

Plexiform arterio-venous malformation of the mandible: A case report

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Abstract

A case of plexiform variant of arterio-venous malformation in the mandible presenting with a persistent profuse bleeding from the gingival sulcus, engorged and pulsatile neck vessels with palpable thrill and bruit on auscultation in the submental and right submandibular region is presented. Intraorally, the patient had blood clots as well as fresh bleeding from the gingival sulcus, mobile teeth, swollen gingivae and poor oral hygiene. An evaluation of the clinical history and examination including radiographs and a massive life-threatening bleeding from the gingivae allowed us to carry out surgery for a total removal of the malformation. The present case report describes this rare condition and discusses the clinical presentation as well as the surgical management of the patient, including the encountered peri and post-operative complications.

Introduction

An abnormal communication between arteries and accompanying veins, arterio-venous aneurysms or fistula, in the oral and maxillofacial region is a rare clinical

condition. The malformation may be present either in the soft tissues or centrally in the bone and occasionally may involve both the soft and osseous tissues.

The plexiform or cirroid aneurysm, a variant of arterio-venous malformation, demonstrate a tortuous mass of intercommunicating small arteries and veins linking a larger artery and vein (Johnson et al, 1991). Such AV malformations are clinically significant as they are vulnerable to hemorrhage and additionally, depending upon the extent and site of the malformation, they may be an extra burden to the heart by shortcircuiting blood from arterial to venous system. Here we present a rare case of plexiform A-V malformation of mandible, presenting with a recurrent gingival bleeding, which also involved the surrounding soft tissues in the submandibular region and neck and the management of the patient discussed.

Case report

A 29-year-old-male was seen at the Cardiothoracic and Vascular Surgery Unit of Bir Hospital on 2054-8-16 with a pulsatile swelling in the neck and recurrent gingival bleeding of a-three-month duration which, according to the patient, had become more severe in the last one month. The patient on clinical examination, otherwise a healthy young-man of an average built, had no history of systemic diseases or bleeding disorders and the laboratory findings, except for

low hemoglobin content, were within normal limits. The patient had engorged and pulsatile vessels with palpable thrill and bruit on auscultation in the submental and right submandibular region and intraorally had blood clots as well as fresh bleeding from the gingival sulcus, mobile teeth, swollen gingivae and a poorly maintained oral hygiene.

Following blood transfusions to compensate the blood loss as well as with use of antibiotics for oral infection, which failed to show any response, angiography was performed to locate the extent of vascular malformation. The angiography, however, did not demonstrate a clear picture of feeding vessels and therefore, right external carotid artery was ligated under general anesthesia on 055-9-10 which also failed to show a satisfactory result. The gingival bleeding continued in such a way that the patient had a number of sleepless nights just to apply manual pressure and pack to stop gingival bleeding and he was then referred to the Dental Department on 054-9-24.

At the dental department, a review was done on the history and clinical signs and symptoms as well as the radiographs. The recurrent and profuse life-threatening bleeding, difficult to control with all the available measures in clinical wards, and multiple radiolucent areas with features of bony destruction of the mandible reasonably allowed us to decide on surgical intervention - to resect the mandible and soft tissues with the AV malformation and reconstruction of the defect.

Following preoperative assessment, 5 bags of fresh human whole blood was transfused to correct the fallen hemoglobin level and on 054-9-25, under general anesthesia resection of the malformation and reconstruction of the defect was carried out. With lower lip split incision from the mid-line

of the lower lip extending below the submandibular region to the angle of the mandible on both sides, the skin and soft tissues over the mandible was excised to expose the mandible. Various large sized blood vessels, few of them entering the mandible through a number of enlarged foramina on the lateral cortical plate, were encountered with profuse bleeding and hemostasis was meticulously performed until the portion of the mandible to be resected was exposed. Well beyond the radiological evidence of bony involvement, the angle of the mandible extending from the left to right was resected and bleeding of the resected stumps through infra-alveolar vessels stopped using bone wax. The attachment of the muscles on lingual surface of the resected mandible was then thoroughly explored for further feeding vessels which showed two large branches of arteries and veins from lingual arteries and veins perforating the lingual plate of the mandible through two large foramen were observed. The vessels were then ligated and the resected mandible was made free from all soft tissue attachment. This was immediately followed by a complete hemostasis and no engorged pulsatile vessels were observed in the field of surgery.

For reconstruction of the mandible, 6th rib of the right side was taken as a free bone graft, shaped to the contour of the resected mandible and fixed to the remaining mandibular stump using transosseous wiring. The geniohyoid and genioglossus muscles were sutured to the grafted rib to prevent fall back of tongue. The oral layer of oral mucosa was thoroughly closed and the skin wound was closed in layers.

