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CONSERVATIVE MANAGEMENT OF ARTERIOVENOUS MALFORMATION OF PINNA -A CASE REPORT

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ABSTRACT

Arteriovenous malformations (AVMs) are congenital malformations resulting from imbalance between pro-angiogenic and anti-angiogenic mechanisms. These are mainly located intracranially in the head and neck region and, are rarely seen extracranially. These extracranial AVMs are challenging to manage, considering the anatomical site involved and the psychological impact of cosmetic deformity. Surgical excision after super-selective embolization is advocated as the mainstay of treatment. Oral anti-angiogenic drugs have been found to have some role in the management of AVMs. Here, we present a case of Arteriovenous malformation of pinna managed conservatively.

KEYWORDS

Arteriovenous malformations; pro-angiogenic; anti-angiogenic; super-selective embolization; pinna)

INTRODUCTION:

Arteriovenous malformations are lesions originating from aberrations of vascular morphogenesis. An AVM is a direct communication between an artery and a vein without any capillary connections, and is mainly located in the intracranial region, lungs and kidneys.[1] An AVM outside the cranium is uncommon. However, the most common sites are cheek, ear, nose and forehead in the extra-cranium.[2] Around 10 cases of AVM of pinna have been reported in adults.

The behaviour of the peripheral (extracranial) AVM is locally aggressive. The initially quiescent lesion progresses to an expansive mass with cosmetic and functional disturbances. With later progression, the AVM destroys normal tissues and eventually leads to complications such as severe disfigurement, uncontrollable bleeding, ulceration, pain and cardiac volume overload.

Currently, the most commonly employed treatment for AVM is complete removal or ablation of the nidus, with recommended treatment being super-selective embolization followed by complete radical resection.

Here, we present a case of AVM pinna in a young male and our attempt at managing it conservatively.

CASE REPORT:

A 23yr old male patient presented to the ENT OPD with an irregular ear swelling for last one and a half year. The swelling was gradually increasing in size and spreading over the left pinna. There was associated history of recurrent episodes of profuse bleeding from the swelling which was controlled by applying pressure over the site of bleeding for some time.

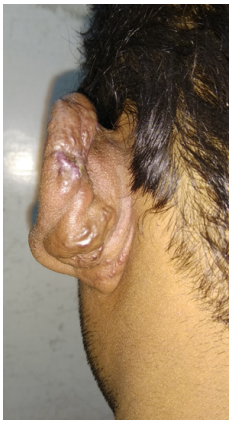

Figure1: AVM of left pinna of the patient

On examination, his left pinna was found enlarged, irregular in appearance, hyperpigmented, scab was seen at the site of last bleed. There was a visible pulsation of the pinna as well.

On palpation, local temperature was raised, the mass was non-tender, soft and compressible, and pulsation was felt.

The patient's routine blood profile was unremarkable for any abnormality.

On USG Doppler, large vessels were seen throughout the left ear lobe and high-velocity low-resistance arterial flow was noted.

CT-Angiography showed dilated vascular channels with multiple arterial feeders- left posterior auricular artery, left occipital artery, left superficial temporal artery, left maxillary artery. Venous drainage was by left internal jugular vein and anterior jugular vein.

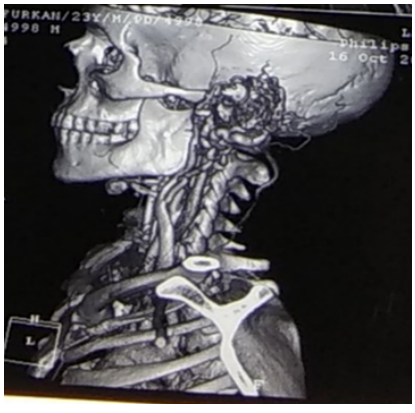

Figure 2: CT Angiogram showing the AVM and its arterial feeders

Finally, a diagnosis of Fast-flow Arterio-venous malformation of left pinna in Schobinger Stage III was made. The patient was explained about the need for surgical excision for definitive treatment. But the patient was apprehensive and refused to undergo any surgery.

So, the patient was started on 10mg Propranolol tablets twice daily and was kept on follow up.

