Case Report

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Fibrous Dysplasia of Temporal Bone with Secondary Cholesteatoma: A Rare Presentation

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Abstract

Fibrous Dysplasia is a benign, non-familial condition in which normal bone is replaced by fibrous tissue and woven bone. It rarely effects temporal bone. The diagnosis is usually based on clinical, radiographic, and histopathologic features. We here present a case of secondary cholesteatoma caused by external auditory canal occlusion by fibrous dysplasia which was successfully treated by radical mastoidetomy and partial cavity reconstruction by muscle periosteal flap with adequate meatoplasty.

Keywords: Fibrous Dysplasia; Monostotic; Polystotic; Temporal Bone.

Introduction

Fibrous dysplasia (FD) is a congenital skeletal disorder characterized by thinning of the cortex and replacement of the marrow with fibrous tissue that demonstrates characteristic ground-glass appearance on radiography with X- Rays and High Resolution Computerized Tomographic (HRCT) scanning. Von Recklinghausen first recognised fibrous dysplasia as a clinical entity in 1891. The term "fibrous dysplasia" was coined by Lichtenstein in 1938, although, it was first described by McCune and Bruch in 1937. There exists a mainly self-limiting form of fibrous dysplasia classified as monostotic (unifocal), which is characterized by dysplastic bone in a single location and a polyostotic (multifocal) form, which can exhibit aggressive growth placing adjacent structures at risk for compressive sequel.It may sometimes present as McCune-Albright syndrome in which it is accompanied by precocious puberty, endocrine disorders and ''cafe au lait'' skin pigmentation [1,2]. The preferred sites include the diaphyses and metaphyses of long bones, ribs, pelvis, shoulder and craniofacial skeleton. The lesions have been reported to account for 2.5% to 7.0% of all benign bone tumors, with an equal predilection for both sexes [3-5]. In head and neck region, the skull and facial bones are involved in 10-25% cases of monostotic and in 50% cases of polystotic FD. Out of all craniofacial involvement temporal bone is involved in 24% of cases [6,7].

The numbers of case reports of fibrous dysplasia of temporal bone (FDTB) with secondary cholesteatoma are very limited. A fewer cases have been reported from India. We here report a case of FDTB presenting with cholesteatoma and subsequent hearing loss.

Case Report

A 30-year old male presented in outpatient department with the history of canalplasty for fibrous dysplasia in right ear about 12 years back. The patient had lost the complete records of previous treatment. After surgery patient was apparently fine for about 2 years after which he developed complaints of gradually progressive loss of hearing, pain and discharge from right ear. There was no

history of tinnitus, vertigo, headache, facial asymmetry, swelling anywhere else in the body or any other neurological deficit. On external examination of the ear the pinna, pre and post-aural regions were normal. On otoscopic examination, a bony hard swelling was seen obliterating the right external auditory canal. Left ear was normal. HRCT scan of temporal bone revealed the characteristic ground glass opacification mainly of squamous and mastoid portion of the right temporal bone, and to a lesser extent the petrous part. This appearance was consistent with that if fibrous dysplasia. The bony labyrinth was normal. The condition did not involve any other bone of skull. The cortex appeared thinned out and external auditory canalwas obtruded by the bony mass. The canal medial to the obstruction, the middle ear cavity, aditus and the antrum were all expanded, their landmarks were distorted and they were occupied with a soft tissue mass. No ossicles were seen except the head of malleus [Figure 1, 2, 3]. Pure tone audiogram showed severe mixed hearing loss with air-bone gap of 54 decibel in right ear. In left ear hearing was normal. All other haematological and biochemical investigations were normal.

Patient's consent for surgery was taken after he was explained prognosis and the possibility of recurrence. Patient was taken up for surgery by post-auricular approach. EAC was obstructed by a bony hard swelling. Skin over the swelling was raised and the bone underneath was drilled. Medial to the bony swelling, cholesteatoma sac was found which eroded the posterior canal wall and extended into antrum, attic, peri-sinus air cells, hypotympanum, Eustachian tube area and peri-facial cells [Figure 4].

All surgical landmarks in the middle ear appeared displaced. Roof and floor of canal, tegmen tympani, and anterior wall of middle ear were pushed and partially eroded. Only head of malleus was found in attic, incus was completely eroded and stapes footplate present. Mastoid cavity and posterior wall reconstruction was done with inferiorly based musculo-periosteal flap (Singapore flap). Temporalis fascia was placed over foot plate of stapes and type III tympanoplasty was done. Total ossicular replacement was not tried at this stage keeping in mind the extent of cholesteatoma and chances of its recurrence. Wound was closed in layers and post-op period was uneventful. Some of the bone from cortex was sent for histopathologic evaluation which revealed irregular trabeculae of woven bone intermixed with a connective tissue stroma in haematoxylin-eosin, which is consistent with fibrous dysplasia [Figure 5].

