Case Report

High-flow vascular malformations: literature review and case report

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Facial vascular malformations can cause dental emergencies that result in fatal or life-threatening and disfiguring situations.[1-6] Dentists must be aware of the clinical manifestations of these lesions in order to prevent iatrogenically related accidents and to minimize potential spontaneous crises for the patient. Effective treatment depends on the type and location of vascular lesion.

Literature review is difficult because of the variation in terminology of vascular lesions. Terms such as arteriovenous aneurysm, cavernous hemangioma, central hemangioma, pulsating hemangioma, angioma, cirsoid aneurysm, arteriovenous shunt, arteriovenous fistula, and arteriovenous malformations have been used synonymously with vascular malformation.[7,8] Prior to the 1980s, vascular lesions frequently were lumped under the term hemangiomas, which were considered hamartomas — congenital lesions comprised of normal tissues of faulty composition.[9] However, Glowacki and Mulliken[10] suggested that vascular lesions be divided into two major groups — hemangiomas and vascular malformations — based on histologic and clinical presentation. For example, hemangiomas are characterized histologically by endothelial hyperplasia and increased numbers of mast cells during their proliferation phase. During the involution stage, they are characterized by fibrosis, fatty deposits, and multilaminated basement membrane formation beneath the endothelium. The mast cell count during the involution stage is similar to normal tissue.[11]

Hemangiomas are frequently documented at birth or within the first 10 days of life.[12] Almost all show rapid clinical expansion during the first year of life[12,13] and frequently involute slowly during the next 5 years. Excellent cosmetic conditions result in approximately two-thirds of the patients if involution occurs by age 5 years.[12] Fewer than 2% require excision due to excessive proliferation. Rarely do hemangiomas involve the skeleton,[11] although some can cause distortion of a bone, (i.e. nasal deviation or depression of the outer cortex). Proptosis and optic nerve compression also can occur.[9]

A vascular malformation is a morphogenetic abnormality of blood and/or lymphatic vessels with normal ultrastructural characteristics and endothelial hyperplasia.[10] It results from a developmental arrest after the endothelial stage of embryologic vascular development — contrary to hemangiomas, which appear to be a failure of differentiation at the endothelial stage.[14]

Mulliken[15] states that vascular malformations are present at birth and may not clinically manifest until late infancy or childhood. However, patient histories in some case reports[7] suggest that vascular malformations also may be developmental. In contrast to hemangiomas, vascular malformations diagnosed at birth show no signs of involution with age and may expand near puberty.[16] Trauma, infection, or endocrine changes appear to contribute to their expansion.[13,14,17,18]

Vascular malformations frequently are associated with skeletal changes that can be intraosseous and can cause secondary changes of bone size, shape, or density.[19] Although vascular malformations can cause extensive facial deformity, the most severe hazard is potential profuse and uncontrollable bleeding with primary tooth exfoliation,[5,11] dental extraction,[2,6,20] incision of a suspected cyst,[5] or surgical removal.[21] Spontaneous bleeding is also a hazard.[13,14,22]

Subdivision of vascular malformations is based on the vessels involved, (i.e. capillary, venous, lymphatic, arterial). This classification simplifies older and confusing terminology.[10] Thus, a port-wine nevus is now considered a capillary malformation. The term "lymphangioma" is actually a lymphatic malformation and a "cavernous hemangioma" that fails to involute is considered a venous malformation. A more proper term for "arteriovenous fistula" is an arterial malformation with fistula.[19] Although the anomaly can consist of a single type of vessel, combinations also occur.[13,19]

The flow characteristics (low-flow vs. high-flow) of vascular malformation indicate their vascular nature with high-flow usually characteristic of arterial vessels.[11,19] High-flow lesions tend to cause more destruc-

tive skeletal changes in the head and neck area than low-flow lesions, perhaps as a result of the hemodynamic characteristics of the high-flow malformation.[19] The flow rate can be used as an indicator of the proper therapy.[5]

Approximately one-third of all the vascular malformations recorded in one vascular lesion registry are in the head and neck region.[12] Both intracranial (i.e. occipital, dural, auricular, cerebral, or spinomedullary) and extracranial (i.e. maxillofacial) lesions are reported.[4] Lesions occur primarily within soft tissue or osseous tissue.[19] Engle et al.[6] state that fewer than 100 maxillofacial arteriovascular malformations are reported.

