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Management of a ruptured large arteriovenous malformation: A case report

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Case Report

ABSTRACT

Arteriovenous malformations (AVMs) are congenital vascular disorders. In their early phase, they often go unrecognized due to their asymptomatic nature until they develop clinical symptoms such as swelling, skin discoloration, and signs of arteriovenous shunting. The high flow from the arteries directly into the veins may potentially increase the risk of vessel rupture. We reported a case of a 54-year-old male who complained of back bleeding, accompanied by pain and sores that had persisted for the previous three days. The patient had been aware of a mass since birth, progressively worsening with age. A physical examination revealed bleeding from the pulsating mass and measured approximately 17.5 cm x 15 cm x 3 cm on the back. The patient was diagnosed with ruptured AVM by Computed Tomography (CT) angiography. He was managed with endovascular coil embolization, polyvinyl alcohol particles, and skin grafts. AVMs are often asymptomatic; an ultrasound examination can be the initial diagnostic tool, while CT Angiography is considered a gold standard. AVMs are managed through embolization and surgical intervention. Regular follow-up is necessary due to the potential for recurrence.

INTRODUCTION

Arteriovenous malformations (AVMs) are congenital disorders that can arise during embryogenesis or develop postnatally. These malformations are characterized by abnormal fistulas connecting arteries and veins, resulting in direct communication between these vessels. The AVMs can manifest extensively, leading to hemodynamic disturbances and cosmetic issues. The interconnected arteries and veins within AVMs contribute to high blood flow in the veins, increasing the risk of fatal vascular rupture.[1,2]

The most common locations for AVM development are the head and neck area (47.4%) and the upper or lower extremities (28.5%). Although less frequent, AVMs can also occur in other areas such as the heart or lungs.[3,4] Diagnosis of AVM relies on a thorough history, physical examination, and supporting tests. Often, AVMs remain undetected until changes in skin colour, mucosal layers, increased pulsation, or noticeable tissue growth occur.[5–7] Diagnostic tools include Doppler ultrasonography and CT angiography with transarterial angiogram as a gold standard.[8,9]

Treatment options for AVMs include transcatheter embolization using chemical agents and mechanical methods, followed by surgery if necessary. The goal of embolization is to reach the nidus (the central abnormal connection) and to close the arteriovenous shunt. However, proximal artery embolization should be avoided due to a high risk of failure and worsening symptoms.[10–12]

In this case report, we explore roles of diagnostics and gradual endovascular management steps in managing a largely ruptured arteriovenous malformation in a 54-year-old male patient.

We also examine roles of regular follow-up in detecting and managing potential recurrences of the AVMs. This case provides insights into the diagnosis, management, and follow-up of AVMs, particularly those that are large and ruptured. We hope this case report can provide a significant contribution to the health and medicine development in Indonesia.

CASE DESCRIPTION

A 54-year-old male patient came to an emergency room with complaints of back bleeding, accompanied by pain and sores that had persisted for the previous three days. The patient reported a pulsating mass on his back that had appeared since birth and had grown larger over time. Initially, the mass was asymptomatic, but in recent months, pain began to manifest in the mass, accompanied by discoloration, which progressively worsened. A physical examination revealed bleeding from the mass, which was identified as a vascular malformation with pulsation. The mass measured approximately 17.5 cm x 15 cm x 3 cm on the back (Figure 1 A). His shoulder and neck movements, as well as vital signs, were within normal. Doppler ultrasonography of the posterior thoracic region revealed large AVMs with pulsatile arterial and venous flow. A computed tomography (CT) angiogram of the back showed large AVMs overlying the back, with multiple arteries originating from the branches of the subclavian and axillary arteries (Figure 1 B and C).

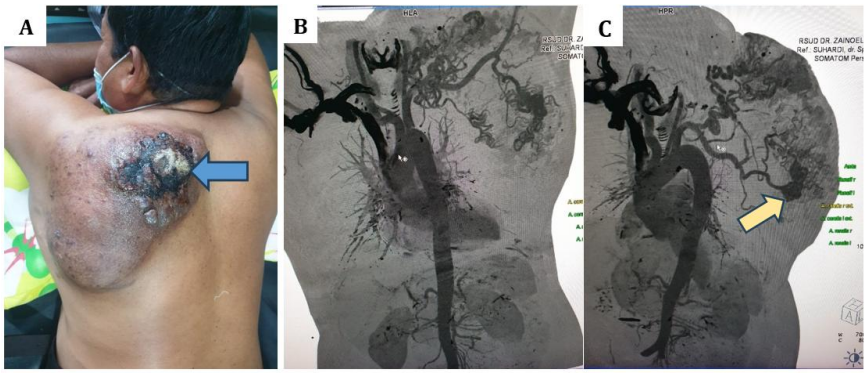

Figure 1. Clinical features of the patient. A) necrotic areas on the back region after arteriovenous malformations bleeding has stopped (blue arrow). B) and C) Computed tomography angiography of the back showing morphology of AVMs with multiple niduses (yellow arrow)

Upon understanding the morphology of the AVMs, their anatomical location, and endovascular accessibility, a gradual management plan was formulated, starting with embolization, followed by vascular and plastic surgery. In the first step, the prominent arterial supply was initially embolized using polyvinyl alcohol particles. Subsequently, coil embolization was employed to maximize the interventions undertaken to completely eliminate the AVMs. Several weeks after the embolization, he underwent surgical debridement followed by a split-thickness skin graft reconstruction of the back wound, using the left thigh as the donor site. He was followed up every three months to assess potential recurrence of AVM via ultrasonography examinations. One-year after this procedure, no recurrence or complaints were found (Figure 2).

