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Pediatric Facial Arteriovenous Malformations: Case Report and Overview of the Basics

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Authors' contributions

This work was carried out in collaboration between all authors. Author PK reviewed the case at initial presentation. Author HC was part of the multidisciplinary team and performed the neurointerventional procedures. Author AB reviewed pre- and post-operative images, and drafted the manuscript. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aims: Describe the multimodality imaging of a large facial AVM in a child and discuss the treatment options of pediatric facial AVMs
Case Presentation: An 11 year old girl developed spontaneous (no prior trauma) enlargement of her left cheek with bleeding from her mouth when eating hard foods. The bleeding was described as pulsatile red bleeding that stopped spontaneously. On physical exam, she had a palpable pulsatile left buccal AVM; auscultation revealing high-flow through it. MRI demonstrated a 2.7 cm heterogeneous vascular mass arising in the soft tissues of the left cheek with involvement of the anterior wall of the left maxillary sinus with a large tortuous ectatic draining vein. CTA demonstrated a large AVM involving the left face with the nidus along the posterior wall of the left maxillary sinus, markedly enlarged draining veins throughout the left face/cheek and prominence of the pterygoid venous plexus. Left common carotid artery angiogram demonstrated an extensive AVM centered in the left maxilla. Left external carotid artery angiogram demonstrates the AVM receiving supply from the distal left internal maxillary, left facial, and left internal maxillary arteries. Angiography of the left internal maxillary demonstrated a high flow AVM with drainage to the left facial vein. The left internal maxillary artery was embolized using Onyx. The left facial, transverse facial, and internal

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maxillary arteries were embolized with PVA particles. Following embolization, blood supply to the AVM was substantially reduced. At surgery, the AVM was resected.
Discussion and Conclusion: Head and neck AVMs have a high morbidity as they tend to be clinically silent until manipulation/trauma results in bleeding. Those that bleed have a high incidence of rebleed. Embolization is utilized pre-operatively to decrease bleeding but not as the primary treatment due to potential for developing collaterals. Treatment remains controversial due to the rarity of the lesion, but typically involves a multidisciplinary approach as in our case.

Keywords: Pediatric facial arteriovenous malformation; embolization; surgical resection.

1. INTRODUCTION

Vascular malformations are one of the most common abnormalities seen in the pediatric population, with an incidence of about 1%; however, arteriovenous malformations (AVMs) are rare [1,2]. Arteriovenous malformations arise from errors in vascular development during embryogenesis, either abnormal vessel remodeling and/or abnormal vasculogenesis [2]. They consist of a central network of abnormal vascular channels, known as a 'nidus,' which shunts blood from the arterial system to the venous system [1,3,4]. We report the case of an 11 year old girl who was diagnosed with a facial arteriovenous malformation, which was successfully treated using embolization and surgical resection.

2. PRESENTATION OF CASE

An 11 year old female presented with gradual enlargement of her left cheek over the course of 2 to 3 months. She also noticed occasional bleeding from her mouth, which was pulsatile, and provoked by eating hard foods. The bleeding would stop spontaneously. There was no history of trauma, and no family history of vascular anomalies.

On physical exam, there was fullness of the left cheek with a pulsatile left buccal vascular appearing mass, which increased in size in the supine position. Auscultation revealed a highflow lesion. A bluish discoloration was noted along the left hemi-palate.

Initial MRI demonstrated a 2.7 cm heterogeneous vascular mass arising from the soft tissues of the left cheek (Fig. 1), with involvement of the anterior wall of left maxillary sinus, and a large tortuous ectatic draining vein. CT angiogram showed a large AVM involving left face with the nidus along the posterior wall of the left maxillary sinus, markedly enlarged draining veins throughout the left face/cheek, as well as prominence of the pterygoid venous plexus (Fig. 2). For further characterization, conventional angiogram was performed with selective left external carotid injection showing the AVM receiving supply from the distal left internal maxillary and left facial arteries. Selective angiography of the left internal maxillary artery demonstrated a high flow AVM with drainage to the left facial vein (Fig. 3).