The prevalence of vascular malformations is difficult to determine. Vascular birthmark registries may receive only the most troublesome or atypical cases.[12] However, vascular malformations comprise approximately 20% of 375 vascular lesions present in 297 pediatric patients recorded in one registry. The remainder are classified as hemangiomas. Other authors[19] report that more than 35% of the vascular lesions studied were vascular malformations. Males and females have equal predilection for a vascular malformation.[12] Caucasians are more likely to be diagnosed with a vascular formation than Hispanics.[12]

Clinically, patients present with symptoms of pain described as ranging from tender to pulsating or throbbing, although some patients report no pain.[1, 23] Other patients report neurosensory changes of the affected area such as numbness.[1, 22] Many patients have mobile teeth.[1] In some cases, a tooth may be extracted because the patient complains of pain and mobility.[1] Other patients will present on an emergent condition with spontaneous hemorrhage or cardiac arrest.[1, 4, 5, 20, 22] A bruit may be discernable[22] although some intraosseous lesions do not produce an audible bruit. Angiomatous discolorations are reported in some patients.[21] Asymmetric facial growth, widened periodontal ligament spaces, and persistent or recurrent oral infections also are reported.[24]

Vascular malformations on radiographs usually are noted as a lytic lesion with a "soap bubble" or "sun ray" appearance.[8] In the mandible, the lytic lesion often is confluent with an enlarged inferior alveolar canal and can replace the central marrow cavity.[21] Unerupted teeth can be displaced and root resorption is reported.[21, 24] An aspirational biopsy can be positive for blood under pressure although it may not indicate whether a high-flow or low-flow lesion is present. Use of a small-gauge needle reduces the possibility of fatal or profuse bleeding reported with biopsies.[2]

A necessary diagnostic tool used to identify the entire vascular malformation and its contributing vessels is selective angiography.[25] Angiography is a radiographic technique monitoring blood flow by placing dye into the vascular system via a catheter near the area of the suspected malformation. Bone densities are controlled to prevent masking of the vascular system.[14] To monitor a lesion in the head and neck area, a catheter usually is inserted into the femoral artery rather than the carotid artery, because this area is considered less traumatic.[14, 26] Angiography in children usually is performed under general anesthesia. Complications, such as hematomas and transient arterial spasms, are relatively rare and usually resolve spontaneously.[14] Bilateral angiography may be necessary to identify either bilateral lesions[21] or contributions from vessels crossing the midline to a unilateral lesion.[23]

Several treatment methods of vascular malformations have been recommended although the success of treatment varies depending on the area involved as well as the flow characteristics of the lesion. Sclerosants, (i.e. sodium morrhuate, boiling water, nitrogen mustard) have been used to manage low-flow lesions,[14] but they are ineffective in high-flow cases because of the rapid removal of the sclerosant from the malformation. Sclerosants also have limited efficacy because more severe and aggressive lesions frequently recur as a result of collateralization. However, they are used to control lesions in inaccessible areas or soft tissue.[24]

Surgical ligation of the external carotid artery may provide immediate relief of an emergent situation, but recurrence is a problem.[1] Ligation of the carotid arteries also prevents subsequent angiograms and embolization procedures.[21] Devascularization alone reportedly is effective in treating low-flow cases but not high-flow cases.[13]

Radical resection of the lesion can result in profuse bleeding and severe facial deformity.[13] Mandibular reconstruction with grafts of rib[13, 23] or iliac bone,[22] metallic prosthesis,[5] and replacement using the same mandibular bone[5, 6] following removal of the lesion are reported. However, infection is a major problem and scars are prominent.[5] Studies reporting the use of prostheses in growing children[5] do not show the long-term effects on growth and development of the jaws. Recurrence is also reported even in cases of radical resection.[5]

