

## CASE REPORT

# Fibrous Dysplasia of the Temporal Bone

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## Abstract

We report a case of monostotic fibrous dysplasia of temporal bone in a 15-year-old male which presented with mass in the temporomastoid region. There was no associated complaint and patient came only for cosmetic correction of the asymmetry of skull. Conductive hearing loss was an incidental finding on audiogram. Computed tomography showed increased bone thickness with ground glass appearance. Shaded surface display of the skull was used in assessing the external bony deformity. Patient was operated for contouring of the bone and canalplasty which improved the cosmetic appearance as well as corrected the hearing loss thereby avoiding an extensive surgery. Patient is under regular follow-up with good cosmetic and functional results. This case highlights the importance of conservative surgery and regular follow-up in selective cases of temporal bone fibrous dysplasia along with use of shaded surface display for planning the correction of external deformity.

**Keywords:** Fibrous dysplasia, temporal bone, contouring of bone.

Fibrous dysplasia is a slow, progressive and benign fibro-osseous disorder of unknown etiology.<sup>1</sup> von Recklinghausen first recognised fibrous dysplasia as a clinical entity in 1891, although prehistoric and Anglo-Saxon specimens have been identified.<sup>2</sup> The term “fibrous dysplasia” was coined by Lichtenstein in 1938, although, it was first described by McCune and Bruch in 1937.<sup>3</sup> Three clinical types have been described namely; monostotic, polyostotic and McCune, Albright syndrome, with the monostotic variant accounting for 70% of the cases.<sup>4</sup> Temporal bone involvement is rare, but when it occurs, it typically presents with conductive hearing loss, mass in the temporal area and external auditory canal stenosis.<sup>5-7</sup> We report a new case of monostotic fibrous dysplasia involving the temporal bone in which swelling over the temporomastoid region as the presenting complaint.

## CASE REPORT

A 15-year-old male presented in our out-patient department with a swelling in the right postauricular region, which was pushing the pinna laterally. The swelling was very slow growing starting about 10 to 12 years back but at the time of presentation it was involving the whole of temporo-mastoid region (Fig. 1). The boy was very concerned about the cosmetic appearance because of the swelling. There was no history of any vertigo, tinnitus, subjective hearing loss or facial weakness. On examination, the swelling was bony hard involving the right postauricular area and mastoid

region. Otoscopy revealed a markedly narrow external auditory canal on right side compared to the left side. Rest of his general physical and systemic examination was normal.

Computed tomography (CT) scan of temporal region revealed increase in the bone thickness of right temporal bone with homogeneously increased ground glass density and loss of trabecular pattern (Fig. 2). Shaded surface display (SSD) showed a marked asymmetry of the skull in mastoid region (Fig. 3) and served as a guide in the surgical contouring of the external deformity. Facial nerve canal and internal acoustic meatus were normal in calibre. Pure tone audiogram showed mild conductive loss in the right ear with normally hearing left ear. Bone scan was showing increased uptake in right temporal bone region.

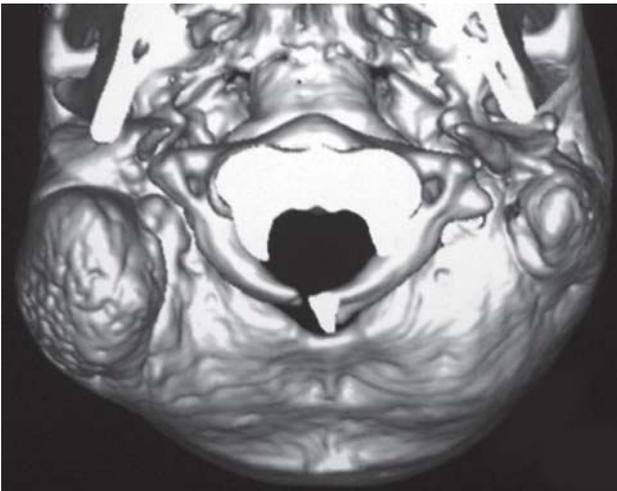
A biopsy was taken from the mastoid region which on histopathological examination showed irregularly shaped trabeculae of bone in cellular fibrous stroma, consistent with the diagnosis of fibrous dysplasia (Fig. 4). All other biochemical tests were normal. Patient wanted a cosmetic correction for the bony swelling and was not finding any hearing disability. The patient was taken up for surgery by a postauricular approach. Partial resection/contouring of bone was done using chisel and hammer and drill along with canalplasty by drilling the posterior bony canal wall using a micro-drill and the epithelial debris in the external auditory canal was cleaned. Postauricular incision was closed in layers over a suction drain and external auditory



