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REVIEW ARTICLE

Arteriovenous Malformation Pinna: Review of Literature

Anuj Goel

Assistant Professor, Department of ENT, Saraswati Institute of Medical Sciences, Ghaziabad, Uttar Pradesh, India

Correspondence: Anuj Goel, Assistant Professor, Department of ENT, Saraswati Institute of Medical Sciences, Ghaziabad Uttar Pradesh, India, e-mail: dranujgoel@rediffmail.com

ABSTRACT

Arteriovenous malformations are rare in the head and neck region and generally arise from intracranial vessels. We present a rare case of arteriovenous malformation related to pinna. The role of color Doppler sonography in the diagnosis and management of such case is discussed along with review of literature.

Keywords: Arteriovenous malformation, Ear.

INTRODUCTION

Arteriovenous malformations (AVMs) are lesions related to defected vascular development. However, they are almost always present at birth, but manifest later in life and require treatment. There is no sex predilection. These must be differentiated from hemangiomas which are neoplastic and affects females more than males and may regress spontaneously. Histologically, hemangiomas show endothelial proliferation in contrast to AVM, which have none.

CASE REPORT

A 22-year-old female presented to the OPD of Department of ENT, Saraswathi Institute of Medical Sciences, Hapur with a history of swelling and skin discoloration in the right postauricular region (Fig. 1), with a marked increase in size over the last 4 years. There had been two episodes of bleeding which stopped upon application of pressure but with difficulty. The patient also complained of disturbing tinnitus of the continuous blowing type. There was no history of trauma or headaches. On examination, a pulsatile swelling with areas of discoloration was seen in postaural area. On palpation, the swelling was soft, noncompressible, spongy, nontender and pulsatile in nature. On auscultation, a continuous bruit was heard which was synchronous with pulse of patient. The ear canal and tympanic membrane were normal. Audiogram was normal. A clinical diagnosis of AVM pinna was made which was confirmed by color Doppler. It was planned to excise this AVM under general anesthesia with repairing of defect by using split skin graft. The histology was consistent with an AVM and the patient recovered uneventfully.

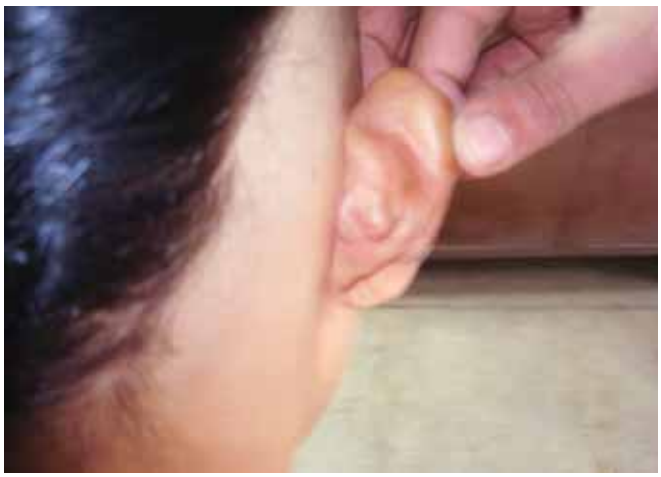

Fig. 1: AVM in right postauricular region

DISCUSSION

AVMs are rare in the auricular region but are common intracranially. They can be divided into two categories: slow flowing and fast flowing lesions. Fast flowing lesions are predominantly AV fistulas, whereas venous, capillary and lymphatic lesions produce slow blood flow malformations.[1] The AVM is composed of a central nidus with anomalous AV shunts and a network of surrounding collateral vessels.[2] The short circuit or shunting between the high pressure arterial and low pressure venous system accounts for much of the clinical presentation, anatomical changes and progression of the lesions. Schobinger described a clinical staging system for vascular malformations, that is, stage I (quiescence)—cutaneous blush/warmth; stage II (expension)—bruit, audible pulsation, expending lesions; stage III (decompensation) cardiac failure. AVMs are usually present at birth but commonly manifest in childhood or adolescence.[3] Their size can increase rapidly secondary to infection, trauma, ligation, attempted excision or via hormonal influences, such as during pregnancy and puberty.[4]

A history and physical examination provide information useful for diagnosing superficial AVMs of the head and neck. Plain radiography and computed tomographic scans have a limited role as diagnostic tools in high-flow vascular malformations; the diagnosis is usually made with Doppler ultrasonography. Magnetic resonance imaging (MRI) has become the investigation of choice due to its ability to depict the extent and lack of invasion of these lesions, provide multiplanar images and differentiate between high- and low-flow lesions.[5] Angiography shows marked hypertrophy and tortusity in the feeding vessels. Collateral vessels usually have a 'cork screw' appearance.[6,7]