After a multidisciplinary discussion with surgery and interventional neuroradiology, the AVM was deemed appropriate for embolization to minimize bleeding during surgery, followed by surgical resection. The patient returned to our institution after 2 months for both procedures.

Distal feeding branches of the left internal maxillary artery were first embolized with Onyx, followed by polyvinyl alcohol (PVA) embolization of the left transverse facial artery and left internal maxillary artery. Immediate post-procedure angiogram demonstrated marked reduction of blood flow from the external carotid artery to the AVM (Fig. 4). Two days later, Onyx was injected using the transmaxillary approach (to cannulate and embolize the presumed nidus) via direct puncture technique in order to achieve further reduction of flow (Fig. 5).

The patient was then taken to the operating room, and the AVM was separated from the masseter muscle and buccal fat pad. The facial artery, terminal external carotid artery, superficial temporal vessels, and venous channels were divided and ligated, and the AVM was removed. Follow-up CT angiogram showed obliteration of the AVM (Fig. 6).

3. DISCUSSION

Vascular malformations were originally classified by Mulliken and Glowacki in the early 1980s, based on their endothelial characteristics, clinical presentation, and biologic behavior [5]. Over time, this classification has been modified, and in 1996, the International Society for the Study of Vascular Anomalies categorized vascular

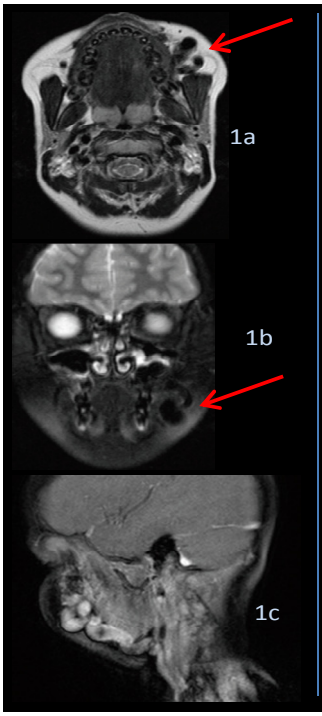

Fig. 1. MRI of the face demonstrates a vascular lesion in the left cheek. A and B, Axial and coronal T2 weighted images demonstrate serpiginous flow voids in the left cheek (arrows). C, Left parasagittal T1 post-contrast image demonstrates contrast within the vessel

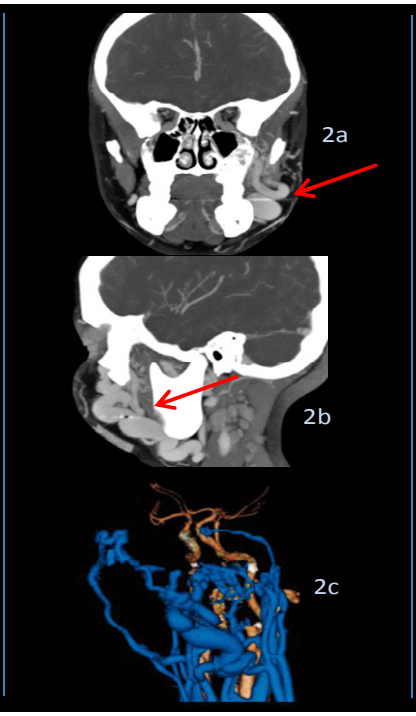

Fig. 2. A and B, CT angiogram (coronal and left parasagittal) demonstrates dilated serpiginous veins in the left cheek. Nidus centered along the left maxillary sinus (arrow). C, 3D volumetric image demonstrates extent of the lesion

