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CASE REPORT

Mandibular Arteriovenous Malformation in an 8-year-old Child: A Case Report

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ABSTRACT

Arteriovenous (AV) malformation is a congenital vascular anomaly in which there is an abnormal connection between the arterial and venous system resulting from developmental arrest during embryogenesis. In children, they are rare and potentially life-threatening. In the present case, an 8-year-old male patient presented with a simple gingival swelling associated with mobility of the corresponding teeth, which appeared relatively simple to excise but had an AV malformation associated with it. Timely diagnosis and investigations revealed its presence and hence prevented a catastrophy from occurring. Conventional method of surgical ligation of the external carotid artery was done as an emergency procedure and surgical resection was done without compromising the associated anatomic structures. This procedure helped control the bleeding as well as achieved an uneventful healing. This case report intends to create an awareness among Pediatric Dentists regarding the condition and thereby preventing an uncontrolled and unmanageable hemorrhage occurring in the Dental Office.

Keywords: Arteriovenous malformation, Carotid artery, External, Hemorrhage.

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INTRODUCTION

Arteriovenous malformations (AVMs) are defects of the circulatory system that are generally believed to arise during embryonic or fetal development or soon after birth. They are composed of snarled tangles of arteries and veins. An AVM is rare, and unawareness of this condition can lead to catastrophic complications. While simple gingival bleeding is seldom taken seriously, massive, pulsating gingival hemorrhage is often dramatic and may be fatal. Precipitated by a simple dental extraction or more violent forms of trauma, control of massive hemorrhage may be difficult for the clinician, particularly when the underlying cause is unknown or involves multiple unlikely differential diagnoses. Mandibular AVMs are recognized as uncommon and potentially life-threatening vascular malformations. The female: male ratio is 2:1, with peak incidence in the second decade of life with a range of 3 months to 74 years. The head and neck region consists of 50% of vascular lesions with only a small percentage occurring in the jaws. The incidence is twice more common in the mandible than maxilla.[1]

EMBRYOGENESIS

In 1987, Yasargil[2] postulated that AV malformations might not be simple structural connection, but 'proliferative capillaropathy'. Several case reports suggested that these lesions are dynamic in nature. Mullan et al proposed that AVMs are formed at the 80-mm stage of embryogenesis. The vasculature consists of primitive vessel morphology, as there are a large number of vessels and an absence of functional architecture. Lasjaunias[2] concluded that large AVMs are formed by a mutation early in embryogenesis, and smaller AVMs by a later causative event.[2]

Pathology

AVMs are lesions that are defined by presence of arterio-venous shunting through a nidus of coiled and tortuous

vascular connections that connect feeding arteries to draining veins.

Histologically

Vascular structures retain the characteristic arterial and venous components, but no capillaries are seen between these two elements, creating direct arteriovenous shunting. Hypertrophy of the walls of both arterial and venous elements is seen. Microscopically, the elastic lamina of the arterial intimal layer is mostly intact, but might show some degradation or deficiencies. The thickened veins can be differentiated by their size and absence of elastic staining. The arterial and venous components show hyperplasia of the smooth-muscle cells in the tunica media. If hemorrhage has occurred, the surrounding parenchyma will have evidence of gliosis and hemosiderin staining.[2]

Clinical Presentation

These lesions frequently appear as high-flow vascular malformations characterized by congenital dysmorphogenesis of the arterial and venous structures in the region involved. Arteriovenous malformations are mostly central lesions that can extend into soft tissues. Their clinical presentations vary from onset of minor gingival bleeding while brushing the teeth to dental loosening, lower lip numbness, facial deformity, malocclusion and even hemorrhagic shock following extraction of teeth.[3]

Radiographic Presentation

The radiographic appearances of these lesions are variable, ranging from a small radiolucency to a markedly obvious osseous erosion of the alveolus with apparently floating teeth. Computed tomography (CT) scans and magnetic resonance imaging (MRI) are helpful imaging tools to assess the extent of the lesions into the bone, soft tissue and major vessels.

A confirmatory diagnostic tool to identify the malformation and the feeding vessels is selective angiography. This radiographic technique monitors blood flow by placing dye into vascular system *via* a catheter near the area of the suspected malformation.[4]

Although AVMs are rare entities, the dentist must consider the possibility and recognize the clinical signs in order to propose proper treatment.[5] We report the case of a benign mandibular gingival lesion that warranted an aggressive approach due to potentially fatal exsanguination.

