Fibrous dysplasia of the temporal bone

A 47 year-old man presented with progressive hearing loss.

Imaging Findings:

A 47 year-old man presented to our hospital complaining of progressive hearing loss. The patient's personal and family history were free. Physical examination depicted extensive stenosis of the left external auditory canal, while the ipsilateral tympanic membrane appeared normal. He had no visible abnormalities of the skull. The audiogram showed a mixed type hearing loss. Hearing on the right side was normal. It should be mentioned that an audiogram that had been performed two years ago, was normal in both sides. Laboratory tests were within normal limits. A plain lateral radiograph of the skull revealed bony overgrowth and sclerosis in the temporal area (fig 1a.) High Resolution Computed Tomography (HRCT) of the skull base demonstrated extensive bone sclerosis and thickening of the left temporal bone, particularly its squamous part and the anterior part of the petrous apex. The affected bones had a ‘ground-glass’ appearance with moderately dense new bone. Expansion of the involved bones with preservation of cortical margins were observed. The left middle ear and the ipsilateral mastoid cells were obliterated probably because of the presence of inflammatory tissue. The external auditory canal was narrowed, but the anatomic structures of the left middle ear (ossicular chain, facial nerve canal) and the inner ear were intact. The anatomic structures of the contralateral, right middle and inner ear, were normal (fig2a,b). The patient underwent MR imaging of the skull base that showed very low signal on both T1- and T2-weighted spin echo images throughout the lesion, which corresponds to the areas of sclerosis and hypertrophy of the affected bones. High-signal intensity on T2-weighted images was observed in the left mastoid cells due to the presence of inflammatory tissue. A region of high intensity on T2-weighted images was depicted in the white matter of the left temporal lobe, representing mild edema, due to pressure phenomena by the bony lesions (fig3a). No contrast enhancement was observed, except moderate meningeal enhancement adjacent to the lesion (fig3b). The patient was operated through a postauricular approach. The external auditory canal was widened by removing bone from its posterior wall. A plastic surgery of the canal (meatoplasty) was performed and the meatus was packed temporarily with Gelfoam. Histology revealed neoplastic fibrous tissue which consisted of spindle cells , with no cytological atypia or any abundant collagen stroma. There were no mitotic figures. Newly formed bone trabeculae (woven bone) were observed within the fibrous tissue. No osteoblasts were identified on the surface of the trabeculae. The patient was discharged the third postoperative day, with no remarkable problems. Follow-up was performed every month for the following year and a complete recovery of the hearing was observed. A year later the patient underwent another CT, which showed extension of the bony lesions of the contralateral temporal bone (fig 4a). These findings suggest progression of the disease. Nevertheless, our patient has not shown any symptoms (or hearing disorders) , three
years after surgical decompression of the left external auditory canal.

Discussion:

