Arteriovenous malformation related to the pinna

Arteriovenous malformations (AVMs) are lesions related to errors of vascular morphogenesis. These are almost always present at birth, manifest late in life and require treatment. There is no sex predilection. These must be differentiated from haemangiomas, which are neoplastic, affect females more than males, and may regress spontaneously. Histologically, haemangiomas show endothelial proliferation in contrast to AVM, which have none.

Case reports

Case 1

A 16-year-old male presented to the otorhinolaryngology out-patient department with a history of a swollen left pinna from the age of 5 years, which had increased further over the last 1.5 years. He also complained of occasional pain around the area but had no associated headache or tinnitus. On examination a soft, compressible, pulsatile swelling was seen on the helix of the left pinna. Colour Doppler sonography showed multiple dilated anechoic areas in the swelling, which was then excised completely using electrocautery. The superior surface of the pinna was degloved, leaving the underlying perichondrium and the cartilage intact. The defect was closed using a split thickness skin graft harvested from the left thigh.

Case 2

A 21-year-old female presented to the otorhinolaryngology out-patient department with a 15-year history of swelling and skin discolouration in the right postauricular region, with a marked increase in size over the last 2 years, following childbirth. There had been two episodes of bleeding which stopped upon application of pressure. The patient also complained of disturbing tinnitus of the continuous ‘blowing’ type. There was no history of trauma to the ear or headaches. On physical examination a pulsatile swelling with areas of skin discolouration and necrosis was seen in the postaural region. On palpation the swelling was soft, non-compressible, spongy, non-tender, and pulsatile with a thrill. A continuous bruit could be heard on auscultation. The ear canal and the tympanic membrane were normal. A clinical diagnosis of AVM was made and confirmed by colour Doppler sonography and magnetic resonance (MR) angiography (Fig 1). An axial angiogram demonstrated enlarged serpiginous structures showing flow-related enhancement in the right retroauricular and pinna region with feeders from the posterior auricular artery and auricular branches of the superficial temporal artery. It was excised using a postaural incision and the defect was repaired using a split skin graft (Fig 2). The histology was consistent with an AVM and the patient recovered uneventfully.

Discussion

Arteriovenous malformations are rare in the auricular region but are common intracranially. They can be divided into two categories: fast-flowing and slow-flowing lesions. Fast-flowing lesions are predominantly arteriovenous fistulas whereas venous, capillary, and lymphatic lesions produce slow blood flow malformations. The AVM is composed of a central nidus with anomalous arteriovenous shunts and a network of surrounding...
collateral vessels.\textsuperscript{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 (expansion)—bruit, audible pulsations, expanding lesions; stage III (destruction)—pain, ulceration, bleeding, infection; and stage IV (decompensation)—cardiac failure. Arteriovenous malformations are usually present at birth but commonly manifest in childhood or adolescence.\textsuperscript{3} Their size can increase rapidly secondary to infection, trauma, ligation, attempted excision or via hormonal influences such as during pregnancy and puberty.\textsuperscript{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.\textsuperscript{5} Angiography is useful in doubtful cases and for embolisation before surgery. It demonstrates the flow characteristics, feeding vessels, and dangerous anastomoses. Characteristic angiography findings are marked hypertrophy and tortuosity in the feeding vessels. The appearance of the nidus (centre) of the lesion varies from large tortuous vessels to innumerable small vessels appearing as an intense blush. Collateral vessels usually have a ‘cork screw’ appearance. When contrast is used, parenchymal staining is generally absent.\textsuperscript{6,7}

If the AVM is small and asymptomatic, no treatment is required, especially in children. For a symptomatic AVM, complete excision with prior embolisation is the treatment of choice.\textsuperscript{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 embolisation.\textsuperscript{9} Proximal embolisation also leads to failure for the same reasons.\textsuperscript{10} Embolisation alone can be used for palliation of lesions located in difficult-to-approach areas or very close to vital structures.\textsuperscript{9} In such cases, complications like stroke, cranial nerve palsy, and blindness can occur.\textsuperscript{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 embolisation.\textsuperscript{12}

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

We have described the above cases because of their rarity. We found only three cases of AVM from the posterior auricular artery in the English literature.10,14,15 In the second case, the increase in size of the AVM following childbirth proves it to be hormone-dependent.

Conclusion

Auricular AVMs are rare entities. We emphasise the role of colour Doppler sonography, MRI, and MR angiography in the management of such cases. Embolisation followed by a wide excision and repair during the same procedure is the treatment of choice. Patients should be given long-term follow-up to detect late recurrences.

References