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Case Reports & Case Series

Venous vascular malformations and compressive neuropathy

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ABSTRACT

Background: Venous malformations (VMs) are slow flowing congenital vascular malformations that demonstrate continuous growth. They are prone to bleeding and frequent invasion of adjacent structures. It is therefore important to differentiate them from schwannomas, which can appear similar on imaging. Currently, the literature demonstrates only a few examples of pathology misdiagnosed as a schwannoma, and of VMs associated with neurological deficits. We will illustrate the characteristics and management of VMs causing compressive neuropathy in the cervical and upper extremity regions and contrast them against the features of peripheral nerve sheath tumors.

Case description: Two case reports are presented, one of a 55-year-old woman with a chronic cervical mass due to a venous malformation (VM) originally misdiagnosed as a schwannoma, and another of a 35-year-old man with a vascular malformation of predominantly venous pathology in the antecubital fossa. Both patients were surgically managed.

Conclusions: Patients presented with chronic pain, weakness and paresthesia. Venous malformations were seen as thinly encapsulated, multilobular, heterogeneously enhancing lesions with internal fluid levels and venous lakes. Post-contrast septate enhancement, intrinsic foci of T1 hyperintensity, phleboliths, T2 hyperintensity, and nodular enhancement of torturous vessels we also noted. MEP, SSEP, and EMG monitoring was used during surgical resection. Both patients had favorable outcomes.

1. Introduction

Venous malformations are uncommon pathologic entities with an incidence rate of 1 to 2 out of 10,000 people per year and prevalence of 0.1–1% [1]. Vascular malformations are thought to arise from congenital defects in morphogenesis and can be subdivided into two categories: slow flow and fast flow. Slow flow malformations include capillary, venous, and lymphatic components, while arteriovenous malformations are an example of fast flow malformations. Venous malformations grow continuously and are second only to hemangiomas in frequency with respect to vascular abnormalities affecting the head and the neck [1]. VMs also occur in the upper extremity [2]. They can be asymptomatic, or they can lead to disfiguration, pain and mass effects on surrounding organs and neural structures [3]. Correct treatment depends on an accurate differentiation between a vascular malformation and a schwannoma [2]. The distinction is paramount, as VMs require a specific approach due to their thin walls prone to bleeding and frequent invasion of surrounding critical structures [4].

Misdiagnoses are relatively common in peripheral nerve sheath tumors. The literature shows that a number of schwannomas have been misdiagnosed as a variety of other growths. For example, a case of a 50-year-old man who presented with neck pain following a motor vehicle accident was diagnosed as a tumor of the fourth ventricle, while it was actually an accessory nerve schwannoma [5]. However, there are fewer examples in the literature where other pathology is misdiagnosed as schwannomas, and only one published example of a VM masquerading as a schwannoma [2]. The importance of describing misdiagnosed cases and their characteristics is to inform surgeons how to avoid potential diagnosing pitfalls, especially when imaging is equivocal.

Here we present a large cervical vascular malformation that was

Abbreviations: VMs, Venous Malformations; VM, venous Malformation; MEP, Motor Evoked Potential; SSEP, Somatosensory Evoked Potential; EMG, Electromyographic; MR, Magnetic Resonance; MRI, Magnetic Resonance Imaging; EVG, Elastic Van Gieson

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originally misdiagnosed at an outside facility as a schwannoma, as the two often appear identical on MR imaging [1], and an antecubital fossa malformation of predominantly venous character. While schwannomas have been commonly misdiagnosed and may mimic a variety of pathology, it is rare that other tumors or growths are misdiagnosed as schwannomas. Neurological deficits due to compression by VMs, as illustrated by our case, are also infrequently reported. This is especially true of deficits in the periphery. An example of the former is a developmental venous anomaly resulting in obstructive hydrocephalus in a pediatric patient [6]. The aim of our case presentations is to aid providers in avoiding diagnostic pitfalls when treating cervical and upper extremity masses.

2. Case one presentation

2.1. History & physical

The patient was a 55-year-old woman without a significant medical history who presented with a right sided neck mass associated with chronic and intermittent pain for 6 years. There was associated right leg numbness, discomfort, headaches, and intermittent nocturnal bilateral hand numbness. She initially noticed the mass after sustaining trauma 10 years ago. Review of systems was noncontributory. Neurological examination demonstrated tenderness around the mass near the occiput. Sensorimotor and cranial examinations were within normal limits, including brisk reflexes throughout and a plantar reflex with downgoing toes. There were no other signs of inflammation or deformity in the cervical region.

