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CASE REPORTS

Retroauricular Extracranial Avm – Case Management And Literature Review

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Abstract

Introduction: Extracranial arteriovenous malformations (AVMs) are rare but challenging vascular anomalies characterized by direct arteriovenous connections that bypass the capillary network, leading to progressive lesion growth, hemorrhage, and functional impairment. The management of these lesions remains complex due to high recurrence rates and the need for multidisciplinary therapeutic approaches.

Case Presentation: We report the case of a 51-year-old female presenting with a left retroauricular pulsatile mass, headaches, and audible bruits. Magnetic resonance imaging (MRI) revealed an extracranial AVM with a 17 × 30 mm nidus. Surgical resection was performed following preoperative evaluation, involving meticulous dissection, selective vascular control, and complete excision of the malformation. The patient had a favorable postoperative course, with no neurological deficits and early hospital discharge.

Discussion: Extracranial AVMs pose significant diagnostic and therapeutic challenges. While embolization serves as an adjunctive measure to reduce intraoperative bleeding, complete surgical resection remains the definitive treatment. However, lesion recurrence is common due to residual microscopic arteriovenous shunts and neovascularization. Advances in molecular pathology have identified key angiogenic pathways involved in AVM progression, opening new avenues for targeted therapies.

Conclusion: This case highlights the importance of early diagnosis, comprehensive imaging assessment, and individualized surgical planning for optimal patient outcomes. Multimodal strategies integrating endovascular techniques and surgical resection remain crucial in managing extracranial AVMs. Long-term follow-up is essential to detect potential recurrence and guide future therapeutic decisions.

Key words: extracranial arteriovenous malformation, vascular malformations, surgical management

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INTRODUCTION

Arteriovenous malformations (AVMs) are high-flow vascular anomalies characterized by abnormal direct connections between arteries and veins, bypassing the capillary network. These malformations disrupt normal hemodynamics, leading to progressive lesion expansion, tissue ischemia, and increased risk of hemorrhage, ulceration, and functional impairment[1]. While AVMs can occur in any vascular territory, extracranial AVMs – particularly those affecting the head, neck, and extremities – represent a rare but challenging subset of vascular anomalies with a high risk of recurrence and significant morbidity[2,3].

AVMs account for less than 3% of all vascular anomalies and are typically congenital, arising from abnormal embryologic vascular development during the 4th-6th weeks of gestation[4]. These lesions may remain asymptomatic for years, often progressing during puberty, pregnancy, or after trauma due to hormonal and mechanical triggers[5]. The estimated prevalence in hospital-based autopsy studies ranges from 5 to 613 per 100,000 individuals, with extracranial AVMs comprising only 4.7% of vascular anomalies seen in specialized centers[6].

While the exact pathogenesis remains unclear, genetic studies have identified somatic mutations in key angiogenic pathways, including:

- RAS/MAPK pathway (KRAS, BRAF, MAP2K1 mutations) – contributing to proliferative vascular remodeling[7].
- TGF-β/BMP signaling dysregulation – leading to endothelial instability and aberrant vessel formation[8].
- HHT (Hereditary Hemorrhagic Telangiectasia) mutations (ENG, ALK1, SMAD4) – seen in familial AVMs[9].

These discoveries suggest potential targets for future molecular therapies, offering promising avenues for pharmacological interventions in AVM management.

Extracranial AVMs exhibit a wide spectrum of clinical manifestations, depending on their size, location, and hemodynamic characteristics. Common signs and symptoms include: localized swelling and pulsatile masses, audible bruits and thrills, cutaneous discoloration (pink-blush or erythema), pain, ulceration, or spontaneous hemorrhage, progressive tissue destruction and functional impairment[10].

The Schobinger classification is commonly used to stage AVMs:

- Stage I (Quiescent phase) – Warm skin, cutaneous blush.
- Stage II (Expanding phase) – Audible bruits, pulsations, and growth.
- Stage III (Destructive phase) – Ulceration, hemorrhage, necrosis.
- Stage IV (Decompensated phase) – High-output cardiac failure[11].

Diagnosis relies on a combination of clinical evaluation and imaging. Color Doppler ultrasound provides an initial assessment, but MRI and MRA (Magnetic Resonance Angiography) are the gold standard for evaluating nidus size, flow characteristics, and soft tissue involvement. Digital Subtraction Angiography (DSA) remains essential for detailed vascular mapping and planning endovascular interventions[12].

