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CASE REPORT

Temporoparietal Fibrous Dysplasia Complicated by Cholesteatoma: A Case Report

Rifat Karli

Department of Otorhinolaryngology, School of Medicine, Ondokuz Mayıs University Samsun,Turkey

Fibrous dysplasia (FD) is an uncommon benign bone disorder of unknown aetiology. It rarely affects the temporal bone. We report a case of fibrous dysplasia of the temporoparietal bone with cholesteatoma in the mastoid cavity. The patient's complaints included progressive hearing loss, ear fullness and headache. Fibrous dysplasia closed the external ear canal and this stenosis caused the cholesteatoma. We performed tympanoplasty and meatoplasty. We compare this interesting case with similar cases appearing in the literature.

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Introduction

In fibrous dysplasia (FD), normal or fully undeveloped bone tissue shows displacement or irregular osteoids characterized by the formation of fibrous tissue.[1] Fibrous dysplasia is a slow-growing skeletal developmental anomaly that especially involves the long tubular bones, ribs, skull and facial bones. A disturbance of normal bone development occurs: the bone structure is poorly differentiated and defects in osteoblastic differentiation and saturation originate in the mesenchymal precursor of the bone.[2,3] This is a benign process in which bone expands due to abnormal development of fibrous tissue.

FD is a congenital, nonfamilial metabolic disorder that produces 2.5% of all osseous tumours and more than 7% of all non-malignant bone tumours. Involvement of single or multiple bones may occur. Common sites of involvement in the head and neck region are the frontal, ethmoid, sphenoid and maxillary bones. The temporal and occipital bones are least affected.[4] In this paper, we report the case of a patient with fibrous dysplasia of the temporoparietal bone that was complicated by cholesteatoma.

Case Report

During the case management, the current ethics standards were considered.A 65-year-old male patient complained of progressive right ear hearing loss, ear fullness and headache on the right side that had been present for many years. Examination revealed a prominent facial asymmetry with swelling in the right temporoparietal region. The external ear meatus was completely closed. An audiological test, including

Corresponding address:
Rifat Karli
Ondokuz Mayıs Universitesi Tıp Fakültesi , KBB kliniği, Samsun / TURKEY
Tel: +90 530 327 73 67
E-mail: rifatkarli@yahoo.com

pure-tone audiometry, revealed a normal left ear but a right ear with a bone conduction threshold of 45 dB and an airway threshold that could not be measured. Systemic examination of the patient was normal and his medical history was unremarkable. Laboratory investigations were all normal. Paranasal Computerized Tomography (Paranasal CT) (Figure 1) and Magnetic Resonance Imaging (MRI) demonstrated massive thickening of the right temporal bone and parietal bone. A soft tissue density filled the right temporal cavity. There was no external ear canal. The bony overgrowth expanded towards the brain temporal lobe . The patient underwent a right meatal stenosis operation under general anaesthesia. A postauricular incision was made.

The cholesteatoma had filled the entire auricular cavity and the posterior meatal wall was fully destroyed. After cleaning out the pathological tissues, the mastoid segment of the facial canal to the entrance of stilomastoid foremen could be observed. The facial canal showed a two-point partial defect which exposed the facial nerve. The tympanic membrane was centrally perforated and the middle ear showed polypoid tissue. The incus and stapes were defective and the malleus was partially intact. The tympanic segment of the facial canal was intact. The Eustachian tube was open and appeared normal in width. The removed malleus was placed on the oval window. Myringoplasty was performed using the temporal muscle fascia. The mastoid cavity and middle ear cavity were separated into two individual compartments. A wide meatoplasty created a new large external meatus to prevent recurrent cholesteatoma. The pathological diagnosis of FD was made after a punch biopsy of the bone mass.

