CASE REPORT

Fibrous Dysplasia of the Temporal Bone

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SUMMARY

Fibrous dysplasia is a benign disease characterized by a progressive replacement of normal bone elements with fibrous tissue and the temporal bone involvement is uncommon. It has a male:female ratio of 2:1 and is seen more commonly in the first two decades of life. Diagnosis is made based on radiological findings and the modality of treatment is mainly conservative. However, surgery is reserved for preserving function and preventing complications. Fibrous dysplasia in the region of craniofacial bones is of particular interest to the otolaryngologist as it causes deformity and dysfunction that can be debilitating. We present a 49 year old Chinese gentleman with complaints of chronic dizziness over the last one year and had no obvious hearing impairment. Computed tomography of the mastoid revealed fibrous dysplasia of the right temporal bone. This case is of particular interest due to the late presentation as it is more commonly seen in the first two decades of life.

KEY WORDS:
Fibrous dysplasia, Temporal Bone, Management

CASE REPORT

A 49 year old Chinese gentleman presented to our specialist clinic with complaints of persistent dizziness that has been plaguing him for the past one year. He was involved in a motor vehicular accident nine months back where he sustained cerebral concussion and was treated conservatively. He however gave no history of hearing loss, pain or tinnitus and a review of his medical as well as personal history proved to be uneventful. Clinical examination revealed grossly normal ear, nose and throat findings with intact cranial nerves. Detailed hematological and biochemical analyses were performed which included full blood picture, fasting blood sugar, fasting lipid profile, renal profile, lever function test, thyroid function, serum calcium and phosphate levels. Normal values were obtained.

Pure Tone Audiometry revealed bilateral mild sensorineural hearing loss involving the higher frequencies. We proceeded with a High-resolution computed tomography of the temporal bone which demonstrated a focal expansion of the right petrous temporal bone with complete sclerosis of the mastoid air cells and multiple lytic changes. The right middle ear cavity, ossicles and external ear canal were uninvolved (Figs 1 & 2). In comparison, the left temporal bone appears well pneumatized with no apparent abnormalities. Hence a diagnosis of fibrous dysplasia was made based on the above findings.

Currently, as the patient's main complaint is of dizziness, we are treating him symptomatically and will proceed to manage him conservatively. As the natural progression of this disease is self-limiting, he will be kept on regular follow-up and will be offered possible surgical intervention if his hearing becomes further impaired or develops any obstructive symptoms.

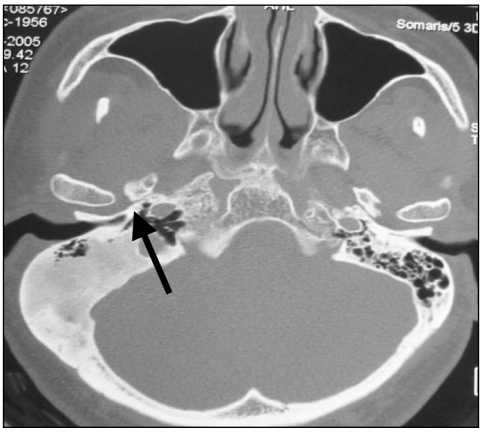

Fig. 1: Axial CT demonstrating bony overgrowth involving the right temporal bone with sclerosis of all the mastoid air-cells. However the right external and middle ear cavities are spared as showed by the arrow. The left temporal bone appears grossly normal.

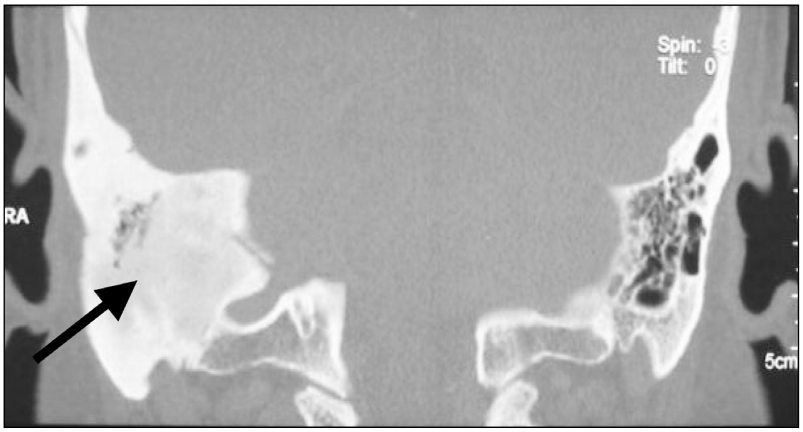

Fig. 2: Coronal High-resolution CT which also demonstrates the bony expansion of the right temporal bone with lytic changes within the mastoid air-cells as shown by the arrow.

