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Case Report

Vascular lesions: Hemangioma or venous malformation?☆

Brandon Thinh Duc Dang, MD, Alan Victor Krauthamer, MD*

Department of Radiology, Columbia University College of Physicians and Surgeons, Harlem Hospital Center, New York, NY 10037, USA

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ABSTRACT

We present a case of a 62-year-old female who was incidentally found to have a venous malformation. Venous malformations are part of a larger category of slow flow vascular malformations and are associated with various familial syndromes and localized intravascular coagulation. Venous malformations were often misdiagnosed as hemangiomas; however, the treatment modalities of vascular malformations and hemangiomas vary significantly. Here we elucidate the imaging findings of venous malformations from various vascular tumors and other malformations.

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Introduction

Venous malformations are the most common type of congenital vascular malformation with an estimated incidence of 1-2 in 10,000 and prevalence of 1% [1]. Clinically, venous malformations can present as thrill-less soft tissue masses, often bluish in hue when superficial and may increase in size with valsalva, bending over, or crying. Intraluminal thrombus, local hemorrhage, or stasis of the lesion can lead to pain in some patients. Historically, venous malformations were often erroneously termed cavernous hemangiomas. Venous malformations are by definition not true vascular tumors, but rather grow in parallel with patients age. Despite having similar vascular roots, the treatment modalities of these very distinct entities are far from similar.

To remedy this confusion, in 2018 the International Society for the Study of Vascular Anomalies (ISSVA) created a classification system to differentiate various vascular tumors and vascular malformations from one another using cellular and molecular characteristics [2]. Vascular anomalies according to the ISSVA can be divided by the type (tumor or malformation) or by the flow (low and high flow). Venous malformations lie within the low flow category, which also includes lymphatic malformation, capillary malformations, and the hybrid combinations of each respectively. While this system is effective for diagnosis of vascular tumors and malformations in the laboratory setting, there is no standardized imaging

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* Corresponding author.
E-mail addresses: dang_brandon@sbclglobal.net, dangb@nychhc.org (B.T.D. Dang).

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classification system to differentiate between the 2 groups. Here we describe an incidental vascular malformation and differentiate between various imaging findings of vascular tumors and vascular malformations.

Case report

A 62 year old female with history significant for hypertension, adrenal incidentaloma and hyperactive sympathetic symptoms presented with dizziness and headache. Patient reported that her usual sympathetic episodes results in high blood pressure upward to 180s with pounding headache and chest discomfort, improved with lying down. Initial vitals on admission revealed a systolic blood pressure of 157/96 and heart rate of 105. Initial labs showed D-dimer of 154 (0-243 ng/mL), platelet of 217K (150-450K/mcl), troponin of <6 ng/L (0-14 ng/L), BNP <36 pg/mL (1-125 pg/mL), and 24 hour free metanephrine of 106 ug (36-209 ug). Physical exam did not identify any external mucocutaneous lesions. Prior CT imaging was significant for round calcification in the left anterior temporalis muscle. Repeat CT scan demonstrated no acute intracranial abnormalities; however, a small rounded calcification was noted in the left buccal space, unchanged in comparison to prior CT studies (Fig. 1). Noncontrast MRI brain was then performed which revealed an oblong T2/FLAIR hyperintense, T1 isointense focus within the anterior left infratemporal fossa, immediately posterior to the left maxillary sinus. Follow up contrast enhanced MRI of the face demonstrated avid enhancement of the lesion respectively, with fluid-fluid levels, as well as a smaller satellite lesion with similar characteristics just caudal to the dominant lesion (Fig. 2). Findings were suspicious for venous malformation or lymphangioma. After discussion with ENT, the patient opted for nonoperative management with analgesics as needed.