If the AVM is small and asymptomatic, no treatment is required, especially in children. For a symptomatic AVM, complete excision with prior embolization is the treatment of choice specially in a large or recurrent AVMs.[8] Surgical ligation of proximal feeding vessels should be avoided, as it not only aggravates the lesion by establishing new collaterals but also precludes later embolization.[9] Proximal embolization also leads to failure for the same reasons.[10] Embolization alone can be used for palliation of the lesions located in difficult to approach areas or very close to vital structures.[9] In such cases, stroke, cranial nerve palsy and blindness- like complications can occur.[11] Total resection which requires a wide field resection of all the involved tissue is necessary to prevent recurrence; however, cosmetic and functional issues might limit the extent to which tissue can be removed. Partial excision usually leads to rapid recurrence; so in these cases, the remaining AVM tissue must be obliterated using intravascular embolization.[12]

Reconstruction during the same procedure, using a split thickness graft and pedicle or free flap, is necessary for good cosmetic results. Patients should be followed up at regular intervals with a clinical examination and noninvasive imaging like color Doppler. Magnetic resonance angiography should be reserved for those requiring additional embolization therapy. A minimum of 10 years follow-up is needed before a patient can be considered cured.[13]

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Editorial Inputs

Arteriovenous Malformation (AVM) of Pinna

Saurabh Varshney

Professor and Head, Department of ENT, Himalayan Institute of Medical Sciences, Dehradun, Uttrakhand, India, e-mail: drsaurabh68@gmail.com

AVM are rare in the auricular region but are common entity intracranially (Fig. 1). These must be differentiated from hemangiomas, which are neoplasm-affecting females more than males and may regress spontaneously. Histologically hemangiomas have endothelial proliferation in contrast to AVM where there is none. The role of MRI (magnetic resonance imaging) and color Doppler sonography (Fig. 2) in diagnosing and management of such cases is very important.

We at our institute have an experience of treating about 26 Arteriovenous malformations (AVM) of head and neck in last 8 years, out of which 10 (38.5%) were involving

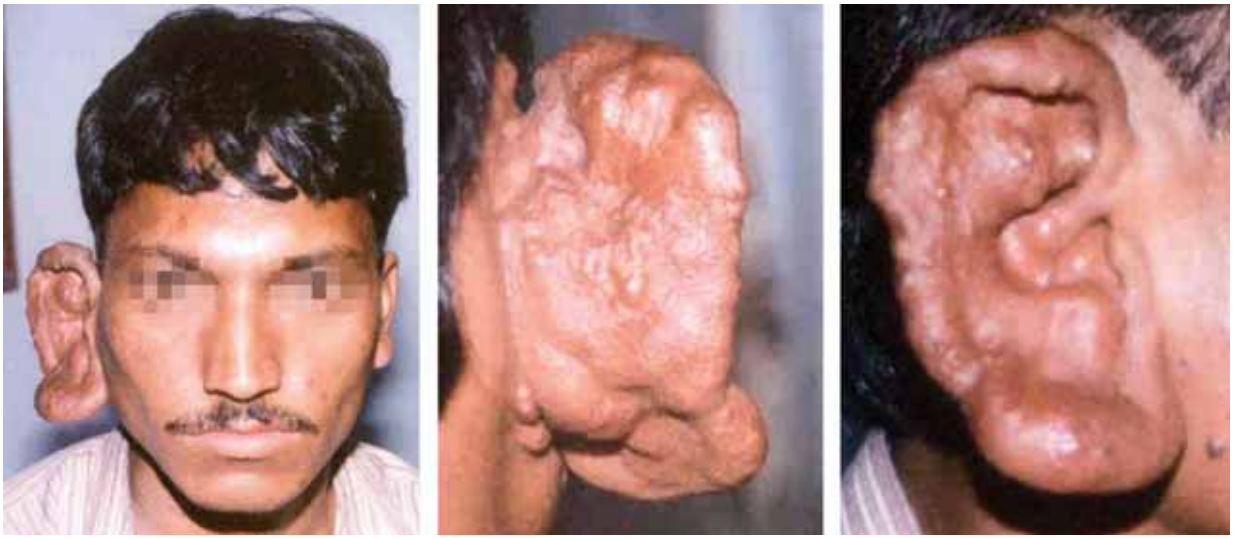

Fig. 1: AVM right pinna (high-flow lesion)

