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Case Report

Endovascular embolization for managing a massive shoulder arteriovenous malformation ☆☆☆

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ABSTRACT

Arteriovenous malformations (AVMs) are congenital high flow pathologic linkages between arteries and veins of different sizes that may occur in any part of the body. The clinical presentation is largely dependent on the size and location of AVMs and can range from an asymptomatic birthmark to congestive heart failure in extreme cases. In this report, we describe a 20-year-old male who presented with a large AVM of the right shoulder that resulted in significant cosmetic and physical impairment and treated with several sessions of endovascular embolization with good clinical outcomes. This case highlights the complexity of diagnosing and managing these AVMs. Most of these anomalies require a multidisciplinary approach that integrates both trans-catheter and surgical interventions with trans-arterial lesion embolization being the cornerstone of the treatment.

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Introduction

Arteriovenous malformations (AVMs) are rare vascular anomalies that are characterized by abnormal linkages between the arteries and veins anywhere upstream the capillaries [1]. Triggered by certain environmental and hormonal factors, these AVMs can grow massively resulting in hemodynamic, and cosmetic sequelae [1]. The hemodynamic effect is attributed largely to the shunting phenomenon of the high velocity blood flow from the arterial to the venous side causing venous hypertension, edema, inflammatory

changes, and ischemic changes seen in advance stages [1,2]. Often a congenital disorder, AVMs can present in the head and neck, lungs, heart, or in the upper and/or lower extremities. The clinical diagnosis is confirmed by Doppler ultrasonography and/or computed tomography (CT) angiography with trans-arterial angiogram remaining to be the gold standard. Management of AVMs is often challenging due to the anatomic and morphologic complexity. Further, it often requires a multidisciplinary approach that consists of both endovascular, and surgical interventions. In essence, these lesions are de-vascularized via trans-catheter embolization using mechanical, and chemical agents followed by surgical intervention as clinically warranted [1,3].

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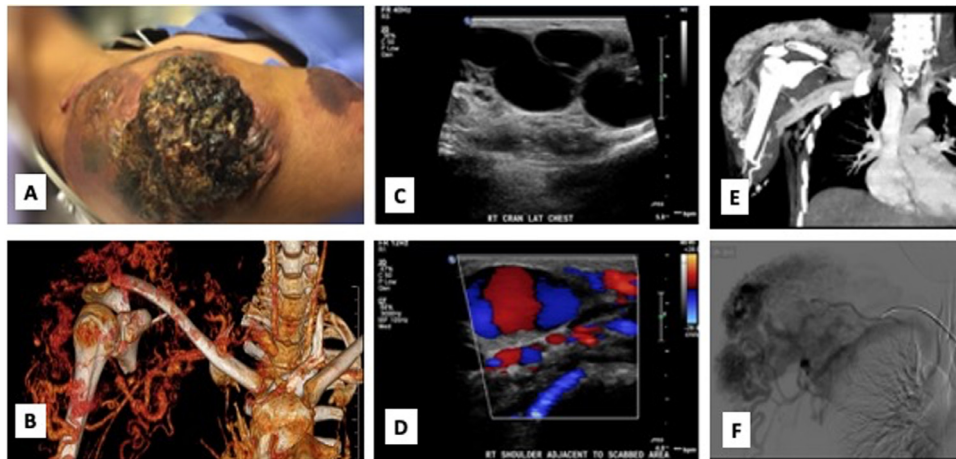


Fig. 1 – (A) Clinical appearance of the right shoulder arteriovenous malformation (AVM) showing necrotic areas accompanied with inflammatory changes. (B) CT angiogram of the chest with post-processing 3-dimensional (3D) view. (C and D) Doppler ultrasonography of the right upper extremity. (E) CT angiogram of the chest shows the complex morphology of the AVM. (F) Selective arteriogram of the shoulder showing complex vascular structure with multiple niduses.

