Case Report

Pediatric Intraoral High-flow Arteriovenous Malformation: A Diagnostic Challenge

Roy Petel, DMD[1] • Malka Ashkenazi, DMD[2]

Abstract: *Arteriovenous malformations (AVMs) are rarely reported in the dental pediatric literature. They may develop adjacent to primary molars and can be life-threatening due to their potential for massive bleeding. The most common symptom associated with documented cases of AVMs is spontaneous gingival bleeding. Other clinical signs include pain, erythematous gingiva, resorption and mobility of teeth, soft tissue discoloration, facial swelling, and asymmetry. Radiographically, AVMs are osteolytic lesions. The purpose of this report was to describe the challenge of diagnosis of a high-flow arteriovenous malformation located in the primary maxillary molar region, which was misdiagnosed as a dentoalveolar abscess adjacent to previously treated primary molars. A decision to extract a tooth with gingival swelling and associated spontaneous bleeding should be made after the differential diagnosis of a vascular malformation has been ruled out. (Pediatr Dent 2014;36: 425-8) Received November 5, 2013 | Last Revision January 30, 2014 | Accepted January 31, 2014*

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Arteriovenous malformations (**AVMs**) of the head and neck present abnormal connections between arteries and veins that do not pass through a capillary network. AVMs result from developmental arrest of blood vessels after the endothelial stage of embryologic vascular development, in contrast to hemangiomas, which appear to be a failure of differentiation at the endothelial stage.[1-8] AVMs are classified according to their hemodynamic features (i.e., high-flow versus low-flow lesions); such features can serve as indicators for appropriate therapy.[9] High-flow lesions, which include arterial anomalies, tend to cause more destructive skeletal changes in the head and neck area than do low-flow lesions.[2] The latter include capillary, lymphatic, and venous malformations.[2,7,8,10]

AVMs are uncommon malformations, with the majority located in the mid-face. Only five percent are located in the maxilla-mandibular regions,[11] and they occur two times more frequently in the mandible than the maxilla.[6] AVMs are present at birth, may enlarge near puberty, and may be unnoticeable until later in life.[8] Most lesions are detected between ages 10 and 20 years.[5] Trauma, surgery, infection, and endocrine changes appear to contribute to their expansion.[12] No genetic transmission patterns for AVMs are known; however, they are more commonly diagnosed in Caucasians than Hispanics.[1]

The most common signs of AVM's presence in the maxilla-mandibular regions are spontaneous gingival bleeding, swelling, and loosening of teeth.[13-16] Other clinical signs and symptoms may include: pain; erythematous gingiva; root resorption; mobility of teeth; displaced unerupted teeth[9,17]; widened periodontal ligament spaces; and persistent or recurrent oral infections.[9] Soft tissue discoloration, angiomatous discoloration,[17] facial swelling, and facial growth asymmetry present in some patients.[3,18,19] Pulsations can be detected in large lesions.[20] The intensity of pain associated with these lesions ranges from no or tender pain to pulsating or throbbing pain.[16,21] Numbness has also been reported.[16,22]

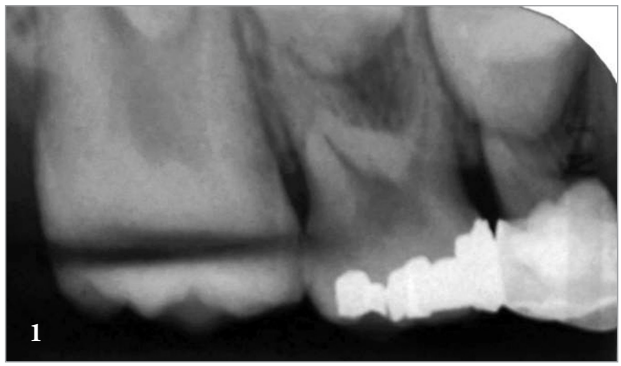
Radiographically, AVMs appear as varying-sized osteolytic lesions with a "soap bubble" or "sun ray" appearance,[23] often with indistinct borders.[3,6,9] In the mandible, the lytic lesion is often confluent and associated with an enlarged inferior alveolar nerve canal.[3,17] Also, root resorption may create the radiographic appearance of teeth floating within the adjacent area of the osteolytic lesion.[24] The varied radiographic manifestations, together with the rarity of these lesions in the jaws, make the diagnosis of AVMs very difficult.

The most severe hazard of AVMs is their potential for uncontrollable bleeding when performing dental procedures such as tooth extraction,[7,9,10,24-27] biopsy, or incision of a suspected cyst,[10,28,29] or during the natural exfoliation of a primary tooth.[3] AVMs are sometimes discovered when a tooth is extracted.[27] Nevertheless, spontaneous bleeding can also be a hazard.[4,22,30] A facial vascular malformation may present as a dental emergency that may result in fatal or life-threatening and disfiguring situations.[10,16,26,31]

The purpose of this report was to review the literature and describe the challenges associated with the diagnosis of an arteriovenous malformation, especially during the mixed dentition period.

