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CASE REPORT

Giant scalp arteriovenous malformation in a 29-year-old male: case report

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ABSTRACT

Arteriovenous malformation (AVM) is rare, and extracranial AVMs comprise only 8.1% of all AVMs. Scalp AVMs may present with headache, local pain, and tinnitus. They may also remain clinically silent until a bleeding episode ensues. We report the case of a 29-year old male with an extremly large scalp AVM in the right temporoparietal region. We initially wanted to do conventional angiography to map the nidus, identify the feeders, and plan for a preoperative embolization. Our patient needed prompt surgery to control an episode of profuse bleeding coming from the mass before a conventional angiography could be done. The surgery incurred a significant amount of blood loss, and a non-resectable mass was left after the procedure. We were able to successfully perform serial embolization of a part of the patient's residual AVM two years after the surgery.

Keywords. embolization, conventional angiography, computed tomographic angiogram, magnetic resonance angiogram

INTRODUCTION

Vascular formation within the brain develops through angiogenesis and vasculogenesis.[1] Arteriovenous malformation (AVM) results from a disruption of normal vascular differentiation and growth during fetal development.[1] The pathogenesis of AVM may be due to a persistent congenital vascular plexus, proliferative capillaropathy, or cellular and molecular differentiation.[1] In the general population, the prevalence of AVM is 15-18 per 100,000 adults.[1] The different treatment strategies for scalp AVMs include transarterial embolization, transvenous embolization, direct percutaneous embolization, electrothrombosis, and surgical excision or ligation.[2] Extracranial AVMs account for 8.1% of all AVM cases.[3] Scalp AVM is rare, and is about 20 times less common than intracranial AVMs.[4] We present the case of a 29-year-old male with a large scalp AVM, the clinical features of the disease, and our diagnostic and therapeutic approaches.

CLINICAL FEATURES

A 29-year-old male came to our hospital due to a scalp mass on the right temporo-parietal area. The mass was noted since birth. It had been gradually enlarging with no associated signs and symptoms until 5 years ago, when the mass had minimal bleeding, which was easily controlled by direct manual compression. A year after, the patient started complaining of on and off headache, exacerbated by exertion and usually relieved by taking paracetamol. The patient did not consult a physician for his condition until three years ago. He was admitted in 2013 so that thorough diagnostic imaging studies could be done. Past medical, social, and family histories were unremarkable.

When we first examined the patient, we noted a 15 x 10 x 3 cm bulging, soft, pinkish to erythematous mass on the right temporoparietal area. The lesion was scabbed, irregularly contoured, and nodular. Figure 1 shows a photo taken by the patient a few days prior to his first admission. The mass had a palpable thrill and an auscultable bruit. Vital signs were within normal limits. There were no other systemic abnormalities noted. At this point, we were thinking that the

IN ESSENCE

Scalp arteriovenous malformation (AVM) is a rare condition that requires imaging modalities for diagnosis and surgical planning.

We managed a patient with a giant scalp AVM on the right temporoparietal area. Because of profuse bleeding of the mass, we performed a surgical resection of a portion the lesion without preoperative conventional angiography or embolization. The patient had massive blood loss during the procedure.

To manage the progression of the postoperative residual mass, we mapped the lesion through conventional angiography and subsequently performed serial embolization.

