Fibrous Dysplasia Involving The Temporal Bone: Report Of Four Cases

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Citation

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Abstract

Fibrous dysplasia of bone is a fibro-osseous tissue disease which has unknown etiology and characterized by replacement of normal bone by a variable amount of fibrous tissue and woven bone. The disease may be unifocal, multifocal or as seen in McCune Albright Syndrome. Fibrous dysplasia rarely involves temporal bone and diagnosis is made with the displacement of neighboring structures. The aim of treatment is cosmetic and functional. We report four cases.

INTRODUCTION

Fibrous dysplasia (FD) is a benign, chronic, slowly progressive disease of fibro-osseous tissue. The disease is characterized by replacement of normal bone by a variable amount of fibrous tissue and woven bone. The term fibrous dysplasia was introduced by Lichtenstein in 1936 ([1]). FD involves one or more bones but never all bones and it extends across a suture line into an adjacent bone. The preferred sites include the diaphyses and metaphyses of long bones, ribs, pelvis, shoulder and craniofacial skeleton. Rarely affects temporal bone ([2], [3], [4]).

The disease may be seen as unifocal (the monostotic form of the disease, MFD), multifocal (the polyostotic form of the disease, PFD) or part of McCune Albright Syndrome. In MFD, single bone like a rib, the tibia or a facial bone, especially a jaw bone may be the site of a lesion. In PFD, two or more bones are involved especially in a lower extremity. McCune Albright Syndrome is characterized by abnormal skin and membrane pigmentation, endocrinological disorders, precocious puberty and associated with PFD ([4]).

The borders of bone lesions are well circumscribed. The lesions grow slowly and unilaterally. The symptoms can be seen because of abnormal extension of bones. The extension of bones causes displacement or compression of neighboring structures or loss of function ([6]). In this article, we reviewed FD and reported four cases.

CASE I

A 22-year-old man attended the outpatient clinic with complaints of a growing mass, pain, and hearing difficulty in his left ear. The history had begun nearly 6 years ago. In physical examination, a solid mass which occupies the mastoid region and pushes the left auricle to outer side was seen. External auditory canal can not be examined because of obliteration. Weber was lateralized to left ear, Rinne was positive at right and negative at left ear. In audiometric examination it is found that right ear, air conduction (AC) 13 dB, bone conduction (BC) 13 dB, left ear AC 70 dB, BC 17 dB. All blood biochemistry was normal. Computerized tomography (CT) scan demonstrated a lesion involving almost all left temporal bone. The lesion had low density and the density was in accordance with fibrous dysplasia (Figure 1). In bone scintigraphy, increased activity was found in left temporal, zygomatic bones, in ramus mandible and in 1/3 upper part of tibia. In other bones the activity was seemed symmetrical and physiological. The diagnosis of FD was made by biopsy under local anesthesia.

Figure 1

Figure 1: Involvement of almost all left temporal bone (case I).

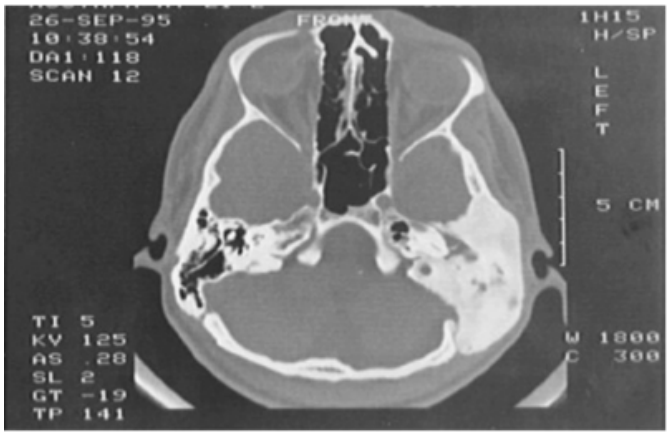
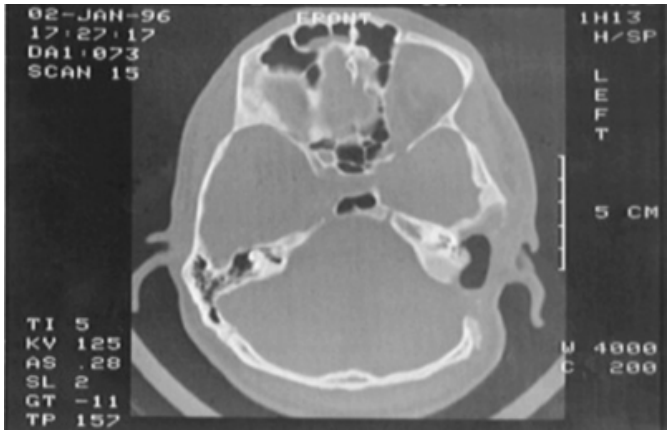