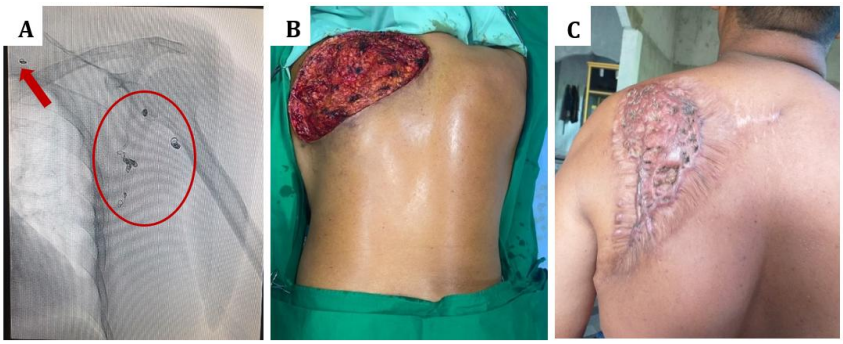

Figure 2. Treatment and follow-up of the patient. A) The position of the embolization coil is placed (Circle and red arrow), B) During debridement and the process for skin grafting, C) Clinical appearance of the back region after a year of skin grafting.

DISCUSSION

AVMs are congenital vascular diseases during embryonic development that can continue to grow postnatally. AVMs are fast-flow malformations characterized by abnormal connections between arteries and veins without interposing capillary vessels. These connections form a network of dysfunctional blood vessels called a nidus. Unlike typical blood vessels, AVMs lack a standard arterial structure, instead comprising multiple arterial vessels that supply the malformation with oxygenated blood. This abnormal configuration leads to arterial blood flowing directly into the venous system under high pressure.[13,14,15] The progression of AVMs is driven by the high-velocity blood flow from the arterial to the venous system, leading to significant anatomical, physiological, and hemodynamic changes. These changes can cause a range of clinical symptoms depending on the location and size of the AVMs.[16-19]

The initial and most critical step in managing AVMs is analysing their angioarchitecture. The diagnosis of AVMs is typically confirmed using Doppler ultrasonography or CT angiography, with trans-arterial angiography considered the most accurate method.[17,18] Angiography provides invaluable insights into the complex vascular structure of AVMs. A comprehensive angioarchitecture analysis of brain AVMs should encompass several key factors, including the composition of the nidus, the types and origins of feeding arteries, the patterns of blood supply, and the types and configurations of venous drainage.[20]

Our patient had peripheral AVMs that affected the back. Both Doppler ultrasonography and CT angiography were used to further delineate the morphology and anatomy of the feeding arteries. In this case, a large AVM located on the posterior surface is supplied by multiple arteries arising from branches of the subclavian and axillary arteries.[20] Subsequently, the information obtained from CT angiography was very valuable in planning a course of treating the AVMs.

Several factors increase the risk of an AVM rupturing, including a fistulous nidus structure, the presence of aneurysms within the nidus or feeding arteries, and venous irregularities such as stenosis or varicosis. When an AVM ruptures, the surrounding connective tissue can resist the expansion of bleeding, acting as the body's natural tamponade. Resuscitation should be delayed until bleeding control is achieved, providing the patient remains conscious. Immediate resuscitation during the rupture, while the patient is conscious, may exacerbate the situation.[22-23] Following an initial haemorrhage, mortality rates range from 10% to 30%, while morbidity rates can be as high as 25% to 60%. Moreover, there is a significant risk of rebleeding within the first year after the initial rupture, with reported rates of 6% to 17%. Due to this high risk of recurrent bleeding, developing a treatment plan is crucial.[20-21]

Due to their complex anatomy and morphology, treating AVMs can be difficult and often requires a team-based approach. The primary treatment options for AVMs include surgical resection and endovascular embolization. Surgery is the gold standard for completely removing the AVMs. The surgery may be sufficient for treating small, easily accessible AVMs. However, for larger, deeper, or more complex lesions with multiple feeding blood vessels, using embolization before surgery can improve treatment success rates while minimizing overall risks.[13-15,17,18]

While embolization used as a standalone treatment or in conjunction with other therapies, its success rates vary. When used alone, embolization has a cure rate of 5-28%, which is lower than surgical resection (80-95%) and radiosurgery (65-85%). Furthermore, standalone embolic therapy is associated with higher morbidity (4-9%) and mortality (2-4%) compared to other treatments. These complications are largely due to the complexity of AVMs, which can make embolization more challenging.[24,25]

