Enlarging Congenital Soft-Tissue Mass: Venous Malformation Mimicking Hemangioma Vs Sarcoma.

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Citation

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Abstract

Background: [L1] Venous Malformations are not uncommon, usually solitary and are often symptomatic.Differential diagnosis includes: infantile hemangioma, other vascular/lymphatic malformation, other vascular tumor, and as this case illustrates, fibrosarcoma or rhabdomyosarcoma. The characteristic physical finding of this vascular birthmark is a soft, blue-purple and easily compressible soft-tissue mass,. Materials and Methods: The presentation described in this case was worrisome because of rapid growth and the possibility of a vascular tumor. After multidisciplinary evaluation and review of the literature, the decision was made to proceed with surgical debulking and obtain a tissue diagnosis.Results: After a combined and coordinated evaluation by plastic surgery and dermatology, as well as ultrasound/MRI evaluation, the patient underwent surgical debulking. [L2] Conclusion: In this case, clinical history was alarming and the physical exam was not diagnostic. Thus imaging studies were employed which aided, but did not make the diagnosis. Due to the sensitive location of this patient's lesion, as well as the unusual behavior and concerning clinical characteristics, surgical intervention was determined to be the appropriate course of treatment. [L1]This is used incorrectly. Cavernous hemangioma is an old term and is confusing, usually it refers to a deep hemangioma of infancy, not a venous malformation. [L2]I would take this out at this point b/c it confuses the picture.

INTRODUCTION

All vascular birthmarks are not angiomas, despite the persistence of this term. Hemangiomas are neoplasms with a typical rapid growth phase characterized by hypercellularity and endothelial multiplication; whereas venous malformations grow commensurately with the child and are true structural anomalies.[1] From a hemodynamic perspective, there are two types of malformations, slow- and fast-flow. Venous malformations (VMs) are classified as slow-flow malformations. VMs arise when the number of smooth muscle cells does not increase with increasing luminal diameter[2]. The molecular mechanism is unknown, but it is unlikely that endothelial hyperplasia plays a role. They are present at birth, although some may not become clinically apparent until later in infancy/childhood or even adulthood. VMs have a wide range of appearances, from superficial varicosities to large complex vascular lesions that may be located in deeper tissues. Bruit is typically not detected on exam. VMs however characteristically expand with Valsalva maneuver and partially decompress with extremity elevation and local compression[5]. VMs are the most common among vascular malformations, with an incidence of 1-4% and no predilection for either sex.

VMs account for more than half of all vascular lesions presenting on the extremities[6]. Most venous malformations occur as a single lesion and present sporadically[1]. Differential diagnosis includes: infantile hemangioma, other vascular/lymphatic malformation, other vascular tumor, and sometimes, soft tissue sarcoma. Tissue biopsy and GLUT-1 assay usually help to distinguish between these etiologies. GLUT-1 is present in typical infantile hemangiomas, but absent in all other tumor types[10]. Although aggressive treatment for VMs is often unnecessary, surgical resection is the definitive treatment for symptomatic or function-threatening lesions as this method can reduce bulk, improve contour and function, and control pain.

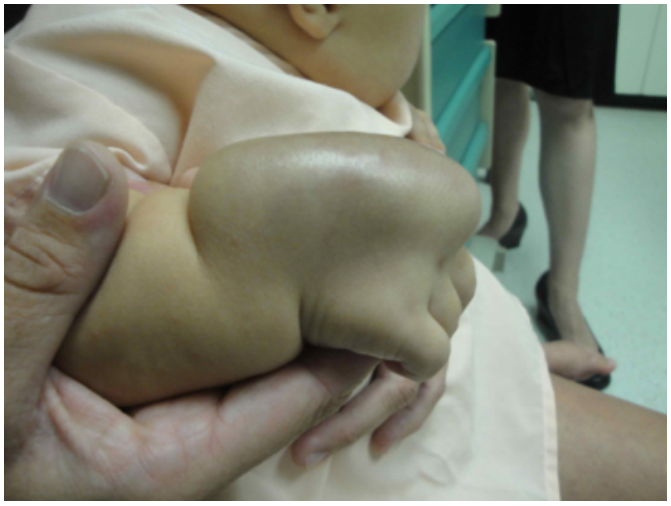
CASE REPORT

A 3-month-old Caucasian baby boy presented at approximately 1 month of age with a congenital mass on the dorsum of the right hand, which had doubled in size since birth. The parents reported a normal pregnancy, and delivery at home without complication. Family history was negative for similar lesions or skin disorders. On exam, there was a 5.5 x 4.5 x 1cm bluish, well-defined and almost squared-off subcutaneous nodule at the dorsum of the hand with no

appreciable epidermal changes (Fig.1)

