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Case Report

Massive craniofacial cystic angiomatosis and extra-skeletal angiomatosis: A unique case report and brief review of literature

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ABSTRACT

Cystic angiomatosis is a rare vascular disorder, characterized by multifocal hemangiomatous and/or lymphangiomatous lesions of the skeleton with possible visceral organ involvement. The aim of the present communication is to report a unique case of massive craniofacial skeletal cystic angiomatosis in a 15-year-old boy with history of progressive enlargement of all craniofacial bones since birth associated with severe anemia secondary to splenomegaly. The computed tomography and magnetic resonance imaging techniques revealed hypertrophy of the craniofacial skeleton secondary to multiple cystic lesions and also disclosed cystic lesions in cervical, lumbar, sacral vertebrae and the spleen. Histologically, it was analogous to cavernous hemangioma. This was an unusual case of classical bilateral, craniofacial skeletal angiomatosis; till date a total of three cases have been reported but the present case was diverse from the reported ones.

1. Introduction

The incidence of intraosseous hemangioma is very rare and accounts for 0.5–1% of all bone tumors [1], whereas venous malformation accounts for 35% of vascular disorders of skeleton. Cystic angiomatosis or skeletal–extra-skeletal angiomatosis is described in literature as a benign vascular proliferation, that lies in the medullary cavity of bone, produces multiple cystic lesions and also affects at least one other type of tissue other than the skeleton.

The first description of 'cranial', 'skeletal and extra-skeletal forms' and the term 'cystic angiomatosis' were given by Jacobs and Kimmelsteil, Charlotte [2] in the annual meeting of the American academy of orthopedic surgeons, at Chicago in 1952. It was reported in maxillofacial literature by Goutoudi et al. [3] in a 10-year-old boy (Table 1).

The aim of the present communication is to report an unusual case of craniofacial and appendicular skeletal cystic angiomatosis associated with visceral organ involvement.

2. Case report

A 15-year-old boy was referred to the department of oral and maxillofacial surgery with a history of painless progressive enlargement of facial and skull bones bilaterally, predominantly involving the zygomatico-maxillary complex, mandible and the occipital bone since childhood. On extra-oral examination, diffuse enlargement of all craniofacial bones leading to excessive exposure of sclera was noticed. Neither hyperemic collateral vessel nor port-wine stain or facial nevus was seen on the face. On palpation, the lesion was found to be hard in consistency and was nontender (Fig. 1). On intraoral examination, bucco-lingual expansion of alveolar process of both the jaws along with generalized distension of gingiva and also displacement of teeth was observed (Fig. 2). No vascular bruit or thrill was detected and aspiration was found to be negative.

Hematological evaluation disclosed that patient was suffering from chronic anemia with a hemoglobin level of 4 g/100 ml. The hemoglobin deficiency was attributed to malnutrition or splenomegaly. No other blood cell structural abnormality was detected by the Department of Hematology. Remaining all biochemical values including serum alkaline phosphatase, calcium, phosphorous and all endocrine hormones were within the normal limits and not conclusive for any diagnosis.

Axial and coronal computed tomography demonstrated bilaterally the presence of multiple cystic lesions in the craniofacial skeleton, which were causing expansion of the cortices and they had intact margins (Figs. 3–6). There was a small breach in the

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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Table 1
Clinical details of reported cases on cystic angiomatosis in the maxillofacial literature (brief review of literature).

	Patients	Age (y)/sex	Clinical features	Radiological features	Histological features	rx done	Author	Year	Journal
1	2 pts	9 y, 6 y, male	Bimaxillary enlargement	Multilocular	Cavernous hemangioma	Resection	Hossein Mortazavi	2011 [4]	J Oral Maxillofac Surg
2	1 pt	10 y, male	Enlargement	Multilocular	Cavernous hemangioma		Goutoudi	1992 [3]	Oral Surg Oral Med Oral Pathol Oral Radiol Endod

Pts, patients; y, age in years.

left buccal cortex of the mandible. Bilaterally, the maxillary sinus was obliterated with polyps. Hypertrophy of middle and inferior nasal conchae on the right side was observed (Fig. 4). The computed tomography of the calvarium revealed widening of the diploic spaces and thinning of the cortices. There was no evidence of encapsulation of the lesion and melded imperceptibly with the surrounding bone. Radiographically, the lesion was granular with 'ground glass appearance'.

