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Image Report

Polyostotic craniofacial fibrous dysplasia with bizarre radiologic finding: Mandible, anterior skull base, frontal, temporal, parietal, and occipital bones involvement

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A 30-year-old man presented with progressive blurred vision and cosmetic facial problems within 2 years ago. Neurological examination demonstrated impaired visual acuity and constricted visual field associated with both forehead and right side craniofacial deformity in the general examination. The craniofacial computed tomography (CT) scan showed expansion and thickening in right mandible, anterior skull base (including sphenoid, orbital roof, and ethmoidal bones), clival part of the occipital bone, right temporal, frontal, and parietal bones and also left frontal bone [Figures 1 and 2]. Figure 1c revealed narrowing of the optic canal and nerve entrapment. He advised for surgical intervention, but denied the surgery.

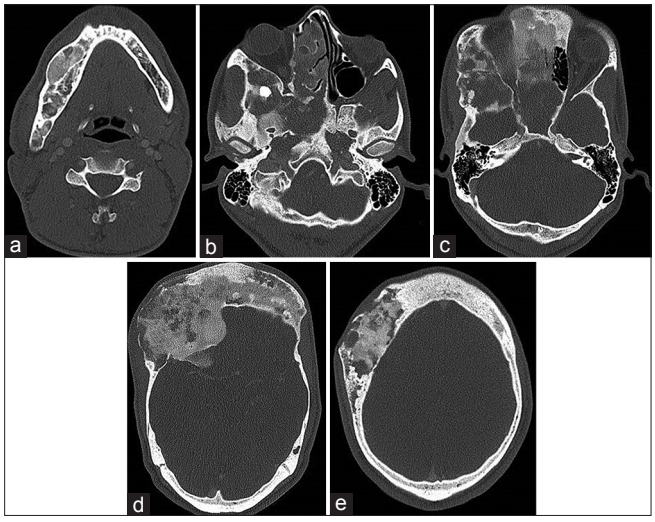

Figure 1: Demonstrated the axial view of CT scan (a) right mandible bone, (b and c) right skull base involvement, (d and e) right and left frontal bone involement

DISCUSSION

Fibrous dysplasia (FD) is an uncommon benign disorder with unknown etiology, although variety causative factors explained. It divides into two primary types. First, monostotic type involves one bone and the second, polyostotic type that involves more than one bone.[3] In this developmental disorder, the normal bone displaced with abnormal fibrous tissue that contains small and irregular bone trabeculae.[2] So, it results to expansion, thickening, and sclerosis of the involved bones.[2] Seventy percent of the FD throughout the body is monostotic type and 70–90% of the craniofacial FD is polyostotic type. Skull and facial bones involve in 10% of the monostotic cases

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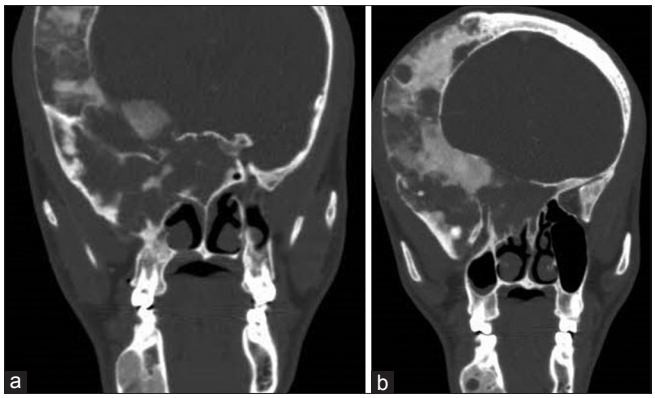

Figure 2: (a and b) the coronal view of the craniofacial CTscan revealed the multiple bones involvement

and 50–100% of polyostotic cases.[1] In the skull, FD can involve any bone but the most common site is the frontal bone followed by sphenoid, parietal, ethmoid, temporal, and occipital bones.[1] The genetic etiology of FD consist mutation over chromosome 20q13. This mutation cause increase in interleukin 6 (IL6); a cytokine which is involved in osteoclast differentiation.[5] Histopathology demonstrated fibrous and osseous components, that substitute normal marrow space. Fibrous component contain benign fibroblastic tissue and show spindle cells with low mitotic activity. The osseous component contain spicules of woven bone with typical osteoblasts embedded within fibrous component.[5]

Clinically FD result to pain, deformity, fracture, and nerve entrapments,[4] but it usually depends on which bone is involved and consist of: Propetosis, facial pain, vertigo, facial asymmetry, headache, cranial nerve dysfunction, auditory impairment and visual impairment, and disturbance.[1] The most important differential diagnoses of FD include Paget's disease, hyperparathyroidism, local reaction to meningioma, osteoma, eosinophilic granuloma, osteochondroma, and sarcomatous neoplasm.[3]

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Conflicts of interest
There are no conflicts of interest.

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