Computed Tomography Findings in Monostotic Fibrous Dysplasia of the Temporal Bone: A Report of Two Cases

TEMPORAL KEMİĞİN MONOSTOTİK FİBRÖZ DİSPLAZİSİNDE BİLGİSAYARLI TOMOGRAFİ BULGULARI: İKİ OLGU SUNUMU

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Abstract

Fibrous dysplasia (FD) is characterized by a progressive replacement of normal bone elements by fibrous tissue. Computed tomography (CT) is the study of choice for diagnosis and follow-up because of its superior bony detail and accurate assessment of the extent of the lesion. In addition, CT can also assist by differentiating FD from other osteodystrophies of the skull base, including Paget's disease, osteogenesis imperfecta, otosclerosis, and osteopetrosis. In this report, we present two cases of FD of the temporal bone associated with CT features with relevance to the literature.

Key Words: Temporal bone; fibrous dysplasia, monostotic; tomography, spiral computed

Özet

Fibröz displazi (FD), normal kemik elemanlarının fibröz doku ile progresif yerdeğiştirmesi ile karakterize bir durumdur. Bilgisayarlı tomografi (BT), lezyonun yaygınlığının doğru değerlendirilmesi ve üstün kemik detayına bağlı olarak tanı ve takipte en uygun radyolojik yöntemdir. Ek olarak, BT, Paget hastalığı, osteogenezis imperfekta, otosklerozis ve osteopetrozis gibi kafatabanının diğer osteodistrofilerinden FD'nin ayrımında da yardımcı olabilir. Bu yazıda, nadir görülen temporal kemik FD'li 2 olguyu BT özellikleri ve ilgili literatür ile birlikte sunuyoruz.

Anahtar Kelimeler: Temporal kemik; monostotik fibröz displazi; spiral bilgisayarlı tomografi

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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 FD 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. It rarely affects the temporal bone.[2-4] The temporal bone is involved in 18% of the cases. FD of the temporal bone is more commonly manifested in the external auditory canal (80%); the inner and middle ear may also be involved. The most common manifestation of temporal bone involvement is progressive conductive hearing loss. The diagnosis depends mainly on radiographic findings. In particular, CT is very useful, assessing the extent of FD within the temporal bone.[5]

We reviewed FD of the temporal bone and reported two cases in this article.

Case Reports

Case 1

A 39-year-old man presented with hearing loss, mass and pain in his left ear for in the left mastoid region. The external auditory canal was severely narrowed. Audiometric examination 6 years. Physical examination revealed a solid mass

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and tuning fork tests revealed left ear conductive hearing loss. The remaining ear, nose and throat examinations were entirely normal. No abnormal skin or cutaneous pigmentation was noted. Cranial nerve palsies were not detected. There was no other bony swelling elsewhere on his body. Blood biochemistry was normal. Audiometry revealed conductive hearing loss of the left ear; the puretone average was 35 dB and the air-bone gap was 12.5 dB. Non-enhanced axial high resolution CT was performed to evaluate the bony involvement and assess the adjacent soft tissue. CT examination of the skull base demonstrated a bony overgrowth with sclerosis of the left temporal bone, particularly in the squamous portion and mastoides (Figures 1 A, B). Diploic space appeared widened. The outer table was either thinned or thickened. Some localized defects of the outer table were demonstrated. The inner table was thinned but no disruption could be found. It produced a stenosis of the left external auditory meatus. The middle ear structures, the jugular foramen, facial nerve canal and temporo-mandibular joint (TMJ) were not affected. The images suggested neither associated cholesteatoma nor facial nerve involvement. The patient had undergone surgical intervention for left meatal stenosis under general anaesthesia, The bony meatus was widened by removing bone from the posterior wall.

Case 2

A 28-years-old woman was referred to our service for a 7 years history of progressive hearing loss without other symptoms. Her personal and family medical history was unhelpful. Clinical examination showed extensive stenosis of the right external auditory meatus. In addition, a solid mass of the right mastoid region was detected. The pinna itself was normal. The rest of the clinical examination was normal without any visible malformations of the head and neck. Laboratory tests were performed and normal values were obtained. Audiometry revealed mixed hearing loss of the right ear; the pure-tone average was 70 dB. Hearing of the left ear was normal. Axial high-resolution CT scan demonstrated a lesion involving almost all of the right temporal bone. CT scan revealed sclerosis and marked expansion of the right temporal bone which had a “ground glass” appearance, along with complete obliteration of the external auditory canal (Figures 2 A, B) and involved the petrous apex, mastoid, and squamous parts of the temporal bone. CT scan demonstrated marked involvement of the middle ear, ossicular chain, and cochlea. Facial