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DISCUSSION

Fibrous dysplasia is a benign bony disorder of unknown origin in which normal bone matrix is progressively replaced by cellular fibrous and connective tissue resulting in abnormal and unstable bone formation with reduced mineralization. The disease was first described by McCune and Bruch and Albright *et al* in separate publications in 1937[2]. However, the term fibrous dysplasia was introduced by Lichtenstein the following year. The disease has been found to have three different variants: monostotic which is the mildest and most common involving a single bone, polyostotic involving several bones usually on the ipsilateral side of the body and McCune-Albright Syndrome which is defined by the triad of café-au-lait pigmentation, polyostotic fibrous dysplasia and hyperfunctioning endocrinopathies, such as precocious puberty, hyperthyroidism, hypoparathyroidism, growth hormone excess, hyperprolactinemia, gonadal hyperfunction and Cushing's syndrome[2]. The monostotic variant involves 10-25% of the craniofacial bones and 50% in the polyostotic type. The bones commonly involved were the ethmoids (71%), the sphenoid (43%), frontal (33%), maxilla (29%), temporal (24%), parietal (14%) and occipital bone (5%)[3]. It has a male: female ratio of 2:1 and is seen more commonly in the first two decades of life[1]. The most common symptoms of temporal bone fibrous dysplasia is progressive conductive hearing loss caused by occlusion of the eustachian tube or external auditory canal. Sensorineural hearing loss can also be attributed to this lesion, which is seen in 14-17% of patients. This 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 either cochlear destruction, internal auditory canal stenosis or vestibular fistulization. Facial nerve involvement is seen in 10% of these cases, cholesteatoma in 40% which can lead to ossicular erosion. Other symptoms include tinnitus, dizziness, atypical facial pain and trismus.

There are three classic plain radiological patterns of fibrous dysplasia described. The pagetoid or "ground-glass", where it appears as a mixture of dense and radiolucent areas of fibrosis. The sclerotic pattern is uniformly dense and is seen in two-thirds of the temporal bone cases. The cystic type is characterized by a spherical or ovoid lucidity surrounded by a dense bony shell. However, in the era of the computed tomography and magnetic resonance imaging the above descriptions have little relevance. On CT Scan, fibrous dysplasia is characterized by focal bony expansion with either a heterogenous density or a homogenous "ground-glass" appearance. A High-resolution CT helps to assess four parameters : 1) the degree of external auditory canal stenosis, 2) the involvement of middle or inner ear structures, 3) the presence of an associated cholesteatoma and 4) the possibility of facial nerve involvement. The differential diagnosis of fibro-osseous lesions of the temporal bone includes fibrous dysplasia, meningioma, aneurysmal bone cyst, ossifying or nonossifying fibroma, Paget's disease, osteochondroma, giant cell tumor and sarcomatous neoplasms.

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[4]. Surgical management for fibrous dysplasia of the temporal bone is only indicated for cosmetic deformities, conductive hearing loss due to external auditory canal obstruction or when there is compressive symptoms of the cranial nerves or vessels traversing the temporal bone in particular facial nerve dysfunction. Surgery can be simple shaving for cosmesis or for widening the external auditory canal. A more radical decompressive or temporal bone resection will be required if there is neurovascular compromise.

CONCLUSION

In the absence of curative medicine for fibrous dysplasia, surgery remains the mainstay of therapy. However, it must be kept in mind that this disease is a benign process and the decision to treat depends on the patient's clinical presentation. Asymptomatic lesions can be monitored regularly by serial CT or magnetic resonance imaging scans. Surgical intervention should be reserved for patients who are symptomatic or in the event that the patient develops complications as a result of the sequelae of the disease process. Due to the unpredictability and tendency for recurrence, it should be kept in mind that surgical intervention should be as conservative as possible. Operative procedures should focus on restoration of function, prevention of complications and the restituition of cosmesis. Radiation should be avoided because of the high incidence (44%) of malignant transformation reported[5].

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