CASE REPORT

Bilateral Fibrous Dysplasia of the Temporal Bone in a Patient with a Unilateral Giant Cholesteatoma

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Fibrous dysplasia, which is a localized pathologic disorder of bone, is characterized by the abnormal proliferation of fibrous tissue that is interspersed with normal or immature bone. Involvement of the temporal bone, which is usually unilateral, develops in only 18% of all patients with fibrous dysplasia and occurs more frequently in a monostotic rather than a polyostotic form. To our knowledge, only 3 patients with bilateral temporal bone involvement have been reported in the literature. We present the case of a patient with minimal symptoms who was diagnosed as having bilateral monostotic fibrous dysplasia of the temporal bones and a concomitant giant cholesteatoma.

Fibrous dysplasia (FD), which is a localized disorder of bone, is characterized by the abnormal proliferation of fibrous tissue that is interspersed with normal or immature bone. FD is associated with precocious puberty in girls. The mandible and the maxilla are the sites most frequently affected by FD.[1,2] In 1891, von Recklinhausen first described the characteristic bony lesion that is known today as FD.[3] Not until 1937 did McCune and Buch describe FD as a clinical entity distinct from other abnormalities of bone formation.[3] The term "fibrous dysplasia" was introduced by Lichenstein in 1938.[3] There are 2 forms of FD: monostotic, which occurs in 70% of afflicted patients, and polyostotic, which occurs in 30%.[4] Craniofacial involvement is found in only 10% of patients with the monostotic variety, and monostotic FD of the temporal bone is very rare.[4]

Involvement of the temporal bone, which is usually unilateral, occurs in 18% of all patients with FD and develops more frequently in a monostotic rather than a polyostotic form.[3] To our knowledge, only 3 patients with bilateral temporal bone involvement have been reported in the literature.[5,6] Although a cholesteatoma (usually in the form of a canal cholesteatoma) causes complications in almost 40% of people with the craniofacial form of FD, there have been to our knowledge no reported cases of minimally symptomatic patients with both a giant cholesteatoma and FD that involved the temporal bone. The patient described in this report, who experienced minimal symptoms, was diagnosed as having bilateral monostotic FD of the temporal bones complicated by a giant cholesteatoma and massive FD on the right side.

CASE REPORT

Fifty-two A 51-year-old male patient was admitted to our clinic with the complaint of progressive hearing loss of 36 years' duration in the right ear and total occlusion of the right external auditory canal (EAC). When the patient was 20 years of age, physical examination for the complaint of right-sided hearing loss revealed a partial obstruction in the right EAC. After an exacerbation of his symptoms 4 years before his visit to our institution, he was told by his physician that his EAC was completely obliterated. This patient had never complained of otorrhea, otalgia, vertigo, or tinnitus.

Otoscopic examination revealed a hard, bony mass that extended from the posterior wall of the EAC and totally obstructed the canal. There was no prominent manifestation in the right temporal region. The left EAC and temporal region were normal in appearance. The patient had had no difficulty with the adjustment of the right temple bar of his eyeglasses for 20 years. His cranial nerve function was within normal limits, but the results of a tuning-fork evaluation suggested a right-sided conductive hearing loss. The thyroid gland was not enlarged. Findings from the remainder of the examination of this patient's head and neck were unremarkable.

A pure-tone audiogram revealed a conductive hearing loss in the right ear with an air conduction threshold of 62 dB and a speech discrimination score of 88%. Hearing in the left ear was within normal limits. Impedance audiometry was not performed on the right side because of the total obstruction, but audiometric findings were within the normal range on the left side. The results of a whole blood count and routine blood chemistry analyses were within the reference range.

A computerized tomography (CT) scan revealed sclerosis and marked focal expansion of the right squamous temporal bone in addition to partial sclerosis and focal expansion of the occipital bone, with the "ground glass" appearance of right temporal bone characteristic of FD. The area affected by FD was filled with a giant cholesteatoma that had eroded both the mastoid cavity and the posterior wall of the EAC. The middle ear cleft appeared to be normal. Massive FD of the entire temporal bone was noted, but the otic capsule and middle ear structures were spared. On the left side, sclerosis and focal expansion that were manifested as a small area of apparent FD were seen only in the temporal bone (Figure 1).

