An arteriovenous malformation of the external ear in the pediatric population: A case report and review of the literature

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The literature regarding arteriovenous malformations of the external ear is sparse. A case of a patient clinically diagnosed with an arteriovenous malformation of the external ear that was managed empirically with surgical excision, without recurrence, is presented. The pathogenesis, clinical presentation, radiological work up and management options regarding arteriovenous malformations are reviewed.

Key Words: *Arteriovenous malformation; External ear; Pediatric*

Une malformation artérioveineuse de l'oreille externe dans la population pédiatrique : Un rapport de cas et une analyse bibliographique

Les publications portant sur les malformations artérioveineuses de l'oreille externe sont rares. Les auteurs présentent le cas d'un patient ayant obtenu le diagnostic clinique d'une telle malformation prise en charge de manière empirique par excision chirurgicale, sans récurrence. Ils examinent également la pathogenèse, la présentation clinique, le bilan radiologique et les possibilités de prise en charge de ce type de malformation.

The literature regarding arteriovenous malformations (AVMs) of the external ear is sparse. Most of the treatment paradigms described in these papers include preoperative angiogram with or without embolization, followed by surgical excision. Here we present a case of a patient clinically diagnosed with an AVM of the external ear that was managed empirically with surgical excision, without recurrence. Our opinion is that these lesions can be excised without preoperative angiogram and/or embolization as long as they are small, well circumscribed and do not have clinical evidence of extensive collaterals.

CASE PRESENTATION

A 15-year-old male patient was referred for evaluation of an enlarging pulsatile mass of the left pinna. The patient noticed the mass two years before presentation. It was described as slow growing with intermittent pain. The patient denied bleeding or drainage from the mass. On examination, there was a 1.5 cm pulsatile, firm, but compressible mass of the inferior pinna, with an audible bruit. The mass extended from the helical rim inferiorly to the inferior sulcus of the ear (Figure 1). There was no evidence of superficial collaterals, ulcerations or active bleeding. Because of the high clinical suspicion of an AVM, the patient underwent excision of the lesion (Figure 2). Postoperatively, the patient was evaluated in the office at one, two and four weeks following resection without recurrence of a mass, pulsation or bruit. Final pathology revealed a benign AVM. To date there has been no clinical recurrence (Figure 3)

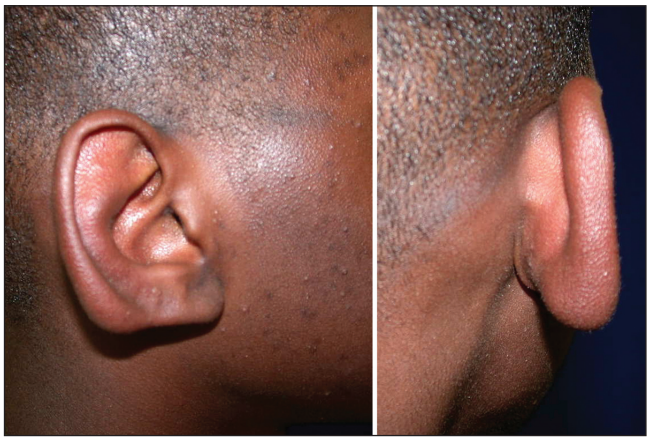

Figure 1) *Preoperative photos of the left ear mass*

DISCUSSION

The arterial origin of external ear vascular malformations can be from the posterior auricular, occipital, temporal and an occipital branch of the extracranial vertebral artery (2). Enlargement of the AVM may be triggered by trauma, infection or hormonal influences, such as pregnancy and puberty. Clinical manifestations of AVM are related to abnormal perfusion and mass effect.

The exact pathogenesis of an AVM has not been clearly defined. One theory states that these malformations arise during fetal development as a result of the failure of regression of arteriovenous channels in the primitive retiform plexus. It has

