



## Clinical case

# Extensive arteriovenous malformation in the face of a pediatric patient – Case report



Matheus Furtado de Carvalho<sup>a,\*</sup>, Natacha Kaline de Oliveira<sup>a</sup>, Eduardo Duailibi<sup>b</sup>,  
Maria da Graça Naclério Homem<sup>a</sup>, Maria Cristina Deboni<sup>a</sup>

<sup>a</sup> Department of Oral and Maxillofacial Surgery, Dental School, University of Sao Paulo, São Paulo, Brazil

<sup>b</sup> Department of Radiology, Dental School, University of São Paulo, São Paulo, Brazil

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### ABSTRACT

Vascular anomalies can be academically divided into two groups: vascular tumors and vascular malformations. Among vascular malformations, we can highlight the arteriovenous malformations, a subtype of anomaly characterized by presenting a high blood flow. The diagnosis is beyond doubt a great challenge for the multi professional team involved in treating these anomalies. Imaging tests such as the ultrasonography, magnetic resonance, CT scan with contrast and angiography are extremely important for the orientation about the characteristics and hemodynamic properties of the blood vessels, for delivering predictable results and for risk reducing of complications. In this paper, we describe a rare case of extensive arteriovenous malformation in the face of a pediatric patient, its clinical characteristics and imaging manifestations as well as the satisfactory clinical results obtained with a combination of sclerotherapy and surgical excision.

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### Extensa malformação arteriovenosa em face de paciente pediátrico – Relato de caso

#### R E S U M O

As anomalias vasculares podem ser didaticamente divididas em dois grupos: os tumores vasculares e as malformações vasculares. Dentre as malformações vasculares, podemos destacar as malformações arteriovenosas, um subtipo de anomalia caracterizada por apresentar um alto fluxo sanguíneo. O diagnóstico é sem dúvida um grande desafio para a equipe multiprofissional envolvida no tratamento destas anomalias. Exames imaginológicos como a ultrassonografia, ressonância magnética, tomografia computadorizada com contraste e a angiografia são fundamentais para orientação quanto as características e propriedades

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\* Corresponding author.

E-mail address: [matcarodonto@yahoo.com.br](mailto:matcarodonto@yahoo.com.br) (M.F. de Carvalho).

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hemodinâmicas dos vasos sanguíneos, proporcionando resultados previsíveis e menor risco de complicações. Descrevemos um caso raro de extensa malformação arteriovenosa de face em paciente pediátrico, suas características clínicas e imagiológicas bem como os bons resultados clínicos obtidos com a escleroterapia e exérese cirúrgica.

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## Introduction

Vascular anomalies are a set of changes of blood vessels that could be associated, or not, to endothelial cells proliferation that results in a vascular hamartomatous growth.<sup>1</sup>

There are many discussions about the classification of these anomalies and for this reason, the correct diagnosis is important to establish the most appropriate therapeutic treatment. Although most vascular abnormalities can be clinically diagnosed, it is common knowledge that severe conditions require complementary tests for the differential diagnosis between soft tissue or bone neoplasms.

In 1996, the International Society for the Study of Vascular Anomalies (ISSVA) adopted and revised the classification system created by Mulliken & Glowacki in 1982,<sup>2</sup> being considered since then the official classification system of vascular anomalies. They proposed the division of these anomalies in two great groups: tumors (infantile hemangioma, congenital hemangioma, tufted angioma, kaposi hemangioendothelioma) and malformations. The vascular malformations can be subdivided into low-flow (capillary, venous or lymphatic) and fast flow malformations (arterial malformation and arteriovenous fistula).<sup>3</sup>

Vascular malformations (VMs) are disorders of benign origin, unusual to the maxillofacial region. They represent about 6.4% of lesions found in the oral cavity. However, studies on prevalence of these lesions are still scarce. The upper and lower lips along with the oral mucosa have the highest incidence.<sup>4</sup>

Although the VMs in the head and neck region cause significant esthetic defects, nasal obstruction and interference



Fig. 1 – Initial extraoral photography (facial deformity).

on speech and dentition, the main concern of patients is the possibility of severe and recurrent hemorrhages.<sup>5</sup>

Complementary methods such as computed tomography, Doppler ultrasonography and magnetic resonance imaging, in association with clinical findings can provide information regarding blood flow characteristics and lesions extent. Generally, low-flow vascular malformations are percutaneously treated with sclerosing agents injection, while in high flow lesions the approach is endovascular, with permanent liquid or solid embolization agents.<sup>6</sup>

The aim of this rare case report of extensive arteriovenous malformation in a pediatric patient's face is to show the importance of a precise diagnosis obtained by imaging exams that assisted the choice of treatment, proving an excellent long-term clinical result.

