Short Communication

Massive juvenile angiomatosis of maxilla and mandible

Abstract

Angiomatosis is a complex vascular malformation of infancy and childhood consisting of proliferating blood vessels with accompanying mature fat, fibrous tissue, lymphatic's and nerves, which may involve skin, subcutaneous tissue, skeletal muscle and occasionally bone. It is extremely rare and benign, but a clinically extensive vascular lesion of soft-tissue, which usually becomes symptomatic during childhood or adolescence. We report a rare case of massive juvenile angiomatosis of maxilla and mandible in a 15-year-old male patient.

Key words:
Angiomatosis, intraosseous hemangioma, mandible, maxilla

Introduction

Angiomatosis is a diffuse vascular lesion, which involves a large segment of the body in a contiguous fashion involving multiple tissues (e.g., subcutaneous tissue, muscle, bone, adipose tissue, etc.) in different planes. Such lesions usually present in the first two decades of life with female predilection and are commonly seen in lower extremities. It clinically mimics hemangioma or vascular malformation (VM) and its surgical removal is difficult because of its infiltrative nature and thus has a high recurrence rate (90%).[1] Therefore, a precise histopathological diagnosis of angiomatosis is important to achieve a curative resection. Histopathologically it consists of proliferating blood vessels of varying caliber, infiltrating into the soft-tissues. Proliferating capillaries are seen within or adjacent to major vessels. Few cases are reported in the head and neck region.[2] We reported a rare case of massive juvenile angiomatosis of maxilla and mandible in a 15-year-old male patient.

Case Report

A 15-year-old male patient reported to Department of Oral and maxillofacial surgery with a chief complaint of swelling of the lower jaw since 6 years. His parents stated that a small swelling on the lower jaw was noticed 6 years ago, which gradually increased to the present size involving both sides of the jaw and swelling in the right side of the upper jaw was noticed from past 1 year. Patient was reported late due to their economic status and their origin from a rural background. Patient was further complained of difficulty in swallowing and mastication.

Gross facial asymmetry was noticed due to a swelling involving the middle and lower one-third region of the face [Figure 1]. Bilaterally fullness of the cheeks and lower lip with obliteration of the mentolabial sulcus and nasolabial fold was noticed. A well-demarcated swelling was noticed over the right side of maxilla and a massive extensive swelling was present on the entire mandible around 20 cm × 11 cm in size, which extended from the left ramus of the mandible to the right ramus of the mandible with displacement of involved teeth.

On palpation, the swelling was hard in consistency with no accompanying cervical lymphadenopathy. On intraoral examination, a diffuse swelling was seen extending from left to right retromolar region. Bilaterally, vestibular obliteration was noticed due to buccal labial and lingual cortical plate expansion. Patient was advised computed tomography scans, which revealed large extensive radiolucent-radiopaque swelling on the right maxilla with expansion of the cortical plates thus, displacing the

Access this article online	
Website: www.cysonline.org	Quick Response Code
DOI: 10.4103/2229-5186.129345	

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involved teeth along with massive expansile mass of the entire mandible displacing the involved teeth [Figures 2-5]. Correlating the history, clinical and radiographic findings and a provisional diagnosis of benign odontogenic lesion of mandible was given and differential diagnosis of fibrous dysplasia, ossifying/cementifying fibroma and central giant cell granuloma, intraosseous hemangioma was made. Patient was further subjected to hematological, radiological and histopathological investigations. Routine hematological examination, serum protein, calcium, phosphorus and alkaline phosphatase levels were estimated and the values obtained were within the normal range. Incisional biopsy was planned under general anesthesia, which leads to severe hemorrhage intraoperatively and four units blood transfusion was performed to avoid further complications. Patient recovery from general anesthesia was uneventful. There were mild bleeding episodes subsequently, which raised suspicion of vascular lesion. The excised specimen was sent for histopathological evaluation, which led to diagnosis of angiomatosis of maxilla and mandible. Photomicrography showed thick bony trabeculae composed of lamellar bone in areas enclosing vascular channels in the fatty marrow some having thick fibrous wall and contain fibrin thrombi in the lumen [Figure 6]. A final diagnosis of angiomatosis of maxilla and mandible was made based on radiographic and histopathology reports, a multidisciplinary approach for resection and reconstruction of the jaws was planned later.

Discussion

VMs are developmental abnormalities of the vascular system. They should be differentiated from vascular tumors or hemangioma because they have different causes, growth patterns, treatments and outcomes. VMs are localized errors of angiogenic development, whereas hemangiomas

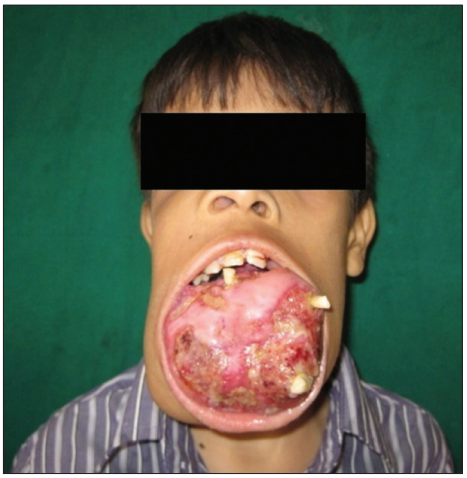

Figure 1: Clinical photograph showing massive extensive well defined swelling on the right maxilla and on the entire mandible