Fibrous dysplasia (FD) is a bone disorder in which normal medullary bone is replaced progressively with abnormal fibrous tissue and disorganized bone trabeculae. The tissue expands and causes weakening of the affected bone, but the inner and outer cortices of the bone remain intact (1). As fibrous dysplasia slowly progresses, may produce bone remodeling and skeletal deformities (1). Fibrous dysplasia is classified as: monostotic, polyostotic and McCune Albright's syndrome. The latter entity is consistent with polyostotic bone involvement along with endocrinopathy, skin lesions and precocious puberty. Craniofacial FD is usually unilateral, and is more frequent in the polyostotic form (40-60%) than in the monostotic form (25%) (1,3). The upper maxilla and the mandible are the most commonly affected bones while the temporal bone is involved in 18% of cases. The typical characteristics of temporal bone FD are the bony expansion of the mastoids and the external auditory canal, the latter being commonly severely stenotic or occluded in approximately 80% of patients (5). The osseous chain may be involved, but the inner ear is spared in the majority of cases. Facial nerve is affected in 10% of the cases. Cholesteatoma may be seen in 40% of cases in the external auditory canal secondary to occlusion (2). CT is the imaging method of choice for radiographically interpreting fibrous dysplasia (5), but biopsy specimen is needed to confirm the diagnosis (3). High Resolution CT of the skull base estimates the grade of stenosis of the external auditory canal, depicts the involvement, if any, of the anatomic structures of middle and inner ear and reveals the possible presence of a canal cholesteatoma or the facial nerve involvement. Computed tomography is also useful for the follow-up of the disease (1). Plain radiography and bone scintigraphy are useful adjuncts (5). Radiographically, FD has a ground glass appearance and is surrounded by a dense cortical bone. Lesions of FD usually expand the affected bones and thus lead to bone remodeling. Craniofacial FD has three radiographic appearances: a) The pagetoid pattern (56% of all cases) is consistent with dense and radiolucent areas, along with bone expansion. b) The sclerotic pattern, in 23% of cases, is depicted by homogeneously dense and expanded bone lesions. c) The cystic pattern (21%) is demonstrated by oval radiolucent areas with sharp dense margins. The two thirds of cases of FD of the temporal bone are represented by the sclerotic pattern (1). Our patient represents a case of unilateral FD of the pagetoid type, affecting the temporal bone. It takes years for the patient to become symptomatic, because the disease develops slowly (1). The most common neurologic symptoms FD involving the temporal bone is hearing loss, which is most often conductive, due to stenosis of the external auditory canal. Sensorineural hearing loss is less frequent (17%) in patients with temporal bone FD (1) and is usually attributed to inner ear involvement, either due to destruction of the cochlea, internal auditory canal stenosis, or vestibular fistulization (5). The present case is unusual in that, the patient also developed sensorineural hearing loss, without having any of the factors mentioned above. It has been reported that permanent sensorineural hearing loss may be the result of entrance of inflammatory elements to the inner ear through the round window (4). In our case it is very probable that such inflammatory products and toxins have penetrated from the mastoids and middle ear cavity, through round window, to the inner ear. The conductive component of our patient's hearing loss can be explained by the extensive stenosis of external auditory meatus and the filling of the middle ear cavity by inflammatory tissue. MR imaging is useful in the assessment of the components of the lesions, but also helps in the evaluation of the effects of the bony lesions on adjacent anatomic structures of the skull base (5). Fibrous dysplasia bone lesions depict low to intermediate signal intensity in T1 and T2-weighted spin-echo images due to the presence of fibrous tissue. Casselman et al. reported gadolinium uptake from the involved tissue in cases of FD, but no histological explanation could be found for this enhancement (3). In our case, there was no evidence of contrast enhancement from the affected bone, in contrast to the adjacent meninges which revealed moderate gadolinium uptake. The presence of white matter edema in the adjacent temporal lobe is attributed to the external pressure by the bony lesion, which is another unusual finding, because fibrous dysplasia typically produces a diffuse expansion of the medullary cavity rather than mass effect. When clinical symptoms such as hearing loss, recurrent infection, dysfunction of facial nerve, are associated with craniofacial fibrous dysplasia, surgery is indicated and includes surgical decompression of the canal when there is severe stenosis of external auditory meatus. Differential diagnosis of fibrous dysplasia from other fibroosseous lesions of the temporal bone includes Paget's disease, ossifying and non-ossifying fibroma, meningioma, osteochondroma, giant cell granuloma, osteoma, sarcomatous neoplasms (1,5). In the literature, 76 cases of fibrous dysplasia of temporal bone have been reported. Nine of these patients developed sensorineural hearing
loss and only one of them restored his hearing after proper treatment (1). Our patient is an infrequent case of FD of the temporal bone, with development of sensorineural hearing loss, despite the fact that the inner ear was not involved. The patient restored completely his hearing after surgical decompression of external auditory canal.

**Differential Diagnosis List:** Fibrous dysplasia of the temporal bone

**Final Diagnosis:** Fibrous dysplasia of the temporal bone

**References:**


**Figure 1**

Description: Lateral plain film shows bony overgrowth and associated sclerosis in the temporal bone.

Origin:
Figure 2

Description: Computed tomography, axial section at the level of carotid canal, shows extensive thickening of the temporal bone and narrowing of the ipsilateral external auditory canal. Origin:
Description: Computed tomography, axial section, at the level of internal auditory canal. Extensive thickening and ground-glass appearance of the squamous part and the anterior part of the petrous apex of the left temporal bone is revealed. The left middle ear and the ipsilateral mastoid air cells are obliterated with inflammatory tissue. Origin:
**Figure 3**

Description: T2-weighted axial image, at the upper pontine level. There is hypertrophy and sclerosis of the squamous part of the temporal bone, which exhibits very low signal. Additionally, an area of high signal intensity at the periphery of the left temporal lobe is depicted, probably representing mild edema, due to pressure phenomena by the bony lesions. **Origin:**
Description: T1-weighted, Gd-enhanced, coronal image. There is marked bone thickening and very low signal intensity of the left temporal bone. A moderate meningeal enhancement is demonstrated adjacent to the lesion, without any evidence of enhancement within the affected bone. Origin:
Description: Axial section at the level of the middle ear shows progression of the bony changes at the left temporal bone, as well as early involvement of the contralateral temporal bone. Origin: