MÆDICA - a Journal of Clinical Medicine

CASE REPORT

MAEDICA – a Journal of Clinical Medicine
2022; 17(2): 524-527
https://doi.org/10.26574/maedica.2022.17.2.524

Fibrous Dysplasia of the Temporal Bone: a Demanding Entity for Radiologists and ENT Surgeons

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ABSTRACT

Fibrous dysplasia (FD) is a rare, benign, chronic, slowly progressive bone disorder characterized by the replacement of normal bone and bone marrow by fibrous tissue, leading to deformity, pain and functional impairment. It was considered a disease of unknown etiology, uncertain pathogenesis and diverse histopathology. It was later discovered that was caused by a non-heritable activating mutation in the α-subunit gene of the stimulatory G-protein coding gene. Temporal bone involvement is the least frequently reported type, especially in children. The purpose of the current manuscript was to report a rare case of fibrous dysplasia of the left temporal bone of a 17-year-old child who came to the emergency room of our hospital with otalgia and progressive hearing loss on the left ear.

Keywords: fibrous dysplasia (FD), skeletal radiology, temporal bone, child, computed tomography (CT).

INTRODUCTION

Fibrous dysplasia accounts for 2.5% of all osseous neoplasms and 7.0% of all benign bone tumors (1, 2). It may occur at any time of life, but mostly before the age of 30 years (3). The terms "fibrous dysplasia" and "polyostotic fibrous dysplasia" were first mentioned by Lichtenstein in 1938, although von Recklinghausen, a student of Virchow, was credited with the first accurate pathological description of the disorder in 1891 (4). It is a rare, benign, localized bone disorder which results from a non-heritable activating mutation in the α-subunit gene of the stimulatory G-protein encoded by the gene GNAS (5, 6). It is characterized by the replacement of normal bone

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Article received on the 24th of May 2022 and accepted for publication on the 15th of June 2022

and marrow by fibrous tissue, leading to deformity, pain, fracture and functional damage (7).

The disease can occur in either one bone [monoostotic FD (MFD)] or multiple bones [polyostotic FD (PFD)] and can develop singly or in combination with café au lait skin macules and hyperfunctional endocrinopathies, including among others early adolescence, macromegaly, Cushing's syndrome, and hyperparathyroidism (8). Cardiovascular and digestive system abnormalities may also develop (1, 9). The most severe type of FD includes the association with one or more of these extraskeletal features and is called McCune-Albright Syndrome (MAS).

Fibrous dysplasia is more commonly seen in Caucasians (>80% of all cases), while in Asians it accounts for only 1% of cases (9). It affects only the temporal bone and was first described in 1946 by Schlumberger (10). Since then, more than 100 cases of FD of the temporal bone have been reported, including 66 cases in children (2, 9). Fibrous dysplasia is a progressive slowly developing condition that tends to subside after puberty and the best treatment remains unclear in many cases (2, 11). ❑

CASE REPORT

A 17-year-old male came to the emergency department of our hospital due to severe ear pain with feeling of hearing loss on the left ear. Initially, one week before, he was diagnosed with external otitis of the left ear by a private otorhinolaryngologist, who had prescribed a combined antibacterial medical treatment of amoxicillin/potassium clavulanate per os and ear drops, without reversing the symptoms. During the clinical examination, the patient reported headache, ear pain, feeling of fullness of the left ear and hearing loss, respectively.

Otomicroscopy was performed, which showed almost complete occlusion of the external auditory canal with mild swelling and redness, but without the image of external otitis. Examination with the stimulus showed conductive hearing loss on the left ear. Laboratory results showed no inflammation and normal vital parameters. Regarding his medical history, no previous medical conditions were reported, not even in his family history. Given the high suspicion for osteoma, exostosis or tumor, the patient underwent a computed tomography (CT) of the visceral skull.

