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Case Report

Arteriovenous malformation of the ear optimized with cinematic rendering images: A case presentation and review of literature ☆

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ABSTRACT

Arteriovenous malformations (AVMs) define rare aberrations in vascular morphogenesis. AVMs are typically present at birth, and unless they are stimulated to grow quickly by trauma, illness, or hormonal effects, they enlarge in proportion to an individual's growth. Clinical manifestations of AVMs are often linked to abnormal mass effects and blood perfusion. In this report, we describe a unique case of AVM of the left ear in a 24-year-old male, employing cinematic rendering along with a review of differential diagnosis and treatment options.

Introduction

Arteriovenous malformations (AVMs) are rare vascular lesions. They are known for their high propensity toward recurrence and bleeding combined with the potential for life-threatening rupture [1,2]. AVMs like all other categories of vascular malformations are considered to originate from developmental errors during embryogenesis (such as abnormal signaling processes that control maturation, growth of vascular cells, and apoptosis), leading to the persistence of vascular plexus cells with a certain degree of differentiation [3]. Unlike most vascular malformations, AVMs may remain quiescent until puberty and in rare cases into adulthood, and are usually triggered by trauma, infection, or hormonal factors [4,5]. Although many AVMs are asymptomatic, they may alternatively trigger severe pain and/or bleeding. The most common symptoms are pulsation (51.2%), bleeding (41.5%), and pain (29.3%) [6].

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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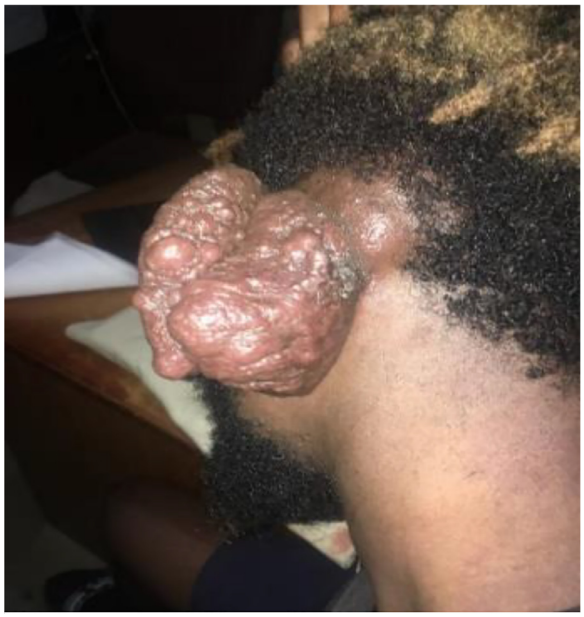

Fig. 1 – AVM of the left ear before treatment.

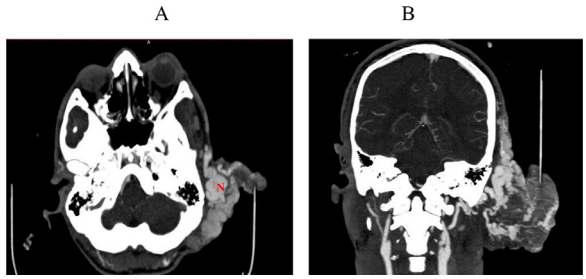

Fig. 2 – Maximum intensity projection (A) axial view showing nidus (N) of AVM of the left ear and (B) coronal view.

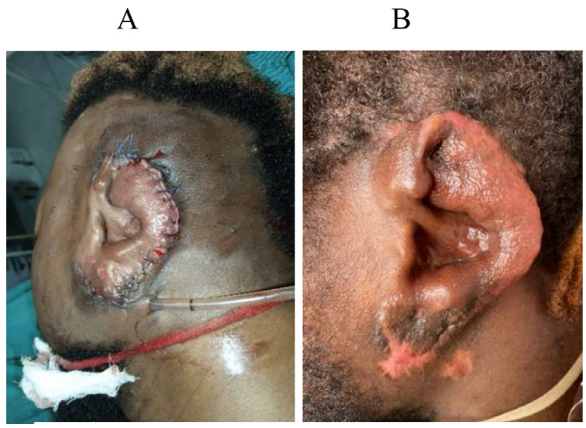

Fig. 3 – (A and B) Postsurgical findings.

