

## CASE REPORT

### Temporal Bone Fibrous Dysplasia: A Rare Mastoid Localization

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**Objective:** To present a rare case of mastoid localization of temporal bone fibrous dysplasia and to give some insight on diagnostic and therapeutic strategies.

**Patients:** A 39 year-old female presented with unilateral, left temporal headache. Imaging diagnostic investigation revealed the presence of a cystic lesion localized at the level of the superficial left mastoid bone, surrounded by a ground glass bony halo and plates of bone matrix in its central portion. Due to the uncertain diagnosis as well as to the persistence of the painful symptomatology, the patient was scheduled for surgery.

**Interventions:** Retro-auricular approach extended superiorly, with cortical mastoidectomy and isolation of the lesion, followed by its full removal.

**Main Outcome Measures:** Clear signs of fibrous dysplasia were found, with typical cystic pattern, associated with mature bone tissue fragments, remodeling aspects and multiple foamy histiocytic cells.

**Results:** The patient fully recovered with subsiding of painful symptomatology.

**Conclusion:** Fibrous dysplasia affecting the temporal bone may also occur at the mastoid level, as it was described in the present report. Hence, this rare localization must be taken into consideration for a differential diagnosis of cystic-like lesions of the temporal bone. Since imaging alone is unable to formulate an appropriate diagnosis, surgical removal and subsequent pathological examination reveal to be crucial for the identification of this rare pathology.

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## Introduction

Fibrous Dysplasia (FD) is a genetic, non-familial disorder encompassing a wide range of phenotypes, including skeletal deformities, endocrine dysfunctions and neurologic involvement.

*Three different types of FD have been described:*

The monostotic form, accounting for almost 70 % of the cases, is the most common and affects a single bone in the body, usually a rib or a cranio-facial bone, in rather young population (between 20 and 30 years of age).

The polyostotic form, accounting for almost 30 % of the cases, has multiple bone localizations, with cranio-facial involvement in 50 to 100 % of the

cases, depending on series. In these last two forms both sexes are affected with the same rate.

McCune-Albright syndrome, the most rare and severe clinical expression, which is characterized by a classic triad of symptoms, i.e. multiple endocrine abnormalities, cutaneous hyperpigmentations and polyostotic FD.

Data regarding incidence and prevalence of the disease are scarce <sup>[1, 2]</sup>. FD temporal bone localizations, as monostotic form, have been described in 10 % of the cases, while association with the polyostotic form seems to be more common.

Histological findings show an identical picture in all three subtypes of the disease. The lesion originates

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from the medullary cavity and expands outward in the surrounding cortical bone, replacing it with an irregular array of spindle-shaped, mesenchymal cells, resembling fibroblasts. Macroscopically, the lesion appears to be white to yellow with a gritty consistency, while at microscopic level is characterized by fibrous stromal cells, variable amounts of collagen and irregular trabeculae. The spindle cells are plump, but bland and mitotically inactive and make woven bone, producing the angulated, irregularly shaped trabeculae, commonly characterized as Chinese letters <sup>[3]</sup>.

Among other TB pathologies to consider for a differential diagnosis, it is worth including eosinophilic granuloma, aneurismal bone cyst, giant cell reparative granuloma, osteoblastoma, meningioma, hemangioma, osteoma, osteosarcoma, Paget's disease, otosclerosis, osteogenesis imperfecta, osteopetrosis and metastatic disease.

The gold standard diagnostic imaging is represented by CT scan that could show three different patterns of the disease: pagetoid (ground glass), the most common form (56 %), with a mixture of dense and radiolucent area of fibrosis; sclerotic (23 %), with homogeneously dense lesions; and cystic (21 %), a spherical or ovoid lucency surrounded by a dense bony shell. CT scan could also show asymmetry of the skull with thickening of the cranial cortex, involvement of particular cranial bones and the presence of cystic changes. It also allows a differential diagnosis with other pathologies mostly owing to the ground-glass appearance, the undefined bony margins and the lack of symmetry. CT scan is also useful for the surgical planning since it may show the demarcation between involved and non-involved bone. MRI is not useful as CT scan. T1-weighted images show a low to intermediate signal, with moderate enhancement after gadolinium administration, while T2-weighted images have an intermediate signal. MRI could be useful for ruling out other lesions, such as meningioma, osteoma, osteoblastoma, aneurysmal bone cyst and mucocele <sup>[1-5]</sup>.