The CT scan revealed expansive thickening of left temporal bone, like ground glass matrix, which is smooth and sclerotic. It concerned the squamous part up to zygomatic process, the tympanic bone with a significant narrowing of the bony part of the external auditory canal, the anterior part of mastoid and the anterior part of the petrous bone, which was shaped like a pyramid. The pneumatization of the middle ear and the tympanic ossicles were normal. The above image is compatible with fibrous dysplasia of the left temporal bone. The cerebral parenchyma and the rest of the bone structures, except for the left temporal bone, are shown to be normal (Figures 1, 2).

After a preoperative evaluation, including hematological and cardiological tests, a biopsy of the external auditory canal of the left ear was performed with a Herman incision. Neutralized lidocaine with epinephrine was used for local anesthesia. The lesion was removed through an excision, followed by haemostasis with bipolar diathermy.

Biopsy was fixed in formalin and sent to a referral laboratory for histological preparation and examination. We used thin skin sutures to fix the skin and a pressure bandage to prevent the occurrence of a hematoma, that remained in place for five days.

Ten days later, the stitches removed safely as the patient followed all aftercare instructions. Histological examination revealed bone marrow

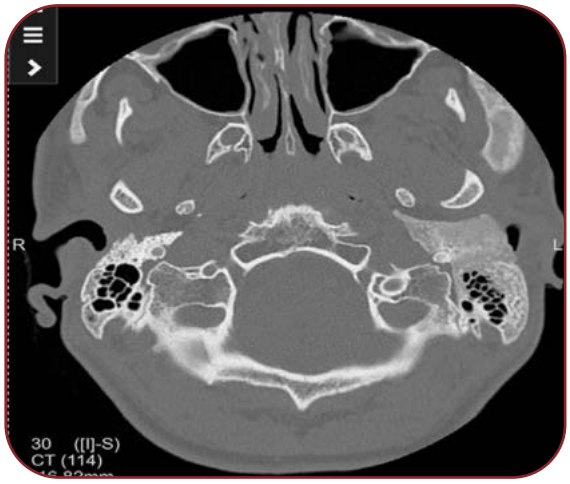

FIGURE 1. Computed tomography showing dense and sclerotic lesions of the mastoid and petrous bone

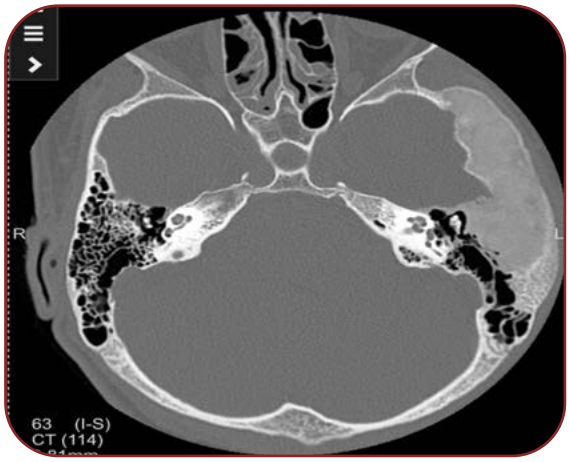

FIGURE 2. Computed tomography showing expansive thickening of the left temporal bone like ground glass matrix

and osseoids surrounded by areas of osteoblasts and osteoclasts compatible with fibrous dysplasia, as expected. The biopsy removed a large piece of tissue resulting in almost complete restoration of hearing and now the patient is under close medical supervision. ❑

DISCUSSION

More than 100 cases of FD with involvement of the temporal bone have been reported in the literature including 66 cases in children (9, 11). Monostotic FD is reported to be the most common manifestation of the disease in 70% of cases, followed by PFD (23%) and McCune-Albright syndrome (MAS) (7%) (7, 12). While the prevalence of MFD is probably greater than PFD, none of these studies reported that patients underwent a thorough screening to determine the full extent of the skeletal and endocrine involvement. In MFD, the zygomatic-maxillary complex is reported to be the most frequently involved region. In PFD and MAS, the craniofacial region is involved in 90% of cases and the anterior cranial base in over 95% of cases (13).