2.2. Surgical treatment

A posterior approach was used for resection. A midline linear incision from C1–7 revealed a vascular malformation in the right-sided paraspinal muscles at C2–4. It was circumferentially dissected out and use of a Doppler probe confirmed the absence of arterial wave sounds within the lesion. Venous vascular malformations demonstrate distinct appearance on Doppler ultrasound [7]. The superior and inferior poles of the lesion were freed from the connecting veins by cutting between two adjacent ties. The firm, round, and well-defined lesion was freed laterally and medially from the surrounding tissue. Patient tolerated the procedure without complications (Fig. 1).

2.3. Postoperative course

The postoperative course and imaging were unremarkable. Neurological examination was at prepathological baseline with no new deficits. Cervical MRI revealed total resection of the vascular malformation. Edema and enhancement were present in the interspinous ligament and soft tissues from C2–3 to C6–7. The patient was discharged with one week of full-time care, but no additional skilled acute physical therapy needs (Fig. 2).

3. Case two presentation

3.1. History & physical

A 35-year-old man with a history of left vestibular schwannoma treated by suboccipital craniotomy in 2014 complicated by left sided facial paralysis in 2016 presented with a recurrent left antecubital fossa mass. It was originally excised in 2014 and pathologically identified as a benign vascular malformation. The mass began to return in 2016 without any associated complaints of pain, numbness or weakness. However, it doubled in size and became painful, especially with hot showers, a week prior to presentation. No trauma or constitutional symptoms were present (Fig. 3).

3.2. Surgical treatment

Resection of the malformation with decompression of the left median and superficial sensory radial nerve was performed under MEP, SSEP and EMG monitoring without complications. Multimodal monitoring has greatly improved sensitivity and specificity [8]. The patient was placed in a supine position, and the prior incision was extended. Bipolar cautery was used for dissection near the neurovascular bundle. The mass was located deep to the pronator fascia. To gain access, some pronator muscle was transected, while neuromonitoring was used to identify areas of crossing nerves. The brachial artery was separated from the deep medial aspect of the mass. The median nerve, which was located deep to the mass, was preserved. Small crossing vascular structures were tied off, and the mass was removed en bloc. Motor evoked potentials remained the same throughout the case. Sensory monitoring displayed a temporary dip in the radial sensory nerve, which begun to normalize towards the end of the case. EMG analysis of the median nerve revealed a threshold of 0.8 mA.

3.3. Postoperative course

At three weeks follow-up, the patient endorsed little to no pain. Decreased sensation over the anterior aspect of his forearm distal to the incision and over the palmar aspect of his thumb was present, but has been improving. The rest of the sensory examination was noncontributory. No constitutional complaints were endorsed, and a full range of motion was present at the elbow.

4. Discussion

This report described a patient who presented with a cervical venous malformation associated with neurological deficits, and a patient with a vascular malformation of predominantly venous character in the antecubital fossa. The purpose of describing these cases is to demonstrate that vascular malformations, though relatively rare, should be included in differential diagnoses of palpable, symptomatic cervical and antecubital growths. Examples of vascular malformations misdiagnosed as tumors are rare in the literature. Parmar et al. described an upper extremity vascular malformation that mimicked a schwannoma on imaging. The patient's mass had been palpable for 2 years with associated shoulder and chest wall pain that began one year prior. However, it was not identified as a venous malformation until the time of surgery and no neurological symptoms were noted [2], unlike our presentation.

Vascular malformations and peripheral nerve sheath tumors can appear similar on imaging. Indeed, the initial working diagnosis for our cervical mass patient was a schwannoma. Slow flow vascular malformations typically appear as hypo- and hyper-intense masses on T1 and T2 weighted MRI sequences, respectively, and display contrast enhancement [1]. Peripheral nerve sheath tumors might be confused with vascular malformations, as they may also appear hypointense on T1 and hyperintense on T2 weighted imaging, and enhance with contrast [2]. Nerve sheath tumors may hemorrhage and display hematocrit levels found in vascular malformations. Lastly, they also possess slow growth and are common head and neck tumors [9]. However, vascular malformations are frequently poorly circumscribed and include phleboliths [2], while peripheral nerve sheath tumors are well demarcated and do not demonstrate these inclusions.

Venous malformations are not often confused with lymphatic malformations or hemangiomas, as they have a distinct appearance on imaging. The former will accumulate contrast in the intercystic septa, as opposed to enhancing the content of the lesion [3]. Proliferative phase hemangiomas also have a lobulated appearance, but they display a moderately intense signal on T2 [10].