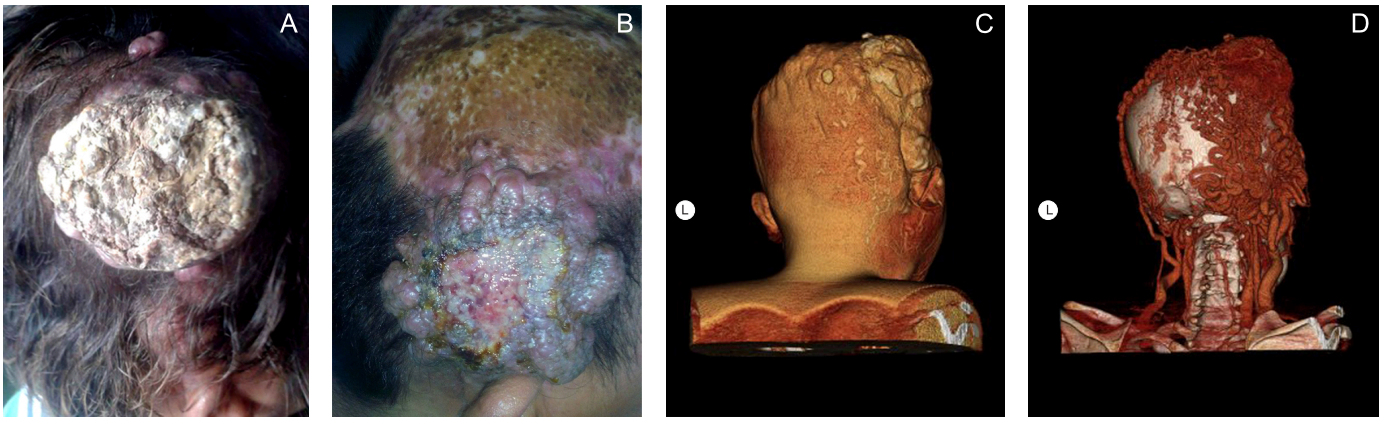

Figure 1 Photos of the right temporo-parietal scalp lesion taken by the patient in 2013, prior to surgical excision (A) and in 2015, two years post-surgery and prior to embolization (B), showing a large, bulging, nodular, pinkish to erythematosus mass. The lesion had varying external appearance. In the preoperative photo, the lesion had a scabbed, coarse and dry surface (A). In the postoperative photo, the residual mass had more irregular contours with areas of necrosis, ulcerations, and minimal bleeding (B). Computed tomography 3D reconstruction from images taken in 2013 demonstrates the location of the external lesion (C) and the extent of the arteriovenous malformation (D). L — left.

patient could have either a lymphatic malformation, a venous malformation, or an arteriovenous malformation.

DIAGNOSTIC AND THERAPEUTIC APPROACHES

A diagnosis of AVM was considered based on history and physical examination. We did a cranial magnetic resonance imaging (MRI) to confirm our diagnosis. The results showed multiple tortuous and serpiginous flow voids in the scalp, predominantly in the right temporo-parietal region (Figure 2). Computed tomographic angiogram (CTA) (Figure 3A) and magnetic resonance angiogram (MRA) (Figure 3B) were also done, which revealed tangled clusters of tortuous, serpiginous, irregularly dilated vessels surrounding the cranial vault, as well as in the scalp, predominantly on the right side, with feeding vessels commencing at the right external carotid artery 5 cm above the common carotid artery. Draining vessels appear in the ipsilateral external jugular vein, with spongiform enhancement located in its communicating vessel. There is consequent thickening of the involved scalp tissue, with nodularity growing extrinsically. The smallest vessels are seen surrounding the scalp with no intracranial insinuation.

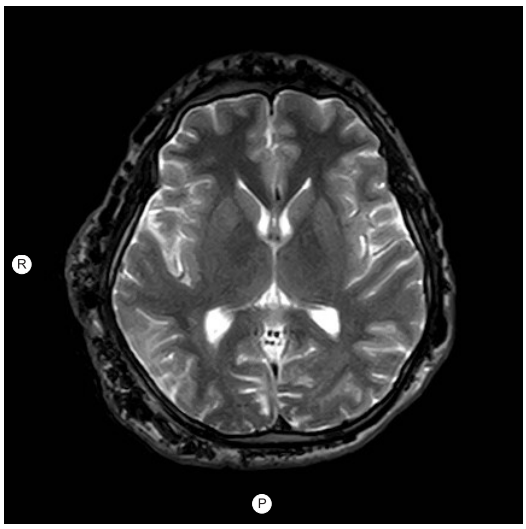

Figure 2 Magnetic resonance imaging on axial view, T2 sequence, showing thickened scalp with multiple curvilinear flow voids, predominantly in the right temporo-parietal region.

