# Arteriovenous Malformation of the Mandible: Review of Literature and Case History

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## Abstract

*Vascular malformations of the jaws can lead to disastrous complications, but there seems to be no consensus as to their treatment. The literature presents the pathophysiology and clinical aspects of these lesions, as well as the divergent views of the authors. Treatment by catheterization and embolization, with direct transosseous injection of cyanoacrylate, appears to be the least harmful and most permanent treatment in certain conditions, as evidenced by the case of this 9-year-old patient having a high-flow mandibular vascular malformation.*

***MeSH Key Words:*** *arteriovenous malformations/diagnosis; arteriovenous malformations/therapy; case report; mandible/blood supply*

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Intraosseous vascular malformations (VMs) of the maxillofacial region sometimes give rise to dental emergencies and may cause disfigurement, morbidity, even death.[1-6] The proximity of the teeth can prove disastrous. A review of fatal cases by Lamberg and others[1] shows that in most instances, exsanguination is the result of dental extractions, the dentist having been unaware of the existence of the arteriovenous malformation. Although VM is rare, the dentist must always consider the possibility and be able to recognize the clinical signs in order to propose proper treatment.

Many terms are used to refer to this lesion, notably, arteriovenous aneurysm, cavernous hemangioma, central hemangioma, pulsatile hemangioma, angioma, arteriovenous shunt, arteriovenous fistula, vascular malformation and arteriovenous malformation.[7] Before the 1980s, vascular lesions were referred to as "hemangiomas."[7] Thereafter, they were subdivided into hemangiomas and vascular malformations.[8]

## Etiopathogenesis

These lesions are the result of an embryologic abnormality of the vascular system. Hemangiomas are caused by a failure of differentiation in the early stages of embryogenesis.[7] Usually extraosseous, they more commonly appear in childhood and tend to regress or disappear in adolescence. They are rarely associated with fatal hemorrhages.[9]

VMs, on the other hand, are caused by a disturbance in the late stages of angiogenesis (truncal stage) and result in the persistence of arteriovenous anastomoses present during embryonic life.[3] They may be capillary, lymphatic, venous, arterial or mixed. VMs of arterial or arteriovenous origin are often referred to as "high-flow vascular malformations" and are often the cause of massive, sometimes fatal hemorrhages.[9]

VMs, which usually present as developmental anomalies from birth, develop in proportion to physical growth.[10] The increase in size of these VMs, asymptomatic and imperceptible at an early age, is promoted by local hemodynamic factors. Areas of low vascular resistance cause a shunting of the blood with decreased perfusion of the peripheral tissue in favour of collateral flow, gradual dilatation of the nutrient arteries with atrophy of their musculo-elastic wall and decreased resistance, and dilatation and arterialization of the draining veins, owing to the increase in intraluminal pressure.[3] The blood shunted to the malformation causes the lesion to grow, which in turn causes increased shunting of the blood; hence, a vicious circle.

## Clinical signs

VMs are attributable to hormonal, infectious or traumatic factors[11]— which would explain their late detection

in later childhood. The theories concerning traumatic factors[12] are still, however, much debated.

Mandibular VMs usually appear during adolescence, with extremes at 3 months and 74 years of age.[3] Some authors have noted a predominance in women (2:1), while others have reported equal prevalence among men and women.

Intraosseous VMs near the alveolar bone are often present with pericoronal bleeding, mobile teeth, and sometimes occlusal anomalies.[10] Gingival bleeding seems to be a symptom common to most documented cases.[2] Many instances of massive hemorrhage, even exsanguination, have been documented following the extraction of teeth associated with these VMs.[2,9]

More central lesions are painful and produce an alteration of facial morphology, a bruit sometimes accompanied by a thrill, and neurosensory deficits.[10] Vascular naevi or phlebectasias may discolour the adjacent mucosa or skin.[2,3] At the level of the nose, sinuses or eye sockets, there may be nasal blockage, epistaxis, rhinitis, sinusitis, proptosis or diplopia.[10] Cardiac symptoms (cardiomegalia, heart failure, murmur) are rare.[2,3]

## Radiological signs

In the mandible and maxilla, the lesion produces a poorly defined, radiolucent image,[10] often having the appearance of a honeycomb or soap bubbles, with small rounded and irregular lacunae.[2,3,13] Root resorption has been observed, creating an appearance of teeth floating in the adjacent alveolar osseous erosion.[3,9]

The spread of the lesion may mimic the appearance of certain odontogenic cysts. As the radiological image varies and suggests numerous diagnoses, further tests are essential.