Discussion

Venous malformations (VM) are thought to be due to congenital errors of normal angiogenesis. Histologically, there present as a labyrinthine network of venous channels with mitotically inactive endothelium and absent internal elastic lamina. This is contrary to hemangiomas and other vascular tumors, which will demonstrate mitotically active endothelium. Phleboliths and intraluminal thrombi may also be seen. Immunohistochemical markers include negativity to GLUT1 and PROX1 and positivity to CD31 and CD34 [3]. The radiologic differential diagnosis of VMs includes lymphatic malformations, infantile hemangiomas, arteriovenous malformations, neurofibroma, dermoid/epidermoid, and soft tissue sarcomas.

The most common location of venous malformations are the head and neck (40%), extremities (40%), and trunk (20%) [4]. On CT, VMs may be seen as multilobulated, solitary, or multiple soft tissue masses with fluid attenuation and variable enhancement. Other CT findings also include remodeling of adjacent bone, adjacent fat hypertrophy, or intralesional thrombus. Phleboliths are virtually pathognomonic for VMs, showing up as rounded calcifications within the lesion. Phleboliths are nonexistent in pure lymphatic malformations as a useful differentiating tool, but may be present in mixed venous lymphatic malformations. Additionally, dermoid cysts can have round calcifications that mimic that of phleboliths, but additional findings of intralesional fat and echogenicity on ultrasound are not found in VMs. Localized bone destruction is also

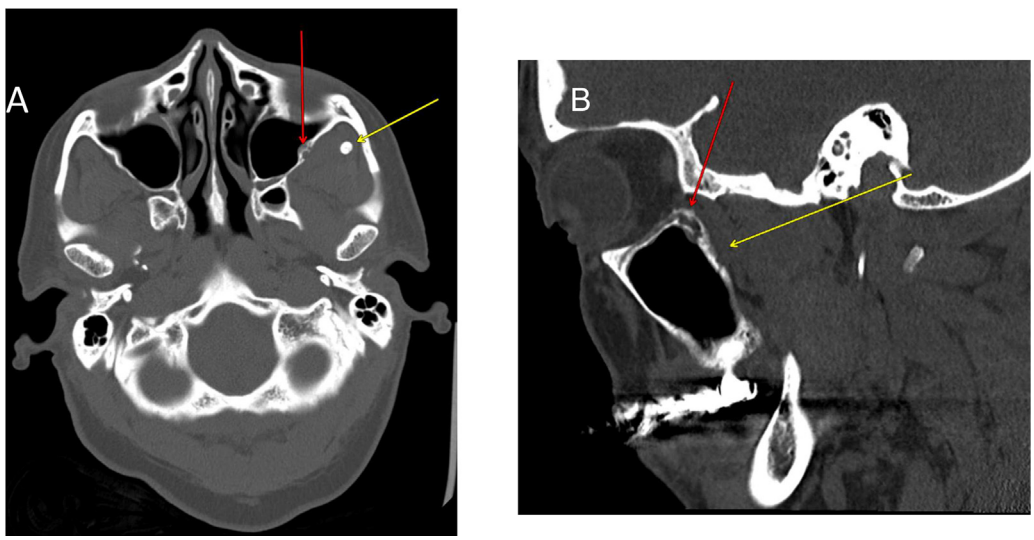

Fig. 1 – Axial CT images at the level of the adenoid tonsils (A) and sagittal CT images at the level of the left maxillary sinus (B) demonstrate bony remodeling of the posterior wall of the left maxillary sinus (red arrows) and isodense lesion within the buccal space, with obliteration of the normal fat planes (yellow arrows). Note round calcification, likely phlebolith within the lesion on axial images.