The patient's history might be diagnostically useful. For example, elevated hydrostatic pressure within the malformation when the patient is positioned supine may exacerbate symptoms [1]. The decreased

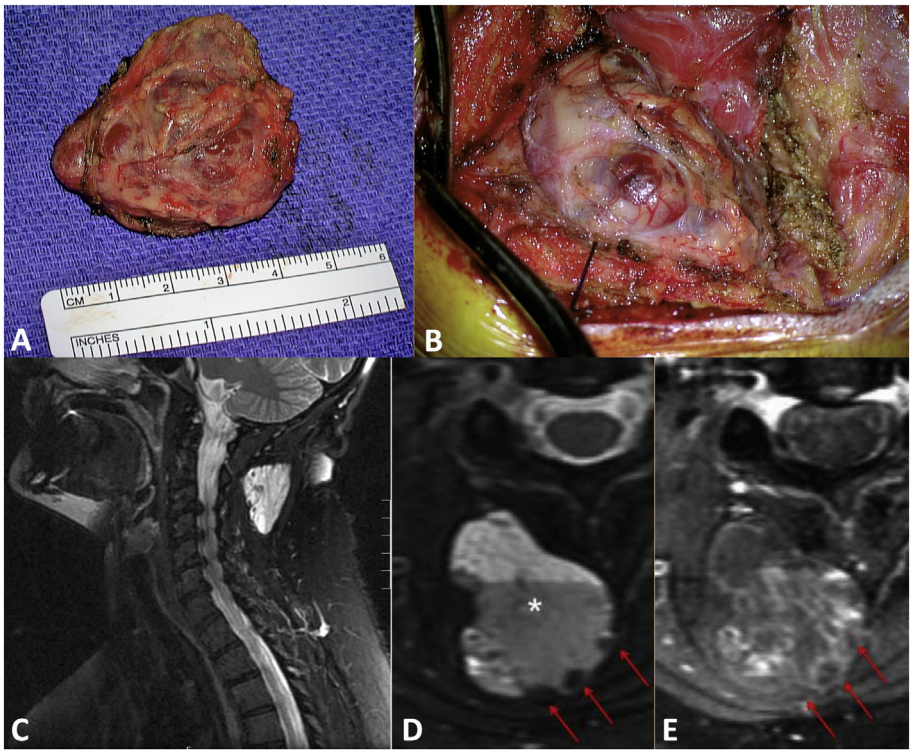

Fig. 1. Gross, MR Neurogram, T2 and T1 Post-gadolinium contrast images of the posterior cervical venous malformation mass of Case One. A: The resected mass was encapsulated and filled with deep red and brown blood breakdown products. It had thick and thin walls, and measured 2.6 × 3.6 × 4.1 cm B: Dissected mass in the field C: Sagittal T2. D: Axial T2. E: Axial T1 post-gadolinium fat saturated sequence. Images demonstrate a multilobular mass involving the posterior cervical triangle. The internal fluid-fluid level is compatible with hemorrhage (asterix). Multiple phleboliths (arrows) are seen as signal voids in the lesion and are nonenhancing. Septate enhancement with characteristic nodular enhancement of tortuous vessels is present. Taken together, these findings indicate a venous vascular malformation. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

gravity pull leads to a physical engorgement [3], which worsens the offending mass effect. This might explain why our patient experienced bilateral hand numbness when lying down at night. An increase in hydrostatic pressure from hot temperatures might also explain why the second illustrated patient experienced pain especially during hot showers. Vascular malformations can be exacerbated by sepsis, trauma or hormonal changes [1]. Our cervical mass patient likely possessed the malformation since birth, but only noted it after it might have been potentiated by her traumatic event. Similarly, our second patient experienced difficulties after the onset of his vestibular schwannoma.

This case report presented a patient suffering from a chronic cervical mass due to a venous malformation misdiagnosed as a schwannoma that caused neurological symptoms, and a patient with an antecubital vascular malformation with a predominantly venous character. The purpose of this presentation is to encourage other providers to consider venous malformations in their differential diagnosis of cervical