CT scanning and magnetic resonance imaging help mostly to clarify the extent of the lesion, bone erosion and the involvement of major vessels.[10]

Super-selective arteriography remains an essential tool in the identification of the VM and contributory vessels.[13] This technique consists in injecting a radiopaque substance into the vascular system through a catheter near the region. The image is processed by computer, and the bone densities are substracted for a clearer illustration of the vascular system.

Super-selective arteriography of the external carotid must be done bilaterally, given the importance of the collaterals and multiple anastomoses of the maxillary artery.[14]

## Management strategies

Various sclerosing agents (sodium morrhuate, boiling water, nitrogen mustard, etc.) have been used to treat high-flow lesions, but have proven ineffective because they were displaced from their site of action by the speed of the blood flow.[7] Other, less recent solutions have been abandoned because of their limited success and their side effects.[3]

The ligation of the external carotid is widely cited as an adjunct to many approaches, but most authors strongly advise against it,[3,4,7,11,13,15-17] since many anastomoses (internal carotid, ophthalmic, vertebral, cervical, and contralateral external carotid) promote the rapid appearance of a collateral circulation. Moreover, it rules out any control angiography or future embolization.

Embolization, which consists in occluding the vessels contributing to the lesion, has been used for some time.[7] Several materials, usually inserted by means of femoral catheterization,[3] have been used: polyvinyl alcohol particles,[10,17,18] muscle,[19] Gelfoam,[2,11,13] cyanoacrylate,[3,7,16,17,20,21] metal coils,[15,21,22] collagen.[4,23] Some authors present this technique as a preliminary and indispensable adjunct to excision and reconstructive surgery,[9] while others use it as the sole, definitive approach.[16] Its limitations are a function of the nature of the occluding material and the characteristics of the lesions.

The main nutrient arteries of the malformation being embolized, the blood flow is redirected to the collaterals which, angiographically invisible owing to limited perfusion, dilate as a result of the hemodynamic change and reirrigate the malformation.[2,3,11,22] A recurrence follows.

The use of fluid materials appears to be more effective in occluding the distal nutrient arteries and reducing the risk of a recurrence.[21] Multiple embolizations are sometimes necessary, and the venous access route may supplement conventional arterial embolization.[15,21]

Recently, the direct transosseous puncture of the vascular bed has been proposed.[16,24,25] The core of the malformation and all associated vascular pedicles are then, theoretically, embolized. Rodesh and others[16] did an interesting study reporting a success rate of 100%. The 9 patients treated with cyanoacrylate were stabilized over the long term (33%) or cured (67%) without further treatment.[16]

Embolization, combined with surgical treatment, is still the most conventional modern approach.[2,4,5,9,11,18,22,26-28] This procedure controls the acute hemorrhagic phase, but does not eliminate the risk of a recurrence, owing to the appearance of a collateral circulation. It does, however, reduce the blood flow, allowing for excision surgery to be performed within anywhere from 48 hours to 2 weeks.[11] Resection of the mandibular fragment containing the lesion has long been considered essential to complete healing.[18,22,27] Curettage of the resected fragment with immediate reimplantation does, however, reduce the morbidity associated with the procedure and the difficulty of reconstruction.[23,27] Lesional curettage without resection preserves good bone support, but the excision is often deemed inadequate.[3]

## Case history

A 9-year-old patient was referred to us in January 2000 for assessment of an asymptomatic left mandibular