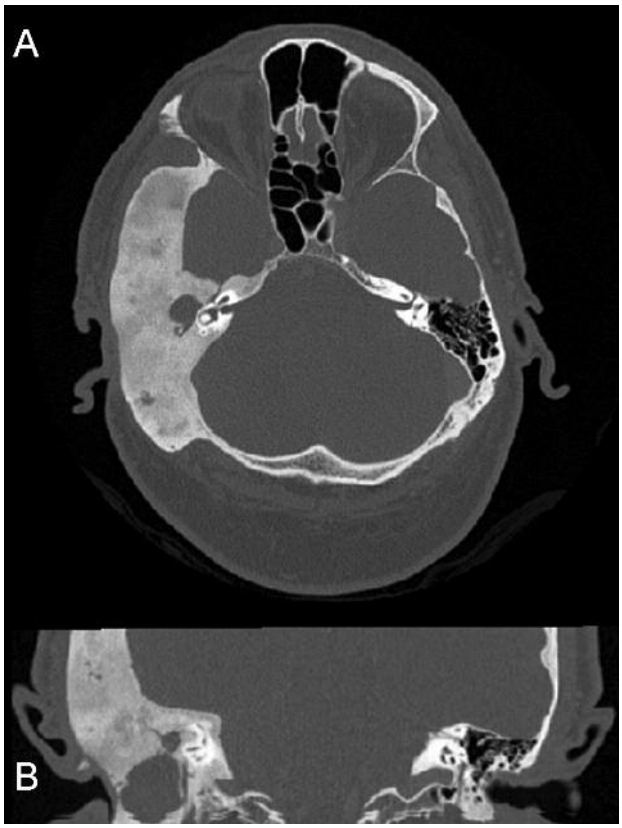

Figure 1. (**A-B**): Coronal (**A**) and axial CT (**B**) shows temporoparietal bone ground-glass appearance.A soft tissue density filled in the right temporal cavity. Inner ear structures and its around bones appear to be normal.

Discussion

Fibrous dysplasia is a disorder in which normal bone is slowly replaced by abnormal fibrous tissue and irregularly arranged bony trabeculae. FD is not a hereditary disease and most commonly occurs between the ages of 3-15 years. The precise etiology of FD is currently unknown.[2,4] FD preferentially occurs in the long bones in the body; however, any bone can be affected. Pouwels and Cremers reported that approximately 18% of cases of craniofacial fibrous dysplasia involved the temporal bone.[5] Even without a priority, the condition may affect the skull, facial and temporal bones. Therefore, this condition is of interest to otolaryngologists. Three clinical forms of this pathology are recognised: 1) the monostotic form (70-80%) that affects only a single bone; 2) the polyostotic form (20-30%) that affects more than one bone either unilaterally or bilaterally; and 3) the polyostotic form that is accompanied by signs of cutaneous and endocrine involvement including goiter, hyperthyroidism, Cushing' s disease and acromegaly. FD in combination with precocious puberty and cafe-au-lait spots is called McCunne-Albright syndrome.[6] We present a case of a patient with polyostotic FD with involvement of both temporal and parietal bone and complications from cholesteatoma.

Fibrous dysplasia itself is a benign disease, but because of its expansive nature, it has the potential to cause severe morbidity when it occurs in the

craniofacial bones. Craniofacial involvement of FD affects the temporal bone in 18% of cases. These cases may be unilateral or bilateral and may affect the facial and neural cranium. Facial nerve involvement is seen in 9% of patients with FD of the temporal bone and cholesteatoma occurs in almost 40%.[7] The most common clinical symptom of temporal bone FD is progressive conductive hearing loss, which is caused by an occlusion of the Eustachian tube or external auditory canal. However, advanced cases may show sensorineural hearing loss and other complications, some of which can be life threatening. In our case, the bone conduction threshold was 45 dB and an airway threshold could not be measured because the external ear canal was completely closed. Facial nerve function was intact but there was facial asymmetry due to asymmetric bone growth.