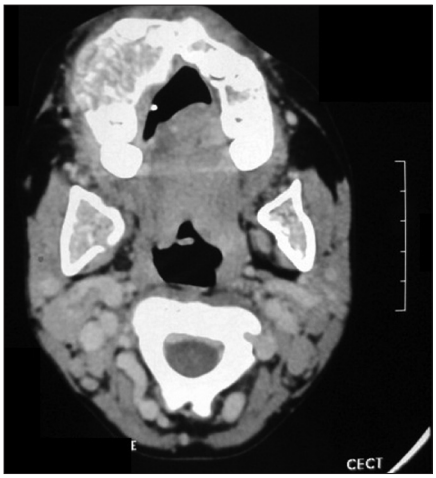

Figure 2: Axial computed tomography scan view shows a large radiolucent-radiopaque swelling on the right maxilla with expansion of the cortical plates thus displacing the involved teeth

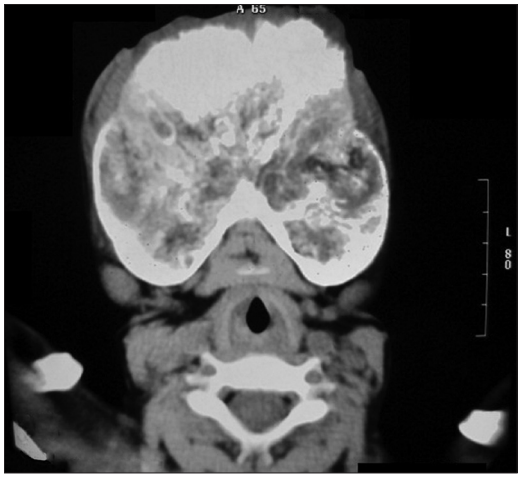

Figure 3: Axial computed tomography scan view shows a large expansile radiolucent-radiopaque swelling on the entire mandible thus displacing the involved teeth

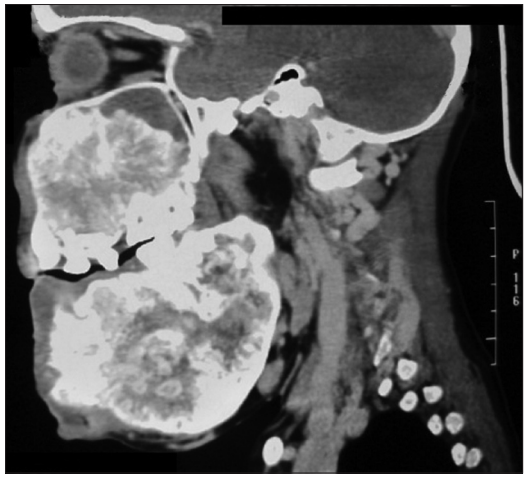

Figure 4: Sagittal computed tomography scan view showing radiolucent-radiopaque swelling on both maxilla and mandible thus, obliterating maxillary sinus

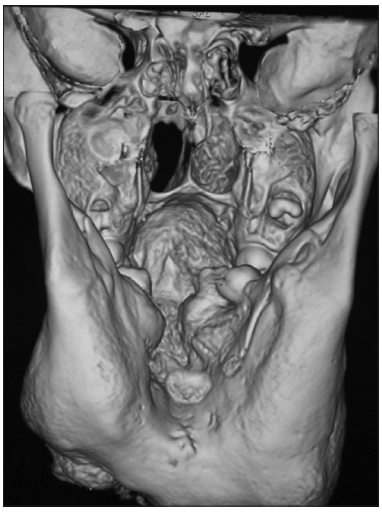

Figure 5: 3D computed tomography scan view showing massive extensive well defined swelling on the right maxilla and on the entire mandible

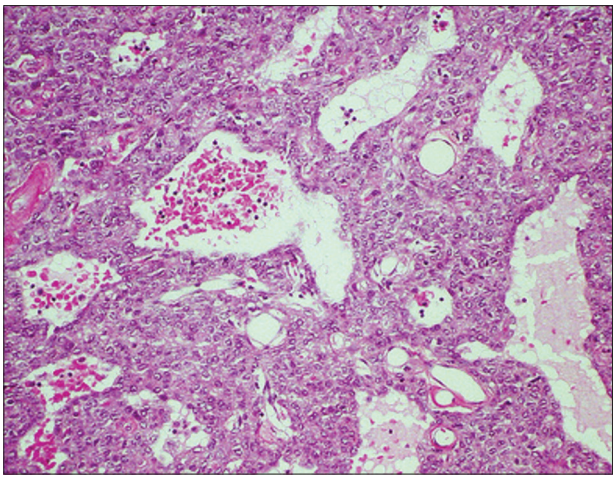

Figure 6: Photomicrography shows thick bony trabeculae composed of lamellar bone in areas enclosing vascular channels in the fatty marrow some having thick fibrous wall and contain fibrin thrombi in the lumen (H and E, ×400)

