MAFFUCCI'S SYNDROME

Case 1 Presentation

A 4-year-old boy was brought to a physician because

of leg length discrepancy. His antecedent medical history was unremarkable. Radiographs obtained at the initial visit revealed multiple enchondromata and arteriovenous malformations (Figure 1). The patient was diagnosed with severe involvement of Maffucci's syndrome. The disease did not progress for 4 years. When he was 8 years of age, however, the child began to develop deformities of both hands, as well as multiple soft lesions of the skin that appeared blue to purple in color. The deformities of the fingers became increasingly severe, requiring multiple debulkings and, finally, amputations (Figure 2). Results of pathology tests indicated that the bone lesions were characteristic of hypercellular enchondromata, and the skin lesions exhibited dilated vascular channels with thrombosis and phlebolithiasis. The patient's health progressively deteriorated, and he died at age 18 years of intracerebral bleeding caused by an arteriovenous malformation.

Epidemiology and Pathogenesis

Maffucci's syndrome is a rare disorder characterized by multiple hamartomas including enchondromata and subcutaneous hemangioma.⁵⁻⁷ First described by Maffucci in 1881, the syndrome has been reported to affect children as a deformity or pathologic fracture after normal birth and infancy.^{1,5} The 2 major hamartomatous proliferations appear to occur independently.⁷ Maffucci's syndrome is reportedly associated with a high risk for malignant transformation of enchondromata.⁵ Other reported associated malignancies include hemangiosarcoma, lymphangiosarcoma, fibrosarcoma, glioma, mesenchymal ovarian tumor, and pancreatic carcinoma.⁵ The vascular component of the disease is classically a diffuse, cavernous hemangioma, with frequent thrombi and phleboliths.⁵⁻⁷ These subcutaneous hemangiomas may cause secondary changes to adjacent bone through a slow, sclerotic erosion from an extrasosseous location.⁷

Dr. Pollard is a Resident, Department of Orthopaedic Surgery, Hamot Medical Center, Erie, PA.

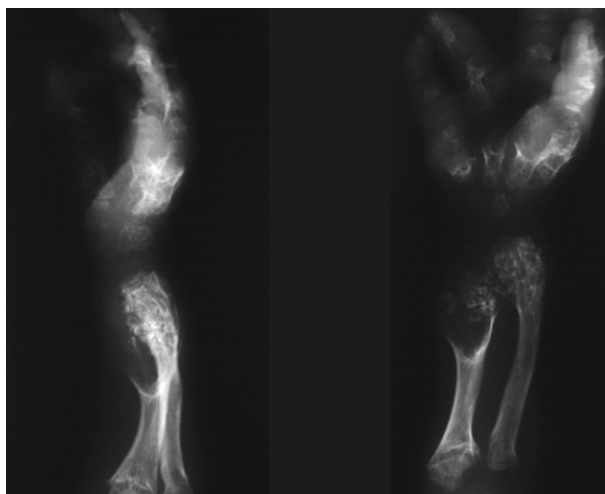


Figure 1. Radiographs showing multiple enchondromata in patient 1.



Figure 2. Clinical photograph showing severe deformity of the hands and fingers in patient 1.

Clinical Features and Management

The classic appearance of the subcutaneous hemangioma in Maffucci's syndrome is a blue or purple mass that may be pulsatile and painful.⁵⁻⁷ Radiographic appearance of these hemangiomas may reveal the aforementioned local bone erosion but more often demonstrates phleboliths.⁵⁻⁷ Histologically, the cavernous hemangiomas exhibit large vascular channels that contain erythrocytes and are lined with flat endothelial cells.⁵ Occurring frequently on the hand or forearm, these hemangiomas may become symptomatic.

Treatment is generally focused on relieving the symptom of pain caused by mass effect or by nerve or vascular compression^{5,7} and usually involves excision with clean margins.^{5,7} Sarcomatous degeneration of hemangiomas in Maffucci's syndrome is rare but has



Figure 3. Clinical photograph showing an enlarging mass of the hypothenar region of the left hand in patient 2. Scars from previous surgeries are visible.

been reported. Clinicians should maintain a high index of suspicion of malignancy for rapidly enlarging hemangiomas.⁵

POSTTRAUMATIC RECURRENT HEMANGIOMA

Case 2 Presentation

A 43-year-old man visited his physician because of a rapidly growing, painful mass on his left hypothenar eminence (**Figure 3**). Approximately 20 years earlier, while in Vietnam, the patient had sustained a shrapnel injury to his left hand. An arteriovenous malformation in his ulnar artery had developed, requiring numerous surgeries. Physical examination revealed a firm, pulsatile, bluish mass present in the area of the hypothenar muscles. Results of magnetic resonance imaging and an arteriogram showed a vascular lesion involving much of the hypothenar mass, apparently arising from the ulnar aspect of the superficial palmar arterial arch (**Figures 4 and 5**). The mass was surgically excised, and pathologic examination of the lesion showed congeries of arterioles and venules, with prominent endothelium (**Figure 6**). Some vessels were observed to compress the peripheral bordering skeletal muscle fibers. No evidence of either malignant transformation or inflammation was found. The patient had an unremarkable postsurgical course; he retained protective sensation but experienced decreased function and range of motion secondary to excision of local musculature.