## Case report

A female child, 9 years old, was referred to the oral and maxillofacial surgery service at Baleia Hospital – Belo Horizonte – Brazil, complaining of painless swelling in the chin region. The mother reported that soon after the child's birth a small purplish spot appeared in the chin. Evolution was slow, progressive and associated with recurrent bleeding episodes, sometimes spontaneous. Facial asymmetry and concern about the possibility of bleeding limited the patient's social life and practice of physical activity.

The extra oral clinical examination showed an increased volume of firm consistency, with well-defined margins and blue-purplish coloration on mental region. Blood vessels superficialization was clinically suggested (Fig. 1). It was possible to observe, during intraoral clinical examination, that the lesion extended to the right side of the rear region of the mouth. The anterior lower teeth were crowded and lingualized (Fig. 2).

The intravascular puncture aspiration by an intrabuccal access, revealed the presence of pulsatile blood content. Initially, an ultrasonography was requested to evaluate the characteristics of the soft tissue volume, the distribution of vascular structures and the intensity of blood flow present in the lesion. The ultrasound examination showed an intense arteriovenous flow suggesting the hypothesis of hemangioma.

There was a suspicion of bone tissue involvement due to dental misalignment. Then, we decided not to use resonance imaging exam due to its limitation in analyzing the hard tissues, if compared to the benefits of CT scan with contrast. The CT scan identified a small calcification in the soft tissue, with no bone continuity circumscribed by an intense vascularization area suggesting a phlebolith (Fig. 3).



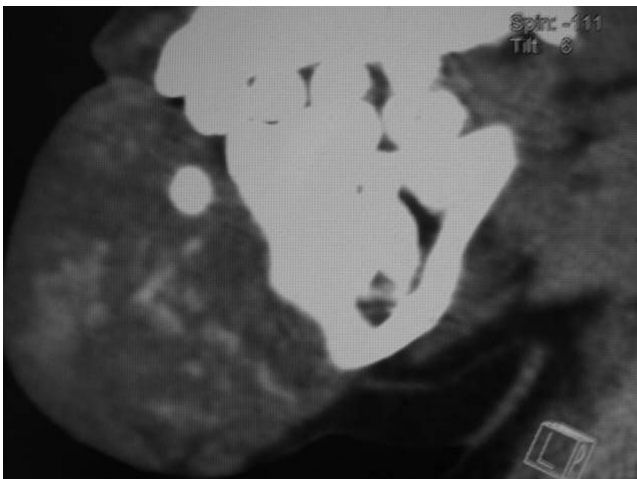
**Fig. 2 – Initial photography (intraoral dental malposition).**

After local anesthesia (2% mepivacaine hydrochloride with 1:20.000 corbadrine, DFL®, Rio de Janeiro, Brazil) an intravascular 1cc ethanolamine oleate diluted in 4 cc of distilled water was injected, in the extra-oral lesion, and another 1 cc of the solution with the same concentration was injected in the intraoral region. The same regimen was repeated at regular intervals of 7 days. This drug, when injected into the vessel produces a dose-dependent inflammatory response, resulting in vessel wall fibrosis with possible occlusion of the same.

During the seven weeks we observed a continuous lesion regression. After six sessions a significant decrease of the lesion was observed and, the aspiration of blood content was negative, suggesting fibrosis of the lesion.

Nimesulide were administered at a dosage of 100 mg of 12 in 12 h, for three days, after each application aiming the reduction of the swelling and the healing time of the area that received the injection.

Phlebolith and fibrosis were surgically removed with no major bleeding. The surgical procedure was performed under local anesthesia, using an intraoral access to the chin. The



**Fig. 3 – Initial CT scan with contrast (vascularization and phlebolith).**



**Fig. 4 – Extraoral photography after a 3-year follow-up period (correction of facial deformity).**

patient was treated with Nimesulide at a dosage of 100 mg, twice daily, during three days. We recommended icepack on chin region in the first 72 h and maintenance of extra-oral pressure dressing for two days to prevent swelling.