Despite advances in interventional radiology and microsurgery, extracranial AVMs remain difficult to treat, primarily due to high recurrence rates (up to 80%) - Incomplete removal or embolization leads to rapid revascularization and aggressive regrowth[13], complex vascular architecture – AVMs contain a nidus of multiple arterial feeders and early venous drainage, making total obliteration difficult, and the risk of functional and aesthetic complications – Surgical resection may require extensive tissue excision, leading to disfigurement or functional loss[14].

Current treatment strategies include:

Embolization – Often performed preoperatively or as palliative therapy, using agents like ethanol, Onyx, and cyanoacrylate glue[15].

Surgical resection – The only definitive treatment, but associated with high morbidity if nidus removal is incomplete.

Multimodal therapy – Combining embolization and surgery has shown improved outcomes by reducing intraoperative bleeding and recurrence rates[16].

This case report presents a rare extracranial AVM, emphasizing its clinical evolution, diagnostic challenges, and therapeutic outcomes. By integrating recent advancements in vascular imaging, endovascular therapy, and molecular research, we aim to highlight innovative strategies for optimizing patient management and long-term prognosis.

CASE REPORT

A 51-year-old female, without a significant personal history, was admitted to our clinic for headache and

audible bruits and thrills on the left ear with an onset of 2 years. Clinical examination revealed a swelling and pulsatile mass posterior to the left ear. Cerebral MRI revealed a pseudotumoral formation, which was subsequently confirmed as a left retroauricular extracranial AVM with a 17 × 30 mm nidus.

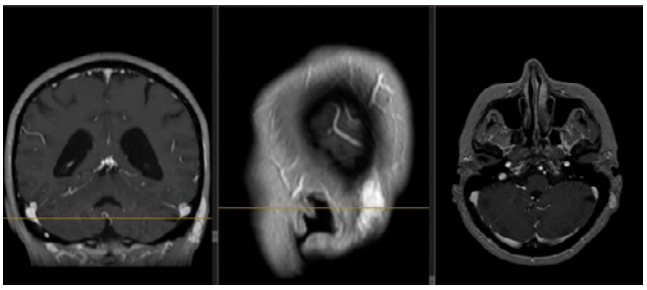

Figure 1. Contrast-Enhanced Head MRI Scan – left retroauricular pseudotumoral formation

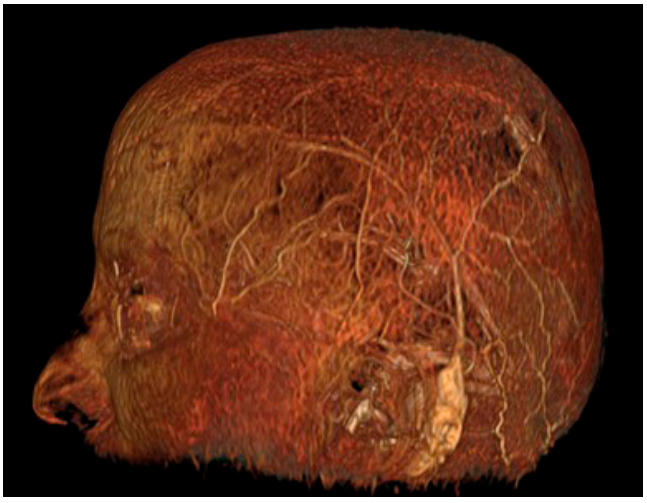

Figure 2. 3D reconstruction emphasizing the drainage system of the left retroauricular extracranial AVM

After discussing the treatment options and associated risks with the patient and her family, surgical intervention was performed according to the hospital's protocol for AVMs.

A left retroauricular arcuate incision is performed (Figure 3). The subcutaneous tissue is dissected (Figure 4), and the inferior pole of the malformation is identified. The vascular connections between the malformation and the adjacent tissues are carefully isolated (Figure 5). The primary arterial feeder, located caudally, is identified (Figure 6), dissected, and secured using a 7-mm straight Yashargil clip (Figure 7), followed by coagulation (Figure 8). Dissection is continued in a cranial direction (Figure 9), leading to the identification and coagulation of the draining vein at the cranial pole (Figure 10). The malformation is circumferentially dissected (2133) and macroscopically resected in its entirety. Meticulous hemostasis is achieved. The surgical wound is closed in anatomical layers, followed by sterile dressing application.