Etiology

Posttraumatic hemangiomas and arteriovenous malformations are acquired, benign vascular lesions.^{1,8-10} Their etiologies are similar, because both types of lesion

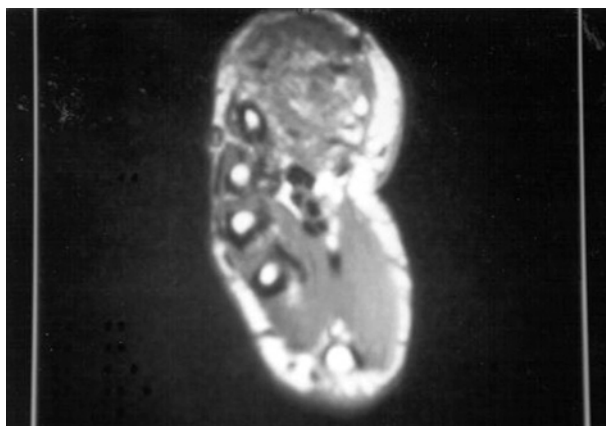


Figure 4. Magnetic resonance image showing a vascular lesion involving most of the hypothenar mass in patient 2.

are apparently the product of aberrant vascular healing following injury.¹ After injury of adjacent arterial and venous structures, healing of a hematoma may follow an irregular pattern, which can result in a circumvention of the capillary system by anastomosis of an arterial structure to a venous structure or in proliferation of vascular tissue in a hemangioma.¹

Clinical Features and Management

These acquired lesions present as enlarging, painful blue or purple masses.¹ Histology is similar to that of the subcutaneous hemangiomata of Maffucci's syndrome; however, a foreign body caused by a prior penetrating trauma may also be found.¹⁰ Recurrence may take place if resection is incomplete.¹⁰ Before surgical resection, the surgeon must thoroughly examine the affected limb to ensure that adequate perfusion will be present after tumor removal.¹ Treatment should consist of early resection of the tumor before permanent damage to the vessel walls occurs, with possible vascular grafting and repair to ensure adequate postresection blood flow.¹

AGGRESSIVE HEMANGIOENDOTHELIOMA

Case 3 Presentation

A 13-year-old boy was evaluated by his family physician for discomfort and swelling of his left hand. He reported a recent history of mild trauma to his hand after a fall at his family's barn. He initially had presented to an emergency department at another facility. At that time, physical examination had revealed a small abrasion over the dorsum of an edematous hand. Radiography had disclosed osteopenia of the index, long, and ring metacarpals, with no evidence of fracture or other abnormalities. The patient had been

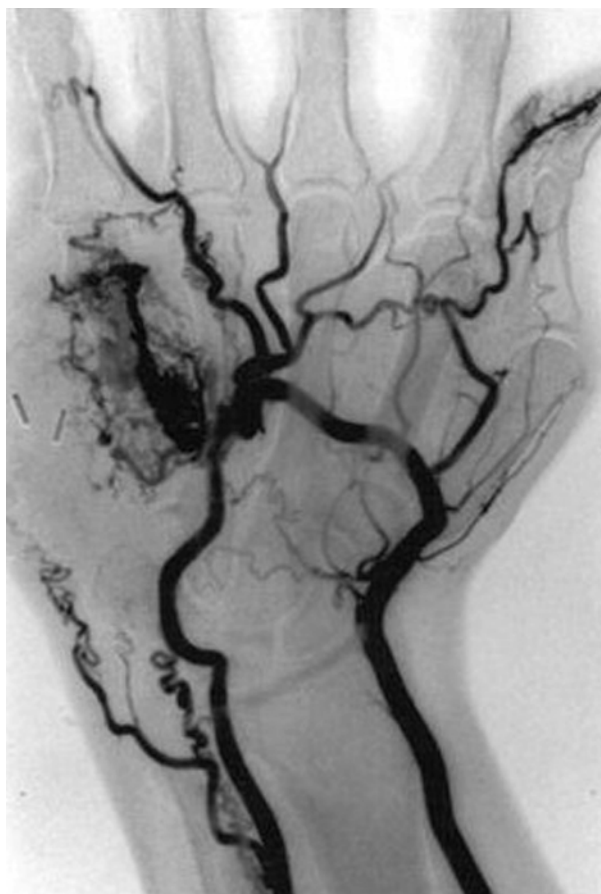


Figure 5. Arteriogram showing a vascular lesion in the region of the hypothenar mass in patient 2.

treated with application of a splint and was instructed to see his family physician in 2 weeks. Repeat radiographic evaluation at this 2-week follow-up revealed progressive destruction of the metacarpals and carpus (**Figure 7**). Routine laboratory measurements obtained at this time included antinuclear antibody, C-reactive protein, erythrocyte sedimentation rate, rheumatoid factor, and sickle cell preparation. All results were within normal limits.