Figure 2

Figure 2: Postoperative temporal bone CT scan of case I.



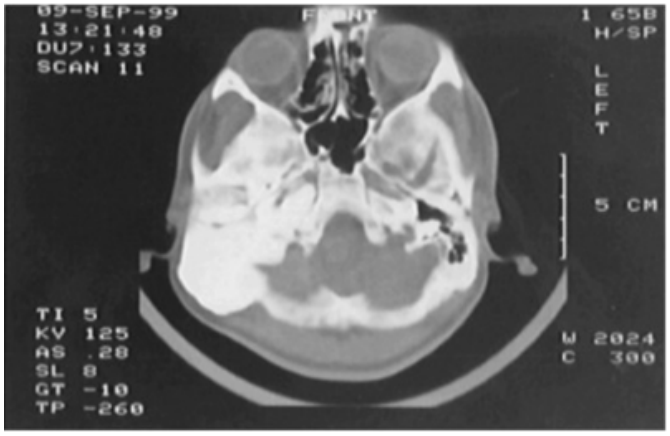
The patient underwent the postauricular canal-wall down mastoidectomy with wide canalplasty under general anesthesia (Figure 2). In the operation we saw that the mass invaded the squamous part, obliterated the external auditory canal. The antrum and mastoid cells were full of granulation tissue and cholesteatoma. The ossicles were eroded and the tympanic cavity was filled with cholesteatoma. The anatomical structures were covered with dysplastic bone, so we could not find an anatomical landmark. The compact structure of petrotympanic suture, facial canal and semicircular canals were not seen. Antrum was in normal place but was partially obliterated. During working on facial matrix, the facial nerve was exposed and the decompression was made carefully. The glenoidal fossa was involved by the fibrotic bone. Abnormal bony structures were debrided without opening the temporomandibular joint. All portions of the temporal bone (mastoid, tympanic, petrous, and squamous) were almost found to be involved. There was not any postoperative complication with the exception of slight facial paresis.

CASE II

A 21-year-old man admitted with the complaints of hearing loss, mass and pain in his right ear for four years. In physical examination a solid mass in the right mastoid region was seen. External auditory canal was severely narrowed. Audiometric examination and tuning fork tests revealed represented right ear conductive hearing loss. All blood biochemistry was normal. CT scans demonstrated a low density lesion involving the whole right temporal bone which is in accordance with FD (figure 3). Bone scintigraphy scans showed increased activity in right temporal bone. The patient underwent canal-wall down mastoidectomy with wide canalplasty under general anesthesia. Entrapment of keratin debris medial to a stenotic canal caused cholesteatoma. The anatomical structures were covered with dysplastic bone and granulation tissue. Because of that we could not find any anatomical landmark for bone conduction recounstruction. Abnormal bony structures and keratin debris were debrided. There was not any postoperative complication.

Figure 3

Figure 3: The low density lesion involved the whole right temporal bone (case II).



CASE III

A 25 year-old-man was complaining for lateral deviation of his eye and a mass on his forehead nearly for 6 years. In physical examination, the extension of frontal bone which was causing an asymmetry in the left side of forehead was seen. The otoscopic examination and audiological tests were normal. Left orbit was pushed downward. All cranial nerves were normal. All blood tests except serum alkaline phosphatase level were normal. CT scan demonstrated a low

density lesion which is in accordance with FD and involved whole left frontal sinus, ethmoid sinuses, lateral orbital wall, frontoparietal calvarium, lateral wall of right frontal sinus, lateral orbital wall, both superior squamous parts of temporal bone and sphenoid sinus. FD caused decreasing of aeration in right ethmoid sinus and sphenoid sinus. (Figure 4 and 5). In bone scintigraphy, increased activity was found in cranium, in upper part of tibia and in pelvis. The patient was not operated because he did not have any otoneurological symptoms and the lesion was diffuse and large.

Figure 4

Figure 4: Involvement of left frontal sinus, ethmoid sinuses, lateral orbital wall and lateral wall of right frontal sinus is seen (case III).

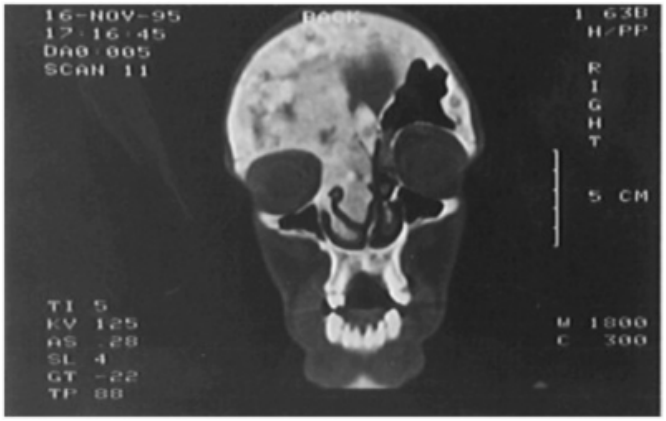
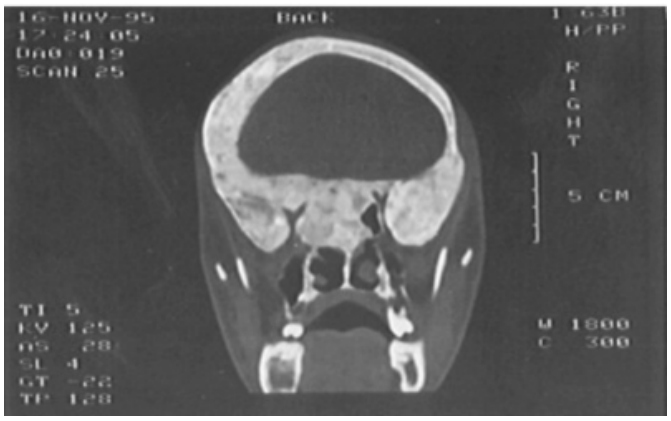


Figure 5

Figure 5: Involvement of both superior squamous parts of temporal bone and sphenoid sinus (case III).