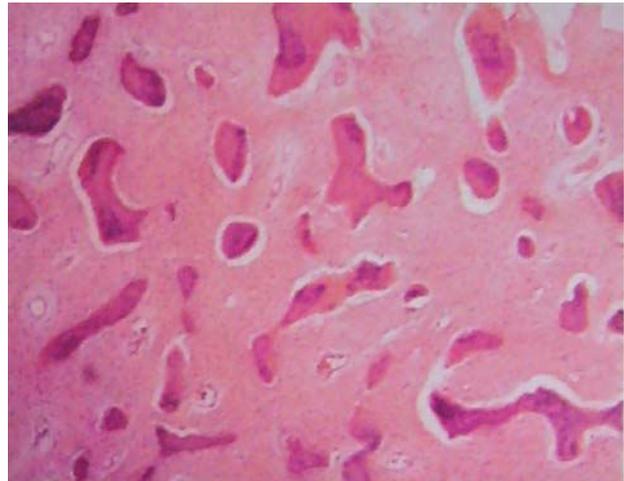
**Fig. 1:** Showing smooth bulge in the right temporomastoid region with shifted hair line and pinna pushed laterally



**Fig. 2:** Computed tomography showing an increased bony thickness with homogenous increased ground glass density and loss of normal trabecular pattern



**Fig. 3:** Shaded surface display (SSD) of the skull base showing gross asymmetry in both mastoid regions



**Fig. 4:** Photomicrograph showing irregularly shaped trabeculae of bone in cellular fibrous stroma (H & E X40)



**Fig. 5:** Follow-up picture after one year showing normal position of pinna

canal was packed with antibiotic soaked pack. Postoperative period was uneventful (Fig. 5) and the patient is symptom free during the follow-up period of more than one year.

## DISCUSSION

Fibrous dysplasia is a genetic but nonfamilial disorder encompassing a wide range of phenotypes, including skeletal deformity, neurological compromise and endocrinologic dysfunction. Early workers believed it to be caused by arrest of bone at an immature stage of development or a disturbance of bone maintenance and turnover.<sup>8</sup> The actual cause has recently been defined as a set of postzygotic missense mutations in the GNAS1 gene, located on chromosome 20q13.2, that normally codes for the alpha subunit of the Gs<sub>α</sub> second-messenger protein.<sup>9,10</sup> Gs<sub>α</sub> is a ubiquitous heterotrimeric G-protein responsible for linking cell-surface receptors to the effector enzyme adenylyl cyclase, promoting intracellular generation of cyclic adenosine monophosphate (cAMP).<sup>11</sup> Fibrous dysplasia results from a constitutive activation of the protein. Fibrous dysplasia develops as a result of abnormal growth and differentiation of marrow stromal cells<sup>12</sup>, with the mutation stimulating proliferation and inhibiting differentiation of bone progenitor cells partially through cAMP-dependent activation of the c-fos gene.<sup>13</sup> The lesion is never inherited and is probably lethal in germline cells.<sup>11</sup> The male to female ratio for the disease in general is equal,<sup>14</sup> but among younger patients males predominate.<sup>15</sup> Three types have been described: monostotic, polyostotic and polyostotic with associated endocrinal abnormalities (McCune-Albright syndrome). Monostotic is the most common form present in 70% of the cases. The temporal bone is involved in 10% of the cases.<sup>5,7</sup>

Fibrous dysplasia of the temporal bone often begins as progressive, painless swelling of the mastoid or squama which may be accompanied by problems relating to the temporomandibular joint or external auditory canal.<sup>5</sup> The most common complaint of patients with fibrous dysplasia of temporal bone is progressive hearing loss, usually conductive, although 17% have sensorineural loss. Involvement of middle ear structures, including erosion of the ossicles, otic capsule, or fallopian canal, can lead to labyrinthitis or, in 10% of patients, facial palsy.<sup>6</sup> Patients may also complain of pulsatile tinnitus, otalgia, or otorrhea. All the areas of the temporal bone, including the otic capsule can get involved with the disease and any cranial nerve can potentially be affected.<sup>5</sup>

In the present case, a painless swelling which gradually progressed to cause a huge bulge in the temporomastoid region was the presenting complaint. There was no

associated complaint but pure tone audiometry revealed mild conductive hearing loss. Clinical and CT scan findings suggested a diagnosis of fibrous dysplasia which was confirmed by biopsy and histopathological examination. SSD of the skull was used in this case which gave exact extent of external bony deformity.