Figure 1

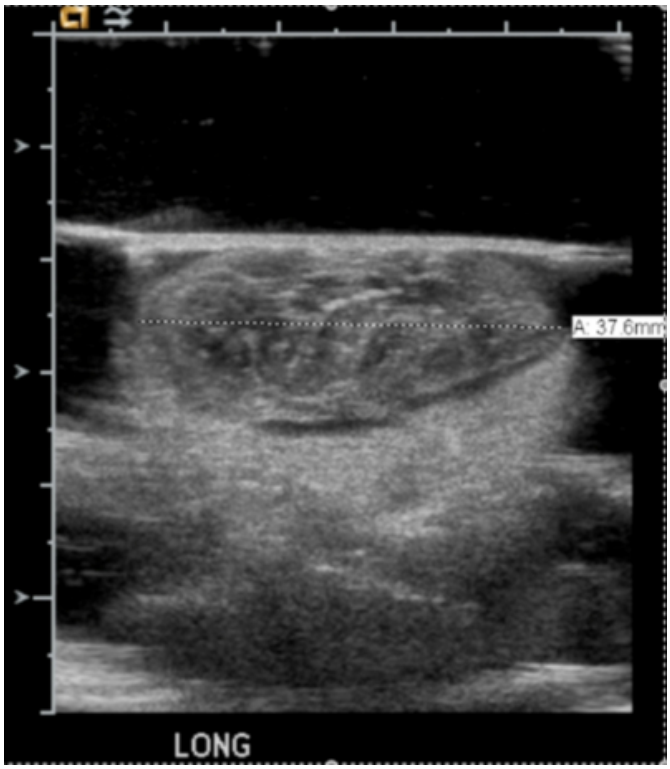
Fig. 1: Gross examination of vascular lesion over dorsum of right hand/wrist



The affected hand was otherwise normal in appearance, with strong pulse and good capillary refill. The remainder of the cutaneous exam was normal. Differential diagnosis at that time included vascular malformation or deep infantile hemangioma, given the reported growth. Ultrasound evaluation was performed and was suggestive of a partially thrombosed venous malformation (Fig. 2).

Figure 2

Fig. 2: Ultrasound evaluation of vascular lesion



The patient was seen in follow-up at 2 months of age. The lesion now measured 8.5 x 5.5cm, was very firm to palpation, and the overlying skin had a deep bluish discoloration. No bruits were appreciated. Differential diagnosis now consisted of deep infantile hemangioma, in its proliferative phase, vascular tumor, thrombosed vascular malformation or less likely, soft tissue sarcoma. Given the rapid growth of the lesion, the family was referred to our institution's multidisciplinary vascular lesions clinic for further evaluation. At this visit, the lesion measured 8.5 x 6.5cm, with deep blue coloration and increased vascularity appreciated. MRI was performed specifically to rule out sarcoma, and was consistent with a venous malformation with thrombus and phlebolith (Figs. 3 & 4).