Complications may develop in two ways: 1: Enlargement of the temporal bone can cause mechanical obstruction and compresses structures; and 2: Cholesteatomas can cause complications. Sensorineural hearing loss can occur secondary to involvement of the cochlear destruction, internal auditory canal stenosis or vestibular fistulisation, which occurs with mechanical obstruction in 14 to 17% of patients.[7] In our case, CT scans indicated involvement of the right temporoparietal bone and showed total stenosis of the external ear meatus and cholesteatoma tissue filling the right mastoid cavity, but the inner ear structures were normal.

The lesion growth in the external auditory meatus leads to progressive stenosis, which results in keratin debris accumulation within the closed external ear canal. Cholesteatoma develops from the accumulating keratin debris over time. Full erosion can occur secondary to cholesteatomas and can result in further complications. Cholesteatomas themselves can also cause complications or can contribute to the spread of infection and products of inflammation (proteins, toxins, local inhibitors, bacteria). These complications lead to permanent sensorineural hearing loss, lateral semicircular canal fistulas, facial paralysis, and intracranial infection, as well as thrombophlebitis. In addition, exposure of the dura can lead to meningitis or lateral sinus thrombosis.[8,9]

Other symptoms include tinnitus, dizziness, pain, trismus and neurological signs related to the involvement of the middle or posterior cranial fossa. A case of presented by Marti and colleagues indicated polyostotic FD with involvement of the temporal bone that was first diagnosed when the patient was 9 years old. His condition eventually became complicated by cholesteatoma and thrombophlebitis of the left transverse and sigmoid sinuses, and he died of his disease at the age of 19 years.[10] This case is an important example that shows the progression of FD.

In our case, dense cholesteatoma tissue throughout the mastoid cavity and external ear canal was massively eroded to the level of the facial nerve canal. The facial canal mastoid segment showed a two-point partial defect that exposed the facial nerve. The centre of the tympanic membrane central was perforated, but its other areas were intact. No middle ear cholesteatoma was evident. Thorough clinical and intraoperative observations did not indicate any evidence for inner ear and neurologic involvement. His facial nerve function was intact.

The differential diagnosis of fibro-osseous lesions of the temporal bone includes FD, meningioma, aneurysmal bone cyst, unicameral cyst, ossifying or nonossifying fibroma, Paget's disease, osteochondroma, giant cell tumour, eosinophilic granuloma exostosis, osteoma and sarcomatous neoplasms.[11] Radiological examination has an important place in the diagnosis and differential diagnosis. CT and magnetic resonance imaging (MRI), rather than direct radiography, are helpful. Craniofacial fibrous dysplasia is usually described as having a ground-glass appearance on CT, with asymmetrical involvement of the cranium and thinning of the cortical wall.[12] CT diagnosis should be the method of first choice as it shows the depth of the lesion. Follow up is particularly important. MRI is a supportive element in the diagnosis. It is especially useful in the differential diagnosis of meningiomas, osteoma, mucocele and soft tissue involvement.[13]

FD is first seen at an early age; therefore, keeping these cases under follow-up is vital. When FD is accompanied by significant clinical symptoms, surgery is planned. Indications for surgery include prominent narrowing of

the external auditory canal, as well as conductive hearing loss, secondary canal cholesteatoma and recurrent infection. Surgery should be aimed at prevention of complications and restoration of function. Non-surgical treatment is not recommended. Radiation therapy should be avoided because of the high incidence (44%) of malignant transformation.[14]

By the time our patient underwent treatment, significant destructive effects of cholesteatoma had already occurred. We saw a mastoid bone structure that was more rigid and much thicker in dimension than normal. Therefore, during the operation, the surgeon must be aware that illusions of depth perception and anatomical cooperation might occur.

Conclusion

In this paper, we have presented a case of temporoparietal bone fibrous dysplasia that became complicated by cholesteatoma and we described its clinical and pathologic features. FD can cause severe morbidity when it occurs in the temporal and craniofacial bones. Early diagnosis and follow-up is important. This is a clinical rarity, but it should be kept in mind by otologists.

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