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MONOSTOTIC FIBROUS DYSPLASIA OF THE TEMPORAL BONE: A RARE CASE FROM A TERTIARY CARE HOSPITAL.

Otorhinolaryngology

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ABSTRACT

Fibrous dysplasia (FD) is a disorder of bone wherein the normal bony architecture is replaced by abnormal bone of increased fibrous tissue. Monostotic Fibrous dysplasia of temporal bone is a rare occurence with only a few case reports available in literature. This case, reports a rare occurence of Fibrous dysplasia of temporal bone in which there was an entrapment of Squamous epithelium with secondary acquired Cholesteatoma. The evaluation and management of the same has been discussed.

KEYWORDS

INTRODUCTION:

The term "fibrous dysplasia" (FD) was first described by McCune and Bruch in 1937[1]. A rare disorder of the bone characterized by replacement of normal bone architecture with fibrous tissue [2]. It results in abnormal bone expansion and the resultant bony tissue is fragile and the site of the skeletal lesion is weakened and thus prone for fractures and deformity, causing pain and functional impairment[3]. Prevalence of FD is 1-2 cases for every 30,000 births [4]. Incidence in the Temporal Bone is <10%. It can affect a single bone (monostotic – 70-80%) or multiple bones (polyostotic – 10-20%) [5]. The most frequent symptom in fibrous dysplasia is a gradual, painless enlargement of the involved bone/bones in the craniofacial region, clinically seen as facial asymmetry. Diagnosis is based on results of the radiographic examination and histopathological findings [6].

FD is most commonly diagnosed on imaging studies where ground glass opacifications, with alternating dense sclerotic and radiolucent fibrotic areas are visualised[7]. Tissue biopsy confirms the diagnosis [8].

The aim of the surgical treatment in patients with FD is to prevent pathological fractures, control the pain and to reduce bone deformities. Recommended treatment options can be divided into 4 categories:

1. Observation
2. Medical therapy
3. Surgical remodelling
4. Radical excision and reconstruction

Small asymptomatic lesions are best managed with observation[9]. Medical therapy has not occupied a prominent role in the management of fibrous dysplasia to date. Biphosphonates like Pamidronate (60 mg/day) through an intravenous route and Calcitonin drug that is mentioned in literature have been tried.

Conservative surgery involves a remodelling procedure aimed at achieving reasonably acceptable aesthetics and can result in recurrence. [10]. This case report discusses the clinical experience in managing a case of monostotic Fibrous Dysplasia of the Temporal Bone complicated by secondary acquired Cholesteatoma.

Case Report :

A 39 year old male patient came with complaints of swelling in the right side of the face, right ear blockage foul smelling ear discharge for the past 1 year. There were no complaints of giddiness or headache. On clinical examination, a 2 cm swelling over the right side of the face in front of the right temporo-zygomatic region in the pre-auricular region was noted. Stenosis of the External Auditory Canal and keratin debris in the Right EAC was present. The Vestibular and the Facial nerve functions were intact. Tuning fork tests showed a Conductive Hearing loss. PTA was done which reinstated the same.

Investigations:

HRCT scan of the Temporal bone showed diffuse expansion with ground glass matrix of temporal bone involving Mastoid, Tympanic and petrous part with preservation of the bony labyrinth giving an impression of Diffuse Fibrous Dysplasia with lytic lesions within. Large cavity 24x24 mm with soft tissue density and irregular foci in Mastoid and EAC was seen . Occlusion of the EAC was also observed. Bony erosion of sigmoid sinus plate, mastoid cells and mastoid segment of facial nerve was seen. Diffuse soft tissue density of the Middle ear and Expansion with lytic areas involving the posterior zygomatic arch was observed. Erosion of Long process of Incus was present. T2 weighted MRI reveals fluid attenuation within the right mastoid air cells. He was diagnosed to have Fibrous Dysplasia of the temporal bone complicated by secondary acquired cholesteatoma. Histopathological examination showed irregular shaped trabeculae of woven bone in a background of moderately cellular fibres which was suggestive of fibrous dysplasia.

Treatment :

Right Modified radical mastoidectomy with sauserisation of fibrous dysplasia under general anesthesia was done. There was no adverse event during and following surgery.

Follow Up:

We followed up the patient for period of 24 months. He was followed up monthly for 6 months, 3 monthly for 18 months. Pure Tone Audiometry (PTA) was done at end of 18 months which showed Conductive deafness of 40dB Air bone gap. At the end of 24 months, there was no sign of recurrence of Cholesteatoma was noted.

DISCUSSION:

FDTB is a rare entity, although its prevalence can be underestimated due to a percentage of asymptomatic cases [11]. The first notable series was reported by Megerian et al. in 1995, summarizing 43 previously published reports and 10 new cases. They found that 70% of patients had monostotic disease, the majority of whom presented with conductive hearing loss and bony ear canal stenosis[12]. Similarly in our case , the patient has External auditory canal stenosis , Conductive hearing loss along with a single swelling over the right side of the face in front of the right temporo-zygomatic region and the vestibular function and the facial function were intact. Male is to female ratio is 2:1, with prevalence in first 2 decades of life[13]. According to Gyawali et al in 2020, Common presentations in a patient having temporal bone fibrous dysplasia are swelling in the mastoid region, aural fullness and decreased hearing from the obstruction of external auditory canal and there is always a good chance for the development of cholesteatoma [14].

Investigation of choice is HRCT temporal bone. Various CT scan appearances of the disease have been described so far e.g. pagetoid, sclerotic, and cystic. Pagetoid pattern, being the most common, is characterized by a ground-glass appearance and substantiated further histologically with irregular shaped trabeculae of woven bone in a background of moderately cellular fibres. Similarly, in our case HRCT Temporal bone was done which revealed ground glass opacifications with alternating dense sclerotic and radiolucent fibrotic areas. The differential diagnosis of fibrous dysplasia might be eosinophilic granuloma, aneurysmal bone cyst, giant cell reparative granuloma, osteoblastoma, meningioma, aneurismal bone cyst, hemangioma,

osteoma, osteosarcoma, Paget's disease, otosclerosis, osteogenesis imperfecta, osteopetrosis, and metastatic disease[15]. Diagnosis can often be difficult and might require histopathological evaluation. Major differential diagnosis are ossifying fibroma, dentigerous cyst and chondrogenic lesions [16]. It's association with External Auditory Canal Stenosis and cholesteatoma, form a definitive indication for excision. In our case he was diagnosed with Fibrous Dysplasia of temporal bone complicated by secondary cholesteatoma.

Fibrous dysplasia treatment indications were classified by Chen and Noordhoff in 1990 into 4 zones. According to this classification, temporal zone involvement belongs to zone 2 and Surgical treatment is indicated for cosmetic purposes and for symptomatic lesions[17]. Surgical treatment ranges from bone shaving to more extensive surgery. Indications for surgery include bony encroachment of the external auditory canal, recurrent infection and secondary canal cholesteatoma [18]. The surgeon should be aware that the surgery of the dysplastic temporal bone can be hazardous because landmarks are often aletered and obliterated and intra-operative bleeding can be profuse.

CONCLUSION:

Fibrous dysplasia is a benign disorder that may involve temporal bone uncommonly. Imaging modalities like HRCT temporal bone scan help in making the diagnosis and plan further management. Histopathological examination confirms it. Surgery in the region near the vital structures should be done for patients with functional impairment or a aesthetic deformity and a subtotal resection in the vicinity of critical areas is an acceptable option. This case highlights rare occurence of Cholesteatoma in a temporal bone Fibrous dysplasia and it's management.

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