AVM of the ear in contrast to AVM of other head and neck regions exhibits unique features. This is attributed to the characteristics of the ear anatomy in that a tinny skin envelope shelters a cartilaginous framework, with blood supply received from terminal branches of the occipital arteries, posterior auricular, and superficial temporal. As a result, inadequate management or progressive growth may cause cartilage exposure, infection, bleeding, and ultimately loss of the ear structure [7].

In principle, AVMs can effectively be managed by total resection, usually achieved with surgery combined with embolization or surgery alone. In the case of the ear, the treatment regimen involves embolization followed by surgical resection, aimed at preserving the cartilaginous framework or in extreme cases total amputation of the ear [8–11]. In this case report, we present another rare occurrence of AVM of the left ear in a 24-year-old male, along with a discussion on the diagnosis and treatment options, intending to provide information toward the management of this rare condition.

Case report

A 24-year-old male presented with a left postauricular mass with progressive enlargement (Fig. 1). On physical examination, there was deformity of the left ear in comparison to the right with episodic bleeding from the mass and discoloration. Seventeen years prior (at age 7), the patient visited an outpatient clinic with a progressive enlargement of the left external ear and was initially diagnosed with hemangioma because of the rapid growth (accelerated endothelial proliferation) observed which was predicted to regress later years. Upon realization that the cellular division would not regress after subsequent years of review, and tend to grow with the patient, it was determined that surgical intervention will be required in adulthood.

The patient on presentation at age 24 complained of episodic hemorrhages from the lesion accompanied by pulsatile tinnitus, vertigo, and swelling. Liaising with an interventional radiologist, a computed tomography (CT) angiogram of the head was requested. The results showed a complex hypodense lesion in the left postauricular region extending to the left infra-auricular region and involving the left pinna measuring approximately 8.5 × 7.3 × 4.5 cm (Figs. 2A and B). The angiographic images showed a complex network of tangled vessels making up the mass. There was supply by the left external carotid artery which was enlarged as well as mild enlargement of the left common carotid artery. Drainage appeared to be into the left external jugular vein. There was no extension into the left external auditory meatus. Based on these findings, the history, and the clinical presentations, a diagnosis of AVM of the left ear was made. On the following day, the patient was prepped and a 7-hour procedure to explore and excise the lesion was done with the external ear remodeled and attached to the posterior ear skin in a phase 1 procedure (Fig. 3). Standard volume and cinematic rendering techniques were employed in visualizing realistic 3D images of the AVM to better illustrate the disease condition for easier understanding by referring clinicians (Figs. 4 and 5).

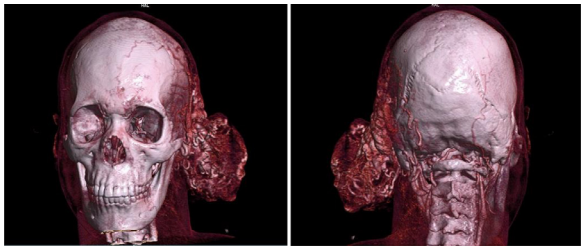

Fig. 4 – Standard volume rendering technique (VRT) showing the AVM of the left ear.

Discussion

AVMs constitute a class of vascular abnormalities that develop from the "nidus" (a recognizable source vessel), which is known to conduct an abnormal connection of venous and arterial systems, with characteristics of being slightly pulsatile and compressible [12]. Normally, this particular type of shunt is congenital but only become apparent in the first or second decade of life. Clinically, AVMs are accompanied by frequent episodes of bleeding, usually in the absence of pain, and can appear in bone or soft tissues [13]. Schobinger further classified AVM into 4 distinct stages along with correlating signs and symptoms. Stage I (quiescence) shows warm and discolored skin; stage II (expansion) is characterized by pulsation, swelling, and bruit; stage III (destruction) is characterized by bleeding, ulceration, and pain; and stage IV (decompensation) involves cardiac failure [14,15]. AVMs are again classified based on their flow rate to better explore management options. Slow-flow AVMs are produced by capillary, lymphatic, or venous lesions whereas fast-flowing AVMs are usually arteriovenous fistulas as in our case [16].