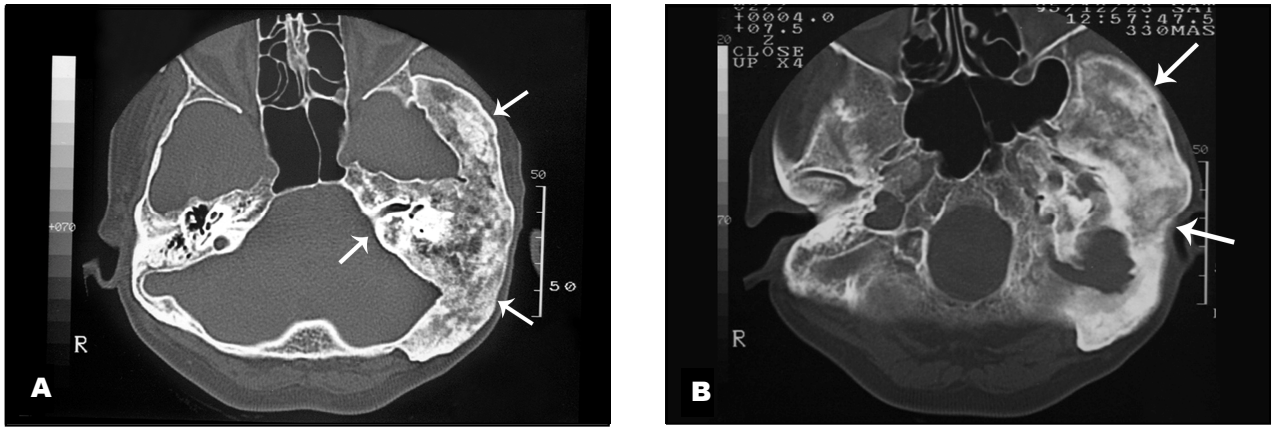

Figure 1. Axial CT scan images of a 39-year-old man who was evaluated for hearing loss. (**A**) Axial image at the level of epitympanic recess shows extensive involvement of the left temporal bone and mastoids by pagetoid FD producing classic ground glass appearance. Note relative preservation of inner ear structures and obliteration of left internal auditory canal (white arrows); (**B**) Axial image at the level of lower mastoid shows extensive bony overgrowth of the left temporal bone with predominantly sclerotic appearance (white arrows).

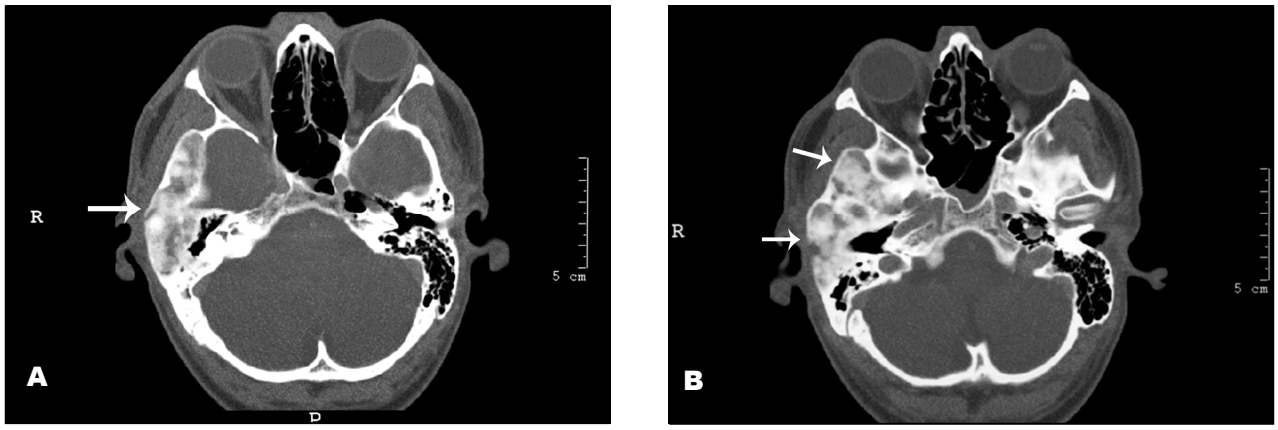

Figure 2. Axial CT images of a 28-year-old woman who had progressive hearing loss due to monostotic temporal bone FD. (**A**) Axial CT section at the level of right internal auditory canal shows mixture of dense and radiolucent areas of disorganized bony trabeculae and fibrous matrix. Lesion reached the right greater wing of the sphenoid bone. It reduced the size of the temporal fossa but did not invade it (white arrow); (**B**) CT scan at the level of the external canal shows obliteration of the right external auditory canal (white arrows).

nerve canal and TMJ appeared normal. In comparison, the left temporal bone was well pneumatized up to the petrous apex. A diagnosis of FD was made based on these findings.

The patient refused biopsy. One year later at follow-up magnetic resonance imaging (MRI) examination, no difference was detected in the imaging findings.

