Craniofacial Fibrous Dysplasia presenting with Visual Impairment

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Introduction

Fibrous or fibro-osseous dysplasia is a developmental disorder of growing bones of unknown aetiology. It occurs in two distinct forms - polyostotic which involves several bones and accounts for 30% and the monostotic form which involves a single bone and constitutes 70% of cases [1]. Craniofacial involvement in fibrous dysplasia occurs in nearly 100% of polyostotic and 30% of monostotic forms [2]. The bones commonly involved are mandible (12%) and maxilla (12%), involvement of the ethmoid, sphenoid, frontal and temporal bones are infrequent [3]. These lesions cause expansion, thickening and sclerosis of the involved bones with resultant visual complications, hearing disturbances, facial asymmetry and tooth displacement depending on the bone involved.

We report two cases of monostotic craniofacial fibrous dysplasia presenting with visual disturbances, near total blindness of both eyes in the first case and impairment of vision of left eye in the second. Both were diagnosed using computed tomography.

Case Report - 1

An 18 year old boy reported with complaints of progressively decreasing vision in both eyes of 4 months duration. General physical and systemic examination revealed no abnormality. Ophthalmologic examination revealed a visual acuity of 6/60 in both eyes, bilateral complete ophthalmoplegia and optic atrophy. A clinical impression of a constrictive lesion causing compression of the optic canal and superior ophthalmic fissure bilaterally, was made. CT scan of the head showed expansile, sclerotic lesions with a density of 433 HU, involving the ethmoid and sphenoid bones on both sides. These lesions were causing obliteration of optic canals, superior ophthalmic fissures and ethmoid air cells on both the sides (Fig 1). Based on the CT findings a diagnosis of monostotic craniofacial fibrous dysplasia causing bilateral optic atrophy and complete ophthalmoplegia was made. A skeletal survey was carried out which did not reveal involvement of any other bone.

Case Report - 2

A 45 year old female patient presented with progressively deteriorating vision of the left eye of 3 months duration. Ophthalmological examination revealed a visual acuity of 6/36 and mild optic atrophy in the left eye. CT scan of head revealed an expansile, hyperdense lesion (HU 358) involving the left pterygoid plate and progressing superiorly to involve the greater and lesser wings of the sphenoid bone on the left side (Fig 2). A diagnosis of monostotic craniofacial fibrous dysplasia causing partial obliteration of left optic canal with compression of the left optic nerve was made.

Discussion

Fibrous dysplasia is a developmental or growth disorder in which normal bone is replaced by abnormal fibrous tissue that contains small, abnormally arranged bone trabeculae. It is considered by some authors to be a hamartomatous malformation that presumably results from an idiopathic arrest in maturation of bone at the woven bone stage [3].

Craniofacial involvement in fibrous dysplasia is seen

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Monostotic fibrous dysplasia has a different skeletal distribution from polyostotic disease and occurs most commonly in the femur followed by tibia, craniofacial bones and ribs [2]. Craniofacial involvement occurs in about 30% of monostotic fibrous dysplasia and typically affects the maxilla, mandible and rarely the calvarium. Polyostotic form of the disease has nearly 100% involvement of the craniofacial bones [3].

Clinical presentation of fibrous dysplasia varies with the primary bone involved and the extent of disease. Visual complications occur with sphenethmoidal complex involvement, as seen in both our cases. Fibrous dysplasia has its onset during early life, usually in late childhood or early adolescence. Patients with polyostotic form of disease are considerably younger. There is an equal sex distribution in monostotic fibrous dysplasia but the polyostotic form has a clear female predilection [3].

Radiographic features of fibrous dysplasia vary depending on the amount of bony and fibrous matrix within the lesion and have been sub-classified into three different patterns: pagetoid type 56%, sclerotic type 23% and the radiolucent type 21% [4]. Both our patients had sclerotic type of lesions; which is the commonest form of involvement seen in facial bones and bones of the base of skull. The lytic and pagetoid types usually involve the calvarial bones.

Radionuclide scanning in fibrous dysplasia shows areas of intensely increased uptake, which is due to diffuse microscopic ossification.

Scintigraphy is helpful in determining the activity and potential multicentricity of the lesion, it is specifically helpful in diagnosing when plain radiographs are equivocal [5].

CT accurately establishes the diagnosis and extent of bone involvement. Involvement of optic canals, orbital fissures, frontonasal ducts and ostiomeatal complex can be best evaluated by CT scanning. CT characteristics of fibrous dysplasia, include expansion of the involved bone with heterogenous pattern of CT densities associated with scattered or confluent islands of bone formation. CT attenuation levels have been reported to range from 34 to 513 HU depending on the fibrous tissue and bone content [6].

On magnetic resonance imaging, fibrous dysplasia exhibits homogenous, moderately low signal intensity on T1 weighted images. On T2 weighted images the tissue usually exhibits very high signal intensity. After intravenous Gd-DTPA, lesions display moderate to significant central contrast enhancement with some rim enhancement. The degree of contrast enhancement on T1 weighted images depends on amount and degree of bone trabeculae and collagen present [7].

Both CT and MRI are excellent imaging modalities in defining the constrictive effect of craniofacial fibrous dysplasia on the orbit, optic canals and adjacent paranasal sinuses. Treatment is surgical with unroofing of optic canal or cosmetic remodelling of the orbit to provide adequate room for the intraorbital contents.

References