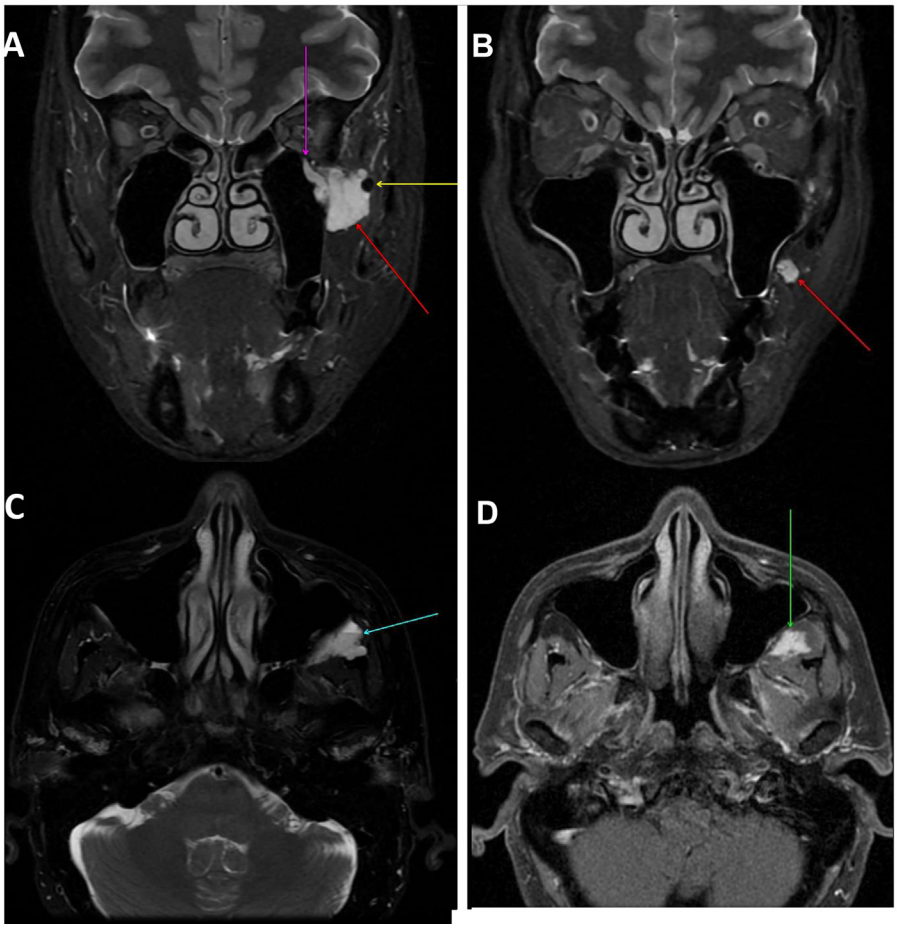

Fig. 2 – Coronal T2 fat-saturated images at the level of the posterior orbits (A) and coronal T2 fat-saturated images mandibular symphysis (B) demonstrating a T2 hyperintense lobulated lesion along the posterior wall of the left maxillary sinus with smaller similar appearing lesion in the left buccal soft tissues (red arrows), venous dilation with adjacent bony remodeling is seen (magenta arrows), and a round phlebolith within the lateral aspect of the larger T2 hyperintense lesion (yellow arrow). Axial T2 fat saturated image at the level of the superior nasopharynx (C) and axial T1 postcontrast images (D) at the level of the adenoid tonsils show fluid-fluid level (cyan arrow) and partial enhancement (green arrow).

a typical finding in soft tissue sarcomas, however fluid-fluid levels, phleboliths, and venous flow signal are not seen.

On MRI, VMs can have variable T1 and T2 appearance, however fluid-fluid levels on T2 can be seen which may represent settling blood products due to stagnation with the plasma remaining T2 bright and the dependent sediment exhibiting an intermediate T2 signal. Lymphatic malformations may share similar findings of a soft tissue mass with blood-fluid levels; however, lymphatic malformations will demonstrate thin enhancing internal septations. High flow lesions such as arteriovenous malformations may show a tangle of tortuous vascular structures, but the presence of flow voids or arterial flow on MRI is not seen in VMs. Neurofibromas can exhibit a bag of worms appearance much like a tangle of vascular structures, however neurofibromas usually present with a target sign of a T2-hypointense center and T2 hyperintense rim and association of the nerves, making this diagnosis less likely.