The Radiology and Surgery services co-managed the patient. Conventional angiography, the preferred imaging modality for AVM diagnosis, was requested by the managing teams to map the AVM nidus, which refers to the tangled blood vessels of the AVM, and the feeders, which are the arteries that supply the AVM nidus.[5] AVM mapping provides useful information for planning embolization and possible surgical excision of the AVM. During the course of the diagnostic procedures however, the patient accidentally scratched a friable portion of the mass, which subsequently bled profusely.

A radical surgical procedure was subsequently done on the patient to ligate major blood vessels on the right scalp. The patient lost approximately 4,500 mL of blood during the procedure which consequently required blood transfusion. A non-excisable residual lesion was left on the patient's right temporal region. A few days after the procedure, the patient was discharged improved and was given instructions to come back regularly for

Table 1 Timeline of the patient's signs and symptoms, with corresponding diagnostic and therapeutic approaches and clinical outcomes.*

Years	Signs and symptoms	Diagnostics	Therapeutic approach	Outcome
1986-2010	Gradually enlarging mass on the right temporo-parietal area	---	---	---
2011	Minimal bleeding of the mass	---	Manual compression	Control of bleeding
2012	Intermittent headache on exertion	---	Oral NSAIDs	Relief of headache
Early 2013	Admission for diagnostics	MRI/CTA/MRA: Scalp AVM with feeding vessels arising from three branches of the right external carotid artery	---	---
---	Profuse bleeding of the mass after a friable portion was accidentally scratched	---	Partial surgical excision	4,500mL blood loss; residual lesion at the right temporal area
Late 2015	Gradually enlarging residual scalp mass, intermittent headache and intermittent bleeding of the mass	Repeat CTA and MRA; Conventional angiography	Successful embolization of the right occipital artery; Successful embolization of the right posterior auricular artery four days after the first embolization; Attempt to embolize the right superficial temporal artery was unsuccessful	30mL blood loss; Discharged improved with instructions for regular follow up
Late 2016	Significant decrease in size of the AVM	---	---	Instructions for regular follow-up

*All signs and symptoms, diagnostics, therapeutics and outcomes pertain to the right temporo-parietal lesion.
AVM — arteriovenous malformation; CTA — computed tomographic angiography; MRA — magnetic resonance angiography; MRI — magnetic resonance imaging; NSAID — non-steroidal anti-inflammatory drugs.

monitoring of the lesion. The patient, however, did not immediately return for follow-up.

Two years after the surgical intervention, the patient came back to our institution due to headache and intermittent bleeding episodes in the area of the scalp lesion. The residual mass in the patient's right temporal region also gradually enlarged within the time that he did not seek any medical attention. Except for hypokalemia at 3.32 mmol/L, which resolved spontaneously, all laboratory findings for complete blood count, creatinine and electrolytes were within normal limits. Repeat CTA (Figure 3C) and MRA (Figure 3D) revealed that the mass now measured 7.3 x 4.3 x 1.6 cm. There was substantial reduction in the number of multiple tortuous dilated vessels surrounding the cranial vault, face and scalp predominantly on the right. The residual lesion in the temporal region had feeding vessels all arising from the right external carotid artery—superficial temporal, posterior auricular and occipital arteries. We did a conventional angiography (Figure 4), and floor mapping of the AVM showed clear demarcation of the feeding vessels. The plan of the Radiology and Surgery services at this point was to do serial endovascular emboliza-tion prior to another surgical intervention to reduce the size of the lesion and prevent recurrence of hemorrhage.