Case report. A seven-year-old Caucasian female was admitted to a public voluntary dental clinic [Dental Volunteers for Israel, Jerusalem, Israel] because of a sensitive gingival swelling with occasional excessive and spontaneous bleeding in the primary maxillary right molar area, which had persisted for approximately three years. A clinical intraoral examination revealed a solitary oval swollen and bleeding lesion measuring one by one centimeter, located between the primary maxillary right first and second molars. The swelling was soft in consistency with a smooth surface. Clinically and radiographically, the primary maxillary right second molar had an intact proximal amalgam restoration, and the primary maxillary right first molar was treated previously by pulpotomy and a preformed crown (Figure 1).

[1]*Dr. Petel is a pediatric dentist, Department of Pediatric Dentistry, School of Dental Medicine, The Hebrew University of Jerusalem, Jerusalem, Israel; and* [2]*Dr. Ashkenazi is a senior lecturer in pediatric dentistry and pediatric dentist in private practice, Petach-Tikva, Israel.*
Correspond with Dr. Petel at roy@dental-dvi.org.il





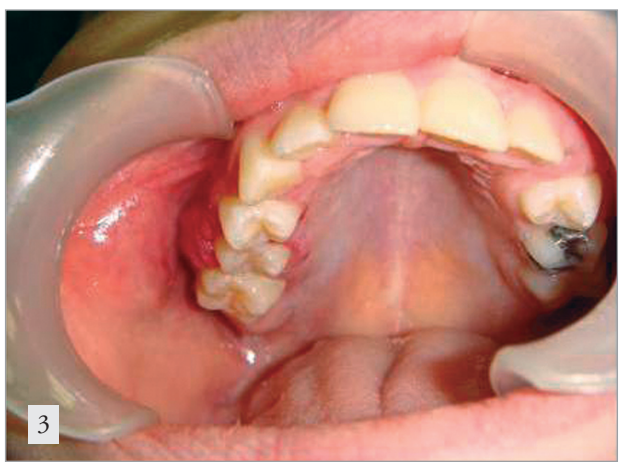




Figure 1. Periapical radiograph of the lesion at the first appointment, when the patient was seven years old, involving the primary maxillary molars.

Figure 2. Preoperative extraoral picture, taken two years later at age nine years, showing asymmetry in the right maxillary area.

Figure 3. Clinical intraoral appearance, at age nine years, of the arteriovenous malformations lesion in the maxillary right extending from the permanent second premolar and permanent first molar.

Figure 4. Preoperative periapical radiograph showing a radiolucent lesion extending from permanent maxillary canine to the permanent maxillary first molar, with disappearance of the lamina dura of the permanent maxillary canine and premolars.

To further investigate the etiology of the swelling, the pediatric dentist accessed the pulp chamber of the primary second molar to evaluate its clinical condition and found that it looked normal regarding color and amount of bleeding. Therefore, a pulpotomy was performed and the child was scheduled for a re-evaluation the following week. When no improvement was observed concerning the gingival swelling, the dentist referred the patient for an oral pathology consultation. The differential diagnosis was a pyogenic granuloma of unknown etiology. A senior specialist in oral medicine diagnosed the swelling as a side-effect of a thumb-sucking habit and recommended a follow-up with no further intervention.

Two years later, the girl returned to the clinic with a chief complaint of nonpainful swelling associated with intermittent bleeding on the right side of the maxilla. Her mother claimed that the lesion had continued to grow since the previous appointment and even after the natural exfoliation of the associated primary maxillary molars. During this period, the girl often suffered from bleeding nocturnally and during sneezing. The girl was so embarrassed from the bloodstains on her sheets that she laundered them herself. She suffered from anemia and several fainting episodes. She was treated by oral ferric iron without significant improvement of the anemia.

A clinical extraoral examination revealed face asymmetry in conjunction with a purple discoloration of the right cheek (Figure 2). No palpable lymph nodes, bone expansion, or soft tissue were noted.

An intraoral examination revealed a purplish swollen and bleeding lesion with three oval-round lobules on the buccal aspect of the maxillary right jaw extending from the permanent maxillary second premolar to the permanent maxillary first molar (two by four centimeters in size; Figure 3). The swelling was soft, with a smooth surface. The associated teeth were intact, without any carious lesions. A periapical radiograph revealed a diffuse radiolucent lesion extending from the permanent maxillary canine to the permanent maxillary first molar with disappearance of the lamina dura of the permanent maxillary canine and premolars (Figure 4). A panoramic radiograph revealed a large osteodestructive lesion with a well-defined, multilocular radiolucency without corticated borders in the

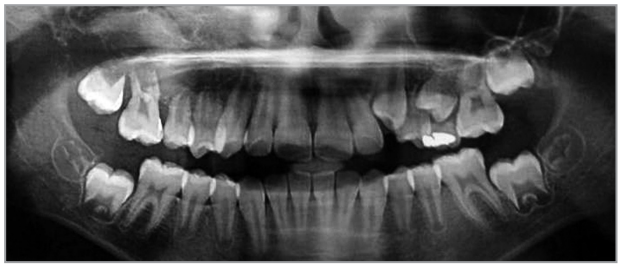


Figure 5. Preoperative panoramic radiograph showing a big osteodestructive lesion with well-defined multilocular radiolucency without corticated borders in the right side of the maxilla, extending from the permanent maxillary canine to the permanent maxillary second premolar.

right side of the maxilla, extending from the permanent maxillary canine to the permanent maxillary second premolar (Figure 5).

