A Rare Case of Fibrous dysplasia of the External Auditory Canal : Case Report and Literature Review

Abstract :

Introduction: Fibrous dysplasia is a rare pathology characterized by the abnormal growth of fibrous tissue, which can manifest as an isolated condition or as part of craniofacial disorders. When it occurs in the external auditory canal, it presents unique clinical challenges. This slowly growing benign tumor is known for its potential to recur, which raises concerns regarding treatment strategies and patient management.

Case presentation : A 21-year-old male patient , was referred for right unilateral progressive hearing loss evolving since one year, without any associated signs , the otoscopic examination found a nearly complete stenosis of the right external auditory canal, preventing the visualization of the eardrum. The audiogram revealed a right conductive hearing loss of 60 db. the temporal CT scan showed a significant thinning of the EAC suggesting right fibrous dysplasia of the external auditory canal. The patient was discussed at a staff meeting, the decision of surgical management was made. which improved his auditory comfort and enhanced his hearing, with no recurrence observed during his follow-up.

Conclusion : the main points emphasize that imaging plays a crucial role in diagnosis, often allowing for confirmation without the need for biopsy. Surgical treatment remains a topic of debate, highlighting the importance of selecting the right indications and timing for intervention. A tailored approach to each patient is essential to optimize outcomes and minimize recurrence.

Keywords : external auditory canal stenosis , external auditory canal fibrous dysplasia, fibrous dysplasia , canaloplasty .

Introduction :

Fibrous dysplasia is a rare condition characterized by a narrowing of the external auditory meatus to less than 4 mm. (1) it can be presented as an independent anomaly; but, it is often associated with microtia and other syndromes. Additionally, it can be an acquired condition that develops following trauma or radiation exposure.

Fibrous dysplasia arises during the second decade of life and the mean age at diagnosis of temporal fibrous dysplasia is 25 to 35 years according to various authors.

The sequelae of this condition can include conductive hearing loss (CHL), cerumen impaction, and cholesteatoma formation.

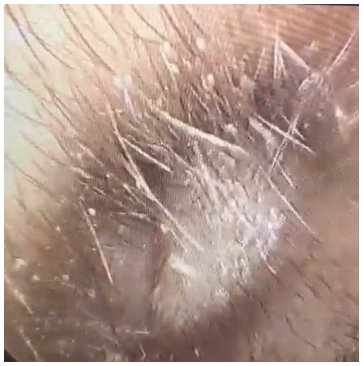
The Treatment in the pediatric population is still debatable. And has historically included monitoring, anticipating growth, or extensive surgery (2)

Patients with fibrous dysplasia must be managed on a case-by-case basis after multidisciplinary consultation. Clinical and radiological monitoring adapted to the site of the dysplasia is systematic.

Case Report :

A 21 years old male patient , with no significant medical history , who was referred to our department for right unilateral progressive hypoacusis evolving since 1 year ,without otorhoea or tinnitus or vertigo or facial paralysis or any other associated signs . all evolving in a context of conservation of the general state .

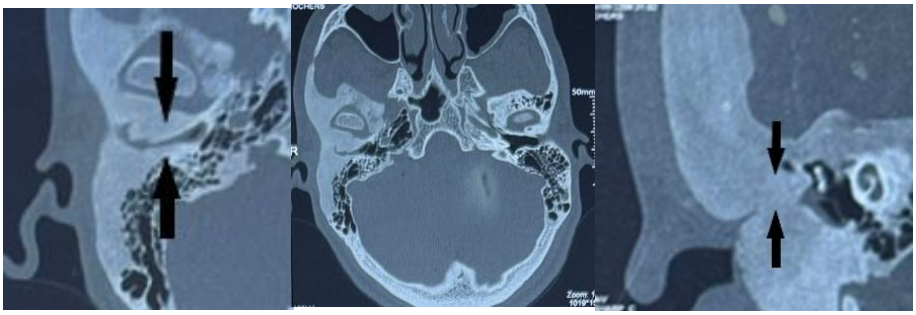
The otoscopic examination revealed an almost complete stenosis of the right external auditory canal, preventing the otoendoscope from passing through, and preventing visualization of the eardrum. In addition, the skin of the meatus appeared healthy and non-inflammatory, ruling out the diagnosis of right otitis externa. The left ear hearing is normal. And the rest of general clinical examination is normal.



Otoscopic image of the external auditory canal stenosis

A tonal audiometry was performedandfound: a right conductive hearing loss of 60 db and an open ear gaps at 45.

The temporal bone CT scan was prescribed and revealed a bone enlargement with a ground-glass appearance of the right temporal bone, significant thinning of the external auditory canal, filling of the mastoid cells, intact ossicular chain, all of which may be related to a right fibrous bone dysplasia of the EAC. The left ear is normal.