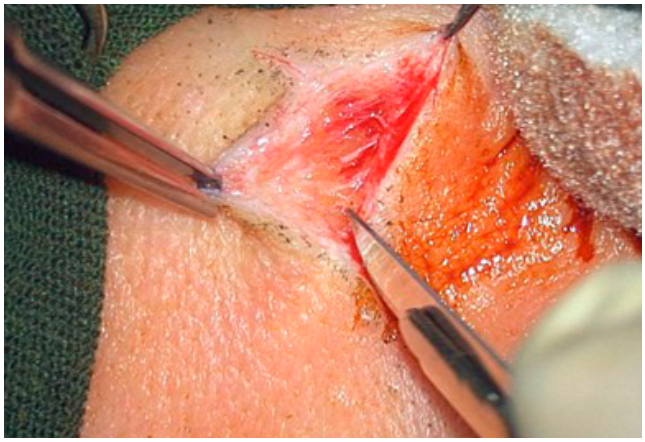

Figure 3. Left retroauricular arcuate incision

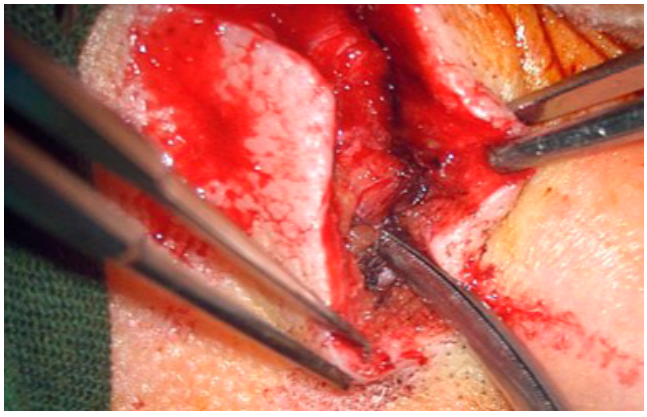

Figure 4. Dissection of subcutaneous tissue

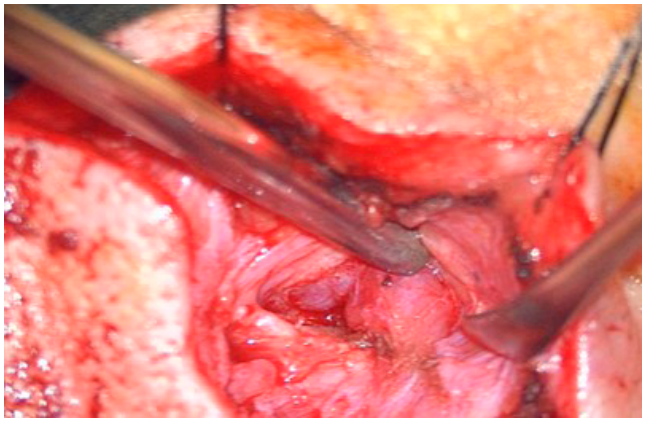

Figure 5. The vascular connections between the malformation and the adjacent tissues are carefully isolated