Figure 3

Fig. 3: Transverse MRI evaluation of vascular lesion

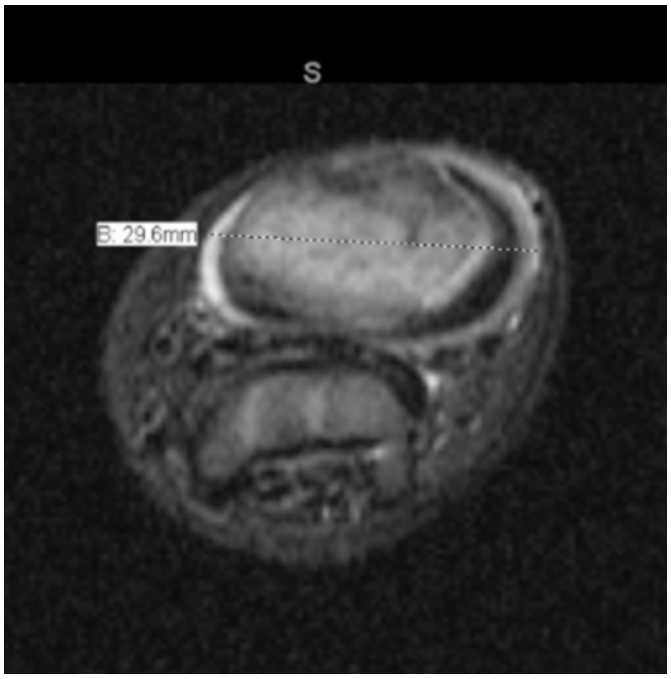
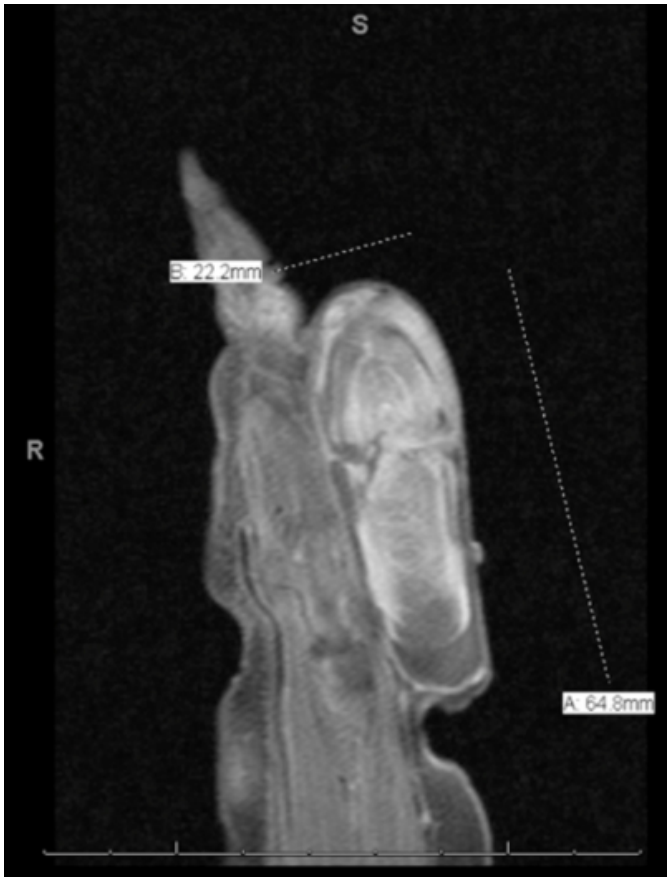


Figure 4

Fig. 4: Sagittal MRI evaluation of vascular lesion

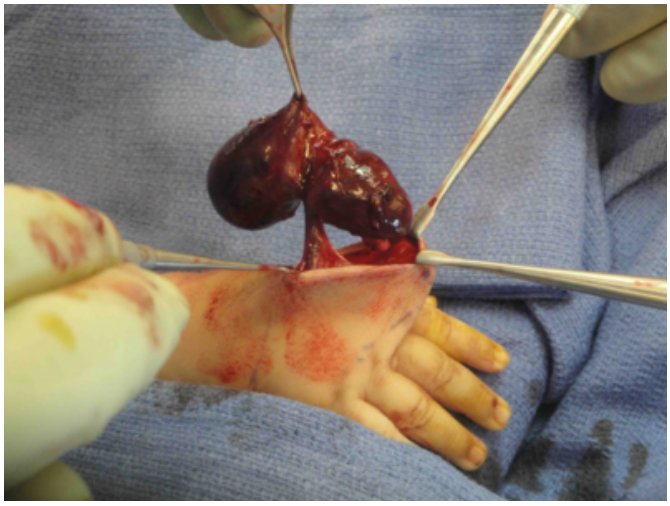


After discussion among the group and with the family, the decision was made to proceed with excisional biopsy/debulking with bloodwork to be drawn the day of surgery. Laboratory evaluation the day of surgery revealed a platelet count of 253x10E3, PT 14.1s, INR 1.09, PTT 43s, and fibrinogen of 168mg/dL.

After general anesthesia and regional block, the extremity was prepped with sterile tourniquet. A 4cm line of incision was made at the metacarpal phalyngeal levels as well as the wrist crease. The entire lesion was then easily separated out from the surrounding tissue (Figure 5).

