

Case-Report

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Maxillary Arteriovenous Malformation - A Case Report

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Abstract

Arteriovenous malformations (AVM) are fast-flow congenital vascular malformations, consisting of anomalous communications between the arterial and venous system. They are very rare, accounting for 1.5% of all vascular anomalies with 50% occurrence in the oral and maxillofacial region. They are the most aggressive form of vascular malformation which can lead to significant deformity and functional impairment. Treating AVMs is challenging and relies on surgery, embolization or both.

A 23-year-old female patient with a personal history of congenital angioma of the right hemiface treated with laser in her childhood, was referred to the Stomatology department due to recurrent episodes of spontaneous high-output bleeding at home after dental scaling at her dental provider in the previous week. The diagnosis of maxillary intraosseous AVM was confirmed by angiography and embolization was performed in two sessions, with a liquid embolic agent obstructing four arterial afferences. The patient reported no episodes of hemorrhage or other symptoms on follow-up.

AVM can easily be misdiagnosed and produce substantial oral bleeding in the operating room, leading to severe life-threatening complications. Due to the rarity and significance of the symptoms in the oral and maxillofacial region and the potentially fatal outcome after surgery on undiagnosed patients, this case report can be very relevant to the clinician.

Highlights 1 - Arteriovenous malformations (AVM) are very rare, accounting for 1.5% of all vascular anomalies, and can easily be misdiagnosed, producing substantial oral bleeding in the operating room, leading to severe life-threatening complications. 2 - Early diagnosis and treatment of AVM is important, as the natural history of these malformations is progressive expansion and associated morbidity. 3 - Doppler ultrasound, MRI and CT scan are required to develop an appropriate treatment plan and prevent complications. 4 - A multi-disciplinary approach is necessary for planning individualized treatment using embolization and surgical resection. 5 - Patients require a tight follow-up with physical examinations and imaging.

1. Introduction

Arteriovenous malformations (AVM) are fast-flow congenital vascular malformations, consisting of anomalous communications between the arterial and venous system, thus causing shunting of blood through a primitive vascular nidus into tortuous dilated outflow veins. [1] The blood shunted to the malformation causes the lesion to grow, which in turn causes increased shunting of the blood; hence, a vicious circle.[2]

Vascular lesions can be divided into vascular tumors or vascular malformations. These last ones are caused by disturbance in the late stages of angiogenesis, resulting in the persistence of embryonic arteriovenous anastomosis.[2] The malformations have a normal rate of endothelial cell turnover and are present at birth, usually becoming noticeable over the years. The enlargement of these lesions is due to the change in pressure and flow, dilatation of vascular channels, shunting and collateral proliferation, rather than cellular proliferation as seen in vascular tumors such as hemangiomas.[3]

Vascular malformations are extremely common lesions, accounting for approximately 7% of all benign tumors. Although the head and neck constitute less than 14% of the total surface area of the body, approximately 50% of all vascular malformations occur in this region.[4]

AVMs are the most common high-flow lesions, frequently identified in the limbs, head, neck and lungs. However, lesions of the oral and maxillofacial regions are very rare, accounting for only 1.5% of all vascular anomalies. [1] They are the most aggressive form of vascular malformation which can lead to significant deformity and functional impairment. [3] AVMs have been reported in the maxilla, but at half the frequency of AVMs in the mandible. [5] AVMs of the jaws are relatively rare, with fewer than 200 cases reported in the literature. [6] AVMs usually appear in adolescence but have an age range of 3 months to 74 years. Some authors noted predominance in females (female-to-male ratio, 2:1) while others have reported equal prevalence among males and females. [7] Kohout *et al.* studied 81 patients over a period of 20 years and found that 55 AVMs (69%) were in the middle of the face, 11 (14%) in the upper third and 14 (17%) in the lower third of the face. [8]

Clinical signs and symptoms of AVM can vary depending on anatomical location and may include pain, slow-growing soft-tissue enlargement with an audible bruit or palpable thrill, mucous membrane or cutaneous pigment changes, erythematous gingiva, spontaneous gingival bleeding, resorption and mobility of teeth, soft tissue discoloration, facial swelling and asymmetry. [9]

Radiographically, AVMs are osteolytic and frequently have indistinct margins. Computed tomography can demonstrate enhancement of the lesions, while angiography can depict distended feeder vessels and arteriovenous shunts. Magnetic resonance imaging can visualize flow voids in high-flow abnormalities. [10]