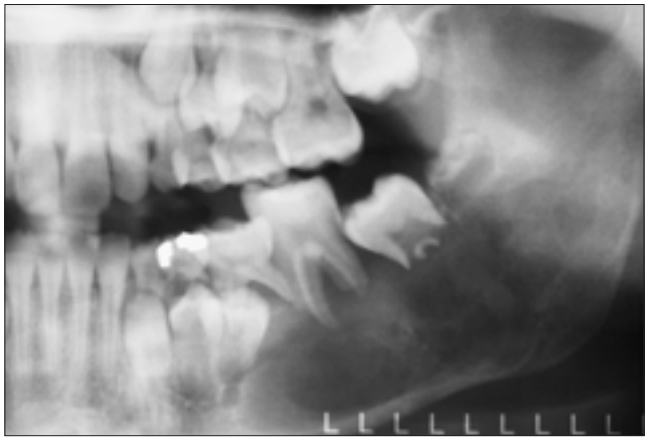

***Figure 1a:*** *Initial panoramic radiograph.*

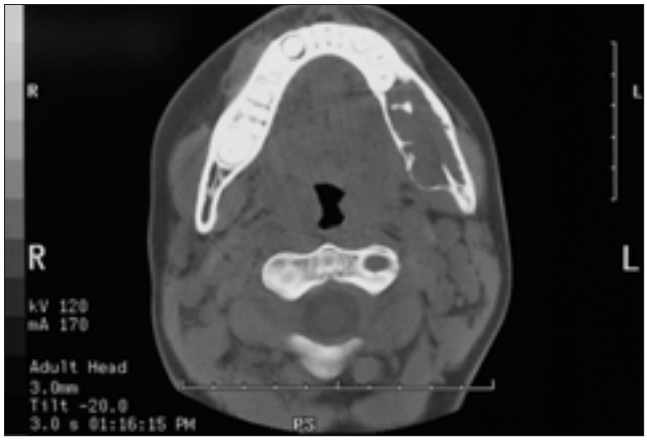


***Figure 1b:*** *Initial CT scan (axial section).*

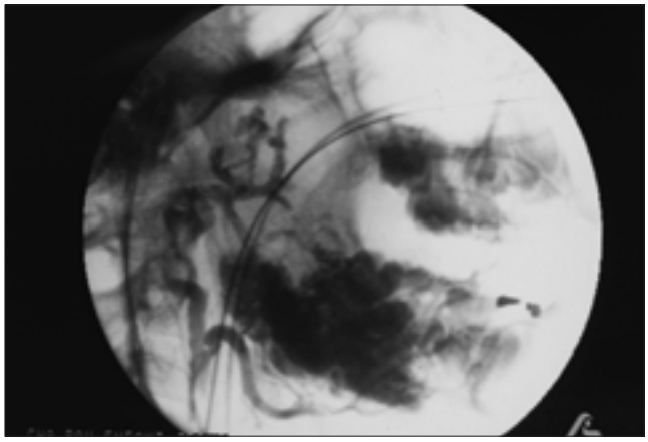

***Figure 1c:*** *Initial angiography.*

intraosseous lesion. Her dentist had noticed the extrusion with gradual mobility of tooth 36, with no other clinical signs.

No abnormality or asymmetry was noted during the facial examination. A slight swelling of the left mandibular buccal cortex was observed. Slight extrusion and dental mobility were confirmed. A non-hemorrhagic, gingival granulomatous swelling was noted. No vascular bruit was detected.

The initial panoramic examination showed extensive multilocular radiolucency of tooth 34 extending to the region of the lingula and the neck of the condyle. Root resorption was noted in tooth 36 (**Fig. 1a**). The mandibular CT scan confirmed the extent of the lesion, with erosion mainly at the expense of the lingual cortex (**Fig. 1b**). The differential diagnosis suggested: follicular cyst, central giant cell granuloma, ameloblastic fibroma, aneurysmal cyst, ameloblastoma.

An incisional biopsy under general anesthesia was suggested on an outpatient basis. The simple extraction of tooth 36 caused a massive hemorrhage. Hemostasis was achieved by digital pressure and displacement of a buccal flap. The patient was discharged once proper hemostasis was observed, but was readmitted in emergency the following day for treatment of massive and spontaneous gingival hemorrhage, controlled by sustained digital pressure. A cerebral and facial angiography was done under general anesthesia (**Fig. 1c**). This examination revealed the arteriovenous malformation of the left mandibular body, fed by the facial and lower left alveolar arteries.

Working with a radiologist, we performed an embolization by direct puncture of the lingual mandibular cortex and injection of a mixture of cyanoacrylate and Lipiodol, under constant digital pressure at the site of the hemorrhage. Complete occlusion of the malformation and cessation of the bleeding were achieved after 3 injections of solution under fluoroscopic and angiographic examination (**Figs. 2a, 2b, 2c**).

Post-operative pulmonary radiography showed the presence of radiopaque material at the level of several distal pulmonary arteries bilaterally, evidence that the embolization material had entered the veins without clinical incident. Antibiotic therapy and anti-inflammatory treatment were prescribed.

Over the 6 months following the embolization, 2 local infectious episodes were treated with antibiotics. Dehiscence of the mucosa, exposing the embolization material at the level of the alveolus of tooth 36, was noted.

Radiographic examination 6 months after the embolization showed changes suggesting a revascularization of the lesion. Chronic osteomyelitis was suspected, given the considerable thickening of the left mandibular buccal cortex (**Figs. 3a, 3b**). Both these diagnoses were ruled out after a gallium scintigraphy and a control arteriography proved normal.