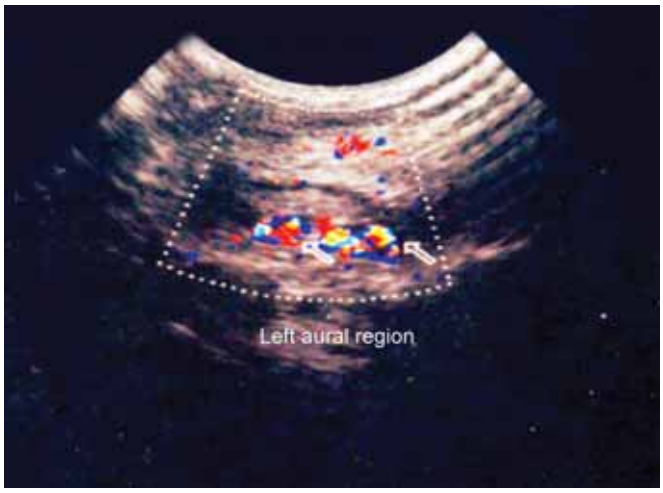

Fig. 2: Doppler USG showing multiple dilated anechoic areas in the swelling with multiple dilated tortuous vessels arising from posterior auricular artery

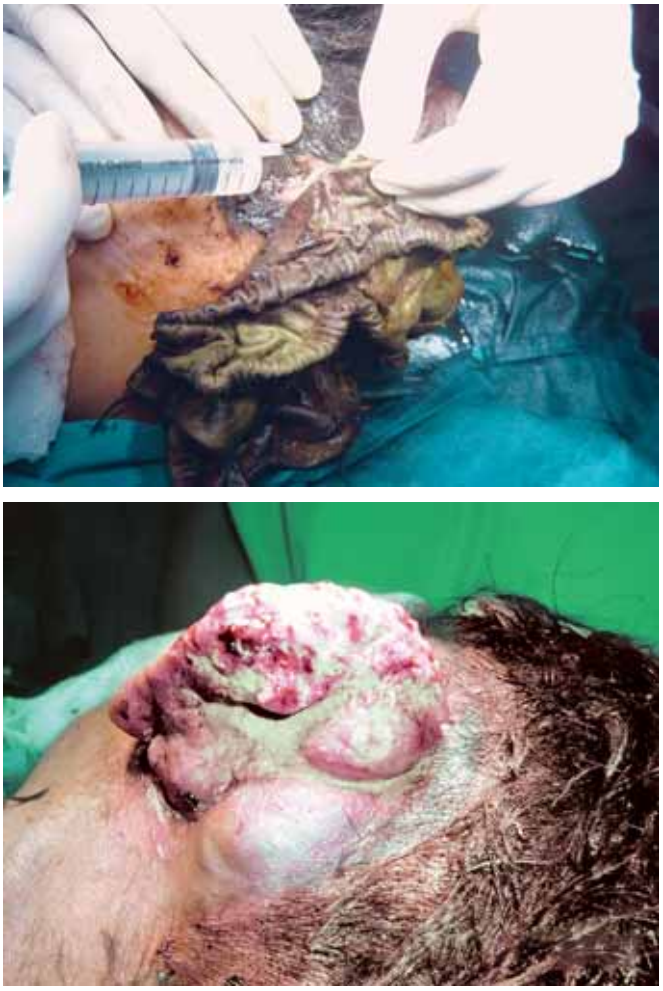

Fig. 3: AVM right pinna (Patient came to emergency, dressed with undergarment)

pinna. Out of 10 cases of AVM pinna 6 (60%) were females and 4 (40%) were males. Age of patients ranged from 16 to 31 years (mean age as 21.2 years). Most of the patients (6, 60%) had lesion since childhood and with increasing age lesion increased. Presenting symptoms were pain, ulceration, hemorrhage, cosmetic deformity, persistent warm sensation and deformity. Physical examination frequently revealed a soft, ill-defined pulsatile mass with overlying skin or mucosal discoloration and sometimes ulceration. Two patients presented to emergency with sudden profuse bleeding which could not be managed elsewhere. One female patient presenting to emergency had sudden profuse bleed which she tried to stop with undergarment (Fig. 3) and dressing over it. All the cases at our center underwent color Doppler and MR Angiography (Fig. 4) as radiological investigation. None of the case had intracranial extension. All patients underwent wide surgical excision under general anesthesia (Fig. 5) with reconstruction as split skin graft. Three patients with fast flowing lesion and ulcerated skin had embolization prior to surgical excision (Fig. 6). The follow up of patients operated ranged from 1 to 8 years (average 3.6 years) without recurrence (Fig. 7).