We were able to successfully do endovascular embolization of the right occipital artery. Four days after, we also did the embolization of the right auricular successfully. For each vessel, we performed cannulation first with a French 3 (1 mm diameter) Progreat® microcatheter, primed with dextrose 5% in lactated Ringer's solution, then subsequently embolized a piece of Histoacryl® tissue glue to the vessel lumen to achieve distal infarction. An accumulated 30ml blood loss was noted during the two procedures. We attempted embolization of the right superficial temporal artery, but it was unsuccessful due to technical difficulties in cannulating the vessel. The plan then was to monitor the size of the right superficial temporal artery and attempt another embolization when the artery lumen enlarges to a size that can accomodate a 1-mm-diameter microcatheter.

OUTCOMES

The patient came back for follow-up consultation five months post-embolization. On physical examination, there was significant decrease in size of the remaining scalp AVM, which now measured 6.6 x 1.8 x 4.9 cm. There was no recurrence of spontaneous bleeding and headaches post-embolization, and since the scalp mass regressed in size, we

decided to temporarily forego the repeat embolization of the superficial temporal artery and to continue monitoring the patient's condition.

DISCUSSION

AVM of the scalp is a rare condition that may involve the frontal, parietal and/or temporal regions. AVM is a consequence of abnormal hemodynamics causing progressive dilation of normal vessels in the region (i.e. supraorbital, carotid and occipital arteries). The vascular lesion, which is seen as a large pulsatile mass or a subcutaneous scalp lump, commonly presents as headache, local pain, and—for some—tinnitus.[6,7] Aside from these symptoms, hemorrhage, although uncommon, may develop in the event of large vascular malformations. Our patient presented with a gradually enlarging mass associated with intermittent headache and bleeding from the lesion.

Differential diagnoses for scalp AVM, like the one found in our patient, include congenital hemangioma (CH) and vascular malformations (VM) such as venous malformation, lymphatic malformation and arteriovenous malformation. We used the modified International Society for the Study of Vascular Anomalies (ISSVA) guideline to differentiate CH and VM according to age of occurrence, sex predisposition, course of the lesion, auscultation, and palpation.[3] Both CH and VM are present at birth with no sex predisposition. In CH, the growth of the lesion is complete at birth or the lesion grows in proportion to the child's growth. Involution may occur rapidly within 6-12 months of life or not at all. In VM, the course of lesion growth may also be proportional to the child's growth, but the lesion does not usually involute. Thrills, bruits, or pulsations are not appreciated in CH, but are common in VM. Our patient's lesion did not involute, the enlargement had been proportional to his growth, and we were able to appreciate thrill on palpation and bruit on auscultation of the mass, so we were inclined to diagnose the lesion as a vascular malformation, particularly an arteriovenous malformation.

The choice of imaging modality for scalp AVM affects the quality of the diagnosis and the therapeutic management. Conventional angiography is the preferred modality for understanding the angioarchitecture of AVM lesions and for ruling out any intracranial component.[8] The procedure, however, remains underutilized because it is costly, time-consuming, and invasive. It also requires operator experience, and is associated with a 1.5-2% morbidity and mortality risk.[9]

CTA and MRA are excellent non-invasive and economical alternatives to visualize AVMs.[10] Both modalities can be carried out for the differential diagnosis of vascular lesions such as cavernous hemangioma, venous malformations, sinus pericranii, and aneurysms. Advantages of CTA include high image resolution, retrospective creation of thinner sections from source data, improved 3D rendering with minimal artefacts, and shorter procedure time. CTA can also demonstrate related bony structures.[9] MRA,