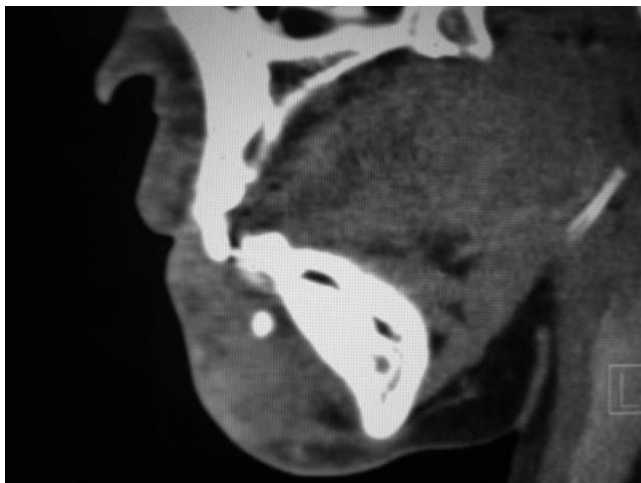
After a 3-year follow-up period, facial asymmetry was significantly decreased, there was no episode of spontaneous bleeding or disturbance of teeth alignment (Figs. 4 and 5). A new CT scan (Fig. 6) showed less local vascularity as well as decreased calcification dimensions.

## Discussion and conclusion

The history of vascular anomalies presents classifications that include overlap of terms. This diagnostic complexity can produce sequels to patients because of misguided choices of treatment. Vascular tumors present distinctive clinical characteristics that are generally the contrary to malformations. VMs are present since birth and never disappear; in addition,



**Fig. 5 – Intraoral photography after a 3-year follow-up period (natural alignment of teeth).**



**Fig. 6 – CT scan with contrast after the last session of sclerotherapy (involution of vascularization and phlebolith).**

they may increase in size over the years as it was in this case report.<sup>7</sup>

VMs may be associated with the development of the skeleton, presenting extra or intra osseous location, and cause some bone changes. In this case, the extra osseous lesion did not cause changes in the size or density of the jaw. However, we observed a change in the contour of the alveolar ridge caused by the expansion of soft tissue injury. Although VMs may involve an extensive facial deformity, the hazard is a possible abundant and often, uncontrollable bleeding, mainly in arterial malformations.<sup>8</sup> They can be classified as high or low blood flow. The former are relatively uncommon. Although they generally happen randomly, they may have a familial characteristic and be genetically determined.<sup>9</sup>

The nature of vascular channels, capillary, arterial, venous or lymphatic, may also be categorized vascular malformations. It is common the coexistence of many vessels types in the same lesion.<sup>10</sup> This situation has been observed in the clinical case presented, confirmed by the ultrasound exam.

The proper diagnosis of these malformations should be initiated from an early history interview and a detailed clinical examination of the lesion. Imaging exams such as ultrasound, MRI, CT scan with contrast and angiography are crucial.<sup>11</sup>

The ultrasonography presents advantages such as low cost, easy availability and it spares the patient's exposure to ionizing radiation. However, some limitations should be highlighted such as low resolution of the image that limits accurate identification of the lesion, especially in depth. Furthermore, with this exam is difficult to identify possible bone involvement.<sup>12</sup>

Magnetic resonance imaging exam has been used extensively for precise lesion delimitation and adjacent soft tissue association.<sup>7</sup> Magnetic resonance imaging is effective both in discovering and as well monitoring vascular anomalies and can be considered a non-invasive method.<sup>13</sup>

Computed tomography and magnetic resonance imaging are recognized standard tools in oral and maxillofacial surgical practice. However, the CT scan with contrast is able to identify

bone disorders such as the presence of phleboliths, calcifications arising of the inflammatory processes in the veins.<sup>14</sup>

Angiography in turn, has limited indication. It is used generally to investigate the origin of blood supply and the dynamics of arteriovenous flow, especially in cases of endovascular interventions.<sup>12</sup>

Histopathological exam was not used for surgical planning given that definitive diagnosis is clinical, through imaging investigation. However, histology can contribute to confirm the nature of the vascular channels.<sup>7</sup> We emphasize that the procedure of biopsy may expose the patient to surgical risks with blood loss.