There was only one episode of bleeding from the swelling in the initial few days of initiation of the treatment. No bleeding occurred for the next 7 months. However, there was no reduction in size of the swelling. Since, the patient was unwilling to undergo surgical excision, he was offered the alternative option to undergo embolization, which was lesser invasive but with guarded results.

He underwent super-selective embolization of the feeding vessels with NBCA(n-butyl-2-cyanoacrylate) and responded well after the procedure without any adverse effects. He has been on follow-up for the last 2 years. The size of the swelling has reduced and is asymptomatic, so far.

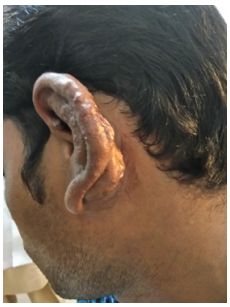

Figure 3: Post-embolization image of the pinna

DISCUSSION:

Arteriovenous malformations, generally, are congenital malformations consisting of a nidus of abnormal capillary beds shunting blood from the arterial system directly into the venous system resulting in high flow vascular abnormality.[3] Arteriovenous malformations of the head and neck (extra-cranial) are high-flow lesions and among the most serious of the vascular malformations because they are difficult to diagnosis, treat and cure. They grow throughout life with frequent aggressive growth spurts. AVMs can be life-threatening secondary to massive bleeding.[4]

The patho-biology of AVMs is related to angiogenesis resulting in the formation of new vessels due to the imbalance between pro-angiogenic and anti-angiogenic factors.[5] The whole process of development and progression of AVMs relies on the dilation of existing vessel as well as on the recruitment of new ones.

Endothelial growth factors such as vascular endothelial growth factor (VEGF) and inflammatory mediators such as tumour necrosis factor alpha (TNFα) are involved in the angiogenesis process.[6]

It has been recently discovered that ß3-adrenergic receptors are involved in favouring a pro-angiogenic molecular environment.[7] It has been demonstrated that the blockade of VEGF and other chemokines can prevent the progression of AVMs.[8]

There are six major classes of drugs that seem promising as therapeutic agents: Thalidomide, Rapamycin (Sirolimus) and derivatives, MMP Inhibitors (Doxycycline, Marimastat), Selective VEGF pathway inhibitors (Bevacizumab), ß3 adrenergic inhibitors (Propranolol) and Interferon.

Propranolol is a non-selective beta-adrenergic blocker used to treat cardiovascular abnormalities such as hypertension and arrhythmias. It has anti-proliferative effect too which is utilised in the treatment of infantile haemangiomas (IH). Mechanism of action of Propranolol include inhibition of angiogenesis by reducing VEGF, FGF and MMP expression on endothelial cells.[9]

Propranolol seems to have anti-angiogenic effects also by reduction in the production of proangiogenic proteins (i.e. ENG and ALK1). Another mechanism which may have a role is that beta3 receptor blockade induces the necrosis of endothelial cells.[10] This mechanism leads to a decrease in tumour weight and volume. These functional effects on the vasculature suggest the potential therapeutic use of Propranolol in other vascular anomalies like AVM. Moreover, Propranolol is cheaper than the other antiangiogenic drugs and its utilisation is characterised by fewer deleterious side effects (like hypotension, hypoglycaemia, bradycardia).[11]

Pre-operative super-selective embolization followed by surgical excision has been advocated as definitive treatment for AVM, but since the patient was not willing, he underwent only embolization with NBCA. Various other materials which can be used for embolization include ethanol, gelatin sponge, polyvinyl alcohol, platinum coil. Embolization is performed as a preoperative procedure before radical excision, in a palliative fashion in children, in surgically unfit patients or in patients not prepared psychologically to undergo radical amputation (as in this case).

CONCLUSION:

Extra-cranial AVMs, like in pinna, pose a unique challenge of management considering the difficulty in treatment and the psychological impact of cosmetic deformity involved. Propranolol may be used as a conservative modality of treatment in AVMs that cannot be radically excised, to prevent their further growth. Embolization appears to be good alternative in cases where radical surgery is precluded. Long term follow-up is necessary to evaluate the efficacy of such conservative approach.

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