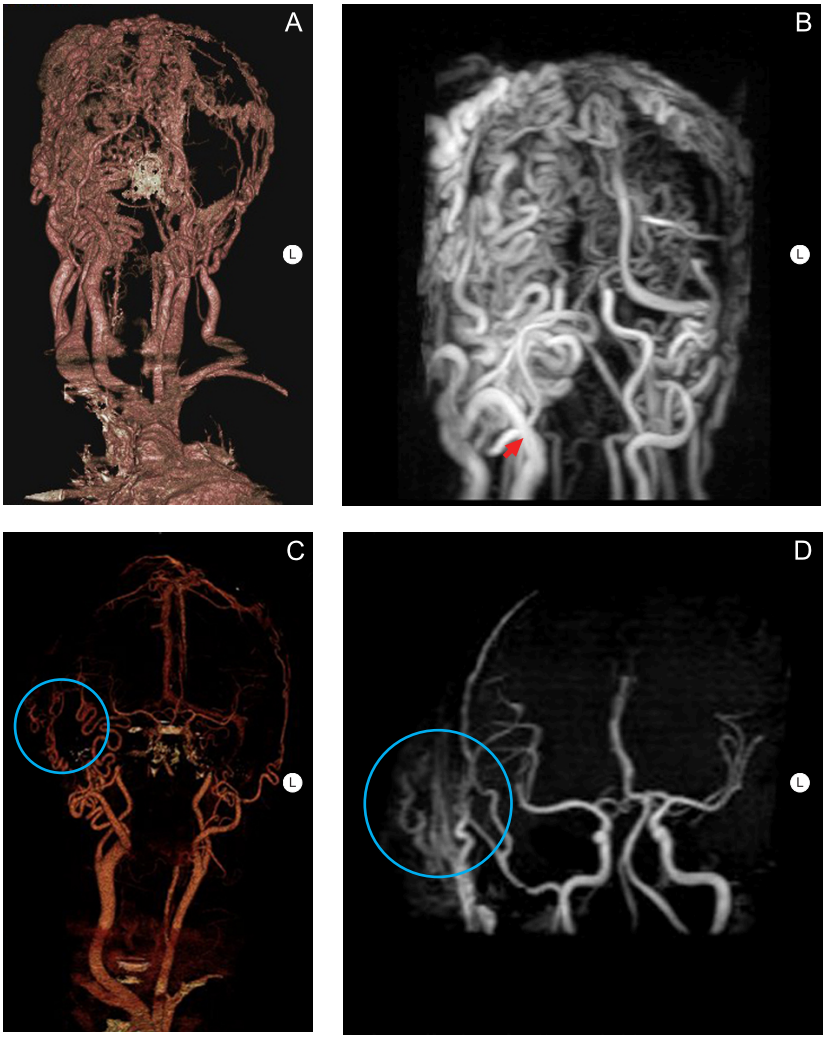

Figure 3 (2013) Initial computed tomographic angiography (CTA) (A) and magnetic resonance angiography (MRA) (B) done prior to surgery, showing extensive dilated and tortuous arteriovenous structures in the right temporo-parietal area. The major feeding vessels arise from the right external carotid artery (B: red arrow). (2015) Repeat CTA (C) and MRA (D) done two years postoperatively, showing significantly lesser arteriovenous structures. A group of tortuous dilated vessels representing the residual mass remains in the right temporal area (C and D: blue rings).