Fibrous dysplasia is usually diagnosed on clinical grounds. The differential diagnosis includes eosinophilic granuloma, aneurysmal bone cyst, giant cell reparative granuloma, osteoblastoma, meningioma, aneurysmal bone cyst, hemangioma, osteoma, osteosarcoma, Paget's disease, otosclerosis, osteogenesis imperfecta, osteopetrosis, and metastatic disease. Surgical biopsy allows histologic examination of tissue, revealing lesions made of mesenchymal cells with the general appearance of spindle-shaped fibroblasts.<sup>16</sup>

Three classical patterns of radiological features have been described for fibrous dysplasia of skull base and facial bones. The pagetoid or ground glass pattern is most common (56%) followed by sclerotic (23%) and cystic variety (21%).<sup>5</sup> CT scan findings are pathognomonic and also aid in monitoring the progress of the disease.<sup>17</sup> Furthermore, CT scan often assists with differentiating fibrous dysplasia from other osteodystrophies of the skull base including otosclerosis, osteogenesis imperfect, Paget's disease, osteopetrosis, hemangioma, meningioma and mucocele. CT scan can identify typical ground glass appearance of the lesions, asymmetry of the skull with thickening of the cranial cortex, involvement of particular cranial bones and the presence of cystic changes.<sup>14</sup> Magnetic resonance imaging is generally not as useful as CT scanning in examining patients with fibrous dysplasia.<sup>18</sup> Diagnosis of fibrous dysplasia is confirmed with a combination of radiographic and histopathologic data.<sup>19</sup> The present case showed the radiological picture consistent with the pagetoid pattern showing a ground glass appearance with marked asymmetry of skull.

Medical treatment for fibrous dysplasia is limited to symptomatic relief. Bisphosphonates have been shown to reduce bony pain and the incidence of fractures.<sup>20,21</sup> Surgery is done for cosmetic or functional reasons<sup>22</sup> with surgical treatment offering the only hope of cure although the decision to operate in areas commonly affected, such as the skull base, requires a careful comparison of the morbidity caused by a surgical resection and that caused by the disease itself.<sup>23</sup> Pathologic bone is spongy and soft and is easily curetted away, although recurrences are common. A subtotal resection in the vicinity of critical structures is acceptable. A definitive indication for surgical intervention in fibrous dysplasia of the temporal bone by an otolaryngologist is stenosis of the external auditory canal with resultant cholesteatoma.<sup>24</sup>

In the present case, surgical intervention was done as the patient wanted a cosmetic correction for the asymmetry of skull and for widening of external auditory canal as pure tone audiogram was showing a conductive hearing loss. Use of shaded surface display in fibrous dysplasia of temporal bone has not been reported in literature. We did not perform the prophylactic decompression of the facial nerve as has been recommended in the literature.<sup>25</sup> Malignant transformation in fibrous dysplasia is rare, occurring in only 0.4% of patients. Radiation treatment of fibrous dysplasia is best avoided because of the tenfold increase in likelihood of sarcomatous transformation.<sup>5</sup> On average, 13.5 years pass between diagnosis and progression to osteosarcoma, fibrosarcoma, chondrosarcoma, or giant cell sarcoma.<sup>24</sup> The risk is tenfold greater, however, in patients with McCune-Albright syndrome and is 100-fold greater in patients who have been irradiated.<sup>24,26</sup> In patients where no definitive surgical indications exist, interval CT scans have been recommended to determine the rate of growth of the lesion.

## CONCLUSION

Fibrous dysplasia is a benign disorder which involves temporal bone uncommonly. Imaging modalities like CT scan help in making the diagnosis and histopathological examination confirms it. Surgery in the region adjoining vital structures should be reserved for patients with functional impairment or a cosmetic deformity and a subtotal resection in the vicinity of critical areas is an acceptable option.

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