Figure 5

Fig. 5: Intraoperative removal of lesion with pedicle visible

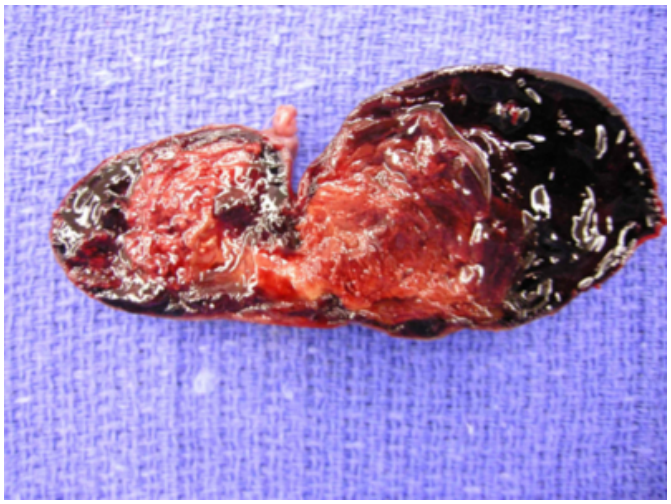


The pedicle was controlled with a #3-0 vicryl tie and the specimen was sent for permanent section. Hemostasis was achieved using electrocautery and bipolar. The patient tolerated the procedure well, was extubated and transferred to PACU in stable condition.

Permanent sectioning demonstrated a preliminary tissue diagnosis of cavernous hemangioma with secondary thrombosis (Figures 6).

Figure 6

Fig. 6: Cross-section of lesion.



GLUT-1 assay returned a negative result. Based on the clinical picture, evolution, imaging results and tissue pathology, our working diagnosis is of a thrombosed VM, now completely excised.

The patient was again seen in follow-up on postoperative day 4. No hematoma was present, the patient was moving all fingers and good capillary refill was present. The scar was healing normally. The patient was again evaluated at 6 weeks postoperatively, and was scheduled for follow-up imaging evaluation at 3 months postoperative

DISCUSSION

Venous malformations, such as in the case described, are present at birth, however they may not become clinically apparent until infancy or early adulthood. As the lesion grows with the child, there is an increased risk of functional impairment and pain as in cases of thrombophlebitis, extensive muscular localization or joint involvement[3]. This case was especially concerning due to the rapid enlargement of the lesion, worrisome for a vascular tumor versus soft tissue sarcoma. Of particular concern were the diagnoses of fibrosarcoma or rhabdomyosarcoma with prominent vascular feed. Angiosarcoma is rare in the pediatric age group, however, cases presenting as young as 3-months of age have been reported and described to rapidly enlarge over a period of weeks[8]. Angiosarcomas can occur in any region of the body, but most arise in the skin and superficial soft tissue, the head-neck region, and often present as a blue or purple lesion[9].

Given this possible diagnosis and the likelihood of complications later in life from a dorsal hand malformation, excisional biopsy and debulking was indicated. Initial ultrasound failed to provide a definitive diagnosis, and the patient underwent MRI, which provides details about the anatomic extent of the lesion, its proximity to vital structures, and involvement of tissue planes[5] and was specifically helpful in this case to rule out the solid tissue mass of a sarcoma. Episodic thromboses often occur in the setting of larger venous malformations, and can result in consumption of fibrinogen and the release of fibrin split products[4]. Clinically, this presents as localized enlargement, such as was seen in this case, and is often associated with swelling and pain within the lesion, and occasional blue or erythematous color change within and adjacent to the malformation. Other complications of venous malformation can include cosmetic disfigurement, bone thinning, demineralization with pathologic fracture and limb underdevelopment[7]. Specific treatment options are often influenced by the location of the lesion.

The two most broadly categorized locations for these lesions are head/neck versus trunk/limb. Treatment of large trunk/limb lesions may be complicated due to their extent and size. Small VMs can be treated with sclerotherapy, although complete resolution is unlikely. Because large VMs may extend into muscles and joints, such patients are often managed conservatively. Such patients often experience chronic localized intravascular coagulation with mild decrease in platelet count. Patients with large VMs are therefore often advised from early childhood to apply compression garments and to treat painful episodes with warm soaks or compresses, nonsteroidal anti-inflammatories, and sometimes oral antibiotics. Surgery plays an important, but limited, role in the treatment of VMs. In addition, surgery may help to limit joint distention from repeated hemarthroses[11].

CONCLUSION

The case of a rapidly enlarging soft tissue mass in an infant raises concerns for sarcoma and other vascular tumors. Definitive diagnosis of infantile hemangiomas, vascular malformations, and even pyogenic granulomas can be difficult on the basis of histology alone. This is important because hemangiomas are self-limiting while vascular malformations are not[12] and thus, the clinical history can often be helpful. In this case, clinical history was alarming and the physical exam was not diagnostic.Imaging studies were employed which aided in diagnosis. Due to the sensitive location of this patient's lesion, as well as the

unusual behavior and clinical characteristics, surgical intervention was determined to be the appropriate course of treatment.

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