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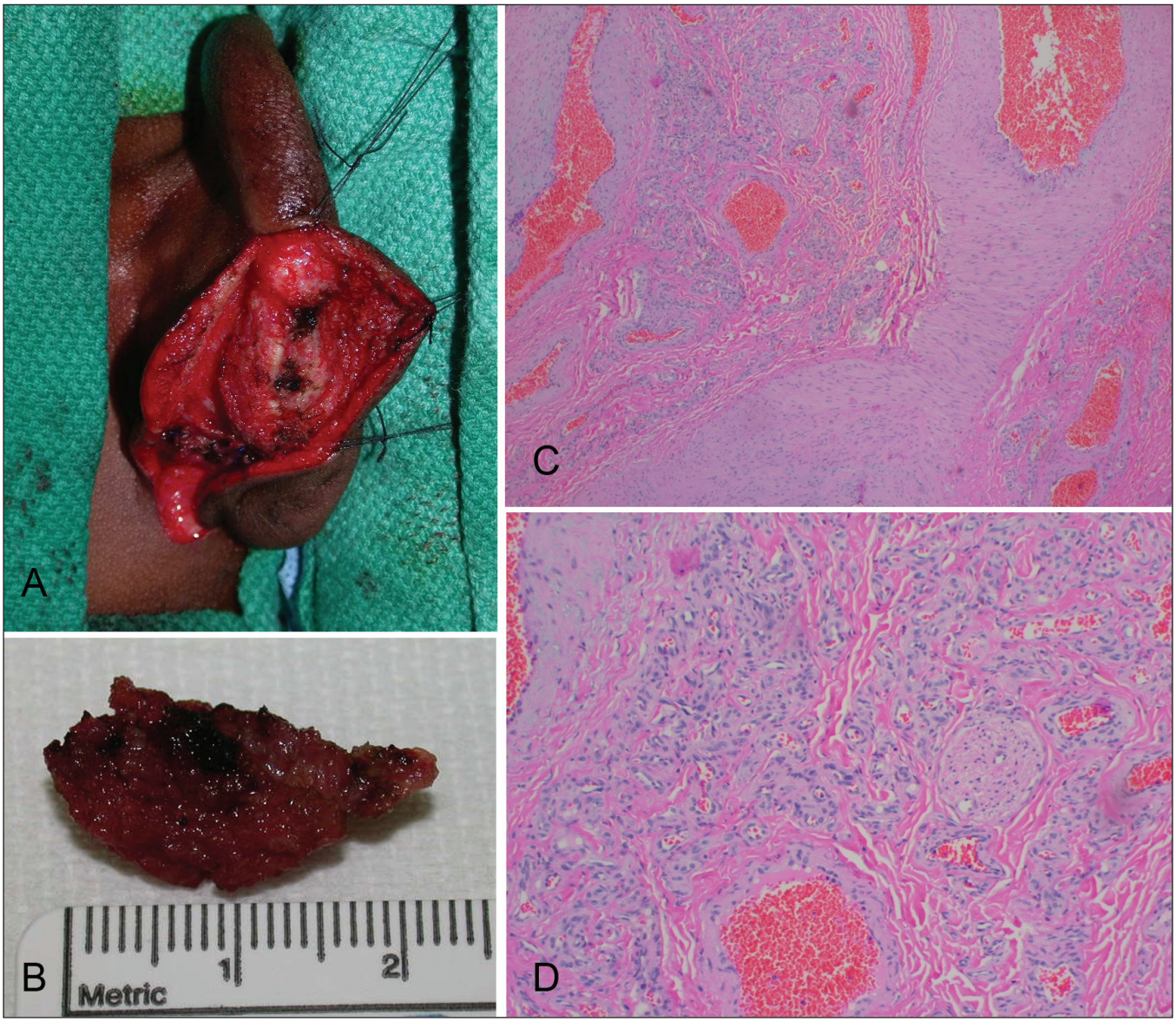

Figure 2) *Intraoperative findings (***A***), soft tissue dissection (***B***) and histology slides (***C***,***D***)*