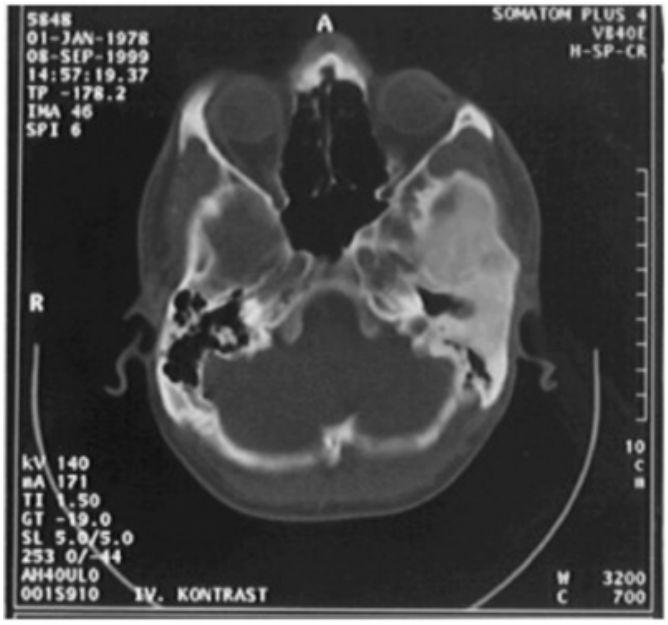


CASE IV

A 21 year-old-man was complaining hearing loss and pain in his left ear for 3 years. External auditory canal was narrowed but not obstructed. Audiometric examination and tuning fork tests revealed left ear conductive hearing loss. All blood biochemistry was normal. In bone scintigraphy, increased activity was found in only left temporal bone. CT scan demonstrated a low density lesion which is in accordance with FD and involved in petrous apex, mastoid, and squamous parts of temporal bone, sphenoid, and whole parietal bone (figure 6). The patient was not operated for his reluctance and little otoneurological symptom.

Figure 6

Figure 6: A low density lesion involved in petrous apex, mastoid, and squamous parts of left temporal bone, sphenoid, and whole parietal bone.



DISCUSSION

The temporal bone may occasionally be involved by fibrous dysplasia. Involvement of the temporal bone results in painless progressive enlargement of the squamosa, mastoid, and external canal. The temporal bone may be the site of monostotic, less frequently, of polyostotic. PFD is differentiated from MFD with severe signs of deformities, frequently spontaneous and recurrent fractures, manifest craniofacial and vertebral affects and tendency to occur on extremities. Diagnosis of MFD is possible with biopsy. On the other hand the typical lesion is enough for PFD ([2], [3]). Radiographic signs were enough for diagnosis our case I, but also supported with biopsy.

The male to female ratio was about 2:1. The ratio of left and right ears affected is approximately equal to each other. In generally the ratio of MFD for about 70%, PFD for about 30%, McCune Albright Syndrome is for about 3% ([3]). The involvement of temporal bones were unilateral in three cases whereas bilateral in other. Our four cases were male.

The radiological features of temporal bone involvement include an increase in size and bone density associated with areas of sclerosis and radiolucency ([3]). There are three different types according to their radiological appearances: pagetoid or ground glass appearance (56%) (Figure-4), sclerotic (23%) and cystic (21%) lesions ([3], [4], [7]). CT scans demonstrated a transition zone between normal and dysplastic bone. The characteristic signs are the expansion of bone, thinned cortical layer and displacement of surrounding structures ([2], [4]). Generally otic capsule is preserved. Labyrinth can be seen as a solitary isle in lesion ([4]). In the first case, otic capsule was normal (Figure 1).

The mass may reach to several centimeters in diameter. A solitary lesion may mimic an eosinophilic granuloma, and multiple lesions may be misdiagnosed as Hand-Schuller-Christian disease ([3], [4]). The incidence of craniofacial involvement is about 10 % in MFD and about 50 % in PFD with moderate skeletal involvement, but reaches to 100 % in PFD with extensive skeletal involvement. Following maxilla and mandible, all the craniofacial bones may be involved but involvement of temporal and occipital bones are unusual. The percentile of temporal bone of involvement between all cranial bones is 18%. As a rule temporal bone is affected unilaterally, but bilaterally involvement may be seen in PFD cases ([2], [3]). Our first and third cases were PFD. In our first case left side of mandible and left temporal bones were affected but in our third case, squamous parts of temporal bones, maxillae, nasal, and parietal bones were affected bilaterally (Figure 1-4, [5]).

Fibroblastic structure in the squamous and mastoid parts of temporal bone are painless and in a tendency of growing ([2]). Sometimes by involving the temporomandibular joint, trismus, and dental occlusions develop. The disease sometimes may cause preauricular pushing or progressive obliteration of external ear canal. The patient complains of a progressive hearing loss caused by increasing narrowing of the external auditory meatus ([3], [4], [7]). By the involvement of sphenoid, ethmoid, nasal and frontal bones, the orbita is invaded and distorted, so the globe displaces to upward and outside then exophtalmos and hypertelorism happens. According to the involved region diplopia, smelling disorders, tinnitus, sensorineural hearing loss or vestibular disorders may happen. Fourth, sixth and seventh cranial nerve palsies may develop as the results of such mass lesions ([3]). Case I, II and IV had conductive hearing loss and pain. In our four patients asymmetrical growing of skull led to cosmetic changes. In case III had orbital invasion caused displacement of left orbita and eyebrow to downward.