Embolization, a method of blocking contributing arteries, has been used since the 1930s to treat vascular malformations.[27] Materials that have been used for embolization include muscle,[27] polyvinyl alcohol particles (PVA), isobutyl cyanoacrylate, dura mater, Gelfoam™ (Upjohn, Kalamazoo, MI), and detachable latex balloons.[4] These materials usually are placed into contributing arteries through the catheter inserted for angiography. Emergent situations with high-flow lesions can be controlled by aggressive embolization.[4] Embolization also is used to manage cases where accessibility prevents surgical removal of a lesion.[4] However, embolization alone without surgical removal of the lesion is not consistently efficacious — particularly in high-flow cases — because of collateralization.[1, 4, 5, 13, 14] High-flow vascular malformations have a central area of low resistance vessels, which are supplied by numerous contributing arteries.[26] Blockage of the contributing vessels results in subsequent enlargement of

additional vessels to the area. Complications include tongue necrosis, hypothesia of nerves,[4] bradycardia,[28] complete facial paralysis,[14] and reflux of the embolizing material into the carotid artery.[28, 29] Embolization is used effectively prior to surgical removal of a high-flow vascular lesion to minimize blood loss.[14] Failure to use embolization prior to surgical removal of a high-flow vascular lesion can result in profuse and extensive bleeding.[21] However, multiple angiograms, embolizations, and surgeries may be required for some patients if the initial angiogram does not reveal all the contributions.[30] Percutaneous puncture and embolization of a mandibular vascular lesion directly through the mandible is reported, although spontaneous bleeding occurred several times prior to apparent remission of the lesion.[24]

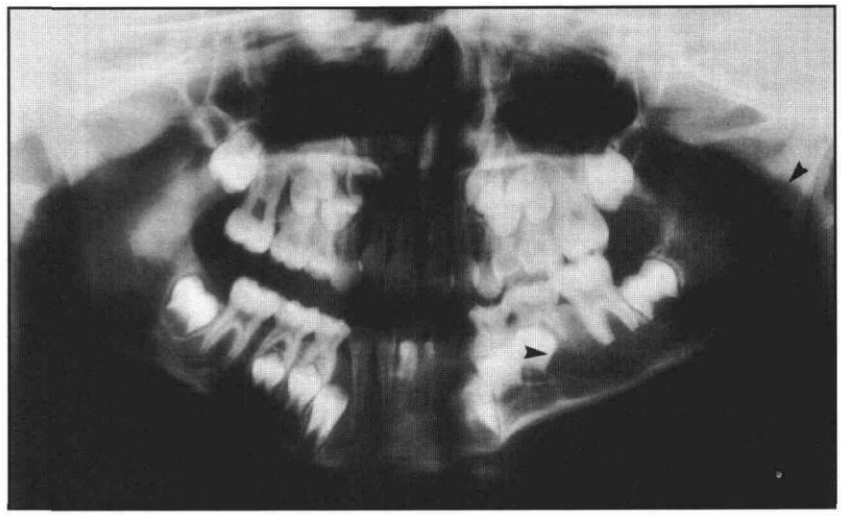

Fig 3. Preoperative panoramic radiograph showing radiolucency extending from mandibular left displaced second premolar (arrow) distal into ramus of mandible (arrow).

The following is a report of a successful clinical case where the diagnosis and treatment of a high-flow vascular lesion in a young child was made prior to any bleeding episode.

Case report

Clinical report

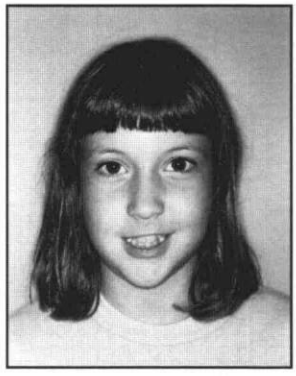

Fig 1. Preoperative frontal photo showing asymmetry in the mandibular area.

A 9-year-old Caucasian female reported "pressure" in the lower left quadrant during a routine 6-month dental examination. The parents' chief complaint was severe crowding and a need for orthodontic evaluation. Examination revealed extraoral asymmetry in the mandibular area (Fig 1), no palpable lymph nodes, no palpable expansion of bone or soft tissue, and no apparent erythema other than generalized gingivitis. Severe crowding with premature loss of the mandibular primary canines was present.

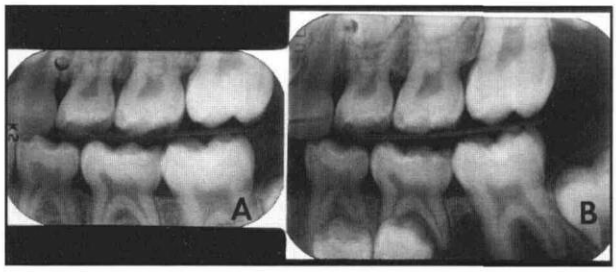

Fig 2. Bite-wing radiographs showing A) no apparent lesion one-year prior to the recall appointment and B) radiolucency apical to mandibular left first permanent and second primary molars and displaced second premolar.