One month after this visit, concern about the interval change of the patient's radiographic results led to a referral to a pediatric orthopaedic hospital for evaluation and treatment. Physical examination revealed marked edema, warmth, and slight erythema over the dorsum of the left hand. No evidence of abrasion or other breaks in skin integrity were seen. Sensory and motor function were intact in the median, radial, and ulnar nerve distributions. All digits were warm, with good capillary refill present. Tenderness was elicited with palpation of the metacarpals, carpsals, and the

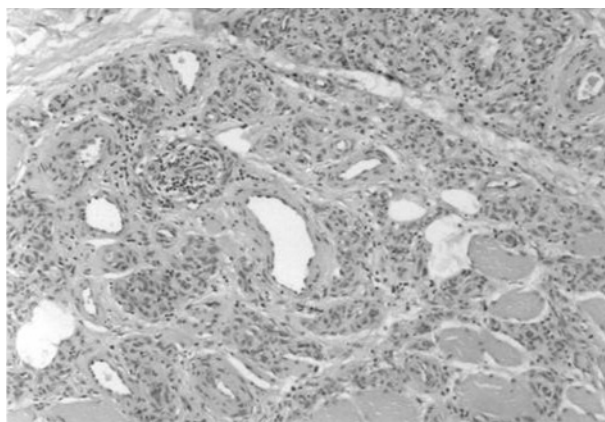


Figure 6. Photomicrograph showing congeries of arterioles and venules with prominent endothelium in patient 2.



Figure 7. Radiographs showing destruction of the metacarpals and carpus in patient 3.

metacarpal joints. No adenopathy of the left upper extremity was noted on examination.

Open biopsy of the involved metacarpals was performed at this time. The only gross abnormality observed was the complete absence of bone. Aerobic and anaerobic cultures obtained at the time of biopsy were negative for bacterial growth. Initial pathology was consistent with Gorham's disease (ie, disappearing bone disease). Two weeks later, the surgical wound from the initial biopsy site developed a hematoma that required evacuation. Within the next 2 weeks, delayed primary closure of the wound was undertaken. The wound required split-thickness skin grafting 2 months after the initial procedure because of delay of healing. Cultures taken at the time of skin grafting grew *Staphylococcus*



Figure 8. Gross specimen showing a well-vascularized lesion with destruction of metacarpals and carpus in patient 3.

aureus and β -hemolytic group A streptococcus, both of which responded to oral treatment with cephalosporins. During the subsequent 2 months, the patient's hand remained edematous and painful. Cyanosis of the index finger developed, which was noted during a visit 2 months after skin grafting. Radiographic surveillance revealed progression of osteolysis. Because of the disease progression, repeat biopsy was performed 5 months after the patient's initial presentation at the emergency department. The pathology report of the second biopsy was consistent with grade II malignant hemangioendothelioma. Repeat laboratory evaluation was negative for any organisms. Results of a bone scan and magnetic resonance imaging showed no evidence of metastatic disease. Six months after the patient's initial presentation, a below-elbow amputation was performed. The gross specimen disclosed a well-vascularized lesion, as well as destruction of the carpus and metacarpals (**Figure 8**). The patient was fitted with a prosthesis; 21 months following the procedure, he showed no signs of local or distant recurrence of disease.

Clinical Features

Hemangioendotheliomas are rare vascular tumors with a potential for malignancy.^{1,2,11-14} Hemangioendothelioma and angiosarcoma are terms used interchangeably to describe the same entity.² They are considered the most common malignant vascular tumors of the hand¹ and are characterized by multifocal lesions in an extremity.² Physical examination and symptomatology are inconsistent, with pain and occasional local swelling occurring most frequently.^{2,4,11} The most common radiographic appearance of the lesion is a radiolucent region, occasionally with a well-defined rim of bone.^{2,11,12,15}

Histology

Histology of hemangioendothelioma is characteristic and has been reported to be the most accurate prognostic determinant. The characteristic appearance of the tumor is that of vascular endothelial cells lining vascular channels. Great variability exists in the number of vascular channels present and in the differentiation of the endothelial cells.