Based on this clinical picture, we did a complete curettage of the occluding material. Teeth 37 and 38 were sacrificed. Bacteriological cultures confirmed major infection with

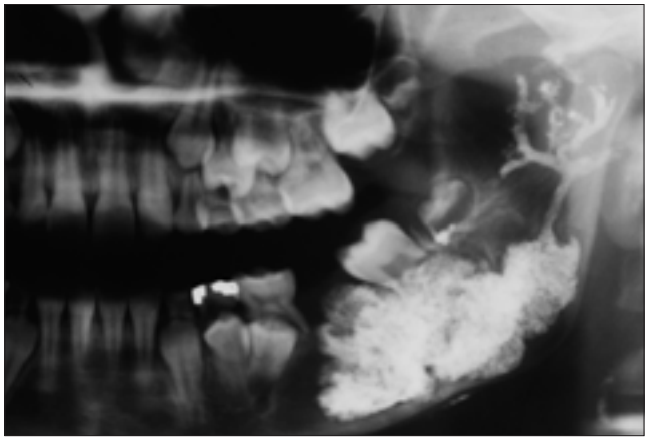

***Figure 2a:*** *Post-embolization panoramic radiograph.*

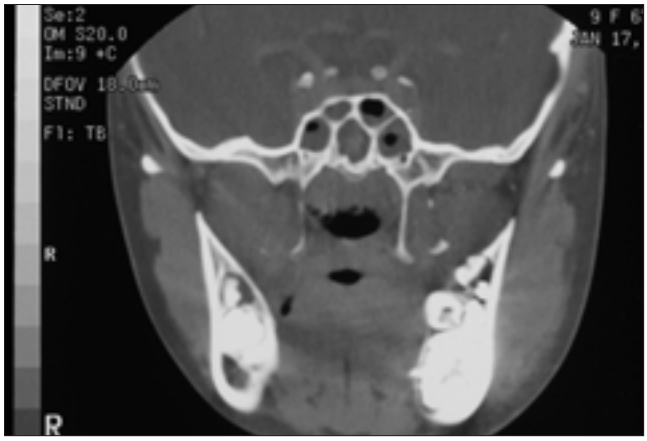


***Figure 2b:*** *Post-embolization CT scan.*

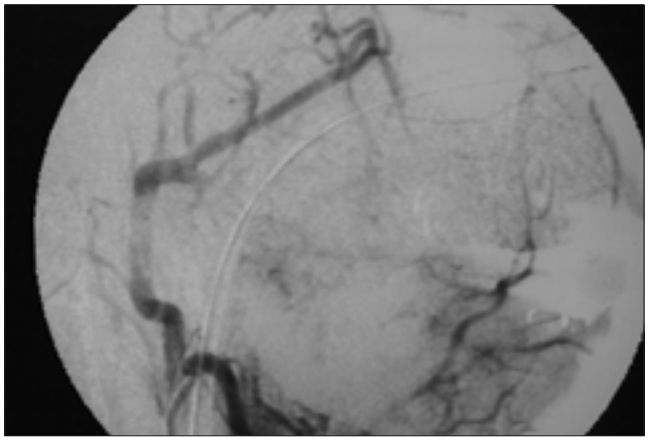

***Figure 2c:*** *Post-embolization angiography.*

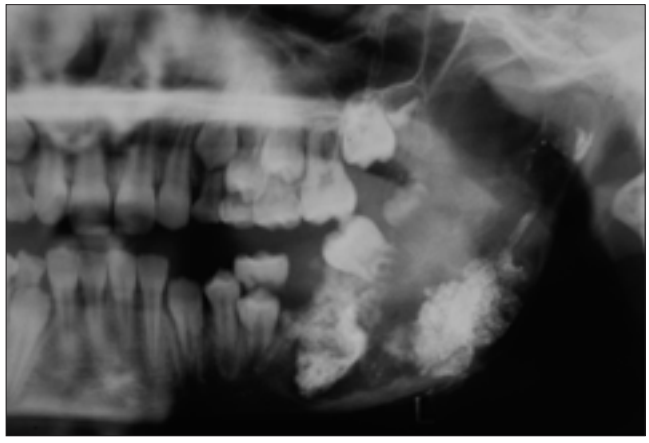

***Figure 3a:*** *Panoramic radiograph 6 months post-embolization.*

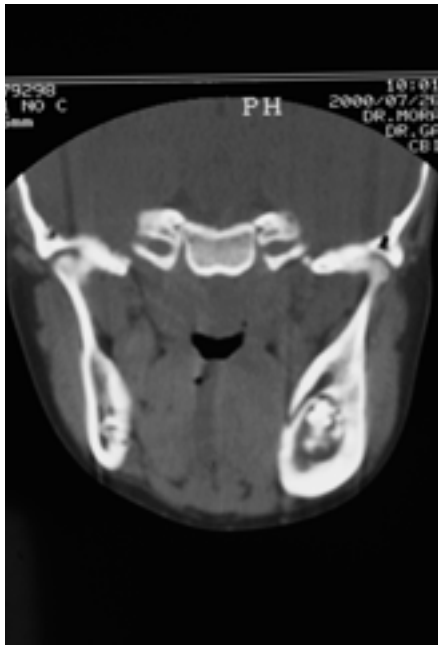


***Figure 3b:*** *CT scan 6 months post-embolization.*

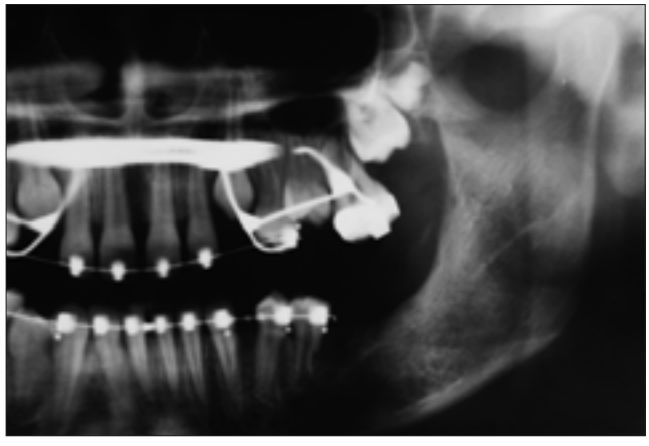

***Figure 4:*** *Panoramic radiograph 14 months post-embolization.*

*actinomyces odontolyticus*. The positive gallium scintigraphy called for 4 months of oral antibiotic therapy (clindamycin). Examination at 14 months confirmed excellent gingival healing, no signs of infection, the maintenance of facial symmetry and complete ossification of the lesion site (**Fig. 4**). Orthodontic management and clinical follow-up are proceeding without incident.

## Discussion

Besides the absence of the usual early signs or symptoms of VMs (gingival bleeding, throbbing tooth, thrill, etc.), the outcome of the present case differs from that of the classic case: there was near-total anatomic restoration, with no residual disability and no apparent impediment to mandibular growth.

Yet fate might have determined otherwise, as the clinical and radiological characteristics favoured an approach that initially ruled out a vascular etiology for the lesion.

This is reason enough to be even more suspicious when making a differential diagnosis in the presence of a radiotransparency of the jaws, especially in the case of young patients and in the absence of the usual signs of VM. In such a context, the investigative algorithm sometimes suggests needle puncture, for which a negative outcome does not completely rule out VM, but for which a positive outcome requires an emergency response. A cautious and preventive approach must therefore be adopted during initial diagnostic procedures.

Once the lesion is confirmed, the therapeutic path does not become any clearer. The multiplicity of approaches contrasts with the rareness of this type of malformation, but is in direct relation to the urgency of the required intervention and the permanency of the measures to be applied.

