CASE REPORT

Rare Bone Disorder Affecting the Temporal Bone

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Summary

Fibrous dysplasia is an uncommon benign disorder of unknown etiology. Rarely, it presents isolated in the temporal bone. We present three cases of monostotic fibrous dysplasia that involved the entire temporal bone.

Key Words: Fibrous Dysplasia, Temporal bone

Case Reports

Case 1

A 16-year-old schoolboy presented with a 2-year history of progressively enlarging left postauricular swelling. There was no hearing loss or facial weakness. On examination the left external auditory canal was narrowed with an intact tympanic membrane. CT revealed an opacity involving the entire left temporal bone, extending to the petrous apex and encompassing the labyrinth. It was radiolucent with ground glass or bubbly appearance (Figure 1). A diagnosis of fibrous dysplasia was made on the basis of these findings. Since the patient was asymptomatic and the swelling was not cosmetically obvious, he was managed conservatively.

Case 2

A 28-year-old gentleman presented to us with a left postauricular abscess that was treated with incision and drainage. There was an associated bony hard swelling occluding his left external auditory canal. The patient has had decreased hearing in his left year since childhood. The facial nerve function was normal. CT scan showed expansile mixed sclerotic and lucent lesion involving the squamous, petrous and mastoid portions of the left temporal bone with stenosis of the bony and cartilaginous segments of the left EAC. MRI revealed mainly intermediate signal intensity on both T1 and T2 images (Figure 2). The patient is currently awaiting surgery.

Case 3

A 24-year-old gentleman was seen in our clinic complaining of intermittent left ear foul smelling discharge with reduced hearing since childhood. There was no history of otalgia, tinnitus, vertigo or facial weakness. On examination, his left vertical ramus of the mandible was prominent and there was a bony hard swelling obstructing his left EAC. He had left conductive hearing loss with an air – bone gap of 60dB.

CT scan of the left temporal bone showed marked sclerosis with soft tissue filling up the middle ear and EAC. The left mandibular condyle was enlarged. Left canalplasty and skin grafting was performed. Intraoperatively, there was a large ovoid bony swelling with copious amounts of keratin flakes expanding the EAC. The tympanic membrane and ossicles were not seen. Postoperatively he recovered well, with the EAC remaining patent at 1 month, intact facial nerve and improvement of the air – bone gap to 30dB.

Discussion

Lichtenstein in 1938 coined the term fibrous dyplasia to describe a disorder characterized by the progressive

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replacement of normal bone elements by fibrous tissue. Theories of etiology for 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].

Fibrous dysplasia can present as monostotic, involving a single bone, or polyostotic, involving two or more bones. In the head and neck region, the skull and facial bones are involved in 10-25% of cases of onostotic fibrous dysplasia and in 50% of the polyostotic variety [2].

In our series, all the patients were adolescent or young adults. Their presentation was varied, with left ear mass, left postauricular abscess and left intermittent ear discharge. Two of the patients had conductive hearing loss and one of them saw an improvement of his air conduction following canalplasty. The facial nerve was spared in all patients. The third patient also had contiguous involvement of the adjacent mandible.

In a review of 43 published cases by Magerian *et al*, the common presenting symptoms included hearing loss in 79.1%, temporal bone mass in 25.6%, unilateral otorrhea in 13.9%, otalgia in 6.9%, and trismus in 2.3% [3]. Malignant transformation is seen in approximately 0.5% of patients [4]. Past experience showed an increase in the incidence of malignant transformation to 44% after radiation treatment [4].

CT scan is the best imaging modality to depict bony changes in fibrous dysplasia. The 3 major radiographic classification of fibrous dysplasia are pagetoid (56%), sclerotic (23%) and cystic (21%) [3]. MRI is a useful adjunct especially in the cystic type where the character of the lesion can be delineated and analyzed. Fibrous dysplasia displays low signal intensity on T1W1 and low, intermediate to high signal intensity on T2W1.

Treatment is conservative and surgery is reserved to accomplish three objectives: reestablishment of function, prevention of complications, and cosmetic restoration. Surgery should be carried out in the temporal bone to re-establish hearing and to prevent secondary cholesteatoma. The patient should be informed about the unpredictability of fibrous dysplasia and its tendency to recur.

In conclusion fibrous dysplasia of the temporal bone is a rare disorder that affects patients in the first and second decades of life. Patients commonly present with stenosis of the EAC, conductive hearing loss and can be complicated by canal cholesteatoma. CT scan is usually characteristic and is usually adequate for diagnosis. For equivocal cases MRI and a biopsy of the lesion would be helpful. Most of them can be managed conservatively with surgery being reserved for cosmetic reasons, restoration of hearing or eradication of canal cholesteatoma. The unpredictable nature of this disease mandates long term follow-up of these patients.

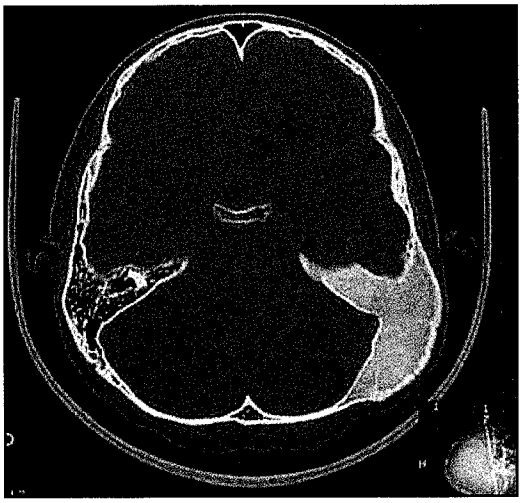

Fig 1: CT showing ground glass appearance of the left temporal bone, extending to the petrous apex. The right temporal bone appears well pneumatized.

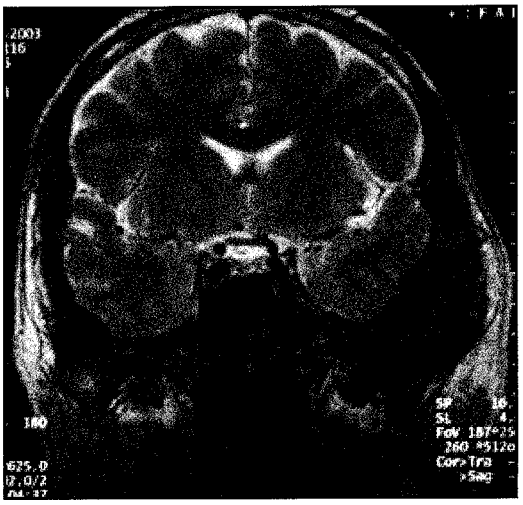

Fig 2: T2 weighted MRI showing intermediate signal intensity in the left temporal bone.

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