The secondary complications of fibrous dysplasia may be secondary external cholesteatoma behind the canal stenosis or obliteration (16-40 %), erosion of the middle ear ossicles inner ear capsule and fallopian canal leading to a labyrinthitis and facial palsy. Involvement of the middle and posterior cranial fossa dura, lateral sinus, jugular bulb and carotid artery may also be seen ([3]). The secondary complication of FD in our series was cholesteatoma in case I and II.

In the differential diagnosis of MFD, solitary unicameral cyst, nonosteogenic fibroma, giant cell tumor of bone, aneurysmal bone cyst, adamantinoma of long bones, eosinophilic granuloma, plasma cell myeloma, fibro-osseous lesions and sarcomatous neoplasm must be considered ([1], [2], [3], [6]). In the differential diagnosis of PFD hyperparathyroidism, polyostotic osteitis deformans, unilateral enchondromatosis, neurofibromatosis and cherubism must be considered ([3]). FD can be differentiated from others with beginning age, distribution pattern and laboratory results (serum calcium, alkaline phosphates, parathormon levels are generally normal). For the real diagnosis of FD histopathologic evaluations are needed ([4]). Typically serum calcium and phosphor levels are normal but if the lesion is active, alkaline phosphatase level may be elevated ([1]). In our case III, the alkaline phosphatase levels was increased but the others blood chemistry results were normal.

FD is a disease of younger people usually manifesting itself in childhood or early adolescence ([3]). MFD is frequently seen in puberty but PFD is generally seen in third decade or later. Generally the first symptom is pain, deformity or repeating spontaneous fractures. If the symptoms are not seen early, it may be diagnosed in late ages ([3]). As in the two cases of Smahua's report, the disease may not be recognized until sixth decade. This may be the result of slow growing lesion ([4]). In our cases the diseases were manifest after 16 ages.

FD disturbs anatomical relationships and causes displacements by proliferating tissue or by covering of this tissue ([4]). The progressive growth of these lesions may cause difficulties in management. The pneumatized cellular structure is replaced by soft proliferative bone, it is impossible to find normal landmarks, so that vital structures such as facial nerve and semicircular canals are lost. These structures are easily vulnerated. Because of these reasons, the operation must be done by experienced surgeons ([3]). In

the operation we performed, we could not find anatomical landmarks. Antrum was in its normal place but was narrowed. Our first patient had slight facial paresis which improved spontaneously in a few days.

The operation must be so conservative that vestibular and cochlear functions can be protected. Secondary complications must be prevented. In the involvement of temporal bone, the treatment depends upon the obliteration of external ear canal. If needed reconstruction must be made for external ear canal. In half of the patients two or more operations are needed ([3]). The patients with FD must be followed up in regular periods. In progressive obliteration of external ear canal, a permanent stent can be placed ([2], [3]).

According to Smauha for creating a wide external ear canal, the abnormal bone must be removed and the cleaned areas must be covered with split thickness skin for preventing soft tissue contraction. The meatoplasty must be wide enough for cleaning the debris and for making postoperative evaluation ([4]). In case I and II, for cosmetic reasons and because of cholesteatoma, postauricular canal-wall down mastoidectomy with wide canalplasty was performed and the cholesteatoma was excised and external ear canal was widened. But silastic stent or split thickness graft was not placed for the possibility that they may not stop the fibrosis. The other two cases were not operated and these patients both are under close follow up.

CONCLUSION

In conclusion, the treatment of FD depends upon the involved area. The patient without any complaint must be followed up. In considering that the anatomical landmarks cannot easily be found, an experienced surgeon must make operation as conservatively as possible.

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