In the present paper a rare localization of FD in the temporal bone, i.e. at the upper mastoid level, is presented, with insights on its clinical course, including the therapeutical strategy.

## Case Report

A 39 year-old woman came to our observation complaining of a 10-years history of temporal headache. ENT physical examination was negative and pure tone audiometry showed normal hearing. CT scan showed a bulging deformation of the temporal squama diploe at its junction with the petrous pyramid, featuring a lithic lesion with a rounded shape, surrounded by a ground glass bony halo (Fig. 1). Inside the lesion, plates of bone matrix were mainly found in the central portion, surrounded by a middle-low dense tissue. Internal planking showed marked thinning with lysis points. MRI showed a roundish mass between the temporal squama and the petrous pyramid inside the diploe, eroding the internal planking (Fig. 2). The lesion was characterized by a peripheral pseudocapsule, with intermediate signal in all the sequences (except for a small anterior, T2-hyperintense and T1-tenuously hypointense area of presumed pseudocystic origin), showing no signals of bony or calcium component inside. After enhancement, the lesion showed thin, slightly intense branches. No enhancement of adjacent meningeal plans after contrast was observed.

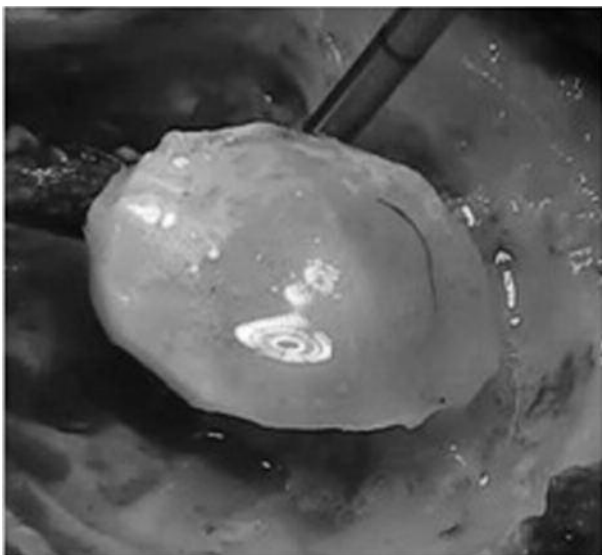


**Figure 1.** Coronal view of CT of the petrous bone. A rounded lesion is localized at the temporo-mastoid fissure, producing lysis of the involved bony walls (arrow). Within the cystic lesion, a few hyperdense structures are present.



**Figure 2.** Enhanced T1-MRI of the brain. This axial view shows a roundish mass between the temporal squama and the petrous pyramid inside the diploë, eroding the internal planking. After enhancement, the lesion showed to be slightly intense, also following thin branches. No enhancement of the adjacent meningeal plans is visible.

On the grounds of these findings, and mostly due to the persistent painful symptom, a surgical management was planned. Via a post-auricular incision, and after the elevation of a musculo-periosteal temporal flap, a slight bulging of about 2 cm on the superficial squamous part of the temporal bone appeared. The lesion (Fig. 3) was isolated by drilling out the mastoid cavity and part of the