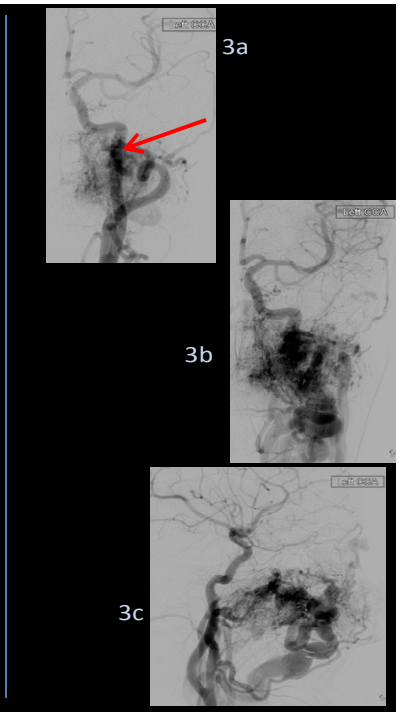

Fig. 3. Conventional angiogram performed for further characterization. A, Early image of a left common carotid artery injection demonstrates early filling of a dilated vein (arrow). B and C, Delayed images show a tangle of vessels and the true extent of the lesion

anomalies as vascular tumors or vascular malformations. Vascular malformations were further subdivided into slow-flow, fast-flow, and complex-combined. Under this classification scheme, arteriovenous malformations are under the fast-flow category [6].

The most common sites for head and neck AVMs are the cheek, then ear and nose; lesions of the mandible and maxilla are rare. They most commonly present in late infancy and early school age, and are slightly more predominant in girls, with a female to male ratio of 1.5 to 1. The lesion enlarges as the child grows, but acceleration of growth can be seen secondary to hormonal changes such as puberty, pregnancy, or even trauma [4,6].

Four stages of progression have been described in literature. The first is the asymptomatic stage, which is characterized by pinkish/bluish discoloration of the skin. The second is the expansile stage, where there is enlargement into a pulsating lesion. It is this pulsation that most commonly causes a patient to first seek physician consultation. If the lesion continues to progress, a destructive stage of bleeding, ulceration, and necrosis ensues due to steal of blood supply from the normal surrounding tissue. In the fourth stage, there is cardiac failure due to a high degree of shunting, known as decompensation [2,4,7].

Typically, silent arteriovenous malformations, those which are not disfiguring or impair function are monitored carefully. Bleeding, ulceration, and pain would be indicators for treatment [8]. Goals of treatment include symptomatic relief, preservation of function (patency of airway, preservation of sight, hearing, and mastication),

