Median Neuropathy Caused by Intramuscular Venous Malformation in the Brachialis Muscle at the Elbow: A Case Report

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INTRODUCTION

Venous malformations (VMs) are a commonly encountered entity in clinical practice, with an estimated incidence of 1 to 2 in 10,000 births and a prevalence of 1% to 2%16,19. VMs are composed of abnormal collections of veins with a variable luminal size and wall thickness, often multiple17. They are often less well-circumscribed than are vascular tumors, such as infantile hemangiommas, and can be interspersed with adipose tissue or within various kinds of atrophic or degenerative muscle18. Although all VMs are present at birth, but they are also identified in adolescence and adulthood19. VMs occur at a frequency of 40% in the head and neck region, 40% in the extremities, and 20% in the trunk20. VMs in the extremities often violate surrounding fascial planes and can infiltrate subcutaneous tissue, muscle, bone, joints, neurovascular structures, and even viscera21.

Intramuscular VMs comprise an uncommon subgroup of VMs15. They are often mistaken for tumors, because of a similar presentation and because of improper nomenclature20. Although most intramuscular VMs in the extremities have been reported to present with a growing palpable mass with or without pain19, the development of neurological symptoms is rare. The authors report a rare intramuscular VM originating from the brachialis muscle and showing symptoms of median nerve involvement.

CASE REPORT

A 58-year-old, right-handed male patient presented with a gradually growing mass in the distal upper arm and mild weakness in left-hand grasping. The mass occurred 6 months prior to the presentation, and there was no pain when it first occurred. The mass gradually increased; in the morning, it became larger, and in the afternoon, it became smaller (Fig. 1A). A month previously, when he touched the mass, he felt tenderness and tingling in the thenar area of his left hand. Two weeks later, the mass got bigger, and his left-hand grip weakened; when he grabbed an object, he began to drop it.

On examination, there was no objective weakness in forearm pronation, wrist flexion, flexion of the first 3 digits and thumb opposition, or abduction, which are innervated by the left median nerve, nor any objective sensory disturbance in the radial 2/3 of the palm that is innervated by the palmar cutaneous branch of the median nerve. There was no tenderness in the mass itself, but when the medial of the mass was pressed, there was pain along with tingling instantaneously in the thenar side of the left palm. It was not mobile. X-ray examination showed a small calcification in the soft tissue in front of the
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Distal humerus (Fig. 1B). Magnetic resonance imaging (MRI) of the upper extremity revealed a 3-cm-sized, lobulated mass within the brachial muscle in the distal upper arm (Fig. 2B).

A T2-weighted fast spin-echo image revealed a well-demarcated, lobulated mass with a hyperintense signal (Fig. 2B). Inside the hyperintense lesion, the heterogeneous appearance with low signal intensities indicated hemorrhage or thrombosis, and often dilated vascular structures were identified. In the T1-weighted image, it was isointense with lesion extent appearing inconspicuous to adjacent muscular tissue (Fig. 2C). Gadolinium administration resulted in heterogeneous enhancement within the mass (Fig. 2C). The veins of the antecubital fossa were dilated and showed ectasia. The left median nerve was not directly involved by the lesion, but was displaced by the swollen brachialis muscle (Fig. 2D). With these unique MR findings, the lesion was thought to be a low-flow VM. Considering the gradual enlargement of the mass and worsening of the pain and neurological symptoms, surgery was planned, and consent was obtained.

After we made a lazy S-shaped vertical incision along the medial antecubital area, we carried dissection down to the antebrachial fascia. After securing the left median nerve, we dissected the swollen brachial muscle under microscopic vision.
Table 1. Classification of vascular anomalies according to vascular dynamics

| Category                  | Subcategory | Description                                                                 |
|---------------------------|-------------|-----------------------------------------------------------------------------|
| I  Hemangioma              |             |                                                                             |
| II Vascular malformations | A Low-flow (venous malformation) |
|                            | B High-flow (arteriovenous malformation) |
| III Lymphangioma           |             |                                                                             |

We did circumferential dissection by identifying a fairly firm venous structure (Fig. 3A). Between the muscle fibers, multiple dilated large venous channels and fibrous septa were identified and ligated with hemoclips. Bleeding venous structures and dilated drainers were controlled by bipolar coagulation. The VM was firmly attached to the surrounding brachialis muscle. However, no difficulty was found in complete excision (Fig. 3B, C). No neurological abnormalities were observed after complete excision, and the mass did not appear again. By one year after surgery, the mass was no longer present, and there was no abnormality in the left hand and arm movements and sensations.

**DISCUSSION**

1. Intramuscular VM

Identification and classification of vascular anomalies were hampered historically by the use of confusing nomenclature⁷). Early classification suggested by Virchow²⁰) and Wegener²¹) classified vascular lesions according to the pathologic appearance of the vessel⁷). Vascular growths were divided into angiomalous and lymphangiomalous without consideration of the biologic behavior and natural history of the vascular lesions⁷). Consequently, there was a tendency to identify any vascular anomaly as a hemangioma⁵⁻⁷,¹³,¹⁶). A variety of terms including “venous angiomalous”, “cavernous angiomalous”, “cavernous hemangiomalous”, and “phlebangiomalous” have been used in the medical literature to describe these anomalies⁵⁻⁷). These terms have led to confusion with the more common proliferating or true hemangiomalous of infancy⁵⁻⁷). For example, capillary hemangiomalous, nevus flammeus, and port-wine stain have all been used in the literature to describe a capillary malformation of the skin⁵⁻⁷).