Current treatment must not lead to the debilitating and deforming sequela that have, in the past, characterized certain surgical procedures, even though they have successfully averted death. Accordingly, embolization has carved out for itself a dominant place in the modern panoply of treatments, as Frame reported in 1987.[29]

An understanding of the pathophysiology of the lesion has removed ligation of the external carotid and direct surgical approaches from the current armamentarium. The endovascular techniques that replace them are not, however, without risks or complications: many minor problems or major sequelea can occur at the puncture site, along the vascular path and during injection of the occluding material. Superselective embolization used in different ways, (as a standalone permanent treatment, as an adjunct to resection-reconstruction surgery, or in combination with a retrograde venous approach) is the current treatment of choice.

Recent accounts[16,24,30] in favour of the direct intralesional injection of isobutyl cyanoacrylate offers us a new perspective. There seems to be less morbidity reported with this approach even though limited outcome data are available.

The greatest advantage of this approach may lie in its intrinsic ability to eliminate the whole vascular latticework feeding the lesion, promoting, especially in children, full expression of the regenerative potential of somatic growth to replace the vascular anomaly. The choice of this approach therefore depends not only on the lesion's size, accessibility or anatomic contiguity to the important structures, but also on the patient's regenerative capacity.

## Conclusion

The rareness of VMs is equalled only by the morbidity they cause and the urgency of the measures to be taken once detected, in all circumstances. A high degree of suspicion leads to their diagnosis and considerably reduces the risks of a catastrophe once identified. Treatment by catheterization and embolization with direct intralesional injection of cyanoacrylate allows for conservative anatomic and functional recovery. It is relatively non-invasive and safe when the anatomy and clinical status permit its use. ➧

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