Although a bite-wing radiograph (Fig 2a) exposed approximately 1 year prior of the patient's left dental arches was unremarkable, a bite-wing radiograph (Fig 2b) exposed at this appointment revealed a large radiolucency apical to the mandibular second primary molar and first permanent molar. The second premolar toothbud was displaced mesially. The contralateral bite-wing was unremarkable. A subsequent panoramic radiograph (Fig 3) showed a large radiolucency confluent with the left inferior alveolar canal extending from the unerupted second premolar into the ramus. An occlusal radiograph showed no apparent cortical expansion. Radiology consult suggested either an odontogenic cyst or a benign tumor.

Oral and maxillofacial surgery consult confirmed prior clinical findings. No bruit or thrill was found. Blood collected during an aspiration biopsy indicated a vascular lesion. Subsequently, an angiogram was scheduled under general anesthesia. The catheter was inserted via the femoral artery and passed through the carotid artery for selective internal maxillary artery injection and lingual facial trunk injection. Nonselective dye placement (Fig 4) showed a large vascular defect extending anteriorly under the second primary molar posteriorly into the ramus. Selective dye placement into the internal maxillary artery showed less extensive vascularization, but with a major defect in the area of the first permanent molar. Selective dye placement into the lingual facial trunk showed extensive contributions of both arteries into the area of the first permanent molar and extending anteriorly around the displaced bud of the second premolar and posteriorly into the ramus. The contribution of the facial and the lingual arteries overpowered the blood flow from the internal maxillary artery, causing turbulence and backwash of blood. The diagnosis was

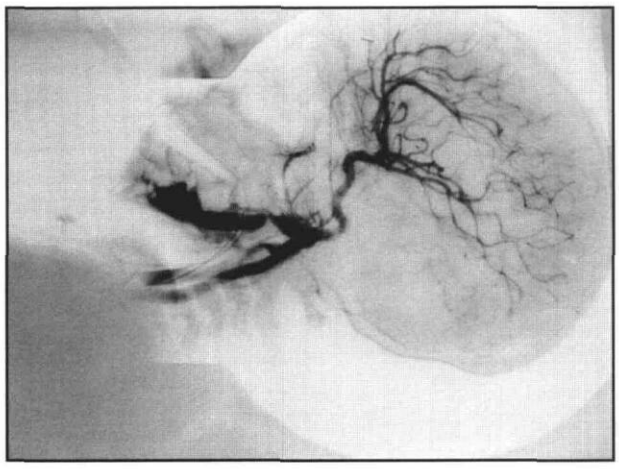

Fig 4. Preoperative nonselective angiogram showing *blood from carotid artery filling the vascular* malformation in mandible.

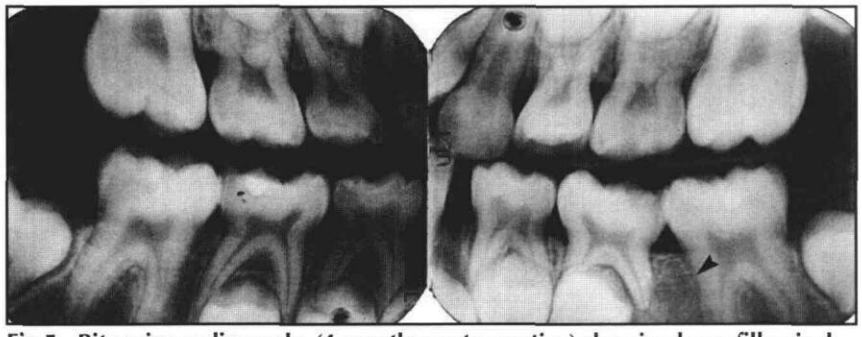

Fig 5. Bite-wing radiographs (4 months postoperative) showing bony fill apical to mandibular left first permanent and second primary molars (arrow).

a high-flow vascular malformation. CT scan demonstrated a lytic defect involving the left body of the mandible. Although the mandibular canal was widened, the outer and inner mandibular cortices were intact, with minimal expansion.