To further clarify the diagnosis, the girl was referred for consultation to the oral surgery department, Hadassah Medical Center. To ensure patient follow-up and to verify final diagnosis, further treatment in the voluntary public clinic was contingent upon performing this consultation.

A computerized tomography (**CT**) scan of the facial bones revealed a bony, destructive, mass-like lesion arising from the right maxilla, measuring 2.2 x 2.5 x 2.6 cm. Accordingly, the differential diagnosis included a large odontogenic cyst and keratocyst. However, other pathological possibilities, including malignancy (although unlikely), were not ruled out. On fine needle aspiration, fresh blood was obtained, and the patient was consequently referred for further evaluation. Magnetic resonance arteriography (**MRA**) of the right jaw revealed an AVM with a high blood flow. The patient was referred for a catheterization under general anesthesia, which confirmed the diagnosis (Figure 6). The treatment consisted of five consecutive embolizations, which resulted in a significant reduction in spontaneous bleeding.

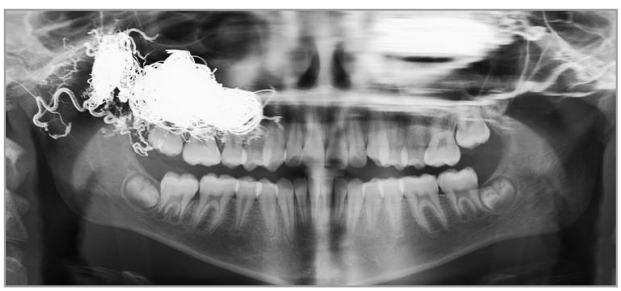


Figure 6. Postoperative panoramic radiograph showing the coils of the AVM at the area of surgery in the right maxilla.

Discussion

Although rare, AVMs of the jaws were reported in more than 200 articles in the medical literature and in fewer than 50 cases in the dental literature. In most of the reported cases, the lesions were detected during childhood and adolescence,[5,27,32] following uncontrolled bleeding and even exsanguination, as a result of primary tooth extraction, a routine procedure performed by a pediatric dentist.[11,13,15,25,33-35] Only three reports were published in the pediatric dental literature during the last three decades.[3,27,32] Moreover, these lesions can easily be misdiagnosed due to their close proximity to the periapical or furcation areas of primary teeth, corresponding permanent tooth buds, and radiographic appearance of resorbed roots.

This case report highlights the complexity of diagnosing AVM lesions located adjacent to treated primary maxillary molars and the difficulty in differentiating between infectious periapical lesions and AVMs. The features of the AVM described in the present case were similar clinically and radiographically to a swelling associated with dental infection near a deep restoration or a pulpotomized primary molar. The key difference between the two is the presence of spontaneous gingival bleeding from an AVM lesion.

Therefore, when evaluating a radiolucent lesion with a clinical manifestation of swelling and spontaneous bleeding, the first step is to rule out a vascular anomaly before any intervention. Secondly, it is important to determine whether the lesion is a tumor or malformation. In most cases, differentiation between the two can be achieved by obtaining a proper history and physically examining the patient.

With the onset of clinical manifestations, intraoral radiographs are an invaluable tool in depicting fine details and are indicated for the examination of most lesions of the dentoalveolar complex and other radiolucent areas of AVM cases.[9,36]

Another diagnostic aid for the evaluation of an AVM is a positive fine needle aspiration that is performed under direct observation, although this may not distinguish between a high- and low-flow lesion. This technique is safe and efficient but usually performed by a specialist. The use of a small gauge (22-25) needle reduces the possibility of fatal or profuse bleeding reported with biopsies. Local anesthesia of the surrounding area of the lesion is optional."[37]

CT and angiography are imaging modalities frequently used to confirm the presence of AVM lesions[38,39] and are helpful in differentiating the type of vascular malformation or the extent of bone destruction. With angiography, it is possible to determine which blood vessels supply blood to the lesion, the characteristics of the relative venous outflow, and the presence or absence of arteriovenous shunts, which are important for determining the appropriate embolization techniques.[19]

Magnetic resonance imaging (**MRI**) is another valuable technique used in cases of AVM, with signal voids sometimes confirming the high-flow nature of the lesions. Another advantage of MRI or MRA is the absence of ionizing radiation, which is a particularly important consideration in pediatric patients. The ultimate decision regarding the use of radiographic techniques, however, must be based on the expected benefit/risk ratio, not solely on the risk of exposure to ionizing radiation.

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