Case Report

An 8-year-old boy was referred to our hospital in April 2014 by a general dental practitioner for evaluation of an unusual mass of tissue in oral cavity. The boy was appearing weak and pale. The father complained of a gingival swelling in relation to lower left back tooth, which appeared 15 days back and recently inflated, and in spite of being painless, was bleeding spontaneously. No significant and relevant medical history was reported.

Extraorally, no gross asymmetry or abnormality was noted. Intraorally, upon inspection, enlargement of gingiva in relation to lower left first permanent molar was noted, which was extending anteriorly up to distal aspect of lower primary second molar and posteriorly up to the anterior border of ramus of mandible. It was reddish in color with corrugated surface and was bleeding spontaneously (Fig. 1). Upon palpation, it was soft and compressible. Left permanent first molar was noncarious with Grade II mobility and compressible in socket. Patient's oral hygiene was satisfactory. The concerned tooth had no plaque or calculus deposits.

Orthopantomogram revealed the presence of bone loss on mesial aspect of left permanent first molar; the root development of left permanent first molar was delayed as compared with that of right permanent first molar and mesial displacement of tooth bud of left permanent second premolar (Fig. 2). The differential diagnoses proposed included hemangioma, central giant cell granuloma, Langerhans's cell histiocytosis, Ewing's sarcoma. For further evaluation, fine-needle aspiration cytology (FNAC) and blood investigations were advised. FNAC revealed the presence of dark red blood, which increased suspicion toward hemangioma. Blood investigations revealed bleeding time, clotting time, white blood cell (WBC), red blood cell (RBC) and platelet counts to be normal, but hemoglobin level was only 9 gm%. After considering the clinical features, radiographic appearance and FNAC, the lesion was

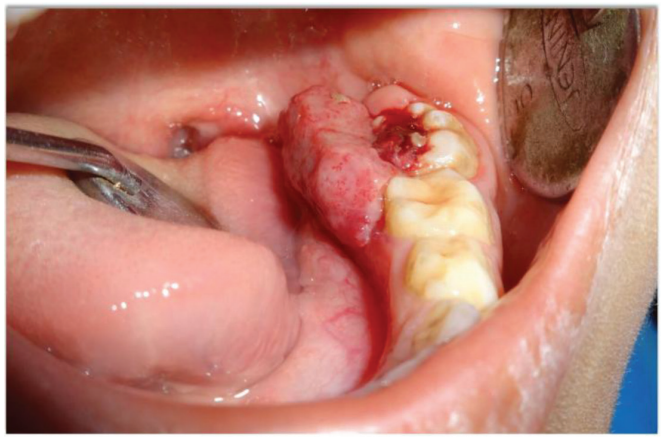

Fig. 1: Intraoral view of the lingual gingival swelling extending from the anterior border of the ramus to the distal of mandibular left primary second molar

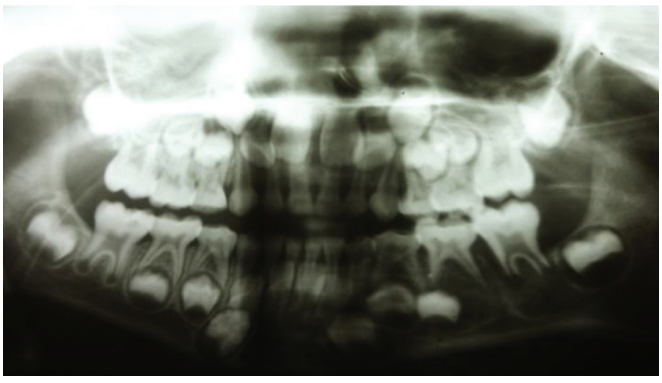

Fig. 2: Orthopantamogram revealing the mesially displaced left mandibular second premolar bud and reduced root growth of the first mandibular molar

suspected to be of vascular origin and the patient was referred for advanced diagnostic investigations such as computed tomography and angiography. The CT and angiography reports were indicative of AVM (Figs 3 and 4). CT scan showed the extent of the lesion, mostly on the lingual aspect, as well as the presence of an artery arising from anterior aspect of external carotid artery that on running antero-inferiorly, was acting as a feeder artery to the anomalous vessels in the affected area and the venous drainage was from anterior division of retromandibular vein and common facial vein draining into the internal jugular vein. The angiography was representative of expansion of the bony cortex by virtue of the remodeling of bone due to the lesion.