2. Case Report

A 23-year-old female patient with a personal history of congenital angioma of the right hemiface treated with laser in her childhood, was referred to the Stomatology department of Hospital Santa Maria, Lisbon, due to episodes of spontaneous high-output bleeding at home, associated with lipothymia. These episodes occurred after dental scaling at her dental provider in the previous week, were very abundant and hard to control and began while the patient was resting. On examination, there was a slight non-bleeding purple swelling in the alveolar region of the tooth 1.6 (Figure 1). A contrast-enhanced CT scan revealed a soft tissue lesion with disruption of the anterior and posterior alveolar cortical bone adjacent to 1.6, associated with multiple vascular structures that were ectatic (Figure 2). Magnetic resonance angiography showed dilated and serpiginous anomalous vascular structures adjacent to the posterolateral and anterior wall of the right maxillary sinus, with multiple flow voids

in T1, T2, that had afferents from the internal maxillary artery (Figure 3).



Figure 1. Intraoral photograph showing swelling in the alveolar molar region.



Figure 2. Contrast-enhanced CT scan showing disruption of the alveolar bone.

The diagnosis of AVM was confirmed by angiography and embolization was performed in two sessions with Squid, a liquid embolic agent made with ethylene vinyl alcohol copolymer mixed with suspended small grains of radiopaque tantalum powder, obstructing four of the malformation's arterial afferents (Figure 4).

On follow-up, the patient reported no episodes of hemorrhage or other symptoms. The patient will maintain long-term follow-up, with the consideration of additional embolization should recurrent bleeding episodes occur.

3. Discussion

Vascular malformations are a form of vascular anomaly caused by abnormal development of vascular ele-



Figure 3. MR angiography showing anomalous vascular structures with arterial afferents.



Figure 4. Angiography showing the occluded vessels with Squid.

ments during embryogenesis and in fetal life. [11] These malformations can be of a single vessel form or in combination of arterial, venous, capillary or lymphatic malformation. Vascular malformations can be a slow flow malformation like capillary, lymphatic or venous; or it can be a fast flow like arterial or arteriovenous malformation. [12]

Various diagnostic tools are available to diagnose vascular lesions such as ultrasound with color Doppler, computed tomography (CT) scan, MRI, and magnetic resonance angiography. Type of malformation can be confirmed by ultrasound with color Doppler examinations. CT scan with iodinated contrast identifies AVM as a highly enhancing lesion and can demonstrate soft tissue enhancement as well as dilated feeding and draining vessels. [13] MRI can be used as an excellent technique in the diagnosis of vascular malformations either by itself or before angiography. It shows a good depiction of vascular structure, permitting the differentiation between

high-flow and low-flow lesions. High-flow lesions show typical signal flow voids both in T1-weighted and T2-weighted sequences with the appearance of serpentine images. [14] In the present case, MRI was able to identify the kind of flow, i.e., high flow as well as the origin of the vessels, i.e., internal maxillary artery.

Management of AVM is most difficult due to the replacement of normal tissue by the diseased vessels and high-flow rate of recurrence. This management mainly consists of surgery, vascular embolization, or a combination of both. Surgical treatment consists of wide resection which is difficult and potentially hazardous due to significant blood loss during surgery and also because of severe deformation of the maxilla and the face. The purpose of embolization is diminishing the blood supply, occluding the vessels contributing to the lesion. The combination of both is used to control acute hemorrhage allowing for excisional surgery to be performed afterwards, if needed. In the present case, the patient was embolized with Squid, controlling the symptoms and avoiding major tissue resections. She was scheduled for yearly follow-up examinations, as well as CT and MRI controls. In case of new episodes, she was instructed to come to the Stomatology Emergency Department. [15]

4. Conclusion

Early diagnosis and treatment of AVM is important, as the natural history of these malformations is progressive expansion and associated morbidity, especially during surgical procedures. Detailed investigations including Doppler ultrasound, MRI and CT scan are required to develop an appropriate treatment plan and prevent complications. Despite that, AVM can easily be misdiagnosed and produce substantial oral bleeding in the operating room, leading to severe life-threatening complications. Due to the rarity and significance of the symptoms in the oral and maxillofacial region and the potentially fatal outcome after dental procedures on undiagnosed patients, this case report can be very relevant to the clinician.

Appropriate management is best achieved via a multidisciplinary approach. Combined embolization and surgical resection, if possible, is the most successful treatment for well-localized AVM. However, these patients must still be followed for years with regular physical examination and ultrasonography and/or MRI.

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