Discussion

FD is a disorder characterized by progressive replacement of normal bone elements by fibrous tissue. Theories about the etiology of this disease include aberrant differentiation of the mesenchyme during bone formation, an arrest of bone at the immature woven stage, or a disturbance of cancellous bone maintenance.[1,2] The mode of transmission has not yet been established. If it is genetic, it is unclear whether FD has an autosomal-dominant or an autosomal-recessive character. It was suggested that monostotic fibrous dysplasia (MFD) occurred secondary to an arrest of bone maturation.[6] In addition, FD was reported to be associated with increased levels of steroid hormone receptors (receptors of estrogens or progesterone).[7]

The disease may manifest 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 jawbone, may be the site of a lesion. In PFD, two or more bones are involved especially of a lower extremity. McCune Albright Syndrome is characterized by abnormal skin and membrane pigmentation, endocrinological disorders, praecox puberty and is associated with PFD.[4] The borders of the bone lesions are well circumscribed. The lesions grow slowly and unilaterally. The symptoms may appear due to the abnormal extension of bones. The extension of bones causes displacement or compression of neighboring structures or loss of function.[5]

MFD usually involves the ribs and femurs. They grow slowly and usually appear to be stationary after puberty.[6] In contrast; PFD frequently becomes evident late in childhood. Patients experience multiple bone involvement, most often of the shoulder, pelvis, vertebral column, and craniofacial skeleton.[7] In PFD, the craniofacial skeleton is affected (usually unilaterally) in 40 to 60% of all cases; in MFD, craniofacial lesions occur in approximately 25% of cases.[8] PFD commonly affects the sphenoid (43%), frontal (33%), maxillary 29%), and ethmoid bones (71%). The occipital (5%) and temporal bones (24%) are less frequently involved. PFD is associated with endocrinopathies in 3 to 5% of all cases.[9] Temporal bone involvement was unilateral in our cases.

The most common symptoms of temporal bone FD are visual impairment and progressive conductive hearing loss caused by occlusion of the eustachian tube or external auditory canal. Sensorineural hearing loss attributed to this lesion, which occurs in 14 to 17% of patients, is the result of either cochlear destruction, internal auditory canal stenosis, or vestibular fistulization.[10] The external auditory canal is involved in 80% of cases. Cholesteatoma occurs in 40% of cases with temporal bone FD.[5] Middle and inner ears are involved after a long period of external auditory canal stenosis.[5,11]

In an extensive review of 69 cases by Nager et al, the male to female ratio was 2:1 and the most common presenting symptoms were progressive hearing loss (56%), increasing size of the temporal bone (50%), and progressive occlusion of the external auditory canal (42%).[3] Some patients even had multiple cranial nerve involvement.[6]

The radiological features of temporal bone involvement include an increase in size and bone density associated with areas of sclerosis and radiolucency. There are three different types according to their radiological appearances: pagetoid or ground glass appearance (56%), sclerotic (23%) and cystic (21%) lesions. CT scan demonstrates a transition zone between normal and dysplastic bone. The characteristic CT signs of FD are ground-glass appearance, thinning of the cortical bone, ballooning of the affected bone, and displacement of surrounding structures. On CT, the petrous bone, the otic capsule, the inner ear, the external auditory canal, the jugular foramen and the TMJ involvement should be studied.[5] Generally, the otic capsule is preserved. The labyrinth may appear as a solitary isle in lesion.[4,5,9,12] In the first case, otic capsule, middle and inner ears were normal. CT scanning, the primary mode for radiographically evaluating FD in our experience is the best way to display the bony changes. It is useful to evaluate soft tissues and fibrous components, and to assess the effect of these primary lesions adjacent to the soft tissue structures of the skull base, jugular vein and brain stem.[5] Plain films and MR imaging are useful adjuncts. Particularly in cases of cystic FD, MR imaging may be useful to assess the soft tissue and fibrous components and to evaluate the effect of these primary bony lesions on adjacent soft tissue structures of the skull base, such as the jugular vein and brain stem. In addition, three-dimensional (3D) CT images are also useful for evaluating the extension of lesions. Conventional CT scan may be used for 3D imaging, but it yields little additional information in proportion to the cost and time required. Reconstructed 3D helical CT images may provide further visual recognition of the fine structure of the temporal bone with much less effort.[13] CT findings may be used for surgical planning to minimize surgery and finally to prepare esthetic bone reconstruction if necessary. Although there is no established rule, the first postoperative CT examination may be performed 6 months later if surgery was complete. If the result is deemed satisfactory, no subsequent CT is scheduled except in case of clinical recurrence. If surgery was incomplete, CT is performed earlier and more frequently depending on clinical and surgical data.[14]

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. The differential diagnosis of PFD include hyperparathyroidism, polyostotic osteitis deformans, unilateral enchondromatosis, neurofibromatosis and cherubism. FD may be differentiated from others by the age of onset, distribution pattern and laboratory results (serum calcium, alkaline phosphates, parathormon levels are generally normal). For the definitive diagnosis of FD, histopathologic assessment is needed. Typically serum calcium and phosphor levels are normal but if the lesion is active, alkaline phosphatase level may also be elevated.[5,6,9,10] In our cases, the alkaline phosphatase levels and the other blood chemistry results were normal.

In conclusion, FD of temporal bone should be one of the differential diagnoses in young patients with progressive hearing loss and external auditory canal stenosis during physical examination. CT

scan is extremely useful for demonstration of the gross bony changes as well as anatomical details. In addition, periodic CT examinations are recommended to follow the progression of the disease and assess the need for any surgical intervention.

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