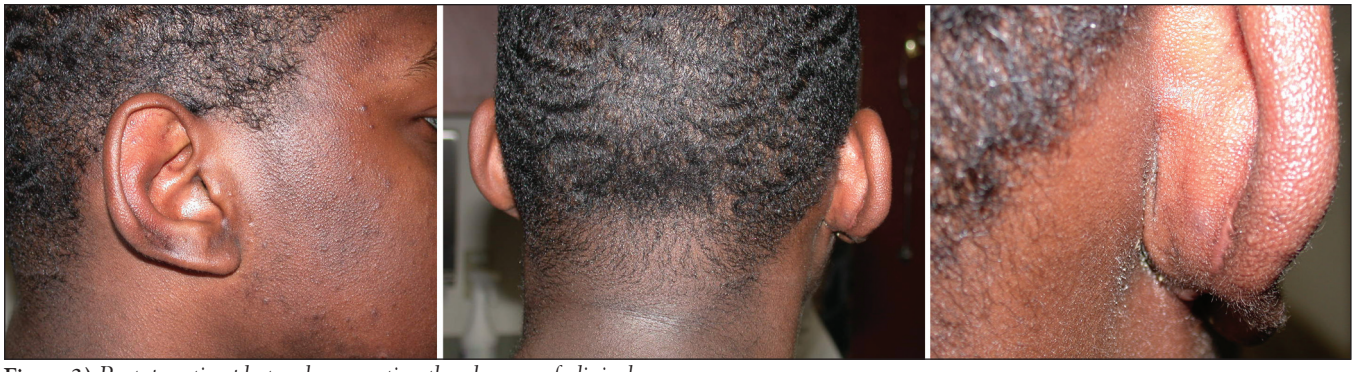

Figure 3) *Postoperative photos documenting the absence of clinical recurrence*

also been postulated that local ischemia plays a role in pathogenesis. It is well known that an arteriovenous malformation enlarges rapidly following proximal ligation. It is thought that this may be the cause of enlargement of the lesions following trauma (1).

In AVMs the endothelial cells have a normal rate of cellular division and do not regress. These malformations are morphogenic anomalies of the vascular structure that are present at birth and tend to grow with the child. Histologically, AVMs may have arteriovenous shunts with reactive, hypertrophic, thick-walled

arteries and veins because of the increased blood flow (2). It is believed that expansion is the result of increased blood flow rather than cellular proliferation. Puberty, pregnancy and trauma are factors which may affect the growth of an AVM (1).

One of largest series of arteriovenous malformations of the head and neck is from Kohout et al (1). They report a 16% incidence of lesions involving the ear. The overall cure rate was 60%. Cure rate for small malformations was 69% with excision only and 62% for extensive malformations with combined embolization-resection. These lesions can be classified into four stages as described by Schobinger (1), I: cutaneous blush/warmth; II: Bruit, audible pulsations, expanding lesion; III: Pain, ulceration, bleeding and infection; and IV: Cardiac failure.

Jackson et al (3) provide a classification of vascular malformations predicated upon the degree of blood flow within the lesion. These definitions are radiographic findings indicating the speed of flow through the lesion and the rate of shunting between the arterial and venous components. Low flow lesions are venous in origin, found in the head and neck, limbs and trunk, and were treated with surgical resection (52%), sclerotherapy (42%) and embolization. High-flow lesions are arteriovenous in origin, found in the head and neck exclusively, and required angioembolization plus surgical resection (n=10 of 16), angioemolization/resection/sotradecol injection (n=3), emolization/sotradecol (n=1), compartmentalization/sotradecol (n=1) and embolization (n=1).

Radiological evaluation of these lesions can be accomplished via various modalities. Magnetic resonance imaging can be used to define the extent of soft tissue involvement, as well as display flow dynamics of the lesion. Computed tomography may determine skeletal involvement. Angiography is the most vital tool in evaluating the AVM because it identifies the vascular supply and allows selective embolization of these collateral vessels before operative resection (1,2). Although superselective embolization may limit intraoperative bleeding, it is essential that the procedure is not delayed more than 48 h after the embolization because of the development of extensive collaterals (3).

The current literature provides a variety of substances to embolize these vascular lesions. N-butyl-2-cyanoacrylate is a permanent agent that polymerizes rapidly and forms a cast that instantly reduces flow through the malformation (4). Absolute ethanol, as well as sodium tetradecyl sulfate (3) have been used as embolizing agents for peripheral AVMs. Complications of sclerosing agents include acute or chronic nerve palsy and recurrence of the lesion (3,5).

AVMs of the external ear are an uncommon entity, often presenting as an enlarging pulsatile mass. The exact pathogenesis of these lesions has not been clearly defined, but are thought to arise during fetal development as a result of the failure of regression of arteriovenous channels in the primitive retiform plexus. It has also been postulated that local ischemia plays a role in the development of AVMs. Depending upon the extend of the malformation, the work-up may be limited to a physical examination or include angiography, magnetic resonance imagine and computed tomography to assist in the delineating the extent of the lesion. Treatment may be limited to surgical excision alone, or may include preoperative embolization with a variety of agents including N-butyl-2-cyanoacrylate, absolute ethanol and sodium tetradecyl sulfate.

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