Thus, a final diagnosis could be reached, stating the condition to be AVM. A definitive surgery was scheduled. After placing the submandibular incision and raising the flap, external carotid artery that was acting as feeder artery and anterior division of retromandibular vein and common facial vein just before draining into the

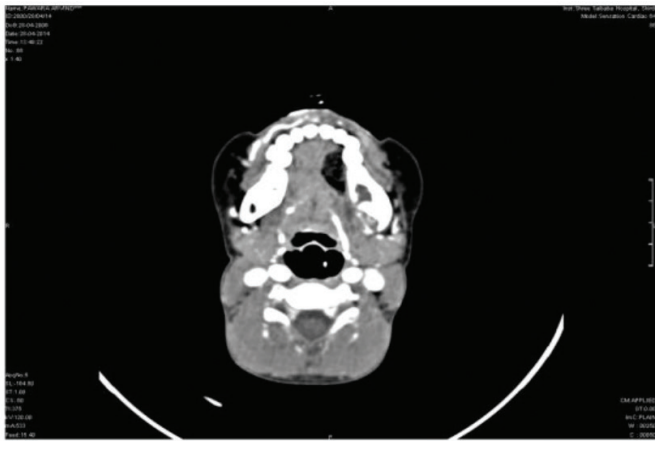

Fig. 3: Multislice computed tomography scan showing well-defined, expansile, osteolytic lesion due to tangled mass of vessels in arterial phase in the body of the mandible in lower left last molar tooth, representing intraosseous AV malformation of mandible

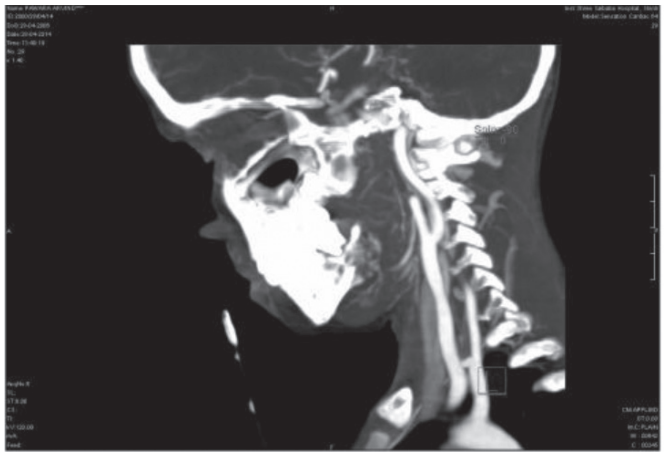

Fig. 4: Computed tomography angiography image showing the anterior branch of left external carotid artery as feeder artery and anterior division of retromandibular veins and commom facial vein draining into internal jugular vein

internal jugular vein were identified and ligated during the procedure. Followed by this, during the intraoral subsequent operatory session, ostectomy of the complete lesion involving left permanent first molar as well as tooth bud of left permanent second molar had to be performed, keeping the safe margins to prevent recurrence. Finally, intraoral and extraoral surgical sites were sutured. The patient was kept on antibiotic regimen and under observation. After 5 days, the patient was discharged once his condition was stable.

A follow-up examination after 6 months showed the boy to be healthy, with no sign of pallor or any weakness. Intraorally, the operated area was appearing to be normal and completely healed (Fig. 5). No episode of recurrence of bleeding or any other abnormal finding was reported ever since the surgery was performed. Nevertheless, the patient has been kept on regular follow-up, as this condition is known to have a high rate of recurrence.

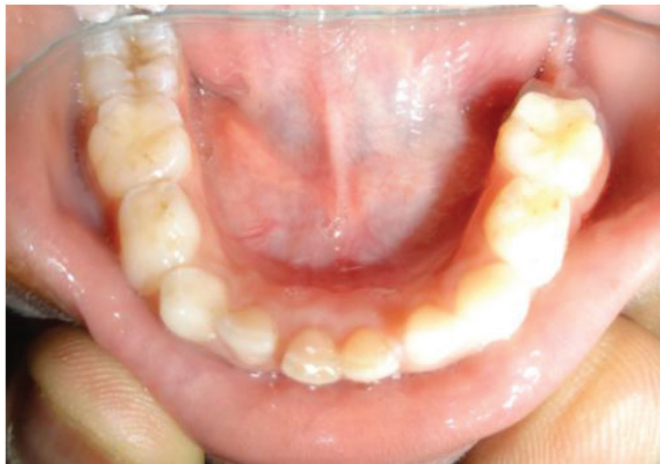

Fig. 5: Postoperative healing after 5 months

DISCUSSION

Vascular malformations are abnormal communications lined by quiescent endothelium. Although these conditions are congenital in nature, they show an additional growth when disturbed by trauma, infections or endocrine fluctuations. In the present case, the child was asymptomatic until 8 years after which a gingival swelling was observed. As reported by Deepa et al,[6] the mean age of presentation of AVM is 19 years with an equal sex predilection and 31% presenting in the maxillofacial region.[6] AVMs of the dental arches occur primarily in childhood. Clinically, they are observed after extractions, biopsy procedures or while brushing.