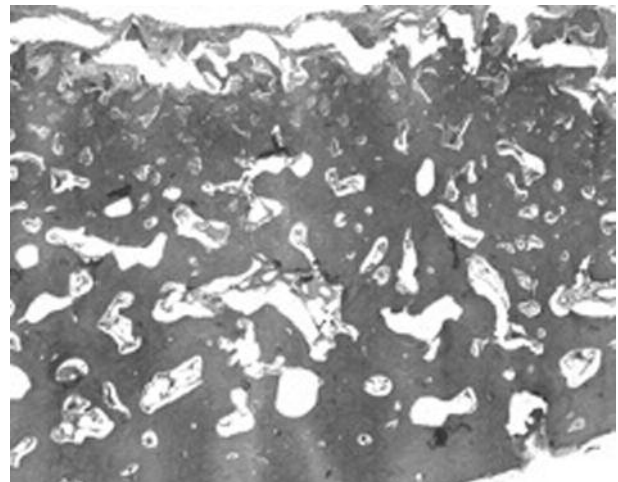


**Figure 3.** Surgical removal of the mastoid FD lesion. The bony mass has been isolated and is just going to be lifted and removed from the underlying dura.

cortex of the temporal squama, where the pedicle of the lesion was identified and eventually detached from the underlying dural layer. The specimen was sent to pathology and the bony defect repaired with bone dust, lyophilized dura and fibrin glue.

Pathology of the specimen showed a cystic pattern, associated with mature bone tissue fragments, remodeling aspects and multiple foamy histiocytic cells (Fig. 4).

One year after the surgical removal, the patient still refers relief from headache.



**Figure 4.** Light microscopy of the FD surgical specimen. Hematoxylin-Eosin staining. Curved and misshaped bone trabeculae, often having a characteristic fish-hook configuration (or Chinese characters), are interspersed within fibrous tissue of variable cellularity.

## Discussion

Owing to the poor clinical symptomatology, the real incidence of cranial FD is difficult to assess. The most involved skull bones are ethmoid (71 %) and sphenoid (43 %). Frontal and maxillary bones can be involved in 30 % of the cases, while temporal and occipital bones are less likely to be affected, i.e. in 24 % and 5 %, respectively. The most common referred symptom in cranio-facial FD is pain, also due to concomitant sinusitis and bone deformity. Regarding temporal bone localization, other symptoms have been described, such as progressive conductive hearing loss due to stenosis of the external auditory canal<sup>[6]</sup>, sometimes leading to a secondary cholesteatoma<sup>[7,8]</sup> and facial nerve palsy<sup>[9]</sup>, while sensorineural hearing loss and vestibular disorders usually follow otic capsule involvement. Neurologic manifestations, such as seizures and, as in the case presented in this report, temporal headache, have also described<sup>[10]</sup>. To our

knowledge, an isolated FD localization at the level of the mastoid bone has only been described by Samani et al.<sup>[11]</sup>, who used the obsolete imaging techniques available in that period, such as conventional x-ray. The present report has therefore overviewed the different aspect of this pathology, enriched by an accurate imaging documentation via CT and MRI.

Different therapeutical attitudes have been proposed for temporal bone FD. Some authors propose the wait and scan behavior, due to the benign nature of the disease<sup>[12]</sup>; others have proposed medical treatment with bisphosphonates to reduce bony pain, especially in the polyostotic or particularly extended forms, where surgery is not indicated<sup>[13]</sup>. Nevertheless, surgery remains one of the main therapeutical options, being able to offer the best clinical outcomes<sup>[14]</sup>.

In the present case surgical removal was mostly selected due to the persisting headache, resistant to painkiller drugs. As a matter of fact, this symptom could be explained by the direct contact of the growing bony mass with the underlying temporal dura, so that, once the lesion was removed, it was progressively subsiding.

In addition, surgical removal has allowed a definite diagnosis of the lesion, since imaging studies were not reliable for differentiating it from other temporal bone lesions. Another important factor in favor of surgical removal in the presented case is represented by the unpredictable natural history and evolution of FD that, in the temporal bone could put at risk vascular and/or nervous structures.

Although malignant degeneration is rare, it has been described to occur in 0.4 % of the patients [15]. This eventful evolution has been raised as a major factor against a possible radiation therapy<sup>[1]</sup>.

Although spontaneous regression of FD has never been described in literature, its recurrence after surgery has been well described<sup>[14]</sup>, thus suggesting the need for a strict longitudinal imaging follow-up of the patient.

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