The perioperative blood loss was nearly 2,000 ml and during the 4 hour long procedure 6 bags of whole human blood were transfused. The recovery from general anesthesia as well as the post-operative events

were satisfactory and based on subsequent hemoglobin estimation, another 3 bags of blood were transfused. The surgical wounds inside and outside of the oral cavity as well as the rib graft healed satisfactorily, however, on 8th post-operative day, a small section of the graft rib was seen exposed in the oral cavity. Attempts to close the intra-oral dehiscence failed and by 4th week nearly 6 cm of the grafted bone was seen exposed in the oral cavity. Therefore, on 2054-12-20, although a satisfactory bony union of the grafted bone was observed at both the resected ends, the graft was removed due to excessive intra-oral exposure and an uncontrolled infection and K-wires were placed to maintain space for future reconstruction. Unfortunately, after two weeks the K-wire as a retainer failed due to excessive mobility. This led us to remove the K-wires and no reconstruction of the mandible was carried out, however, the soft tissue anatomy and the facial contour remain in a highly favourable position. The outcome of treatment during the period after this surgery, nearly 6 months till the writing of this case report and despite of the loss of mandible, was found satisfactory, to our surprise, for speech and oral functions with minimal defect.

Discussion

Hemorrhagic lesions, aneurysms and haemangiomas, in the soft tissues of the maxillofacial region may be reasonably identified in many instances. However, those centrally in the bone may present difficulty in

the differential diagnosis. In the jaw bones, a history of bleeding from gingival sulcus around the teeth, loosening and mobility of the teeth accompanied by bleeding, erosion of root surfaces on radiological examination and changes in the color and contour of the soft tissues, pain or paraesthesia all related with common diseases of periodontal tissues are often associated with these group of rare

hemorrhagic lesions (Yih et al 1989, Sadowsky et al 1981). A palpable thrill or a bruit on auscultation, as seen in our case, may be an important sign of aneurysms but still may not be present in haemangiomas and any of the above signs may be absent. Central lesions, in addition, may not have any specific or consistent findings and therefore may present as a destructive lesion which may be of varying size and appearance and not uncommonly suggestive of a cyst. In addition, they may present with honey-comb

appearance or a multilocular radiolucency as in ameloblastoma and sometimes with radiating spicules at the expanded periphery forming sun ray or sun-brust appearance as in osteosarcomas (Waldron CA 1995).

A-V malformation, in general is classified as plexiform or cirroid which contain tortuous mass of small arteries and veins like a larger artery and vein, a varicose aneurysm which show an endothelial-lined sac connecting an artery and a vein and lastly, an aneurysmal varix with a direct connection between an artery and a dilated vein (Gometz and Bernatz 1970).



(Fig-Post-operative radiograph)

Cases presenting as a plexiform or cirroid variant of malformation in the maxillofacial region, as in the present case, because of tortuous mass of small arteries and veins in the maxillofacial region needs more attention and care in the diagnosis and management as it constitute an absolute contraindication to tooth extraction as well as oral prophylaxis including scaling and root planing. Extraction of teeth in situations in which the root structures are involved with the aneurysm, as also in cases of hemangioma, may produce death on the operating table or dental chair either due to a massive bleeding, develop hemorrhagic shock, or aspirate the high velocity and volume of the blood. Therefore, it is always advisable to attempt to aspirate fluid through aspirating needle before any surgical intervention of the area under suspicion and when the aspirate is blood, it should be further evaluated.

The A-V malformations may be congenital or acquired, where the acquired is usually associated with a history of trauma. We could not describe the etiology of our case as in a number of cases of A-V malformations of the oral and maxillofacial reported in the literature (Gometz and Bernatz 1970).

Management of hemorrhagic lesions, central aneurysms and hemangiomas include surgical excision, irradiation, sclerosing agents, embolization and cryosurgery. Angiographic techniques have led to embolization which precisely occlude the feeding vessels of the lesion (Alling and Alling 1984). Due to the necessity of an immediate intervention in this case, otherwise a delay of even few hours to few days could have been life-threatening, we did not attempt for further angiography when the first attempt failed and the most reasonable modality left behind us was total resection of the lesion and reconstruction. Surgery along with an

immediate reconstruction using autogenous bone graft has shown a highly favorable outcome in aneurysms of the maxillofacial region (Moriyama et al 1994).

Our perioperative events, although associated with a severe blood loss, and immediate post operative events were highly satisfactory. The exposure of the rib-graft in the oral cavity leading to graft-failure, although unions were observed at both the grafted ends in X-rays, may be due to a thin lining or the oral mucosa immediately above the graft. In addition, a long grafted piece of bone and an inadequate immobilization of the graft with trans-osseous wire resulting in the mobility of the graft may be the factors for its failure. Therefore, it is always preferable to use firm fixation of the graft using internal fixation with miniplates and the oral surface covered in more than two layers thick enough to prevent exposure of the grafted bone. Despite the failure of the bone graft which lead to its removal, it is worth to note that the appearance and function remain highly satisfactory.

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