Depending on the type and location of FD, the signs and symptoms vary and include facial deformity and asymmetry, pathological fractures, cranial nerve palsy, visual disturbances, nasal congestion and/or obstruction, pain and paraesthesia. Gradual stenosis of the external auditory canal can also lead to conductive hearing loss, which is the most common symptom of fibrous craniofacial dysplasia in 80% of cases (8). However, another study reports that although more than 70% of patients with craniofacial FD have temporal bone involvement, the majority of them (more than 85%) have normal or near-normal hearing. Middle ear blockage or obstruction of the Eustachian tube can also be an explanation for conductive hearing loss.

Fibrous dysplasia is commonly misdiagnosed as auditory canal exostoses or osteoma. But intraoperatively, in contrast to exostoses, vascularized bone with a cancellous coarse-grained consistency is seen. Most patients are asymptomatic and are diagnosed when a family member or friend who has not seen the patient for a long period of time notices asymmetry or has an abnormality noted accidentally on dental or panoramic X-rays or CT scan of the head and neck (8, 14).

Radiologically, CT imaging is the gold standard and is superior to radiographs. In case of extensive disease, MRI scan is recommended. On CT there are radiolucent areas with soft round borders and sometimes radiopaque areas. Bone lesions can be classified into three main bone patterns: cystic, sclerotic and mixed. An FD defect in the skeleton appears as an area radiolucent ground glass matrix, which is usually smooth and homogeneous, not centrally located within medullary bone. Fibrous dysplasia lesions can range in size from small to large, involving most or all of a long bone. Craniofacial FD typically presents with dense and scleroting lesions, but always maintains a smooth outer contour of the cortex (14).

Regular patient follow-up is the basic principle for FD management. Surgical treatment is used symptomatically or in terms of a biopsy which is done to confirm the diagnosis. Canaloplasty may be useful for the symptomatic treatment of conductive hearing loss or recurrent external otitis. Also, the treatment of FD consists of physiotherapy and treatment of fractures (15, 16).

Patients and their families should be informed of the non-inherited genetic nature of disease and that malignant transformation of FD is rare, occurring in less than 1% of all cases. Malignant changes to osteosarcoma, fibrosarcoma, chondrosarcoma and malignant fibrohistiocytoma have been reported. Risk factors include excess growth hormone and a history of previous radio-

therapy. Worsening of pain and local swelling are suspicious clinical findings (16-20). ❑

CONCLUSIONS

Fibrous dysplasia is a complex unique disease, and knowledge of its pathogenesis includes not only bones but also functional disorders of multiple organs. Radiologists play a key role in diagnosing FD-related bone complications. The craniofacial form of the disease is the most difficult form to manage. Requires clinical, radiological evaluation and follow-up. It is important to properly evaluate these lesions to ensure that a correct diagnosis is made, especially in relation to malignant versus benign mass.

In younger patients, observation and a "wait, see and scan" protocol are recommended until significant functional disturbances or aesthetic problems become apparent. Surgery is not preferred and should be delayed until adolescence because FD tends to stabilize after that. Surgical treatment may be given to patients with severe symptoms, but the safety of this treatment in children remains controversial. Many patients with FD undergo repeated imaging. However, high exposure to radiation is a concern and efforts should be made to reduce the risk of cumulative radiation in these patients. ❑

Conflicts of interest: none declared.

Financial support: none declared.

Ethics statement: The patient provided the written informed consent to participate in this study as well as for the publication of any potentially identifiable images or data included in this article.