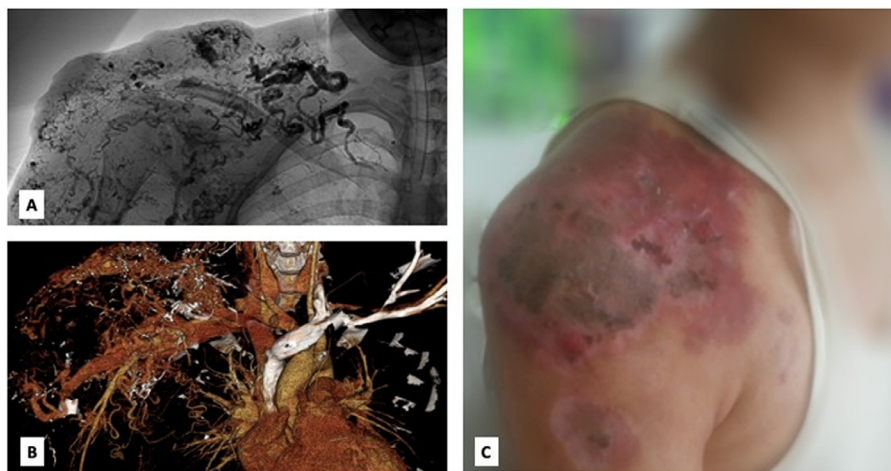


Fig. 2 – (A) Angiographic appearance of the N-butyl cyanoacrylate glue-filled AVM vessels after embolotherapy. (B) CT angiogram of the chest with post-processing 3-dimensional view showing the N-butyl cyanoacrylate glue filling the blood vessels. (C) Clinical appearance of the well-healed shoulder AVM after skin grafting.

In this report, we describe the interesting case of a giant AVM of the shoulder in a young male that required a multidisciplinary staged approach, and resulted in good clinical outcomes.

Case report

A 20-year-old Hispanic male with no past medical history presented to our facility complaining of worsening right shoulder AVM over the last 2 years. More history revealed that the AVM started as a birth mark then grew throughout the course of his life without experiencing any symptoms. However, over the last few months, the patient reported severe pain attacks and decreased range of motion of the affected shoulder. Furthermore, the AVM started to grow with worsen-

ing appearance, and occasional ulceration accompanied with subsequent bleeding. In addition to the poor cosmetic appearance, the patient's quality of life was impaired to the degree that he was not able to maintain working. Importantly, no focal neurologic deficits or seizures were reported. Physical exam revealed a large vascular malformation measuring 8×4 cm involving the soft tissues of the right shoulder, neck, and supraclavicular fossa extending to the mid right arm (Fig. 1A). Further, the range of motion of the shoulder with abduction, and adduction was limited. Vitals signs were within normal limits. On auscultation, a bruit was heard at the front aspect of the right shoulder.

Doppler ultrasonography of the right upper extremity revealed a large AVM involving the anterior right shoulder, supraclavicular region, and upper arm with pulsatile venous and arterial components (Fig. 1C and D). Computed tomography (CT) angiogram of the chest showed a large AVM overlying the

right shoulder with multiple feeding arteries that arise from the brachiocephalic artery (Fig. 1B and E). In order to delineate the exact angioarchitecture of the AVM, diagnostic digital subtraction angiography was performed, which revealed an extensive right shoulder, and neck AVMs with multiple feeding vessels arising from the right thyrocervical trunk, brachial artery, vertebral artery, superior thyroid artery, lateral thoracic artery, and right occipital artery (Fig. 1F). In order to rule out other organ involvement, CT angiography of the brain, abdomen and pelvis was done, and revealed no vascular abnormalities.

After reviewing the morphology of the AVM, the anatomy of the feeding arteries, and the endovascular accessibility, a staged, multidisciplinary approach between interventional radiology and both vascular surgery and plastic surgery was planned to treat the shoulder AVM. Essentially, the prominent feeding arteries were initially embolized and obliterated using a mixture of N-butyl cyanoacrylate glue (Trufill, Cordis, Miami Lakes, FL), ethiodized oil (Guerbet, Princeton, NJ), and tantalum powder (Admat Inc, Norristown, PA). Coil embolization was used to further exclude the large feeding arteries. In order to achieve complete obliteration of the AVM, a total of 4 sessions of embolization was necessitated (Fig. 2A and B). The inflammatory response after embolization was treated symptomatically. A few weeks after embolization, the patient underwent debridement followed by a split-thickness skin graft reconstruction of right shoulder wound using the right thigh donor site. One year later, the patient showed good healing of the AVM site (Fig. 2C), and marked improvement of the clinical symptoms, including the range of motion of the shoulder.

