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Fronto-orbital cystic fibrous dysplasia in Dar Es Salaam: Case report.

*Mboka Jacob[1], Jinna Kim[2]

1. Department of Radiology, Muhimbili University of Health and Allied Sciences
2. Department of Radiology, Severance Hospital, Yonsei University College of Medicine, Seoul, Korea

***Corresponding author**

Dr. Mboka Jacob
Department of Radiology,
Muhimbili University of Health and Allied Sciences
Dar es salaam,Tanzania

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Abstract

Frontal-orbital Fibrous dysplasia (FD) is well documented, however, it rarely present as cystic lesion. Typically occurs in patients between the ages of 5 and 15 years. We report a case of frontal-orbital fibrous dysplasia in a 27 years old man who presented with supraorbital swelling and painless proptosis of the left eye. Computed tomography revealed a disease, mainly cystic and expansile with areas of punctuate calcification encroaching the orbit, frontal and ethmoid sinuses. The diagnosis of FD was confirmed on pathological examination. This case report includes clinical aspect and radiographic appearance. The differential diagnosis with ossifying fibroma, aneurysmal bone cyst, giant cell tumour and osteomyelitis are discussed as well.

Key words: Fibrous dysplasia, ossifying fibroma, fronto-orbital, Computed Tomography

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Introduction

Fibrous dysplasia is a benign idiopathic skeletal disorder that can affect any bone in the body. Typically it occurs in patients between the ages of 5 and 15 years, and has two basic clinical forms; monostotic or polyostotic form, accounting for 70% and 30%, respectively [1]. The skull and facial bones are the most affected sites in 10-25% of patients with monostotic fibrous dysplasia and in 50% of patients with polyostotic fibrous dysplasia. Conventional radiographic and computed tomography (CT) studies reveal characteristic findings of fibrous dysplasia, which consist of the following three varieties; sclerotic (ground-glass) (25%), Pagetoid (mixed lucent and sclerotic) (50%), and cystic pattern (25%) [2].

The aim of this paper is to discuss the uncommon CT imaging features of a cystic form of fibrous dysplasia involving the fronto-orbital area and paranasal sinuses, and an awareness of this imaging finding can help to reduce the possibility of misdiagnosing a craniofacial fibrous dysplasia that might mimic neoplastic disease.

Case summary

A 27-year-old man visited our hospital with complaint of painless swelling of the left eye and headache on left frontal area for two years. On physical examination, there was a hard palpable mass on the left supraorbital area. CT showed a well-defined radiolucent expansile bony mass involving left supraorbital frontal bone with areas of punctate calcifications (Fig. 1 A, B, C). The cortical rim surrounding the mass was thin and remodeled, but intact without disruption. It also extended into the ipsilateral orbit, frontal and ethmoid sinuses, and displaced intraorbital structures including the globe. The mass exhibited strong enhancement after contrast administration.

Partial excision of the mass for establishing the pathologic diagnosis was performed. Histopathologic examination revealed fibrous dysplasia.

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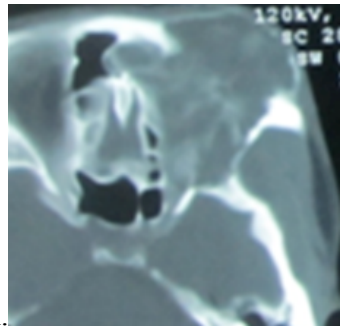
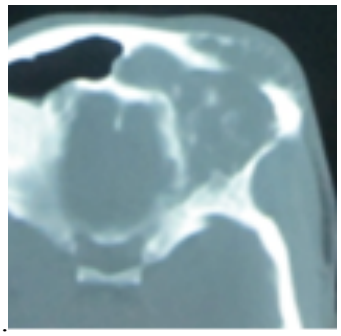
A.  B. 

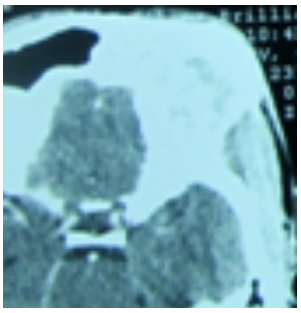
C. 

Figure 1. (A) Axial CT image with bone window setting shows an expansible radiolucent bony mass involving the left supraorbital frontal bone. (B) Axial CT image at another level shows areas of punctuate calcification within the mass. (C) Axial contrast-enhanced CT image with soft tissue window setting demonstrates strong enhancement of the mass.

Discussion

Patients with craniofacial fibrous dysplasia usually present with complaints of progressive facial deformity, pain, paresthesia, proptosis, and visual disturbances [3,4]. Fibrous dysplasia typically appears as an expansile osseous lesion, and the most common radiologic feature is ground-glass appearance. However, some craniofacial fibrous dysplasia demonstrates an expansile cystic mass as shown in our case, which has been scarcely reported in previous literatures published in Africa [5]. Fibrous dysplasia is a congenital fibro-osseous lesion in which normal medullary bone is replaced by weak osseous and fibrous tissue, and its imaging appearance varies with relative content of fibrous and osseous tissue. According to the previous studies, cystic form constitutes up to 25% of all fibrous dysplasia in the head and neck area [2].

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In regard to the radiologic diagnosis CT is an imaging modality of choice for fibrous dysplasia as it define the extent of the disease and shows internal characteristics such as ground-glass texture in detail.
Fibrous dysplasia affects the ocular structures in many ways [4]. Our case showed involvement of frontal bone and sphenoid wing which lead to proptosis and extraocular muscle palsy.

The differential diagnoses for craniofacial cystic fibrous dysplasia include ossifying fibroma, aneurysmal bone cyst, giant cell tumour and osteomyelitis. Forty two percent (42%) of ossifying fibroma present as a cystic radiolucent lesion which can mimic cystic form of fibrous dysplasia. Aneurismal bone cyst is relatively uncommon in the facial bone, and most cases have been found in the mandible.

Conclusion

Cystic fronto-orbital fibrous dysplasia is a rare disorder which is characterized by expansile bone lesion which can lead to a range of clinical presentation. Our case showed involvement of frontal bone and sphenoid wing which caused proptosis and extraocular muscle palsy in this patient. Imaging plays an important role in the diagnosis and management. Less is reported on Cystic fronto-orbital fibrous in Africa.

Acknowledgements and Competing interests

The authors declare that they have no financial or personal relationship(s) that may have inappropriately influenced them in writing this article.

Authors' contributions

M.J. (MUHAS) worked up the patient, wrote the original manuscript and worked on the literature review and on the final document. J.K (Yonsei University), contributed to diagnosis ,writing the legends and preparing images and was involved in the final editing of the document.

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