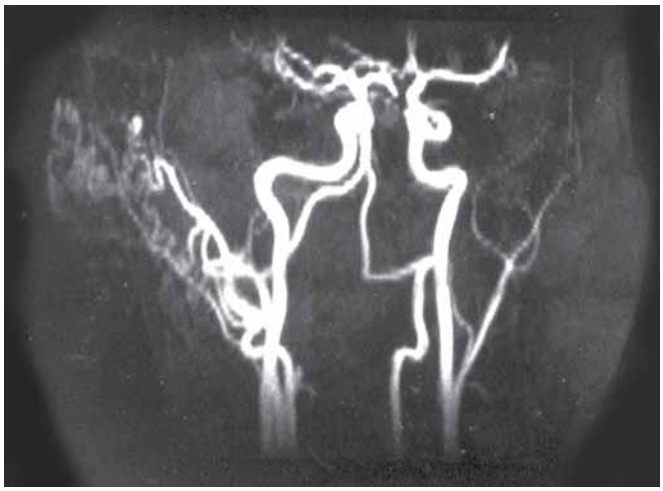

Fig. 4: MR angiography of a case of AVM right pinna, showing "nidus" and feeding vessel as postaural artery

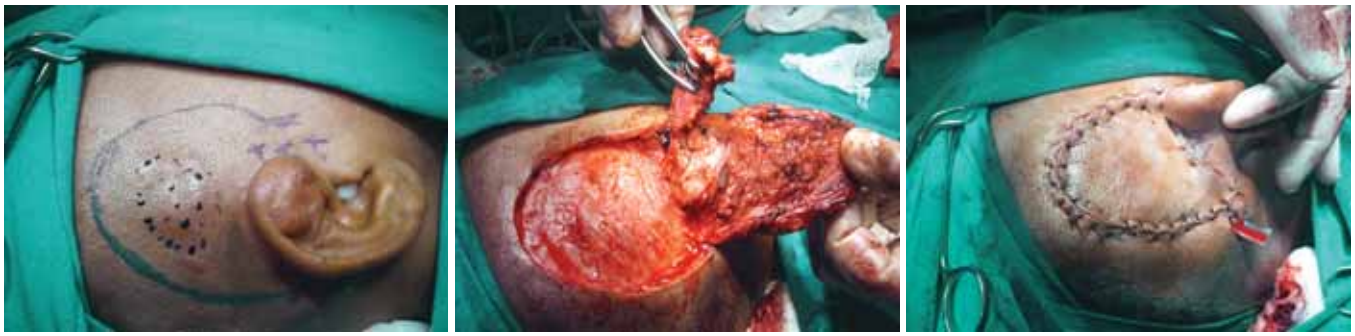

Fig. 5: AVM right pinna (pre, per and postoperative)

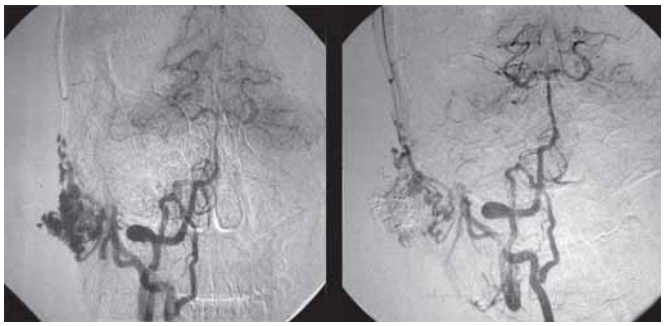

Fig. 6: AVM right pinna (pre- and postembolization)

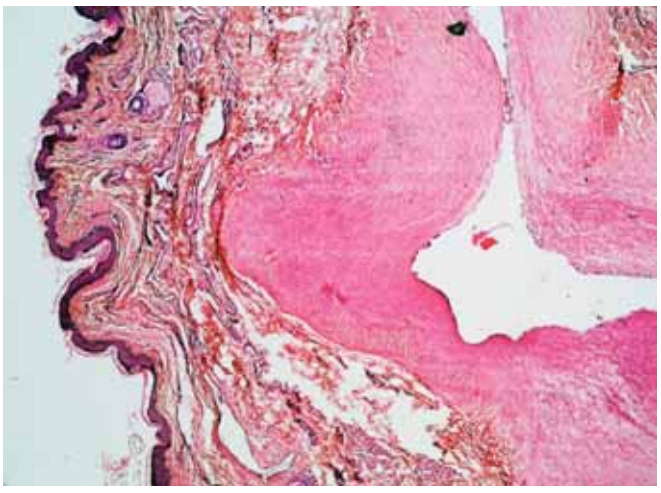

Fig. 7: Histopathology of AVM lesion