References

1. **Couturier A, Aumaître O, Gilain L, et al.** Craniofacial fibrous dysplasia: a 10-case series. *Eur Ann Otorhinolaryngol Head Neck Dis* 2017;134:229-235.
2. **Fattah A, Khechoyan D, Phillips JH, Forrest CR.** Paediatric craniofacial fibrous dysplasia: the Hospital for Sick Children experience and treatment philosophy. *J Plast Reconstr Aesthet Surg* 2013;66:1346-1355.
3. **Camilleri AE.** Craniofacial fibrous dysplasia. *J Laryngol Otol* 1991;105:662-666.
4. **Mierzwiński J, Kosowska J, Tyra J, et al.** Different clinical presentation and management of temporal bone fibrous dysplasia in children. *World J Surg Oncol* 2018;16:5.
5. **Weinstein LS, Shenker A, Gejman PV, et al.** Activating mutations of the stimulatory G protein in the McCune-Albright syndrome. *N Engl J Med* 1991;325:1688-1695.
6. **Landis CA, Masters SB, Spada A, et al.** GTPase inhibiting mutations activate the alpha chain of Gs and stimulate adenylyl cyclase in human pituitary tumours. *Nature* 1989;340:692-696.
7. **Pardo-Maza A, Lassaletta L, Ruiz-Bravo E, et al.** Fibrous dysplasia of the temporal bone secondary to ear surgery: a case report. *Journal of Medical Case Reports* 2015;9:129.
8. **Lee JS, FitzGibbon EJ, Chen YR, et al.** Clinical guidelines for the management of craniofacial fibrous dysplasia. *Orphanet Journal of Rare Diseases* 2012;7(Suppl 1):S2.
9. **Kim YH, Song J-J, Choi HG, et al.** Role of surgical management in temporal bone fibrous dysplasia. *Acta Otolaryngol* 2009;129:1374-1379.
10. **Schlumberger HG.** Fibrous dysplasia of single bones (monostotic fibrous dysplasia). *Mil Surg* 1946;99:504-527.
11. **Megerian CA, Sofferman RA, McKenna MJ, et al.** Fibrous dysplasia of the temporal bone: ten new cases demonstrating the spectrum of otologic sequelae. *Am J Otol* 1995;16:408-419.
12. **Muthusamy S, Subhawong T, Conway S, Temple T.** Locally Aggressive Fibrous Dysplasia Mimicking Malignancy: A Report of Four Cases and Review of the Literature. *Clin Orthop Relat Res* 2015;473:742-750.
13. **Lee JS, FitzGibbon E, Butman JA, et al.** Normal vision despite narrowing of the optic canal in fibrous dysplasia. *N Engl J Med* 2002;347:1670-1676.
14. **Kushchayeva Y, Kushchayev S, Glushko T, et al.** Fibrous dysplasia for radiologists: beyond ground glass bone matrix. *Insights into Imaging* 2018;9:1035-1056.
15. **Weiss N.** Rare Diseases of the Middle Ear and Lateral Skull Base. *Laryngo-Rhino-Otol* 2021;100:S1-S30.
16. **Sadeghi SM, Hosseini SN.** Spontaneous conversion of fibrous dysplasia into osteosarcoma. *J Craniofac Surg* 2011;22:959-961.
17. **Liakos GM, Walker CB, Carruth JA.** Ocular complications in craniofacial fibrous dysplasia. *Br J Ophthalmol* 1979;63:611-616.
18. **Pfeiffer J, Kayser G, Boedeker CC, Ridder GJ.** Posttraumatic reactive fibrous bone neoformation of the anterior skull base mimicking osteosarcoma. *Skull Base* 2008;18:345-351.
19. **Reis C, Genden EM, Bederson JB, Som PM.** A rare spontaneous osteosarcoma of the calvarium in a patient with long-standing fibrous dysplasia: CT and MR findings. *The British Journal of Radiology* 2008;81:e31-e34.
20. **Tsai EC, Santoreneos S, Rutka JT.** Tumors of the skull base in children: review of tumor types and management strategies. *Neurosurg Focus* 2002;12:e1.