Axial / coronal temporal CT scan images: showing the right stenosis of the external auditory canal.

The patient was scheduled for surgical management and underwent a right canaloplasty. This procedure involved elevating a surgical flap to reveal the meatal stenosis. The mastoid was subsequently drilled to remove the stenosis until apparition of the tympanic membrane, then The flap was then repositioned and sutured. The goal of this intervention was to restore the normal anatomy of the ear canal.

In the postoperative follow-up, the patient had no symptoms of recurrence, and the follow-up audiogram revealed remarkable improvement in hearing. The patient also noted a significant enhancement in auditory comfort.Moreover, he didn't experience any recurrence during the followup.

Discussion :

Fibrous dysplasia is a rare condition that can affect any bone of the body, especially the facial bones. The craniofacial bones most commonly affected are the maxilla, zygoma, frontal bone, ethmoid and mandible, while the temporal bone is rarely involved and accounts for only 11 to 12% of all cases of craniofacial fibrous dysplasia. (3)

It is a benign condition, characterized by fibro-osseous proliferation of the facial bone and skull, Bone tissue is transformed into cellular fibrous tissue containing irregular trabeculae. It represents 3 % of bone tumors and 7 % of benign bone tumors. There are two forms: monostotic (70 % of cases) and polyostotic (30 %). (4)

This condition primarily affects young individuals, with an average age of onset around 35 years(5).And presents a high risk of recurrence, highlighting the necessity to systematically inform patients about this risk.(6)

Its physiopathology, described by Weinstein et al. (2) in 2002, involves a genetic mutation on chromosome 20q13.2-13.3, altering protein Gs. The mutation induces abnormal enlargement of cytokine IL6, involved in osteoclast differentiation. (7)

Clinically,the most prominent symptom is the hearing loss. However, symptoms may differ based on the bone'sinvolvement or not, with the primary symptoms of temporal bone involvement being headache and hearing loss. Hearing loss is usually conductive, related to external auditory canal stenosis and/or, more rarely, ossicular chain fixation. (8)

Radiologically, in the temporal CTscan, Diagnosis is indicated by a juxtaposition of "frosted glass" or "ground glass "condensation areas and areas of demineralization. On MRI, the lesion presents as variable isosignal on T1-weighted images, hypersignal on T2 and heterogeneous gadolinium uptake. (7)

MRI can also exclude a retrocochlear lesion in a context of sensorineural hearing loss not explained by fibrous dysplasia (9). biopsy is not systematically performed in the presence of typical CT features in an asymptomatic patient (10)

In our case, a temporal MRI was not prescribed since the patient did not have sensorineural hearing loss. Additionally, the biopsy was not performed due to the clinical context and the imaging findings suggesting fibrous dysplasia.

The differential diagnosis includes osteoma, that must also be systematically considered in view of its relative frequency. However, the diffuse, non-pedunculated nature and bone density lower than that of healthy bone in fibrous dysplasia eliminate this diagnosis. Bone metastasis may mimic the radiological features of fibrous dysplasia and, at the slightest doubt; biopsy and a staging assessment should be performed.
(8)

Temporal bone fibrous dysplasia can lead to complications, including infections and chronic cholesteatomatous otitis. However, this disease generally remains asymptomatic. (8)

Malignant transformation is extremely rare, observed in 0 to 1% of cases but should be suspected in the presence of a rapidly growing lesion. (8)

Although there is no consensus on the treatment of external auditory canal (EAC) stenosis, which remains a controversial topic, the principles of surgical intervention, when the decision is made, include maximal tumor tissue resection, functional restoration, and aesthetic optimization. (5)

Surgical management may also be indicated for aesthetic purposes, to treat external auditory canal stenosis, to eradicate cholesteatoma, to relieve pain or for facial nerve decompression in the case of facial paralysis. (11)

Canaloplasty has a success rate of 56 to 84% and early intervention may allow a better hearing outcome in patients with active lesions. (10)

In children or adolescents, surgery should be performed after puberty whenever possible due to the often rapid growth observed after puberty (12)

Administration of bisphosphonates or calcium and vitamin D supplements can be proposed to stabilize the disease, but the efficacy of these treatments has not been formally demonstrated (12)

Standardized clinical practice guidelines cannot be proposed due to the rarity of the disease, and the unpredictable natural history. (5)

Conclusion :

Fibrous dysplasia of the external auditory canal is a rare condition characterized by a high risk of recurrence. Imaging is a vital tool in diagnosing external auditory canal fibrous dysplasia, often providing confirmation without the need for invasive biopsy. The management of this condition through surgery remains controversial, underscoring the importance of carefully evaluating the indications and timing for intervention. A personalized treatment approach tailored to each patient is key to optimizing outcomes and minimizing the likelihood of recurrence.

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