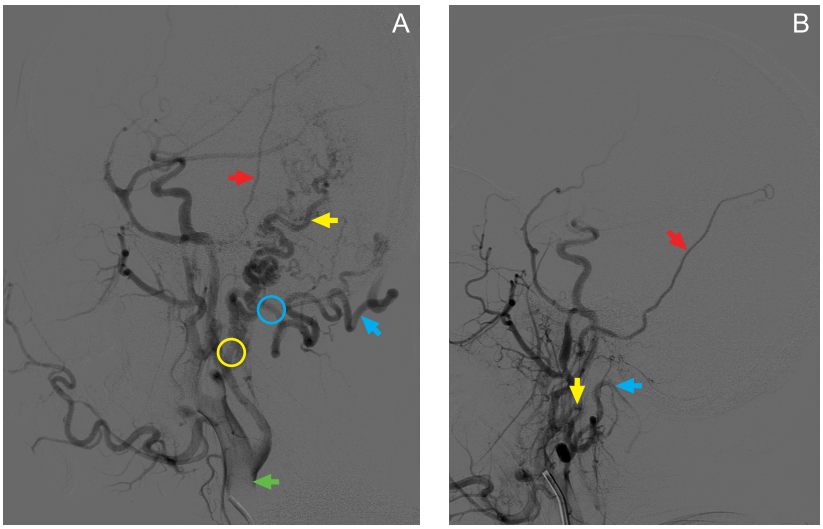

Figure 4 Pre-embolization conventional angiographic image (A), showing the branches of the right external carotid artery (A: green arrow)—the superficial temporal (A: red arrow), posterior auricular (A: yellow arrow) and occipital (A: blue arrow) arteries—supplying the lesion. The points in the posterior auricular artery (A: yellow ring) and the occipital artery (A: blue ring) where Histoacryl® tissue glue was introduced during embolization are also shown. Post-embolization conventional angiographic image (B) demonstrates absence of blood in the areas originally supplied by the right occipital (B: blue arrow) and right posterior auricular (B: yellow arrow) arteries, and consequent dilatation of the right superficial temporal artery (B: red arrow).

on the other hand, can differentiate scalp AVMs from other malformations.[7]

Our patient's CTA and MRA revealed multiple tortuous and intensely enhancing vessels on the right side of the scalp, which is indicative of scalp arteriovenous malformation. We were able to identify the vessels that feed the malformation as three arteries that arise from the right external carotid artery—the superficial temporal, occipital and posterior auricular arteries. There was no evidence of communication of the malformation with the intracranial circulation.

The management approach to scalp AVMs remains a challenge because of the high shunt flow, complex vascular anatomy, and cosmetic changes associated with the lesion.[6 11 12] Cosmetic correction and bleeding prevention are the goals of therapy.[13-15] Ligation of feeding vessels, embolization, electrothrombosis, and introduction of sclerosant into the AVM nidus are the suggested approaches.[7 16] In general, preoperative embolization of the nidus and feeders of an AVM greatly helps in reducing the size of the lesion, decreases the risk of massive hemorrhage, and facilitates subsequent surgical treatment.[7 17] Surgical excision after successful embolization or ligation of the AVM nidus is an effective method in dealing with large scalp vascular lesions.[7] Although embolization followed by surgery has been proven to be beneficial, cure rates are low unless the AVM is focal and located in a safe anatomic area.[18]

For our patient, we planned to initially do conventional angiography, which would guide us in planning for embolization and eventual surgical excision. Our patient underwent radical surgical excision before conventional angiography could be done because of persistent bleeding of the lesion. We were able to surgically control the bleeding and ligate some vessels, but a non-resectable portion of the lesion remained and even grew in size. The patient did not submit to regular monitoring postoperatively and only returned when headache and bleeding of the growing residual lesion occurred.

The actual recurrence rate of AVM is unknown, but recurrence is more common among children and rarely occurs among adults with AVM.[19] Several possible mechanisms of AVM recurrence have been proposed including persistence and proliferation of an initially occult portion of the AVM that was not removed during surgery, and *de novo* AVM formation.[20] One or a combination of these reasons may explain the growth of our patient's resected AVM two years post-surgery.

AVM lesions require preoperative conventional angiography to adequately visualize the lesion and plan for therapeutic interventions. Embolization of the nidus and major feeders of the lesion minimizes blood loss during surgical excision. We had to perform surgical ligation on our patient's scalp AVM before we could do conventional angiography and embolization. This resulted in significant intraoperative bleeding. To properly manage our patient's postoperative residual scalp mass, which later grew in size, we performed serial embolization two years after the surgery. We were able to successfully embolize two of the three identified feeders, and—as of this writing—we are looking to perform another embolization on the third feeder.

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Patient consent
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(http://www.care-statement.org/downloads/CAREchecklist-English.pdf)
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Competing interests
None declared

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