are vascular tumors. Arteriovenous malformation, a term well-founded in the radiologic literature, imply functional arteriovenous communications and may correspond in some instances to what pathologist's term angiomatosis.[2] According to Rao and Weiss,[2] angiomatosis has been defined as a histologically benign vascular lesion that affects a large segment of the body in a contiguous fashion, either by vertically involving multiple tissue types (e.g., subcutaneous tissue, muscle, bone) or by involving similar types of tissues (e.g., multiple muscle). It has a highly characteristic, but not totally specific histologic pattern. It has no malignant potential, but because of its infiltrative nature, it could be misdiagnosed as a malignancy using imaging techniques. For these reasons diagnosis based on clinical and radiographic findings in correlation with histologic features would be desirable. Its diffuse infiltrative nature makes surgical resection difficult; hence, recurrence is a common feature in angiomatosis.[2,3]

Intraosseous vascular lesions are rare conditions, comprising only 0.5-1% of all intraosseous tumors. They mainly occur in the second decade of life especially in women. The most common locations are the vertebral column and skull; nevertheless, the mandible is a quite rare location.[4,5] According to the World Health Organization, hemangioma is a benign vasoformative neoplasm of endothelial origin. Clinically, the patient may be completely symptom-free or may present discomfort, pulsatile bleeding, bluish discoloration, mobile teeth, and derangement of the arch form or accelerated dental exfoliation. Most frequently radiographic finding is a multilocular radiolucent image with honeycombs or soap bubble appearance.[6] Differential diagnosis includes neoplasms such as ameloblastoma, cystic lesions such as residual cyst, keratocyst and fibro-osseous lesions like fibrous dysplasia. There are some therapeutic alternatives, although wide surgical excision remains as the gold standard. The most hazardous complication is uncontrolled bleeding and consequently even death, during a surgical procedure, such as tooth extraction or biopsy.[7] The present comprehensive critical analysis showed that the lesion may present at any age, but was most commonly discovered in the second decade of life.

Angiomatosis can either be congenital or acquired. The congenital form may be seen sporadically or accompanying certain types of syndromes such as Klippel–Trenaunay–Weber syndrome, Sneddon's syndrome or Gorham disease.[8] The acquired form of angiomatosis may be infectious or iatrogenic. Bartonellosis and human immunodeficiency virus are associated with angiomatosis. Because of its infiltrative nature, angiomatosis may resemble many of the vascular tumors and malformations. Histopathological differential diagnosis includes angiolipoma, angiomyolipoma, infiltrating lipoma, angiomyxolipoma and liposarcoma. The treatment of choice in extensive angiomatosis is either radiotherapy or interferon alpha-2a treatment.[8] In localized cases, complete resection is preferred, but there is a risk of local recurrence.

Conclusion

Angiomatosis of the jaws is one of the rarest tumors occurring in the maxillofacial region. Proper diagnosis is very important to avoid mishaps in management and a more cautious multidisciplinary approach is required to treat such type of lesions. It is a surgical challenge for reconstruction and rehabilitation for the patient post-operatively.

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How to cite this article: Chauhan DS, Guruprasad Y. Massive juvenile angiomatosis of maxilla and mandible. Chron Young Sci 2014;5:75-8.

Source of Support: Nil, **Conflict of Interest:** None declared

FORM IV

*Statement about ownership and other particulars about newspaper **(Chronicles of Young Scientists)** to be published in the first issue every year after the last day of February as per Rule 8*

1. Place of publication	:	Mumbai
2. Periodicity of its publication	:	Semiannually (January and July)
3. Printer's Name	:	Hemant Manjrekar
Nationality	:	Indian
(a) Whether a citizen of India?	:	Yes
(b) If a foreigner, the country of origin	:	N.A.
Address	:	B5-12, Kanara Business Center, Off Link Rd, Ghatkopar (E), Mumbai - 400075, India
4. Publisher's Name	:	Hemant Manjrekar
Nationality	:	Indian
(a) Whether a citizen of India?	:	Yes
(b) If a foreigner, the country of origin	:	N.A.
Address	:	B5-12, Kanara Business Center, Off Link Rd, Ghatkopar (E), Mumbai - 400075, India Phone: 91-22-6649 1818/1816,
5. Editor's Name	:	Dr. Himanshu Gupta
Nationality	:	Indian
(a) Whether a citizen of India?	:	Yes
(b) If a foreigner, the country of origin	:	N.A.
Address	:	C/o. Organization of Pharmaceutical Unity with BioAllied Sciences, 21, Jaina Building, Roshanara Road, Delhi-110 007, India
6. Names and addresses of individuals who own the newspaper and partners or shareholders holding More than one per cent of the total capital.	:	Organization of Pharmaceutical Unity with BioAllied Sciences

I, Dr. Himanshu Gupta hereby declare that the particulars given above are true to the best of my knowledge and belief.

Date: 1st March 2014 **Hemant Manjrekar** **Dr. Himanshu Gupta**