Surgical Management of Capillary Malformation-Arteriovenous Malformation (CM-AVM) Syndrome of the left mandible and RASA1 mutation in a Pediatric Patient

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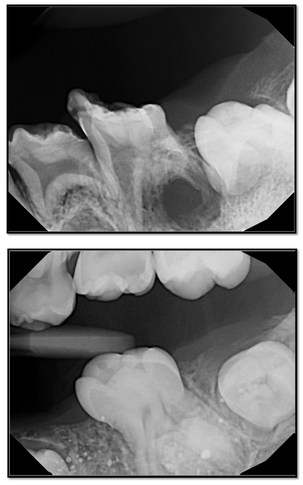
BACKGROUND

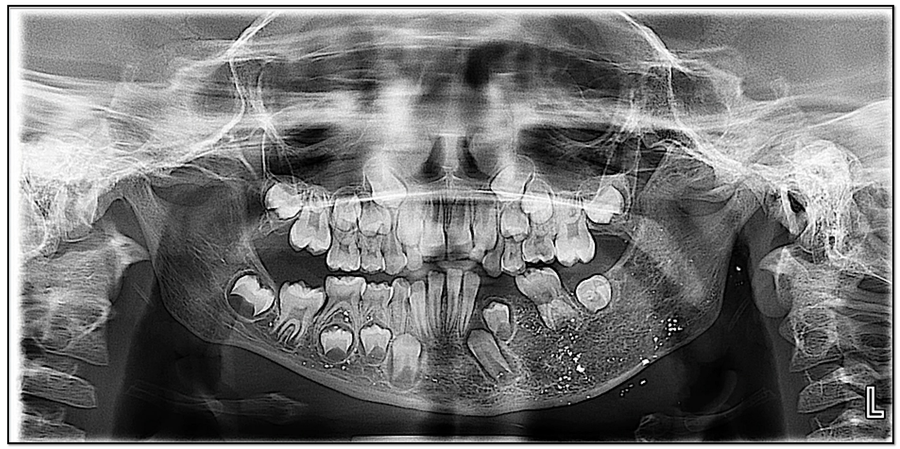
Intraosseous Arteriovenous Malformation in the mandible is a rare congenital vascular anomaly associated with abnormal connection between arteries and veins. Capillary Malformation-Arteriovenous Malformation (CM-AVM) is caused by a mutation of the RAS p21 protein activator 1 (RASA-1) gene, which encodes the protein p120-RasGAP vital for regulating cell proliferation and differentiation . The result of this mutation is the formation of a highly vascularized, irregular, tangled capillary network that is prone to rupture and bleed. Intraosseous CM-AVM causes expansion of the mandible and excessive bleeding post brushing, minor trauma and during surgical intervention. Iatrogenic injuries such as traumatic tooth extractions can have a sequalae of severe bleeding and possibly death which have been reported post extractions due to CM-AVM involvement in the mandible.

CLINICAL PRESENTATION

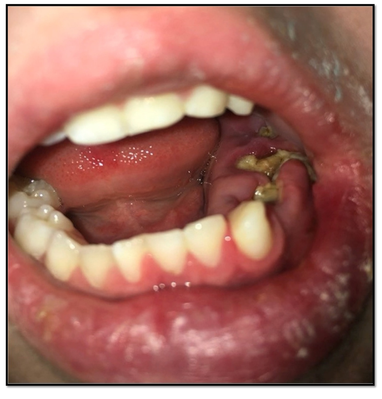
A 3-year-old female presented to Riley Children's Hospital outpatient pediatric dentistry clinic as a referral from the departments of otolaryngology and interventional radiology for evaluation and assessment of her dentition as it related to the malformation. Her medical history includes CM-AVM and RASA-1 mutation, learning delay and breathing problems (non-specific). She had no current medications and no known drug allergies. Upon evaluation, extraoral findings consisted of multiple red hemangiomas present on the lower face and chin, along with slight facial asymmetry and swelling on the left side consistent with the AVM of the mandible. Intraoral findings consisted of a vascular malformation present on the left dorsal tongue displayed as 3cmx3cm red patches. Maxillary teeth and soft tissue were within normal limits. Osteomyelitis of the left jaw secondary to necrotic #K and #L present. #K and #L were severely malformed, hypoplastic and presented with class II mobility. Radiographic evaluation revealed furcal and apical radiolucencies on both #K and #L. A treatment plan was composed for extractions of teeth #K and L under general anesthesia.

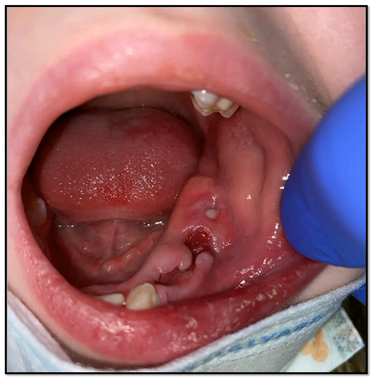
RADIOGRAPHS





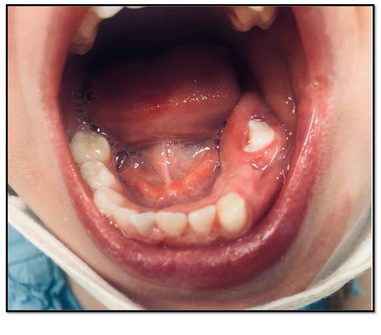
1-week Post op

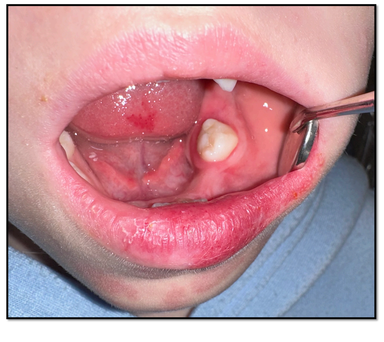




1-month Post op

6-month Post op

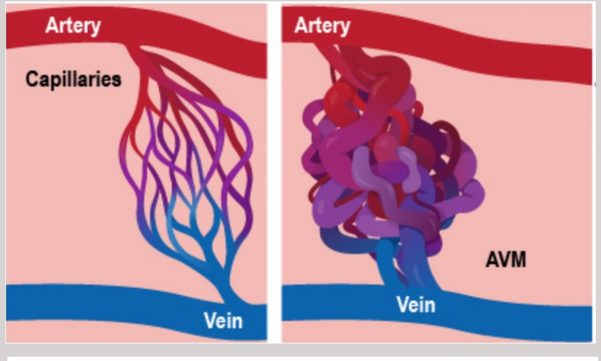




3-year Post op

TREATMENT/MANAGEMENT

After a 6.5-week course of IV Unasyn, the patient was brought to Riley Children's Hospital Day Surgery where dental treatment was completed under general anesthesia in conjunction with Neuro Interventional radiology. Teeth #K and #L were extracted with no complications. Immediately following the extractions, several interventions and injection of N-BCA glue (glue embolization) was used to control bleeding. The patient's blood loss post surgical procedure was approximately 200 ml and a blood transfusion was conducted during the procedure. The patient was then transported to the ICU intubated and admitted overnight for monitoring. The patient was evaluated one day post op, and the extraction sites were sound, and the bleeding had subsided. The patient was then cleared for extubation. During the patient's subsequent routine periodic follow up visits, the extraction sites were evaluated clinically and radiographically and exhibited normal healing. The patient continues to maintain 6 month recall intervals.



REFERENCES