Embolization also can be performed as a neoadjuvant therapy before surgical resection.[13-15] Performing embolization before surgery can enhance the safety of the subsequent surgical procedure. This staged approach may also mitigate the risk of normal perfusion pressure break-through syndrome, a potentially life-threatening complication. A study by Lee et al. revealed that patients with AVMs that could be surgically removed had the best outcomes when treated with a combination of preoperative embolization or sclerotherapy followed by surgery. Meanwhile, patients with AVMs that couldn't be surgically removed (only endovascular treatment) showing a positive results, satisfactory outcomes in 25 patients and fair to good outcomes in the remaining 7 patients.[26] Deruty et al. reported that preoperative embolization may improve outcomes, with a 100% cure rate observed in patients who underwent combined treatment.[27] Furthermore, when embolization is used as part of a multimodal treatment strategy, the associated morbidity and mortality rates tend to be lower, ranging from 4% to 6% and 0% to 2%, respectively.

The primary objective of various embolization techniques in treating peripheral AVMs is to cut off their blood supply. These techniques can be applied through the arterial or venous side of the AVMs, or by directly injecting the nidus. To minimize the risk of damaging surrounding tissues, embolization treatments are typically performed in multiple stages, with each session carefully planned to achieve optimal results while ensuring patient safety.[17,28]

The use of embolic agents to treat vascular malformations (VMs) has evolved significantly since the 1980s. Today, various endovascular therapies are available, offering a range of treatment options. These approaches can be broadly categorized into two groups: solid embolic tools and sclerosant liquid agents. Solid embolic tools, such as particles, plugs, coils, and detachable balloons, induce thrombosis directly, allowing for controlled placement and immediate haemostatic control. However, this approach may lead to the formation of new blood vessels due to the release of angiogenic factors from ischemic endothelial cells.[29,30] On the other hand, sclerosant liquid agents, including bleomycin, ethanol, polyvinyl alcohol, and sodium tetradecyl sulphate, destroy endothelial cells, addressing the issue at a cellular level. Nevertheless, liquid agents can cause severe complications, such as deep vein thrombosis, pulmonary embolization, and local injuries. Ultimately, the choice of treatment depends on the specific characteristics of the VM and the patient's overall health.[31] Polyvinyl alcohol has been chosen since the inception of endovascular treatment of AVMs due to its ability to cause devascularization through endothelial damage of the vessels, serum protein denaturation, and rapid thrombus formation. The most common complications of AVM treatment are skin necrosis and nerve injury. However, 77%-85% of skin necrosis heals independently, and 92% to 100% of injured nerves regain function.[32,33]

In a previous study, complete or near-complete embolization was achieved in 56.7% of cases. However, the success of embolization depends on various factors, including the size and shape of the AVMs, as well as the diameter and complexity of its feeding blood vessels. In this study, the degree of embolization was not a primary objective. Notably, the primary goal of treatment is not necessarily to achieve complete embolization but rather to address the vulnerable aspects of the AVMs that pose risks.[34]

The recurrence rate of AVMs ranges from 8% to 81%, depending on the size of the AVMs. Signs of recurring AVMs include skin discoloration, telangiectasia, swelling, pulsation, bleeding, or Suen's sign (rapid soft tissue rebound upon palpation).[13,16] Follow-up of the patient one year after embolization and surgery showed no recurrence of AVMs.

This present study has some limitations which need to be mentioned. A key limitation of this case report is the high cost of endovascular embolization, coupled with the rarity of ruptured giant arteriovenous malformations, which limits the generalizability of the findings. Patients who

undergo embolization and surgery should have long-term follow-up because AVMs can recur. The other limitation is the low rate of angiographic follow-up. Due to the very low rate of angiographic follow-up, it is inappropriate to compare the pre-and post-operative angiographic characteristics in this series of patients.

CONCLUSION

This case report underscores the potential severity of AVMs, a condition that often remains undetected due to its asymptomatic nature. However, when symptoms do manifest, they can pose a serious health risk due to the potential for vessel rupture. The successful management of AVMs, as demonstrated in this case, typically involves a combination of embolization procedures and surgical interventions. This case also highlights the importance of regular follow-up to assess potential recurrence of the AVMs. The findings from this case have significant implications for the diagnosis and treatment planning of AVMs.

CONFLICT OF INTEREST

We declare no competing interests in this study. The patient involved in this case report had given their informed consent for the publication of the study, respecting their privacy and confidentiality.

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DATA AVAILABILITY STATEMENT

Patient data cannot be published to the public, due to maintaining the confidentiality of patient personal data, to obtain complete patient data, you can write to us, the author, for further information.

SUPLEMENTARY MATERIAL

This manuscript does not contain any supplementary material.

AUTHORS CONTRIBUTIONS

AS: conceptualization, software application, formal analysis and investigation, writing-original draft preparation, finalization; FJ: methodology, validation, supervision

DECLARATION OF USING AI IN THE WRITING PROCESS

Non declare

LIST OF ABBREVIATIONS

AVM: arteriovenous malformations; CT: computed tomography. VM: venous malformations

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