Massive Arteriovenous Malformation Of The Ethmomaxillofacial Region: A Case Report

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Citation

A Gupta, S Bansal, A Gupta, P Tuli. *Massive Arteriovenous Malformation Of The Ethmomaxillofacial Region: A Case Report*. The Internet Journal of Head and Neck Surgery. 2006 Volume 1 Number 1.

Abstract

Paranasal sinuses are a rare site for vascular tumors. But their occurrence in this particular site can be life-threatening because of the bleeding which can be massive and difficult to control. Embolization alone and in combination with surgical resection has been the mainstay of treatment but literature does not mention any particular modality of treatment. We report a case who presented to us with massive arteriovenous malformation (AVM) of the ethmomaxillofacial region to emphasize upon the surgical modality of treatment. We are unaware of any previous reports of an AVM simultaneously involving the ethmoids as well as the maxilla successfully managed surgically in the English literature.

INTRODUCTION

Vascular tumors affect the region of the head and neck commonly, particularly the jaw; but arteriovenous malformations (AVM) are rare[1]. The earliest description of AVM was the reporting of snakes covering the head of Greek God Gordon's head[2]. Though usually benign, sometimes these can prove fatal because of their potential for massive bleeding. Arteriovenous malformations can either be congenital or acquired. Congenital AVMs occur as a result of lack of differentiation of arteries, veins and the capillaries during vascular development[3].There is a persistent communication between them, resulting in short circuiting of blood[4]. The acquired malformations are usually associated with a previous history of surgery or blunt trauma[5]. In either case, an alternate pathway bypasses the normal high capillary resistance region and profuses it with an excessive amount of blood; the incidence of hemorrhage is high[6,7]. The literature does not mention any one particular modality as the treatment of choice. We report a case who presented to us with massive arteriovenous malformation (AVM) of the ethmomaxillofacial region to emphasize upon the surgical modality of treatment. We are unaware of any previous reports of an AVM simultaneously involving the ethmoids as well as the maxilla successfully managed surgically in the English literature.

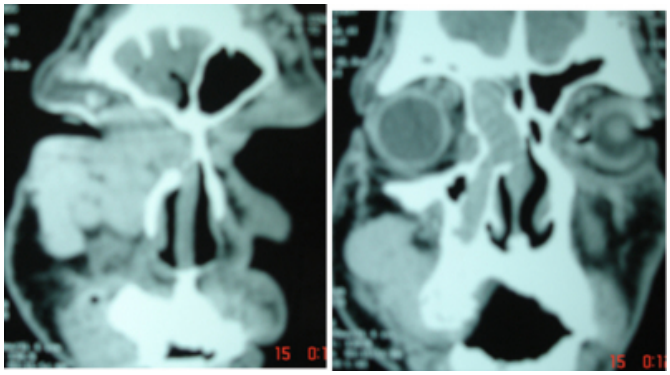
CASE REPORT

A 19-year-old male presented to our department with complaints of progressively increasing swelling and deformity of right side of the face since birth associated with 4-5 episodes of spontaneous bleeding from the oral cavity for the last 10 months requiring transfusion and hospitalization. There was no history of trauma or surgical intervention in the past. Examination of the patient revealed soft to firm pulsatile mass in the region of right side of the forehead, upper and the lower eyelid involving the cheek with fullness in the ipsilateral gingivobuccal sulcus, discolored gingiva in the region with erosion of the palate adjacent to the second molar with malocclusion. Bruit could be heard over the swelling.

The patient was subjected to contrast enhanced computed tomography (CECT) of the area revealing contrast enhancing soft tissue lesion involving above mentioned areas with involvement of ipsilateral ethmomaxillary complex and evidence of frontal recess block by the lesion (Fig. 1a,1b).

Figure 1

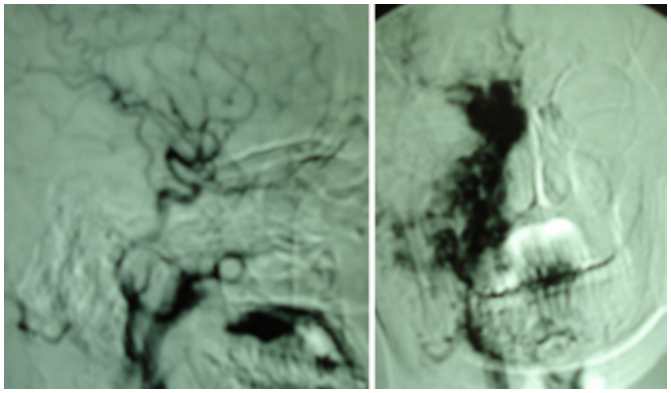
Figure 1ab: CT Scan showing enhancing soft tissue lesion in the ethmomaxillary region blocking the frontal recess (1a) and maxillofacial region (1b).