In the differential diagnosis of vascular malformations as in our case, it is necessary to be able to distinguish between these vascular malformations and hemangiomas, as they are all characterized by the proliferation of endothelial cells [17]. The underlying distinction between the two is based on histological assessment of increased cell turnover. Hemangioma is clinically marked by the rapid growth of endothelial cells during the postnatal period and slow regression over a 5- to 8-year period. The endothelial cells in vascular malformations however have a normal rate of cellular division which does not regress and tends to grow with the child [18,19]. Histologically, due to the increased blood flow, AVM may have arteriovenous shunts with reactive, hypertrophic, thick-walled veins and arteries, resulting in increased warmth, erythema, and edema of the affected tissues. This shunting may further cause ischemic injuries to part of the lesion and surrounding tissues leading to deformity, pain, bleeding, and ulceration as observed in our case [17]. With regard to ear AVM, complications such as conductive hearing loss and tinnitus may be observed. In this patient, episodic hemorrhage compounded by tinnitus, swelling, and vertigo accelerated the need for surgical intervention. This was in line with the observations of Kim et al. [20] in their study of a 60-year-old man and 52 patients with AVM.

Imaging has been known to play important role in the differential diagnosis of AVM. Magnetic resonance imaging (MRI) remains the modality of choice in the initial examination because it provides precise anatomical distribution and flow dynamics of the lesion. CT is ideal for bony AVMs whereas selective angiography is most useful for further investigation of AVMs, to identify the specific arterial supply [17]. In the present case, which involved superficial tissues of the head and neck (the left ear), physical examination and the clinical history were sufficient to determine the clinical diagnosis of AVM. However, for further investigation needed for effective surgical intervention, a CT angiogram was indicated. This was successful in identifying the complex network of tangled vessels making up the mass and the feeding and draining vessels. To further aid in diagnosis, cinematic rendering was employed to create 3D photorealistic representations of the AVM,

Fig. 5 – Cinematic rendering of the AVM with different presets, configurations, and views.

which helped in better understanding of the medical condition's tortuous vascular structure. Additionally, recommending surgeons were really satisfied with the helpful information that volume rendering representations offered.

In general, the treatment of an AVM is based on the concept of nidus obliteration, as this is thought to curtail the growth of the lesion by preventing the recruitment of new vessels from nearby regions. However, if the AVM is small and asymptomatic, treatment is unwarranted. For symptomatic AVM, as in the present case, embolization and sclerotherapy remain the first-line options to allow for safer intraoperative resection with less loss of blood [21]. In all likelihood, ligation of feeding vessels or arterial vessels should never be done as this leads to the formation of new collateral circulations causing further enlargement [22]. Superselective embolization alone is rarely successful due to the formation of new collaterals but remains the only option for surgically inaccessible AVM and may be indicated for palliative care [23]. In considering resection for an AVM, it is worthy of note that these lesions are rarely curable and that the focus should be on disease control. Subtotal resection normally leads to recurrence with lesions often larger than the primary lesion. The optimal approach is based on a comprehensive approach of superselective embolization and a total surgical excision. Selective angiography can delineate feeding vessels allowing for complete excision and greatly reducing the chances for recurrence. Regarding ear AVM as in the present case, embolization followed by surgical resection is recommended [10,24]. In the present case, surgical management formed the foundation of the treatment regimen to manage postembolization necrosis and block the blood flow at the level of the AVM nidus as shown in similar cases [10,24].

AVM is a rare occurrence and causes marked deformities. It is often a source of great concern not only for patients but also for treating physicians. A multidisciplinary approach and employing the right imaging modality for proper identification as presented in this case is key for optimum treatment and achieving better results.

Patient consent

Authors have obtained written informed consent from the patient who is being studied under this subject matter in terms of the use of identifiable images/data and journal publication.

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