Ultrasound will show a heterogenous porous compressible mass with multiple distinct venous channels with variable size. While venous flow can be seen on doppler, a majority of VMs will show minimal internal flow due to the slow flow nature of the malformation. A multimodality approach to diagnosing venous malformations should be considered as the imaging findings of venous malformations are diverse.

Infantile hemangiomas can be differentiated from VMs due to their growth pattern: a small vascular tumor that emerges from the ages of 2 weeks to 2 months with vigorous growth during infancy then eventual involution. Contrary to our patient, whose age range excludes infantile hemangiomas, the lesion's stability over the years also rules against infantile hemangioma.

93% of venous malformation cases are solitary focal lesions, however; around 1% of cases are multifocal [5]. Consider syndromic etiologies if multifocal venous malformations are identified, such as familial cutaneomucosal venous malformations, blue rubber bleb nevus syndrome, glomuvenous malformation, Klippel-Trenaunay, Gorham-Stout, Sturge-Weber, and Bannayan-Riley-Ruvalcaba syndrome [6–8]. Treatment modalities for venous malformations range

from conservative therapies with compression to more invasive procedures such as sclerotherapy, laser ablation, embolization, and surgical excision [9,10]. Management of venous malformations is multidisciplinary with aims for treatment to minimize symptoms and/or cosmetic effect on quality of life. For patients electing for medical nonsurgical management of these lesions, the goals of treatment focus on preventing further venous ectasia, pain management, and complications of localized intravascular coagulopathy (LIC). Compression therapy with clothing, dressings, or compression devices have been described to reduce swelling and thrombophlebitis [11,12]. Mild analgesics and anti-inflammatory medications are typically used for pain symptoms. Disseminated intravascular coagulation (DIC), though rare, is a serious complication portended by localized intravascular coagulopathy which can manifest with elevated D-dimer and pain. Two weeks of low molecular weight heparin is typically used for LIC pain and for prevention of progression to DIC [13].

Sclerotherapy is treatment modality of choice for definitive treatment of VMs or preoperative adjunct therapy. Initial imaging with MRI with the Goyal classification or venous grading via diagnostic venography is useful for determining effectiveness of sclerosant therapy, where smaller lesions <5 cm with minimal venous drainage networks being the most optimal factors for treatment success [14,15]. Sclerotherapy may be still be used for lesions >5 cm and prominent venous ectasia, though with higher rates of repeat treatments and increased risk of complications. Complications of sclerotherapy are dependent on the type of sclerosant, with absolute ethanol possessing the highest rates of complications but lowest rates of disease recurrence [16,17]. Nerve injury, pain, hyperpigmentation, and skin necrosis have been reported. Deep venous thrombosis or pulmonary embolism are rare but important complications when considering sclerotherapy with absolute ethanol, the risk of which can be minimized with correct dose limitations and proper localization of sclerosant agents with radiopaque agents under fluoroscopy while injecting [18].

Surgical excision is typically appropriate for easily accessible and small VMs < 4 cm without involvement of critical structures. While wide excision is preferred for its high potential cure rates, in areas where the complication risk of sclerotherapy is high such as near nerves, partial or complete surgical resection may be performed. For more complex VMs without clearly defined borders or limited access, use of preoperative n-butyl cyanoacrylate glue is helpful in creating a demarcation of the lesion for resection [19]. Less described but promising new therapies for VMs treatment include cryo-ablation and laser treatments with diode or neodymium:yttrium-aluminum-garnet lasers; however, additional clinical trials will be needed to delineate their efficacy [20].

In summary, venous malformations while common have key radiologic features that are needed to distinguish it from other vascular malformations and neoplasms. As the treatment modalities for VMs are diverse, clinicians should consider a multidisciplinary approach based off the clinical presentation, imaging characteristics, and patient goals for therapy.

Patient consent

All relevant patient information was anonymized and the manuscript only includes non-identifiable images. The informed consent for publication has been obtained.

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