Angiography of the lesion was also done revealing the blood supply from ipsilateral anterior ethmoidal vessel, facial, internal maxillary, superficial temporal arteries (fig. 2).

Figure 2

Figure 2: Digital substraction angiography (DSA) showing blood supply to the tumor.

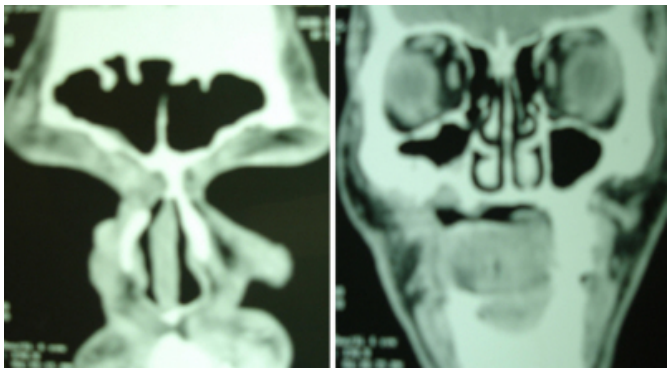


The patient during hospital stay had an episode of major hemorrhage from the erosion in the palate area requiring pressure and transfusion. Embolization was not carried out due to its supply from the ethmoidal vessel. Patient was taken up for surgery.

After taking the control of the major vessels in the neck, Weber Ferguson with lip splitting incision was made and the tumor was dissected from the soft tissues of the facial region and then the inferior partial maxillectomy along with the clearance of the ethmoidal gallery was done and the vessels supplying the tumor were ligated in sequence. The blood loss was around 1200 ml requiring transfusion. The patient had a relatively uneventful post operative period and was discharged on 8th post operative day. A CECT of the area was done two months later to see for the residual disease and there was none (Fig. 3a, 3b). The patient is on regular follow up and is doing well after 10 months of surgery.

Figure 3

Figure 3ab: Contrast enhanced CT Scan showing no residual soft tissue density in the frontoethmoid region (3a) and maxillofacial region (3b).



DISCUSSION

Vascular tumors affect the region of the nasal cavity and nasopharynx more often than the paranasal sinuses, most of which are capillary hemangiomas of osteogenic origin[8]. Arteriovenous malformations are uncommonly reported from maxillofacial region in the English literature. A slowly growing, expansile mass accompanied by spontaneous gingival bleeding near the affected site are the most common symptoms of arterioveuons malformation. Paresthesia, loosening of the involved teeth, gingival discoloration, and hyperthermia over the lesion, a feeling of pulsation, and pain are the associated symptoms in most of the cases. These can present with massive epistaxis or oral bleeding which can be life threatening and rarely iatrogenic bleeding during surgical interventions on the upper jaw such as tooth extraction can occur[9]. Occasionally bruits or pulsations can be detected in large hemangiomas.

Our patient's findings were consistent with the data in the literature. The patient had an AVM affecting the ethmomaxillary complex and the facial region which was managed successfully surgically. Darlow et al has recommended that in order to differentiate a vascular lesion from a highly vascular mucosa caused by an inflammatory process, a routine aspiration of all expansile lesions in the maxillofacial region should be done[6]. Plain radiography can demonstrate multilocular radiolucencies in the involved bone. CT can detect a highly vascular mass with multiple lobulations and angiography is essential to confirm the diagnosis as well as to evaluate the vascular architecture[10].In our patient, CT demonstrated that mass with indistinct

borders was present in the ethmomaxillary complex and the facial region with blocked frontal recess by the tumor. The tumor had invaded the palatine portion of the maxilla. Massive hemorrhage from the gingival mucosa led us to the preoperative differential diagnosis of a vascular tumor. We performed angiography, which demonstrated the high degree of vascularity in the tumor.

Treatment of these lesions is difficult and literature does not mention the best modality of treatment of these. The primary goal of the treatment is to stop further episodes of bleeding and to prevent its recurrence[11].The treatment itself involves either complete surgical excision or complete obliteration by embolization[12]. Complete resection of an arteriovenons malformation following embolization is the safest treatment, but resection of the AVM affecting the facial and the maxillary region can cause functional and cosmetic defects[9,11].Surgical curettage following preoperative embolization or ligature has also been attempted with high incidence of recurrence of the lesion[11].Selective embolization with particles or glue and direct transosseous puncture alone have also been mentioned as treatment modality with high recurrence. Coil embolization has also been successfully used to produce permanent occlusion to arteriovenous malformations[12]. We did not attempt embolization of the AVM as it was getting supply from the ethmoidal vessels as well carrying a high risk of retrograde embolization of internal carotid artery leading on to stroke, so a surgical resection of the lesion was carried out successfully.

Because recurrence or the residual lesion is frequent following embolization or the surgical resection for arteriovenous malformation, follow-up angiography and radiography are important. Follow up CECT in our patient did not reveal any tumor.

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