Post-operative period was uneventful and patient was discharged on seventh day in a satisfactory condition. After 6 weeks fallow up, post-operative cavity was well epithelised [Figure 6], there was only mild hearing improvement, though the remnant air

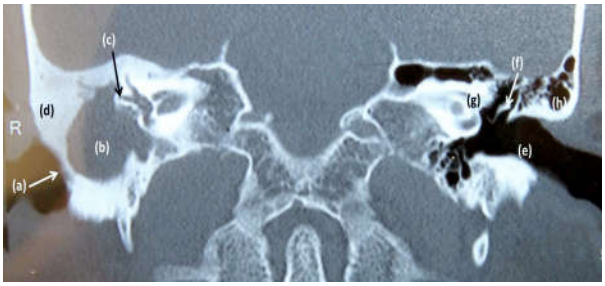

Fig. 1: CT Scan in coronal section showing **(a)** Obstructed external auditory canal of right ear, **(b)** Cholesteatoma cavity, **(c)** Right lateral semicircular canal, **(d)** Fibrous dysplasia in right temporal bone, **(e)** Patent external auditory canal of left ear, **(f)** Incus, **(g)** Cochlea, **(h)** Normal air cells in temporal bone

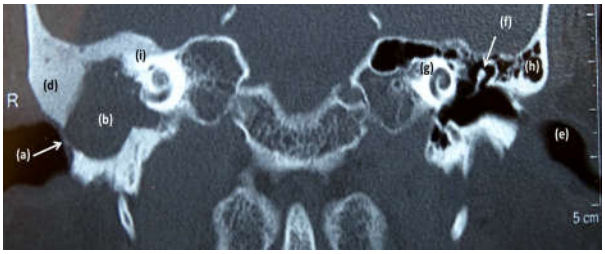

Fig. 2: CT Scan in coronal section showing **(a)** Obstructed external auditory canal of right ear, **(b)** Cholesteatoma cavity, (c) Right lateral semicircular canal, **(d)** Fibrous dysplasia in right temporal bone, **(e)** Patent external auditory canal of left ear, **(f)** Incus, **(g)** Cochlea, **(h)** Normal air cells in temporal bone

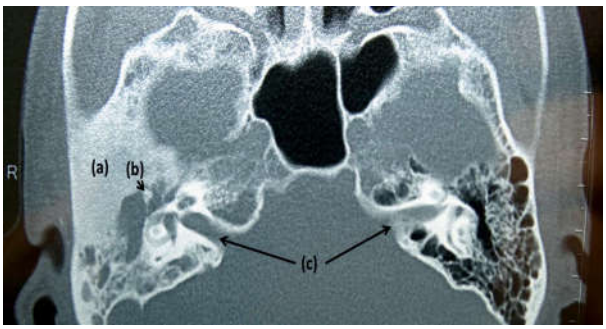

Fig. 3: CT Scan in axial section showing **(a)** Fibrous dysplasia in right temporal bone , **(b)** Head of malleus in attic, **(c)**Internal auditory meatus

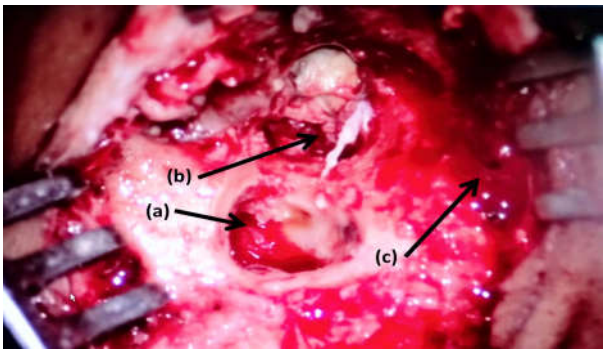

Fig. 4: Per-operative picture showing **(a)** Mastoid Antrum, **(b)** Cholesteatoma in external auditory canal and middle ear, **(c)** Tip of mastoid

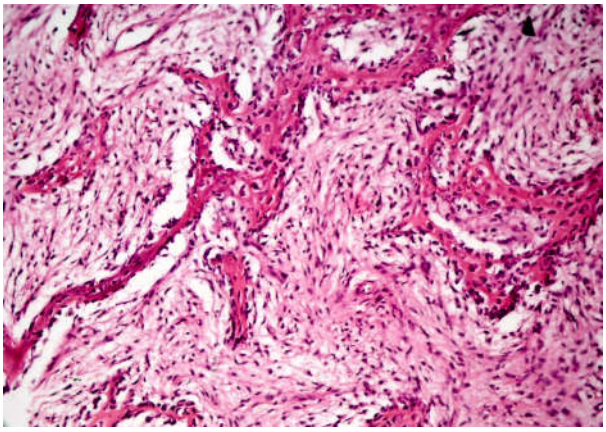

Fig. 5: Histopathological picture showing irregular trabeculae of woven bone intermixed with a connective tissue stroma (haematoxylin-eosin, original magnification X 40)