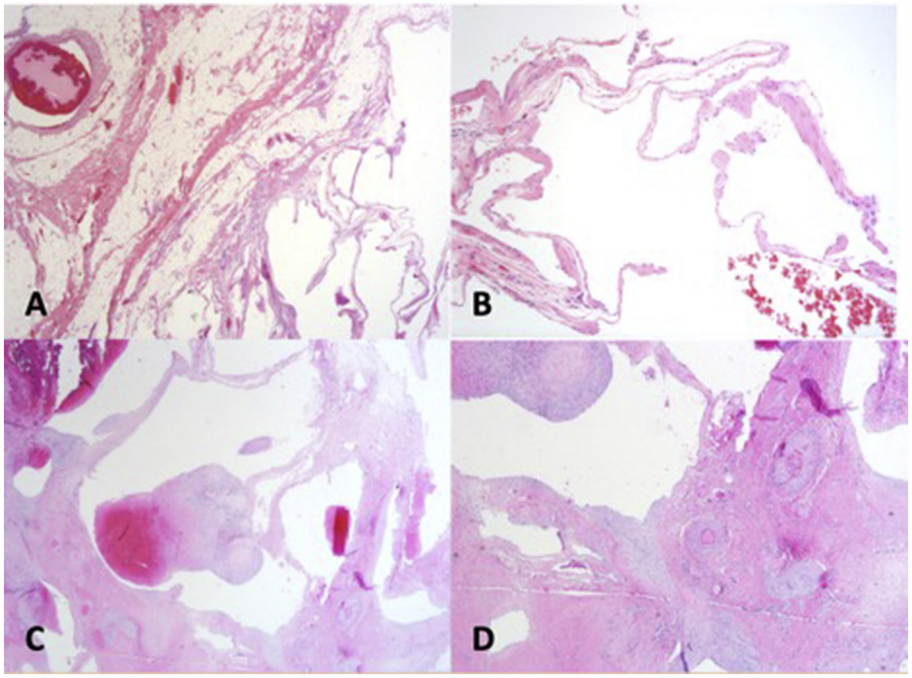

Fig. 2. Representative Hematoxylin and Eosin histologic images of both cases. A &B: Case One at 20× (A) and 40× (B) magnification. Microscopically, the mass was predominantly composed of dense aggregates of thin-walled dilatated vascular structures, and extensively involved adjacent fibroadipose tissue and skeletal muscle. Focal mural thrombus formation and circular laminated structures with mineral deposition consistent with phleboliths were present. Elastic Van Gieson (EVG) and reticulin stains demonstrated that the majority of the vascular structures lacked internal elastic lamina, indicating a paucity of arterial vasculature and excluding the diagnosis of an arteriovenous malformation. Overall, the pathologic findings were consistent with a venous vascular malformation. C &D: Case Two at 20× (C) and 40× (D) magnification. The soft, lobulated mass (5 × 4.3 × 3 cm) demonstrated a smooth, dark brown and gray external surface with a thin capsule. The inner surface consisted of an array of vessels with a varying diameter and wall thickness. Microscopically, it demonstrated variably sized vessels with dilation, hemorrhage and organizing thrombi. The majority of vessels appeared venous in nature, with occasional arterioles. Overall, the mass was best classified as an arteriovenous malformation with a predominantly venous component. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

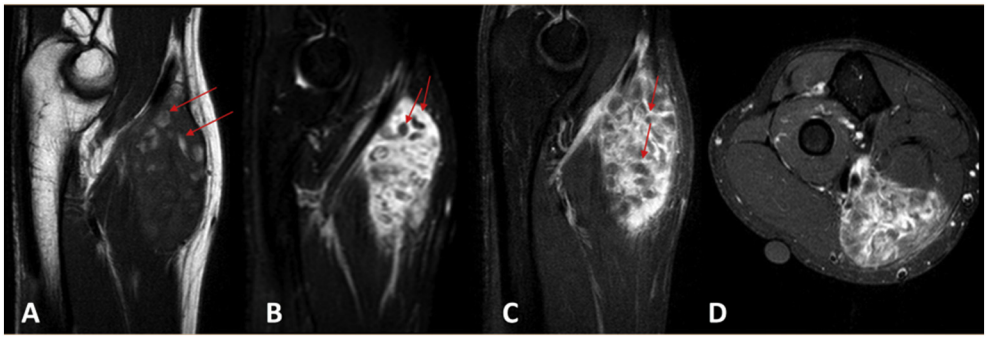

Fig. 3. MR Neurogram, T2 and T1 Pre and Post-gadolinium contrast images of the antecubital mass of Case Two. A $3 \times 4.1 \times 5.6$ cm multiseptated, T2 hyperintense mass with venous lakes separated by thin hypointense septa was identified. The heterogeneously enhancing mass was located in the left antecubital fossa between the pronator teres and extensor carpi radialis longus muscles in a close proximity to the median and radial nerve. The adjacent neurovascular bundle was displaced posteriorly, and the proximal part of the median nerve displayed a mild T2 hyperintensity. The present mass was adherent to the brachial artery and vein A: Sagittal pre-gadolinium contrast T1. B: Sagittal T2. C: Sagittal post- gadolinium contrast T1. D: Axial post- gadolinium contrast T1. The intrinsic T1 hyperintensity may represent flow within tortuous vessels (A arrows). Post-contrast septate enhancement and non-enhancing signal voids representing phleboliths were present. (B, C arrows). The above findings suggest the diagnosis of a low flow vascular malformation, such as a venous malformation.

and antecubital masses.

Disclosures

No disclosures or conflicts of Interest are present for any of the authors.

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