Chronic large symptomatic mass mimicking and compressing peripheral nerve tumor: venous vascular malformation

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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.

Objectives

- vascular malformations should be included in differential diagnoses of palpable, symptomatic cervical and antecubital growths.
- Vascular malformations and peripheral nerve sheath tumors can appear similar on imaging. However, vascular malformations are frequently poorly circumscribed and include phleboliths,, while peripheral nerve sheath tumors are well demarcated and do not demonstrate these inclusions

Case One

History and 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 negative Babinski sign. There were no other signs of inflammation or deformity in the cervical region.

Imaging

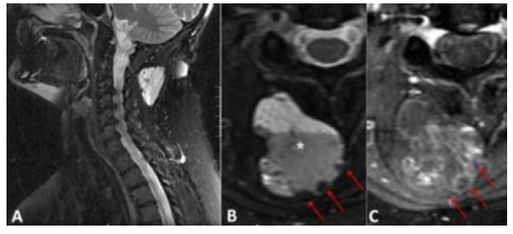

FIG.1. MR Neurogram, T2 and T1 Post-gadolinium contrast images of the cervical mass of Case One. A: Sagittal T2. B: Axial T2. C: Axial T1 post-gadolinium fat saturated sequence. Images demonstrate a multilobular mass involving the posterior cervical triangle. The internal fluid-fluid level is compatible with hemorrhage (asterix). Multiple phleboliths (arrows) are seen as signal voids in the lesion and are nonenhancing. Septate enhancement characteristic nodular enhancement of tortuous vessels is present.

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. 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.

Gross pathological Findings

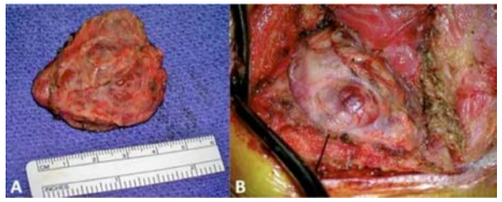

FIG 2: The resected mass (2 x 2 x 1.3 cm) was encapsulated, with thick and thin walls, and was filled with deep red and brown blood breakdown products (Figure 1).

Postoperative Course

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. No epidural fluid collection was observed. The patient was discharged with one week of full-time care, but no additional skilled acute physical therapy needs.

Case Two

History and 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.

Imaging

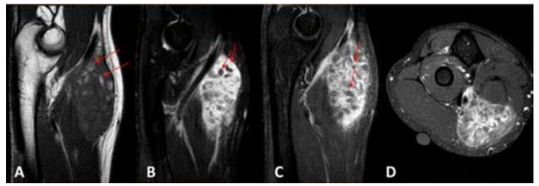

FIG. 3. MR Neurogram, T2 and T1 Pre and Post-gadolinium contrast images of the antecubital mass of Case Two. A: Sagittal pre-gadolinium contrast T1. B: Sagittal T2. C: Sagittal post- gadolinium contrast T1. D: Axial post- gadolinium contrast T1. Images demonstrate a multiseptated heterogeneously enhancing mass with intrinsic T1 hyperintensity which may represent flow within tortuous vessels (A arrows), post-contrast septate enhancement and non-enhancing signal voids representing phleboliths. (B, C arrows).

Surgical Treatment

Resection of the malformation with neurolysis of the left median and superficial sensory radial nerve was performed under MEP, SSEP and EMG monitoring without complications. 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.

Microscopic Pathology

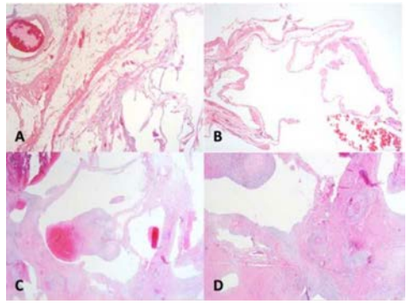

FIG.4. Representative Hematoxylin and Eosin histologic images of both cases. A &B: Case One demonstrating the venous malformation composed of thin-walled vessels, and involvement of adjacent fibroadipose tissue at 20x (A) and 40x (B) magnifications. C &D: Case Two demonstrating the predominantly venous malformation composed of thin-walled vessels with occasional arterioles at 20x (C) and 40x (D) magnification.

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.

Conclusion

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.