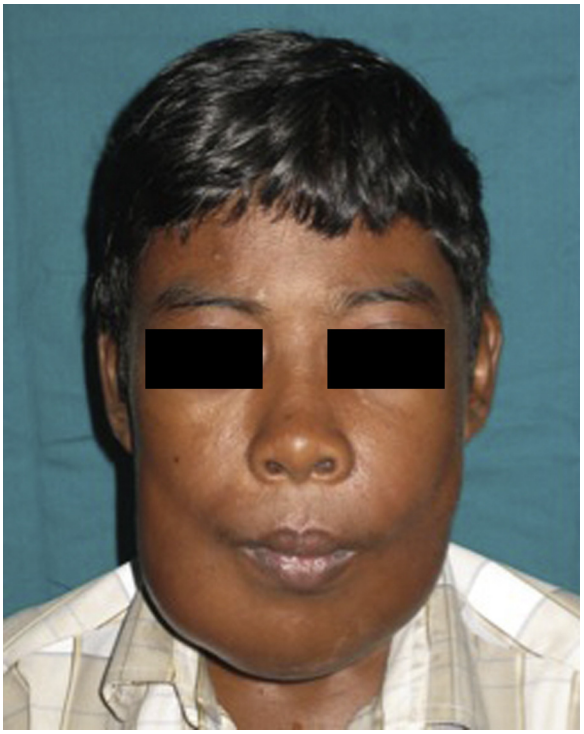

Fig. 1. Facial clinical photograph depicting the generalized hypertrophy of entire craniofacial skeleton with excessive scleral show.

The magnetic resonance image (MRI) of the brain showed normal study without the evidence of calcifications but the MRI findings of the skull revealed hypertense T2 cystic lesions (Figs. 7 and 8) and also depicted sharply defined lesions with low signal intensity related to muscle on T1 weighted images and extremely bright signal much brighter than that of fat on T2 weighted, ST1R and gradient pulse echo and also disclosed hyperintensity images in the right tibia, left femur, lumbo-sacral vertebrae, cervical vertebrae and spleen similar to 'fluid signals of urine', suggestive of cystic lesions (Fig. 9). MRI of the right maxilla showed marked increased in density of soft tissue with displacement of the facial vessels (Fig. 7). The existence of cystic lesions was also confirmed by Ultrasonograph and Angiogram revealed 'normal flow' or 'gush' and disclosed the absence of feeding vessels.

The incisional biopsy was planned from the zygomatic maxillary complex of the right side under general anesthesia. At operation, the mucosal incision was given; brisk bleeding was encountered, but was controlled with hemostatic pack. At some stage, after entering into the bone marrow, brisk and massive hemorrhage was encountered which obligatory immediate resuscitation with 1 unit of whole blood and 2 units of packed red blood cell and hemostasis was achieved with Surgicel® (Ethicon) and bone wax. Because of abnormal, profuse bleeding, only a small piece of the tissue was harvested.

Light microscopy displayed the features of distended blood vessels which were lined by endothelium, comparable to capillary hemangioma or cavernous hemangioma (Fig. 10). By correlating the clinical, serological, radiological and histological findings, the most probable diagnosis of cystic angiomatosis was given. Surgery was not attempted because the patient was suffering from frank anemia and splenomegaly and was referred to local regional cancer center for radiotherapy to suppress the active proliferation of vascular tissue. The patient was reported with anemia related complications after 1 year without increase in the size of the lesion [the growth was stabilized], after that they failed to report for further followup.

3. Discussion

Cystic angiomatosis is a multifocal disorder of bone and viscera, which is composed of either hematogenous or lymphatic deposits or both, and is a rare cause of massive bone destruction. This type of angiomatosis typically presents in two distinct clinical settings: cystic angiomatosis or multiple primary angiomatosis and massive osteolysis (Gorham's disease or Stout syndrome) or diffuse cystic angiomatosis. Till date only three patients [3,4] have been reported in the maxillofacial literature.

Skeletal angiomatosis was classified into aggressive and nonaggressive types based on the clinical behavior and spread (regional and disseminated). It occurs in the form of 'skeletal' or 'extraskeletal' or 'both'.

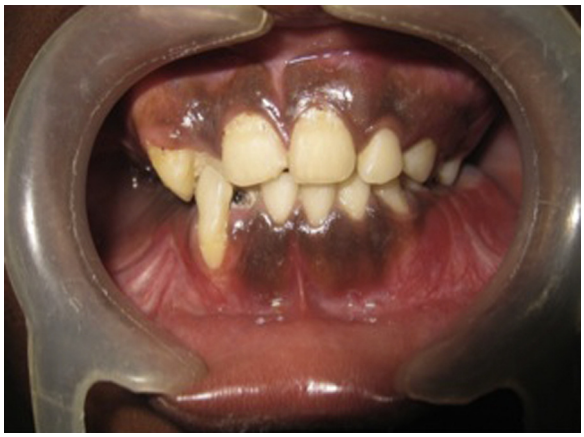

Fig. 2. Intraoral clinical photograph showing the generalized hypertrophy of gingiva, the alveolar process and severe tooth deracination.