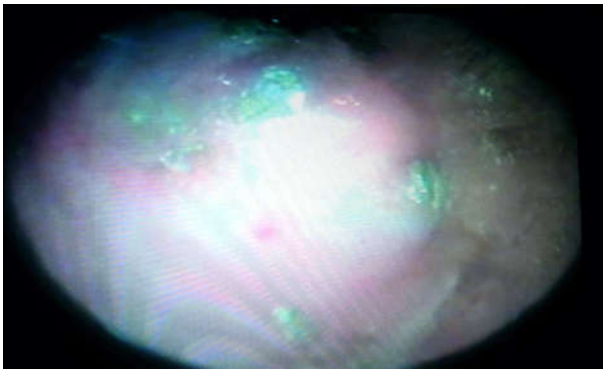

Fig. 6: Well-epithelised post-operative cavity

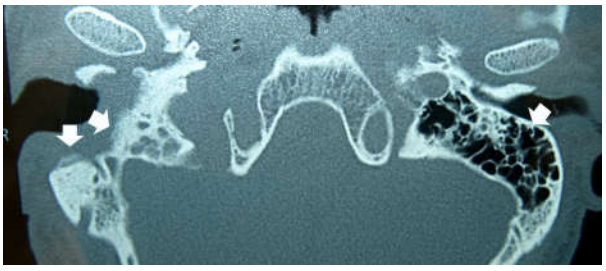

Fig. 7: Dissected mastoid cavity (compare white arrows on both sides) with soft tissue reconstruction of posterior wall

bone gap persisted. CT scan performed after surgeryshowed patent external auditory canal [Figure 7]. Patient was asymptomatic in follow-up forup to 6 months.

Discussion

FD exists in three forms out of which monostatic variety accounts for about 75-80%, polyostotic for about 20-30%, and McCune Albright Syndrome for about 3% cases. Out of all the craniofacial involvement, the ethmoids were the most commonly involved (71%), followed by the sphenoid (43%), frontal (33%), maxilla (29%), temporal (24%), parietal (14%), and occipital (5%) bones [7]. The temporal bone may occasionally be involved, and it may be the site of monostotic, less frequently, of polyostotic FD. The underlying defect in fibrous dysplasia is a mutation of the *GNAS1* gene, which leads to constitutive activation of gene products that preclude the maturation of osteoprogenitor cells and lead to development of abnormal bone matrix, trabeculae, and collagen, produced by undifferentiated mesenchymal cells. Although malignancy is rare (<1%), there is a risk of sarcomatous degeneration, which is increased by exposure to ionizing radiation [1].

The diagnosis of FD is usually based on clinical, radiographic, and histopathologic features. Clinically, the most common presentation is swelling; other manifestations include weakness, localized pain, deformity, fractures, and compromised vision or hearing. Involvement of the temporal bone results in painless progressive enlargement of the squamosal and mastoid, which may manifests as progressive conductive hearing loss due to stenosis of the external auditory canal, sometimes leading to a secondary cholesteatoma and facial nerve palsy, while sensorineural hearing loss and vestibular disorders usually follow otic capsule involvement which occurs due to spreading infection or penetration of inflammatory product and toxins through the round window into the inner ear. This in turn results in cochlear destruction, internal auditory canal stenosis or vestibular fistulisation [7-10]. A clinical staging had been suggested by Barrionuevo et al, in accordance to the progression of the disease. Stage 1 is the latent or asymptomatic phase, where by the management is conservative with regular follow-up. Stage 2 is the symptomatic phase and stage 3 is for those with complications [11]. The case reported here had stage 2 manifestations.

However, the majority are asymptomatic and discovered incidentally on radiographs as lesions with the characteristic ground glass appearance. A radiographic diagnosis is usually sufficient and a subsequent bone biopsy may not be required. However when performed histological examination reveals disorganized bony trabeculae and spindle cells surrounded by a fibrous matrix. A bone scan may be considered to rule out polyostotic FD, and follow-up radiographs recommended every 6 months to ensure there has been no progression. Computed tomography (CT) and magnetic resonance imaging are additional modalities for further elucidating the extent of bony and neurovascular involvement, and total body bone scintigraphy can determine the extent

of skeletal disease and predict functional outcome. 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 [1-4].

Appropriate treatment of FD is often highly individualized and based on presentation. Observational studies report bisphosphonates (like pamidronate) help improve function, decrease pain, and lower fracture risk. Surgical intervention is aimed at preventing functional complications while improving regional asthetics [10]. In the present case, stenosis of the external auditory canal with resultant cholesteatoma was a definitive indication for surgical intervention. The patient underwent radical mastoid exploration with partial reconstruction of the cavity with inferiorly based musculoperiosteal flap and temporalis fascia which healed and epithelized well in the post-operative period.

Conclusion

FD is a benign, non-familial condition which can affect temporal bone and can lead to formation of secondary cholesteatoma. Diagnosis can be made mainly on the basis of radiographs and CT scans, though histology can further confirm the diagnosis. Surgical treatment is directed towards excision of all the resectable bone, choleasteatoma and restoration of as much hearing as possible. Partial obliteration of cavity with inferiorly based musculoperiosteal flapwith adequate meatoplastycan help in achieving shallow and dry cavity. Periodic CT can be used to follow the progression of the disease in post-operative period.

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