In the present case, provisionally the condition was diagnosed as hemangioma. Several differential diagnoses including cyst, ameloblastic fibroma, giant cell granuloma, fibrous dysplasia and malignant tumors might be considered.[4] As the FNAC showed the presence of frank blood, bleeding was expected intraoperatively. As a precautionary measure, blood was arranged. The bleeding could be immediately brought under control once the ligation of the vessels and excision of the lesion was completed. There was an uncontrolled hemorrhagic episode consistent with the findings of Benndorf et al.[7]

Early diagnosis is paramount in the management of AVM. Conventional radiographic methods present the lesion as ill-defined radiolucent with a honey-comb or soap-bubble appearance, with the displacement of the adjacent teeth or resorption of the teeth corresponding with the lesion. Consistent with this finding, the present case shows an irregular radiolucency with displacement of the premolar and reduced root growth of the first molar. Ultrasound technique can be used to detect high flow lesions, but it fails to differentiate between AVM and other vascular lesions. The most sensitive test to diagnose AVM is magnetic resonance angiography (MRA), which is less invasive and safer procedure.[6] Although CT, MRI and MRA may localize the arteriovenous shunt lesion, superselective arteriography remains an essential tool for diagnosis and planning of treatment.[8-13]

In children, the expected treatment outcome is the elimination of the vascular latticework with somatic regeneration to replace the vascular anomaly. In the present case, surgical ligation of the external carotid artery and anterior division of retromandibular vein and common facial vein was undertaken that is considered to be one of the conventional methods of managing the patients with AVM. Considering the recent diagnostic trends and for the better understanding of the pathophysiology of vascular lesions, newer treatment modalities, such as embolization of the feeder vessels, have emerged. Super-selective embolization with occluding materials such as cyanoacrylate is a recent technique for the management of AVM, but this technique demands highly sophisticated armamentarium and an interventional cardiologist, and after the embolization of the feeder vessels, the operative procedure should be performed within 24 to 48 hours to prevent the development of collaterals.[14] Although a conventional approach of ligation of vessels followed by ostectomy of the entire lesion involving teeth no 18 and 19 was considered in the present case, there has been no recurrence of the lesion and the patient was in a stable condition for the 6 months postoperative phase. The surgical correction involved removal of the entire lesion without compromising the facial structures that has been very beneficial in children, who have a lot of growth left in them.

Mandibular AVMs are difficult to treat and the methods are not well defined. The goal of the treatment is to stop the hemorrhage and prevent its recurrence. As these hemorrhages often occur in children, treatment should be as conservative as possible and aid in the growth and maintain the functional role of the mandible. Although many treatment modalities are proposed, ligature of the external carotid artery is now completely proscribed, as it neither treats the AVM nor prevents recurrence of bleeding but increases the difficulty of future treatment. Complete resection after partial embolization is the safest treatment, but it may induce major functional damage to mandibular growth.

Surgical curettage after preoperative embolization has also been proposed. It is a more conservative treatment modality, but its long-term efficacy is tentative and has the risk of recurrence. Superselective arterial embolization with particles or glue has been reported. Embolization with glue seems more effective than with particles.[15,16]

When embolization fails or the access to the AVM nidus is not possible, surgery is indicated. Due to the lack of well-defined margins, high vascularity and presence of major structures, surgery is difficult. Hence, the surgery must be performed by an experienced surgeon with the ability for reconstructive surgery. Resection of the entire nidus is the goal of the surgery. The nidus may be difficult to define because of diffuse feeder vessels and draining veins. Although ligation of major arteries is not the proposed treatment option, it can be helpful in life-threatening conditions. Lasers are less effective and rarely uesd.[17]

CONCLUSION

- In the present case, conventional technique of surgical ligation of the external carotid artery and draining

veins was carried out resulting in complete regression of the lesion without development of collateral anastomosis.

- With limited hospital resources and considering economic condition of the patient, conventional technique can provide equally good results.
- The presence of life-threatening conditions can surface in the treatment of routine dental procedures in children; an awareness of these conditions can prevent major mishaps from occurring in the Dental Office.

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