Three autologous blood units were collected prior to surgery in case of significant blood loss. Three days prior to surgery, selective angiography was performed again to confirm the arteriovenous communications and perform appropriate embolization. At that time, the facial, the lingual, and inferior alveolar arteries were embolized with PVA particles (500–700 μ). Since persistent flow continued in the draining vein following these embolizations, platinum/Dacron™ coils were used also to embolize the arteries. Post-embolization angiography showed continued but slow filling of the defect and associated veins.

Three days later, the exterior mandibular cortical bone was removed through an intraoral approach and the vascular malformation was removed. Embolization, along with hypotensive anesthesia and isovolumic hemodilution, enabled surgery with minor blood loss. Following removal of the third molar tooth bud, demineralized bone was compressed into the bony cavity. The patient tolerated the procedure well.

One-year postoperative dental recall showed calculus, generalized gingivitis, and exfoliating primary molars in the area of surgery. Severe crowding was still present. The patient refused to brush the surgical area despite repeated appointments to reinforce oral hygiene. Otherwise, no pathology was noted intraorally or extraorally. Although bite-wing radiographs (Fig 5) showed fill of the defect in the area between the primary second molar and permanent first molar, a panoramic radiograph revealed a radiolucency apical to the molars. Root formation of the permanent teeth in the area of the enucleated vascular malformation appeared retarded compared with the right mandibular quadrant. A selective angiography series (Fig 6) completed under subsequent general anesthesia showed no evidence of abnormal vasculature. The radiolucency was interpreted as a residual bony defect.

The decision was made to proceed with serial extraction of all first primary molars and maxillary primary canines, and enucleation of accessible first premolars. During removal of the primary teeth, the decision was made to extract only the mandibular left first premolar. The permanent canine and the premolar were allowed to erupt with removal of the second primary molars and first premolars planned for a later date.

A panoramic radiograph (Fig 7) taken 2 years postoperatively showed continued root formation of the left second premolar and first permanent molar. Root formation of the right mandibular permanent first molar appeared complete and the root of the right second premolar showed no sign of retardation as compared with the contralateral tooth. Bony regeneration appeared more extensive throughout the area of lesion than in the radiograph taken at the 1-year postoperative appointment. The patient was scheduled for removal of all remaining primary molars and enucle-

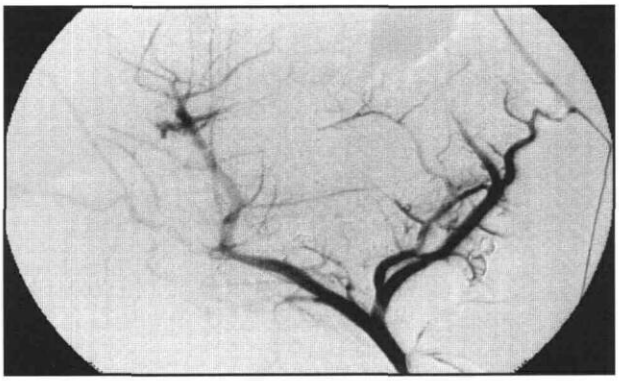

Fig 6. One-year postoperative angiogram showing no recollateralization in area of the former vascular malformation.