After reaching a consensus on a definitive diagnosis, the participation of a multi professional team was fundamental for the establishment of the best suited therapeutic treatment. If the choice is surgical therapy, attention to the size and shape of the lesion should be given. Surgery is more suitable in cases where the lesion is well defined and far from important anatomical structures.<sup>15</sup>

The age of the patient, the degree of partaken with surrounding tissue and the characteristics of that lesion, had led to a more conservative initial treatment. The non-surgical treatment is reported in the use of systemic corticosteroids, interferon-alpha, low intensity laser therapy, radiotherapy, sclerotherapy and trans arterial embolization, complemented or not by surgical treatment. Sclerotherapy associated to surgical treatment has been the most common treatment described.<sup>7,16-19</sup>

Sclerosing solutions can be divided into three categories: detergents, osmotic and chemical (composed).<sup>16</sup> In the case reported monoethanolamine oleate was used because it is considered safe and with a low rate of complications. This drug, when injected into the vessel produces a dose-dependent inflammatory response, resulting in vessel wall fibrosis with possible occlusion of the same. The sclerosing agent spreads through the vessel wall and causes an extra vascular inflammatory and a reactional fibrosis.<sup>21</sup>

Ethanolamine oleate sclerosing agent is composed of 4.23% weight/volume of oleic acid, 0.910% weight/volume of ethanolamine and excipients, namely benzyl alcohol and water for injection. Each ampoule has a volume of 2 cc, with individual dosage in vials of 0.5-2 cc; which must not exceed 6 cc. Its administration should be carried out slowly, avoiding touching the perivascular region, as it may result in ulceration or even tissue necrosis.<sup>22</sup> Ethanolamine oleate is contraindicated in patients with hypersensitivity to any component of the formula, as well as in the following circumstances: varicose veins, voluminous, ostial and valvular insufficiency; cutaneous lesion in the skin to be treated; acute infections; serious systemic diseases; arterial occlusive diseases; deep vein thrombosis; senile or non cooperative patients; pregnant women in the 1st and 3rd trimester or in lactation; stasis ulcer; acute phlebitis and severe edema.<sup>21</sup>

Literature presents many cases of complete resolution of signs and symptoms due to vascular lesion substitution by a fibrosis tissue. Ethanolamine oleate dose and frequency of injections depended on the site and dimension of the lesion.<sup>20,22-24</sup> Another case reports involving the region of

face and great extension were not found in the literature. The maximum dose that has been reported is 0.4ml/kg and fraction injections varying between 0.5 and 10cc per session.<sup>24</sup>

Different concentrations, between 1.5% and 2.5%, have reported equal clinical results around 95% of lesions regression. Therefore it is preferable to use the lower concentration (1.25%) to prevent complications such as kidney damage, reduction of fibrosis, pain, redness and heat.<sup>22,24</sup> In the case reported, we injected the sclerosing agent at 2.5% based on the lesion dimensions at its aggressive appearance. In addition, no complications were either observed in the perioperative or postoperative period.

Despite patients' present pain and edema after injections, they vary in intensity and usually last a short period. Generally it is a safe procedure and well tolerated by patients.<sup>23</sup> In the case described on this paper, the patient had no complications at any time, which allowed us to give the same dose that was chosen with the hematology team and also made us confident about choosing the frequency of applications found in the literature.

Finally, most of the studies on vascular anomalies are retrospective studies. Prospective long term investigations are needed to demonstrate the non-recurrence of these lesions. More specific information about preparation of ethanolamine oleate solution and standardization of the doses applied in each session are lacking.

The participation of a multidisciplinary team and the request a ultrasound exams and the CT scan with contrast allowed proper diagnosis and the correct choice of treatment plan in two stages. The dosage used and the interval between the applications of the sclerosing solutions have allowed the necessary fibrosis of the lesion, without any complication after the injections and/or during the soft tissue-plasty and removal of the phlebolith, therefore, it is an effective method for the treatment of extensive arteriovenous malformations in the face.

### Conflicts of interest

The authors have no conflicts of interest to declare.

### Ethical disclosures

**Protection of human and animal subjects.** The authors declare that no experiments were performed on humans or animals for this study.

**Confidentiality of data.** The authors declare that they have followed the protocols of their work center on the publication of patient data

**Right to privacy and informed consent.** The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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