Discussion

AVMs are rare pathologic connection between the arteries and veins anywhere upstream of the capillaries with an estimated prevalence of 1:100,000 person-years [1]. Embryologically, AVMs stem from birth defects that involve the arterial and venous vessels leading to direct communications between the different-sized vessels, which eventually form undifferentiated dysplastic networks called nidus [1–3]. Outside the head and neck, the upper and lower extremities are the most common locations of AVMs [1]. Remarkably, most peripheral AVMs that involve the extremities have a nidus interpositioned between the feeding arteries, and draining veins [3,4].

The pathophysiologic progression of AVMs is attributed to the blood shunting of high velocity flow from the arterial side of AVM into the venous system. The arterio-venous shunting may progress, leading to significant anatomic, pathophysiological, and hemodynamic effects resulting in different clinical symptomatology based on the AVM location, and size. Several environmental and hormonal factors are proposed as possible triggers for AVM growth [1,4,5].

While most of AVMs are asymptomatic, these vascular anomalies frequently become symptomatic after they are large enough to cause hemodynamic, and local changes. Long-standing AVMs of the extremities are associated with venous hypertension, which potentially could lead to venous aneurys-

mal formations, edema, and chronic inflammatory changes. In rare occasions, AVMs can progress to cause steal syndrome, resulting in high output cardiac heart failure [1,6].

Several diagnostic modalities can be utilized to diagnose AVMs. Non- and minimally invasive modalities include Doppler ultrasonography, computed tomography, and magnetic resonance angiography. Findings on Doppler ultrasonography imaging consist of hypervascular networks of dilated tortuous channels between the arteries and veins. However, in general, contrast-based CT and/or magnetic resonance angiography are usually necessary for definitive diagnosis. Nevertheless, catheter-based angiography remains the gold standard for both diagnostic and treatment planning purposes [1,2,6,8].

Our patient had peripheral AVMs that affected the right shoulder and upper chest while also showing multiple niduses on transcatheter angiography. Both Doppler ultrasonography and CT angiography were utilized to further delineate the morphology and anatomy of the feeding arteries. Subsequently, the information obtained from CT angiography was very valuable in planning the course of treating the AVMs.

With regards to management, it is well acknowledged that most AVMs require a multidisciplinary approach that integrates multiple clinical services including interventionalists and both plastic and vascular surgery [1]. In general, the goal of managing peripheral AVMs is to devascularize these vascular structures using different embolotherapy approaches that employ either the arterial or venous side of the AVM or via direct nidal injection. Importantly, the embolotherapy is generally staged on multiple sessions in order to minimize the risk of ischemic complications of the adjacent tissues [1,7].

Several embolotherapy agents are used in managing AVMs such as N-butyl cyanoacrylate (NBCA), ethanol, and coiling. The primary goal of embolization is to reduce the nidus size, and eventually de-bulking the AVM, so that the surgical intervention can be facilitated [9]. Overall, with the development of endovascular trans-catheter therapy, the role of surgical intervention has shifted from being the first line of treatment to become an adjunctive modality to embolotherapy. Because most peripheral AVMs have complex nidus and morphology, coils are often combined with the polymerizing and/or sclerosing agents to facilitate occluding the feeding arteries. The combination of coils and NBCA was used in our case, given the complexity, and the extension of AVM [1,4,7].

Relevantly, NBCA is a polymerizing liquid adhesive agent that polymerizes upon contacting the blood. NBCA is usually combined with ethiodized oil (lipiodol) in order to slow the polymerization time. The mechanism of action is linked to its mechanical and chemical effects leading to devascularization of the AVM, a merit that helps in controlling the post embolization bleeding [1,7,8].

In conclusion, AVMs are a rare, challenging, and potentially life-threatening form of vascular anomalies that are usually asymptomatic. For symptomatic and aggressive AVMs, the initial diagnosis and assessment are formulated by non- to minimally invasive investigations. However, trans-catheter arteriography of AVM lesion remains the gold standard for diagnostic and treatment planning purposes. The treatment of AVMs often requires a multidisciplinary approach that incorporates both embolotherapy and surgical interventions.

Author contributions

All authors contributed equally to this article. All authors read and approved the final manuscript.

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Patient consent

Written informed consent was obtained from the patient for all procedures and publication of this case and accompanying images.

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