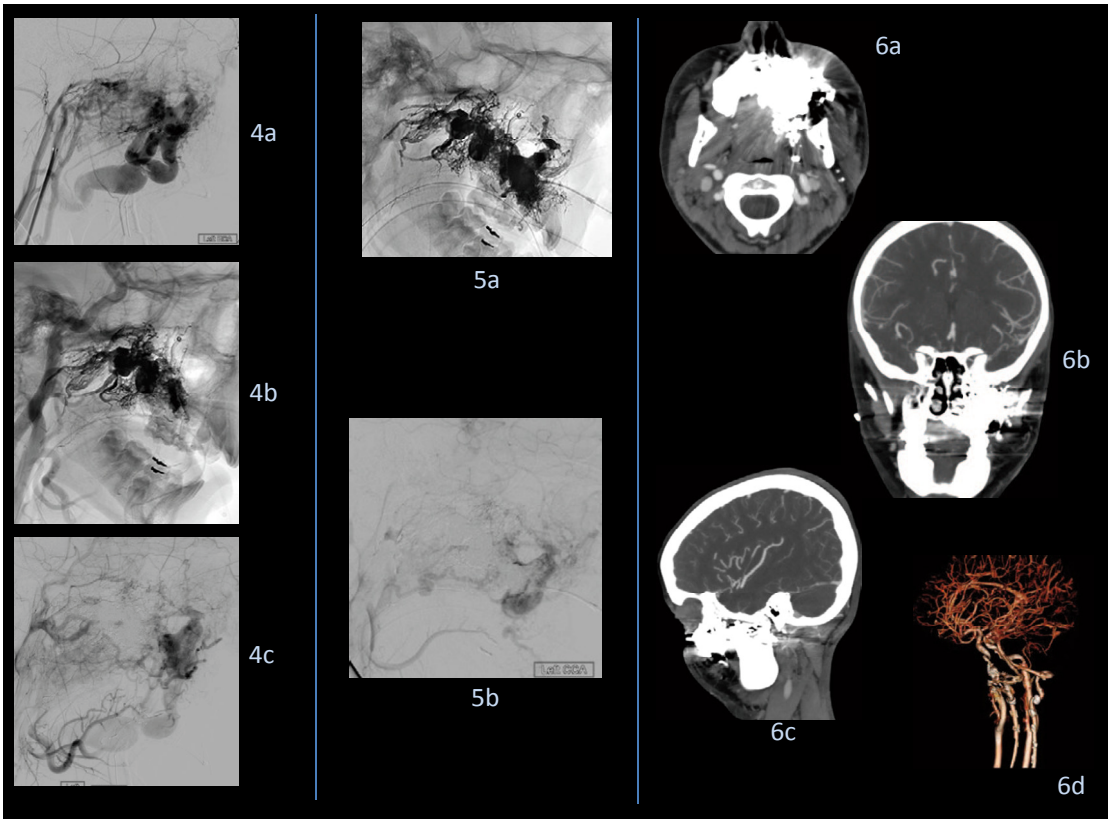

Fig. 4. Pre- and post-embolization. A, Left external carotid artery injection again demonstrates the AVM. B and C, Post-embolization with PVA particles and Onyx shows decreased flow to the lesion

Fig. 5. Further reduction of flow via the direct puncture technique. A, Injection of Onyx into the left maxillary region, presumed region of nidus. B, Delayed image of left common carotid artery injection showing minimal flow in region of AVM

Fig. 6. Imaging after treatment with embolization and surgical resection. A, B, and C, Select images from CT angiogram demonstrate no vascular lesion. Streak artifact from Onyx embolization (an extremely hyperattenuating agent, arrows). D, 3D volumetric image illustrates treatment (compare to Fig. 2C)

prevention of further osseous remodeling, and achieving a reasonable cosmetic appearance [4,9].

Unfortunately, there are no clear guidelines to treatment approach, which makes this topic extremely controversial. Embolization decreases blood flow to the lesion with instantaneous success during angiography, and is able to take out vessels that may be surgically inaccessible. More importantly, it helps prevent complications such as hemorrhage and subsequent cardiovascular collapse, owing to the high flow rates in AVMs. On the other hand, over time, embolization may cause the lesion to increase in size. It is therefore important to super select vessels; embolization of a proximal vessel can cause collateral vessels to 'open up,' thus causing recurrence or even further growth. These smaller vessels also nourish the normal skin of the face, and occlusion may cause skin necrosis, leading to cosmetic deformity.

Embolization may involve the transarterial approach or direct puncture technique [10,11], or as in our case, both. Direct puncture of the arteriovenous pouch has been shown to reduce both the risk of skin necrosis and inadvertent embolization of crucial smaller branch vessels such as the central retinal artery [11]. Despite the various embolization techniques and methods, recurrence is still high, and therefore is usually performed in conjunction with surgery. It has been shown that surgery within 24-48 hours after

embolization is best, which helps decrease blood loss and better define the surgical margins of the lesion [11,12].

Percutaneous sclerotherapy is another treatment option, typically used for surgically inaccessible lesions, most commonly with ethanol. Advantages of using ethanol include strong endothelial damage and subsequent thrombus formation within the vessel; therefore, a higher response rate to treatment. It is also inexpensive and easy to obtain. Unfortunately, there is a relatively high complication rate, ranging from 7.9-27.9%, which includes pain, skin necrosis, and nerve damage, especially worrisome when involving the face and a pediatric patient [13,14]. Most treatments involve a team approach, utilizing embolization, sclerotherapy, and surgical resection [15].

4. CONCLUSION

Pediatric arteriovenous malformations are rare, and therefore, treatment approach can be controversial. This case demonstrates the classic presentation of a pediatric facial AVM. Our patient is a female, who presented in the early school age years, during the destructive phase, with pulsatile bleeding. The maxillary nidus location was not unusual, but one of the more rare locations cited in literature. Treatment included both embolization and resection, which is also typical. Follow-up studies showed no evidence of residual AVM, which is unique because only rarely is complete removal seen on post-treatment imaging.

CONSENT

Not applicable.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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