Grade I hemangioendotheliomas display well-differentiated cells with moderate nuclear size; scarce pleomorphism; thin, dispersed chromatin; and rare mitotic figures (ie, fewer than 5 cells per 10 microscopic fields, magnification $\times 100$). Grade I tumors also display well-formed capillaries.

Grade II tumor cells have larger nuclei, moderate pleomorphism, slight hyperchromatism, and more frequent mitotic figures (5–15 cells per 10 high-powered microscopic fields, magnification $\times 100$). Grade II lesions display more prominent areas of solid angioblasts than do grade I tumors.

Grade III tumors, sometimes called hemangiosarcomas, display undifferentiated spindle cells and angioblastic cells with very large nuclei and are obviously pleomorphic and hyperchromatic, with frequent mitotic figures (20–30 cells per 10 high-powered microscopic fields, magnification $\times 100$). The architecture of grade III tumors is extremely disordered, with large solid areas of angioblastic cords and fields.

Management and Prognosis

It has been suggested that treatment of hemangioendothelioma be based upon these histologic grades. Grade I hemangioendothelioma may be treated conservatively, with curettage often proving curative. Grade II lesions may also be treated with conservative curettage, but bone resection may be preferable. Grade III lesions are reported to carry a high mortality rate in most studies. Treatment should be ablative, with adjuvant chemotherapy.^{3,11}

Controversy exists over the prognosis and malignant potential of hemangioendothelioma. Some authors contend that hemangioendothelioma has limited metastatic potential.⁴ Most authors, however, believe a malignant potential exists that increases with the higher histologic grades.^{1,2,11,13,15} Diagnostic consultation with an experienced musculoskeletal pathologist and an orthopaedic oncologist should be sought for suspected hemangioendotheliomata.

CONCLUSION

Vascular tumors represent an uncommon but important clinical entity to the musculoskeletal surgeon. Whereas many may be benign, certain tumors carry a high potential for malignancy. Recognition of these lesions is essential to the proper care of patients with vascular tumors. **HP**

REFERENCES

1. Acharya G, Merritt WH, Theogaraj SD. Hemangioendotheliomas of the hand: case reports. *J Hand Surg [Am]* 1980;5:181–2.
2. Campanacci M, Boriani S, Giunti A. Hemangioendothelioma of bone: a study of 29 cases. *Cancer* 1980;46:804–14.
3. Lewis RJ, Ketcham AS. Maffucci's syndrome: functional and neoplastic significance. Case report and review of the literature. *J Bone Joint Surg Am* 1973;55:1465–79.
4. Palmieri TJ. Vascular tumors of the hand and forearm. *Hand Clin* 1987;3:225–40.
5. Strang C, Rannie I. Dyschondroplasia with haemangioma (Maffucci's syndrome). *J Bone Joint Surg* 1950;32:376–83.
6. Elmore SM, Cantrell WC. Maffucci's syndrome. Case report with a normal karyotype. *J Bone Joint Surg Am* 1966;48:1607–13.
7. Watson MD, Kaye JJ. Traumatic venous aneurysm presenting as a ganglion cyst. A case report. *J Bone Joint Surg Am* 1988;70:1248–50.
8. Aprin H, Schwartz GB, Valderamma E. Traumatic venous aneurysm. Case report. *Clin Orthop* 1987;(217):243–6.
9. May JW Jr, Atkinson R, Rosen H. Traumatic arteriovenous fistula of the thumb after blunt trauma: a case report. *J Hand Surg Am* 1984;9:253–5.
10. Milgram JW, Riley LH Jr. Hemangioendothelioma of the proximal part of the humerus. A case report. *J Bone Joint Surg Am* 1972;54:1543–7.
11. Larsson SE, Lorentzon R, Boquist L. Malignant hemangioendothelioma of bone. *J Bone Joint Surg Am* 1975;57:84–9.
12. Dunlop J. Malignant hemangioendothelioma of bone: case report of en bloc resection and prosthetic hip replacement. *J Bone Joint Surg Am* 1977;59:832–4.
13. Unni KK. Hemangioendothelioma (hemangiosarcoma) and hemangiopericytoma. In: Dahlin's bone tumors: general aspects and data on 11,087 cases. 5th ed. Philadelphia: Lippincott-Raven; 1996:317–28.
14. Fleming AN, Smith PJ. Vascular cell tumors of the hand in children. *Hand Clin* 2000;16:609–24.
15. Lezama-del Valle P, Gerald WL, Tsai J, et al. Malignant vascular tumors in young patients. *Cancer* 1998;83:1634–9.

Copyright 2003 by Turner White Communications Inc., Wayne, PA. All rights reserved.