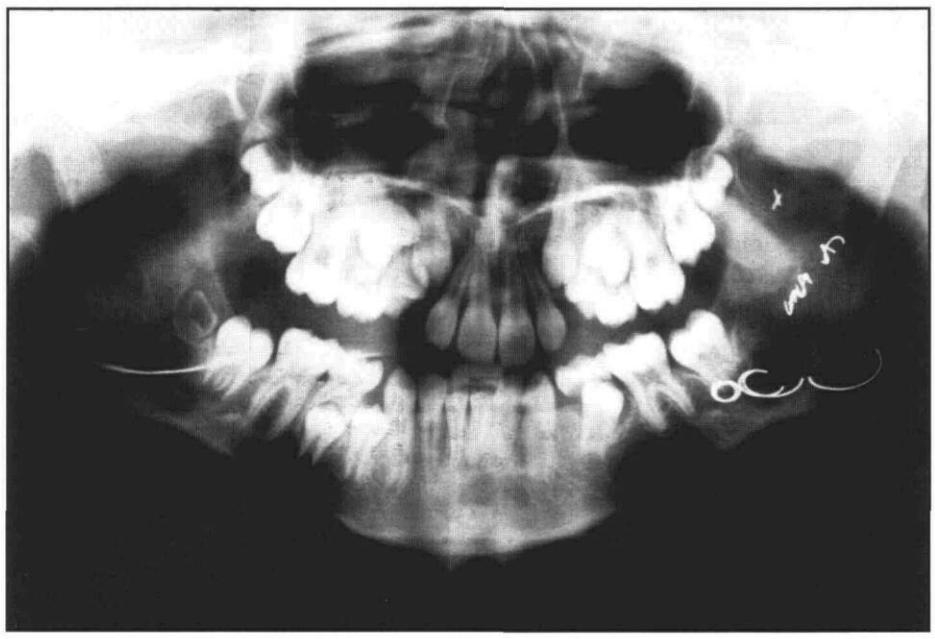

Fig 7. Two-year postoperative panoramic radiograph showing apparent bony fill of the surgerized area.

ation of accessible first premolars. Surgical removal was uneventful.

Discussion

Similar to other reports,[1,5] the vascular malformation in this patient became apparent around the time of primary tooth exfoliation. However, in this case, the lesion was noted radiographically prior to any bleeding episode. Symptoms included vague complaints of pressure in the area, but not pain. The only clinical sign was a facial asymmetry that was similar to many children.

A combination of selective angiography and embolization, aggressive surgical curettage, and demineralized bone implant resulted in a treatment outcome of an intraosseous mandibular vascular malformation in a child that was effective, functional, and esthetically pleasing. Intraoral incisions prevented massive scarring from the surgical procedure. Infection was controlled. These management methods negated the need for mandibular resection. The mandible and the mandibular teeth on the affected side appeared to be developing and erupting normally with the exception of slightly stunted root growth.

Root resorption of teeth forming in proximity to an expanding high-flow vascular malformation is reported.[24] In the present case, the initial panoramic radiograph indicates possible root resorption of the first permanent molar. However, incomplete maturation of the root is also possible. The apparent retardation of the premolar root formation 1 year postoperatively appears to have resolved with the first permanent molar and second premolar continuing root formation by the second year postoperatively. Thus, the developing teeth appear to have retained the ability to recover from the potential influence of the high-flow vascular malformation, the surgical procedure, and the effect of embolization.

Although the lesion was detected on the bite-wing radiographs, the lack of prior diagnostic panoramic or periapical radiographs in this case prevented early detection of the vascular malformation. The malformation appeared to be expanding, based on the size of the lesion on the bite-wing radiographs at the time of discovery, as compared with the lack of evidence on the bite-wing radiographs exposed 1 year previously. Treatment of a smaller lesion would have been preferable. If new radiographs had not been taken, the patient may have bled spontaneously and fatally during exfoliation of primary teeth. Alternately, emergency procedures to control the spontaneous hemorrhage may have resulted in significant facial deformity. Although the patient's orthodontic needs may have necessitated consequent complete radiographic records that would have detected the lesion at a later date, the risk would be higher for massive bleeding due to the potential expansion of the lesion. Although vascular malformations are rare, this case emphasizes the need for pediatric dentists to evaluate all patients with either panoramic or full series radiographs at least when the patients reach the early mixed dentition stage. This is particularly necessary when dental extractions or surgical procedures are indicated. However, careful evaluation of all radiographs is emphasized since the lesion was originally detected on a bite-wing radiograph.

Selective angiography allowed detection of the specific arteries contributing to the lesions and allowed visualization of the placement of the embolizing materials. Angiography also allowed the visualization of the effect of the embolizing materials on blood flow to the area. This prevented profuse bleeding during surgery since the initial PVA particles alone did not adequately decrease blood flow.

A future emergent and, possibly, fatal situation resulting from exfoliation of the primary teeth or extraction of teeth for orthodontic purposes was prevented by a timely diagnosis and treatment of the vascular malformation. Two-year follow-up shows no signs of recurrence. The patient's function and esthetics have not been compromised and preliminary orthodontic treatment (serial extractions) has

been started. Although all signs are currently positive for effective treatment, this child should be monitored on a yearly basis for potential recurrence of a vascular malformation.

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