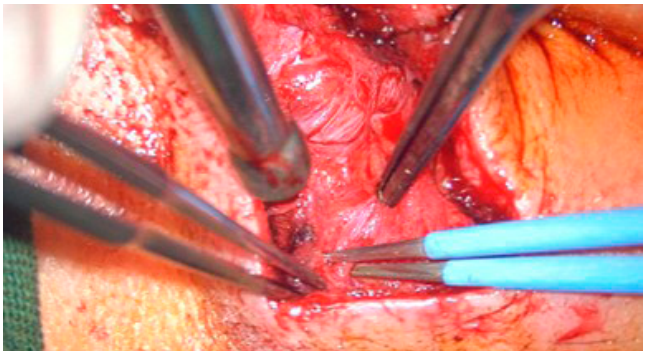

Figure 6. Primary arterial feeder, located caudally

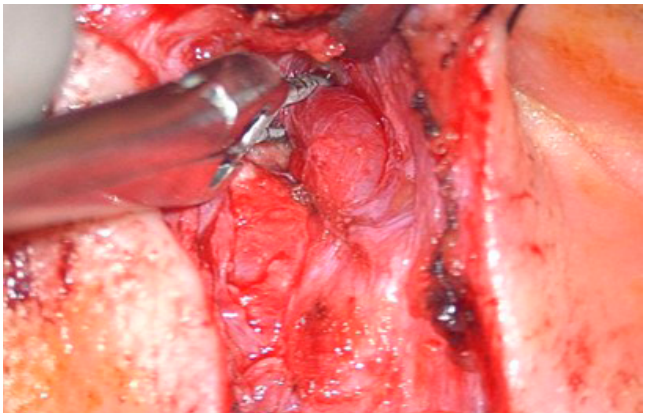

Figure 7. Temporary clipping of the main feeder

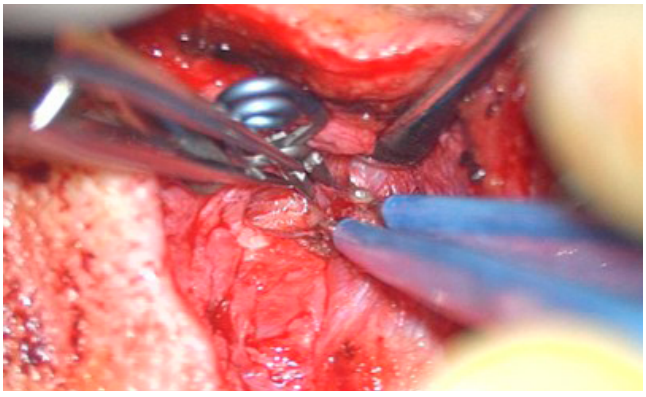

Figure 8. Coagulation of the main feeder

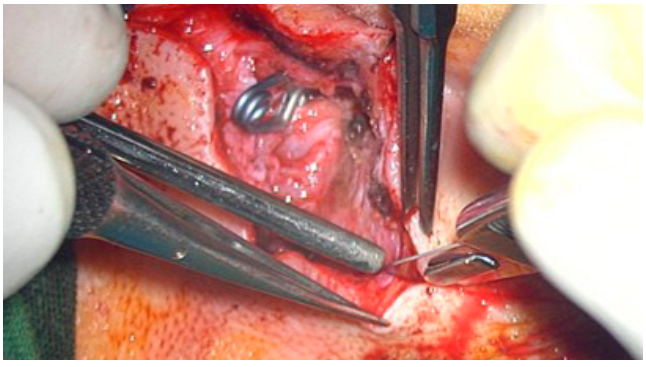

Figure 9. Dissection is continued in a cranial direction

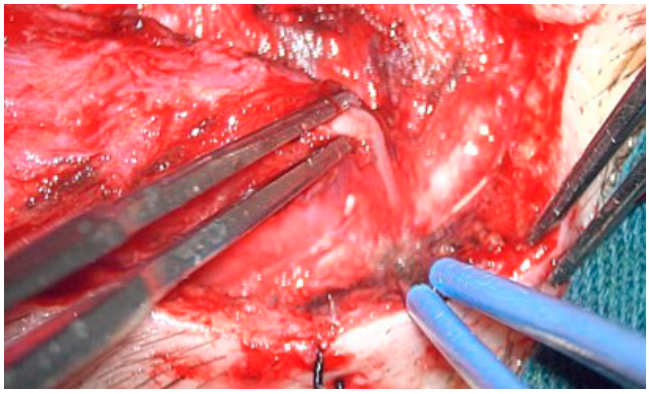

Figure 10. Coagulation of the cranial vein

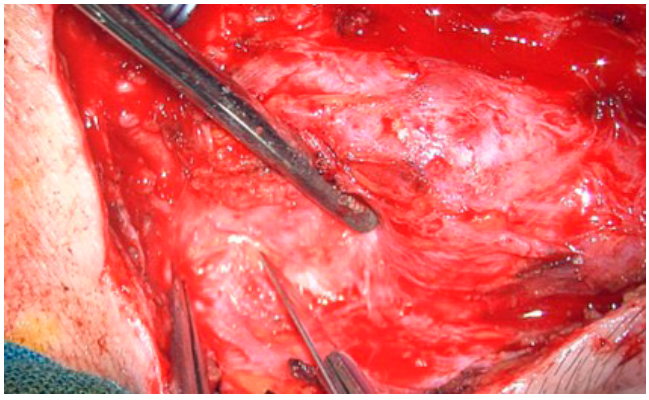

Figure 11. Circumferential dissection of the AVM

The procedure was carried out without complications. The patient's evolution was favorable, and she was transferred to the neurosurgery ward. She was discharged the next day in good condition, without neurological deficits.