In 1982, Mulliken and Glowacki⁶⁰) proposed a modern classification of vascular anomalies according to the lesion’s biologic and pathologic differences; all vascular anomalies were assigned to 1 of 2 broad categories: hemangiomalous and vascular malformations. The former category was later expanded to include vascular tumors. The suffix “-oma” was to be reserved for only those lesions exhibiting increased cellular turnover, the classic example within this category being the infantile hemangiomalous¹³). The term “vascular malformation” was applied to those lesions present at birth growing commen-surately or pari passu with the child¹⁵). The vascular malformations were composed of normal “mature” flat endothelial-lined vascular spaces with a normal rate of cell turnover and were further subdivided into capillary malformations, VMs, arterial (arterio venous) malformations, and lymphatic malformations⁵⁻⁷). In 1993, Jackson et al.⁹) classified vascular malformations according to flow patterns instead of the former anatomicopathologic classification for ease of investigation and treatment (Table 1). They simplified flow patterns within vascular malformations as either low-flow (VMs) or high-flow (arteriovenous malformations), keeping separate categories for lymphangiomalous and hemangiomalous, with the purpose of creating “system directly related to investigation and treatment”⁹).

2. Clinical Manifestation and Diagnosis

The diagnosis of VM and differentiation from other vascular malformations can be usually made by clinical history and
physical examination. Because VMs are congenital lesions, they may be identified at birth. However, they are usually identified from infancy to puberty, which is the period of greatest enlargement of the lesion. Continued linear growth within the malformation despite the end of somatic growth in late adolescence often results in clinical manifestations later in life and is typically the case in deeper lesions.

VMs typically appear as soft, compressible, blue-tinged masses that can enlarge with dependent position and Valsalva maneuver. The blue tinge is considered pathognomonic and is caused by dilated venous channels within the dermis. Forty percent of VMs occur in the head and neck region, and may involve the mucosa of the tongue, palate, orbital, mandibular, or neck region, even direct involvement of the ophthalmic branch of the trigeminal nerve in the face. VMs in the extremities often infiltrate the surrounding tissues. As such, patients commonly experience symptoms as a result of several mechanisms. Venous engorgement secondary to dependent positioning, exercise, after prolonged stasis, or after morning awakening frequently results in significant swelling and pain. Mass effect may cause local compression of the nerve and muscular contracture or restricted range of motion of an adjacent joint. Local hemorrhage and local stasis on a background of chronic low-grade intravascular coagulopathy thromboembolic state within the lesion can occur. In the current case, a VM occurred in late adulthood at the age of 58 and originated from the brachialis muscle. It showed swelling in the morning due to venous stasis and symptoms of left median nerve irritation due to local mass effect.

The imaging modality of choice for VMs, which classically appear as either isointense or hypointense on the T1-weighted sequences. The lesions appear focal, diffuse, or demonstrate lobulated margins. A more heterogeneous appearance can be identified in the setting of hemorrhage or thrombosis, and often dilated and serpiginous vascular structures can be identified compatible with abnormal veins. Lower signal area or signal voids may represent dystrophic calcification or phlebolith on all imaging sequences. In addition to calcification, lower signal areas on T2 can be caused by either vascular channels or fibrofatty septa. Gadolinium administration results in homogeneous or heterogeneous enhancement within the substrate of a VM.

3. Treatment of Intramuscular VM

Treatment is generally indicated if the lesion causes pain, functional impairment, or aesthetic problems, as in craniofacial lesions. With the exception of some superficial VMs where laser therapy is effective, VMs are generally treated with direct surgery and sclerotherapy. Surgical resection is considered preferable if the lesion could be completely removed so as to avoid recurrence. This includes patients with local well-defined VMs that are thrombosed, confined to a single or specialized muscle group, or causing a neurological or compression syndrome, and patients where there is a good possibility of anatomical and functional restoration. Many lesions are infiltrative, however, and involve multiple muscle groups or fascial planes where surgical resection results in an unacceptably high functional and cosmetic deficit. Sclerotherapy has been increasingly incorporated in surgical regimens and now has been considered as an adjunct to surgery or the stand-alone therapy of choice for most VMs. We chose surgical resection in the current case because the lesion was well-localized within the brachialis muscle and caused local neurological symptoms by mass effect. The treatment resulted in complete relief of the symptoms of VM, with no recurrence up to 2 years after surgery.

CONCLUSION

Here, we report on a rare intramuscular VM originating from the brachialis muscle in the left arm, which caused the local mass effect, showing the symptoms associated with irritation of the median nerve. The lesion was diagnosed through a typical MR imaging finding. Because the lesion was well-localized within the brachialis muscle, surgical resection resulted in complete symptomatic relief without recurrence.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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