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# **Case Report**

# Surgical excision of calcified intramuscular arteriovenous malformation of the leg after failed angioembolization

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#### ABSTRACT

Arteriovenous malformations (AVMs) can occur in any location but are more commonly seen in the brain. Surgical excision combined with embolization therapy is the preferred method of treatment. We had a young female with intramuscular AVM of left leg and the patient has already undergone sclerotherapy and embolization multiple times but has not been relieved of symptoms. We have excised the severely calcified stone-like AV malformation with ligation of the feeder artery. In this report, we have discussed the challenges associated with performing surgery post-angioembolization and the importance of titrating excision with the associated risk of functional disability due to loss of muscle tissue.

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#### 1. Introduction

The Peripheral vascular malformations can range from simple hemangiomas to larger arteriovenous fistulas. Vascular malformations can occur at any age. The origin of vascular malformations may be from artery, vein, capillary, or lymphatics. AVM is relatively uncommon and contributes to 7 % of all soft tissue tumors. <sup>1</sup> Most patients with AVM remain asymptomatic and are often treated for cosmetic purposes. In this case report, we discuss a middle-aged female who presented with pain and swelling in the left lower limb with a history of multiple sittings of sclerotherapy and embolization and underwent excision of calcified stone like AVM.

## 1.1. Case report

A 28-year-old female patient presented to our outpatient department with complaints of swelling in the left lower limb for twelve years which was initially small in size

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measuring around 2 X 2 cm when she first noticed it but gradually increased to its current size of about 8 X 12 cm. She has developed pain in the swelling for the past three years after which she had undergone embolization and sclerotherapy multiple times. On physical examination, tender non-pulsatile swelling of size 9 x 5 cm is noted in the posterior compartment of the leg with no skin changes. The swelling was firm in consistency, relatively immobile, and non-compressible, and the skin overlying the swelling could be pinched. The margins were not well defined and its attachment to the underlying bone could not be checked due to tenderness. Routine blood investigations were within normal limits.

The chest X-ray and electrocardiogram were also normal. The Ultrasound Doppler study was suggestive of a low-flow vascular malformation in the intramuscular compartment of the leg. Contrast-enhanced computed tomography showed arteriovenous malformation with calcifications within the medial head of the gastrocnemius muscle (Figure 1). Magnetic resonance imaging showed AVM in the medial head of the gastrocnemius muscle with a feeder vessel

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**Fig. 1:** CECT showing calcified AVM in **A:**) Coronal section **B:**) Axial section **C:**) 3 D reconstruction



**Fig. 2:** MRI showing AVM in medial head of gastrocnemius muscle with **A**) Size **B**) feeder vessel



Fig. 3: Excised AVM specimen

from the posterior tibial artery (Figure 2). Since the patient has already failed previous percutaneous interventions and was symptomatic (Schobinger clinical grade 2), surgical excision was planned after discussing the case in a multidisciplinary forum amongst a vascular surgeon, an orthopedic surgeon, and an interventional radiologist. Under spinal anesthesia, the patient was placed in a supine position with the left leg flexed and externally rotated, and a vertical skin incision was made over the swelling. The subcutaneous tissue was dissected till the gastrocnemius muscle was reached and hard calcified AVM was found. It was dissected circumferentially and excised in-toto (Figure 3). The specimen was sent for histopathological examination. A 12 French closed suction drain was inserted and the incision was closed in layers. The drain was removed on the first postoperative day and the patient was mobilized. Postoperatively, the patient was relieved of her symptoms without any disturbance in the gait. The histopathological report confirmed the vascular malformation and the patient is doing well to date.

#### 2. Discussion

Vascular malformations are of a wide spectrum ranging from simple small hemangiomas to large arteriovenous fistulae. The AVMs comprise 4.7 to 15% of all vascular malformations.<sup>2</sup> These are low-resistance and high-flow vascular channels with a central nidus of interconnected arteries and veins.3 Jackson et al. developed radiological classification based on flow dynamics which includes low-flow malformations such as venous malformations, lymphatic malformation, and mixed malformations and high-flow malformations such as arteriovenous malformation and arteriovenous fistulas.<sup>4</sup> Hamburg classified AVM as truncal and extra-truncal. Truncal AVM is further subdivided into superficial and deep. The extratruncal is further subdivided into infiltrating and limiting lesions. Vascular malformations can be isolated lesions or associated with syndromes like Sturge-Weber syndrome, Klippel-Trenaunay syndrome, etc.<sup>5</sup>

Congenital maldevelopment is the most common cause of AVM. The incidence is 1/100000 population per year. Males and females are equally affected with symptoms mostly occurring between the age groups of 35 and 40. They most commonly occur in the brain, neck, pelvis, and lower extremities. <sup>6</sup>

AVM presents with pain, bleeding, congestive cardiac failure, or ulceration. Schobinger clinical grading help in estimating disease progression. Diagnostic modalities include ultrasound Doppler, computed tomography, magnetic resonance imaging, or conventional angiography. Conventional angiography is the gold standard tool for identifying feeding vessels. Magnetic resonance imaging helps to decide the resectability of malformation.

AVM can be managed both surgically and in non-surgical modalities like trans arterial embolization or sclerotherapy. The peripheral AVMs have been classified angiographically into three types which determine the percutaneous treatment modality. The Arteriovenous fistulas classified as Type I have a single draining vein and no more than three feeding arteries. Multiple arterioles shunt to the focal segment of a single draining vein in type IIa, whereas many arterioles shunt to the venous sac with many draining veins in type IIb. Multiple arterioles shunt along the long element of the draining vein in type IIc. Multiple arterioles shunt numerous draining veins through numerous fine fistulae in Type IIIa and through several enlarged fistulae in Type IIIb. The embolotherapy techniques are based on this classification system. Arteriovenous fistulas of Type I and II AVMs are embolised with coils, either with a direct puncture or a transvenous catheter technique. Trans arterial or direct puncture ethanol injection is used to treat Type III AVMs. Surgery is indicated when percutaneous techniques fail to treat the AVMs or sometimes surgery can be offered straight away. During surgery, it is essential to remove the nidus along with ligation of the feeder vessel.<sup>8</sup> If Nidus is left behind and only ligation of the feeder artery is done, recurrence is common. Rarely reconstructive surgery is needed depending on the location. If a major vessel of extremities is involved peripheral bypass may be needed. Intraarticular AVM needs reconstructive surgeries. Intraoperative hemorrhage can be minimized by preoperative embolization. Preoperative embolization with N butyl cyanoacrylate has shown good outcomes in shortand long-term follow up.9

#### 3. Conclusion

Arteriovenous malformations should be treated early before they land in complications like bleeding, ulceration, and congestive heart failure. Magnetic resonance imaging help in deciding the extent of excision. Conventional angiography helps in identifying the feeder vessels. Surgical excision combined with preoperative embolization is the preferred treatment of choice. Diffuse intramuscular AVMs can also be excised completely and the involvement of a multidisciplinary team including an orthopedic surgeon and a vascular surgeon takes care of complications like bleeding and functional disability.

### 4. Source of Funding

None.

#### 5. Conflict of Interest

None.

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