DISCUSSION

Extracranial arteriovenous malformations (AVMs) pose a significant challenge due to their aggressive nature and complex management. These high-flow vascular lesions are characterized by direct arteriovenous connections that bypass the capillary network, leading to progressive enlargement, hemorrhage, and functional impairment[13]. While AVMs are predominantly intracranial, extracranial AVMs affecting the head, neck, and extremities are increasingly recognized as sources of significant morbidity[14]. Early diagnosis of extracranial AVMs is challenging due to their variable clinical presentation. Patients may present with symptoms ranging from subtle cutaneous discoloration to pulsatile masses with audible bruits, localized hyperthermia, and

spontaneous hemorrhage[15]. Furthermore, AVMs can mimic other pathologies such as soft tissue tumors or peripheral nerve sheath tumors, leading to misdiagnosis and delayed treatment[16]. Given their potential for aggressive progression, prompt recognition and classification using systems such as Schobinger's staging are essential for guiding management strategies[12].

The management of extracranial AVMs remains complex and typically requires a multidisciplinary approach. Treatment options include embolization, surgical resection, or a combination of both. Endovascular embolization using agents such as ethanol, Onyx, or cyanoacrylate glue has emerged as a key strategy in controlling AVM growth and reducing blood flow within the nidus[11]. However, embolization alone is rarely curative, and recurrence rates remain high, particularly when complete nidus obliteration is not achieved[10]. Surgical resection offers the best chance of definitive treatment but is often limited by lesion size, location, and involvement of critical anatomical structures[9]. A staged approach combining embolization followed by surgical excision has demonstrated favorable outcomes by reducing intraoperative bleeding and facilitating complete lesion removal[8]. However, the risk of functional and aesthetic sequelae must be carefully weighed against the benefits of intervention.

Recurrence remains a major concern in AVM management, with rates reported as high as 80% following incomplete embolization or surgical resection[7]. The risk of recurrence is particularly high within the first five years post-treatment and can persist for up to a decade[6]. Inadequate treatment of the nidus has been shown to promote the formation of new arteriovenous shunts, exacerbating disease progression[5]. Long-term follow-up with serial Doppler ultrasound, MRI, or angiography is essential for monitoring disease stability and detecting early recurrence[4]. Advances in molecular research have highlighted potential therapeutic targets, such as TGF-β/BMP and RAS/MAPK signaling pathways, which may pave the way for novel pharmacological interventions[3].

Complex cases, such as AVMs associated with venous malformations, require individualized treatment strategies. In a recent case report, AVM excision led to stabilization of an associated venous malformation, suggesting an interdependent pathophysiological relationship[2]. These findings underscore the importance of a tailored approach, particularly in cases involving multiple vascular anomalies. While most of the cases in the literature are performed by multidisciplinary teams formed of EMT, general surgeons, or plastic surgeons, using embolization and other methods[17], in this particular case, a neurosurgical team used microsurgical techniques for complete removal of the malformation. Thus, recurrence risk is drastically reduced.

CONCLUSION

Extracranial arteriovenous malformations are complex vascular anomalies requiring early diagnosis, multidisciplinary management, and long-term surveillance. The high recurrence rates and potential complications associated with embolization and surgical excision emphasize the need for individualized treatment strategies. Although embolization has revolutionized AVM management, it remains a palliative rather than curative intervention in most cases. Surgical resection, when feasible, offers the best chance of long-term control, particularly when combined with preoperative embolization. However, achieving complete nidus obliteration remains a significant challenge. Future advancements in molecular therapy may offer new treatment modalities aimed at targeting dysregulated angiogenic pathways, such as TGF-β/BMP and RAS/MAPK signaling. As our understanding of AVM biology continues to evolve, the integration of novel pharmacological agents with conventional endovascular and surgical techniques may significantly improve outcomes for patients with these complex vascular lesions. Long-term follow-up remains crucial, given the potential for late recurrence, and further clinical trials are needed to assess the efficacy of emerging therapeutic options. A personalized, multidisciplinary approach remains the cornerstone of optimal AVM management.

Ethics Statement and Conflict of Interest Disclosures
Financial support and sponsorship: All authors have declared that no financial support was received from any organization for the submitted work.
Ethics Consideration: The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national laws. Written informed consent was provided by the patient participant in this study.
Conflict of interest: No known conflict of interest correlated with this publication.
Availability of data and materials: The data used and/ or

analyzed throughout this study are available from the corresponding authors upon reasonable request.

Competing interests: The authors declared that they have no competing interests.

The use of generative AI and AI-assisted technologies: The authors did not use in this article generative AI and AI-assisted technologies.

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