A right-sided modified radical mastoidectomy and meatoplasty were performed after the patient had

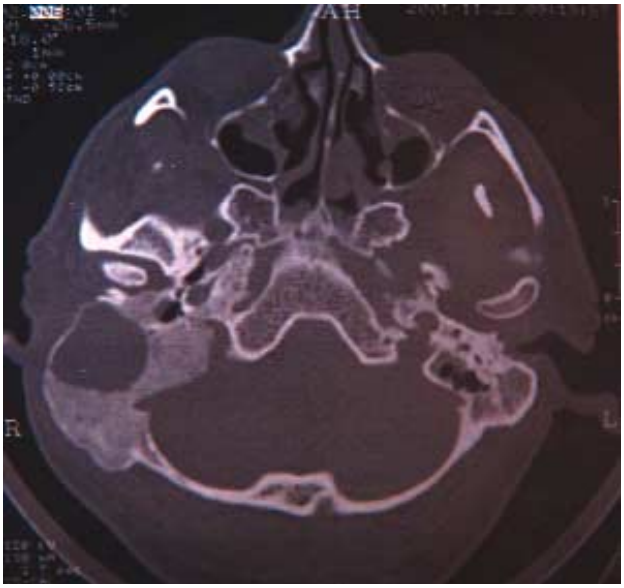

Figure 1: A computed tomographic scan showing sclerosis, the marked focal expansion of the right squamous temporal bone, and the partial expansion of the occipital bone, as well sclerosis and a small focal expansion consistent with fibrous dysplasia in the left temporal bone.

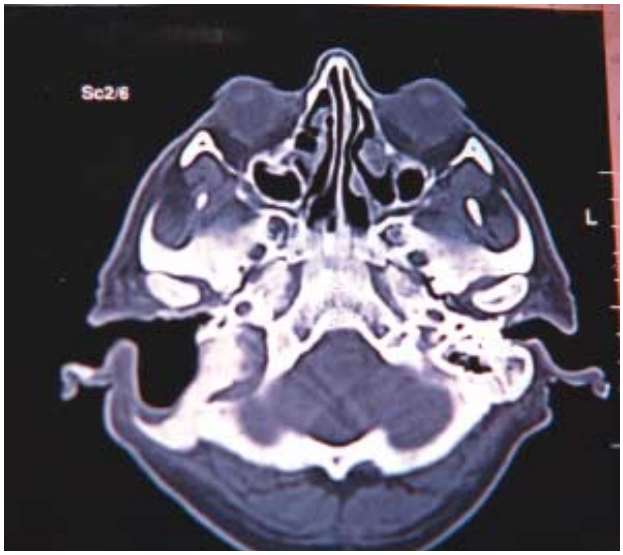

Figure 2: A computed tomographic scan of the right mastoid cavity during the third postoperative month.

received a general anesthetic. After the postauricular incision was made, the mastoid cortex was drilled. We noted that the mastoid cortex was very thin and that a huge cholesteatoma appeared just beneath that layer. After the removal of the cholesteatoma, the volume of the mastoid cavity was 15 mL, as indicated by the measurement of saline solution in a syringe. We also noted that the cholesteatoma had eroded the posterior wall of the external auditory meatus. The tympanic membrane and the ossicles were normal in appearance. The fibrotic tissues in the temporal bone were saucerized. At the conclusion of the operation, the volume of the mastoid cavity was 29 mL. A wide meatoplasty was performed. No fibrotic proliferation was noted on the CT scan performed during the third postoperative month (Figure 2). At that time, the pure-tone average of the 3 conventional frequencies (0.5, 1, and 2 kHz) was 37 dB.

DISCUSSION

FD of the bone is widely recognized as a skeletal disease of unknown origin. FD involving the temporal bone has been well documented in the literature. Three types of FD involving bone (monostotic, polyostotic, and Albright's syndrome) have been reported to date. In our opinion, those types represent the extent of disease at the time of diagnosis. To our knowledge, only 1 patient with FD of the bone who was monitored via long-term follow-up has been reported in the literature. In that case, monostotic FD of the temporal bone extended into the sphenoid bone after 23 years.[7] This development suggests that what had been diagnosed as the monostotic form of FD might instead have been an early phase of the polyostotic form.[7]

At the time of his examination at our institution, our patient had had FD for 32 years. Although FD involved both the temporal and occipital bones on his right side, only a small focus of FD had developed in the left temporal bone (ie, the monostotic form), according to the results of a CT scan. We suggest that these findings demonstrate the extension of FD from the right temporal bone into the occipital bone.

Barrionuevo and colleagues[8] classified FD of the temporal bone into the following 3 stages according to the progression of that disorder: stage 1, latent phase; stage 2, symptomatic phase; and stage 3, complication phase. The diagnosis of FD depends primarily on radiographic findings. When compared with conventional radiography, high-resolution CT provides

greater detail of the extent of FD in the temporal bone[9] and is useful in assessing the degree of EAC stenosis, the involvement of middle ear structures, the presence of an associated cholesteatoma, and the extent of the involvement of facial nerve and inner ear structures.[10] Because our patient had not undergone a CT scan when the partial obstruction of his right EAC was assessed 32 years earlier, his FD was not identified. At that time, he had refused surgery because his symptoms were not severe. However, when he was admitted to our clinic for the treatment of hearing loss caused by complete obstruction of the EAC FD was correctly diagnosed via high-resolution CT and was confirmed by histopathologic examination.

The hallmark of FD is the intact state of both the inner and outer cortices of the involved bones.[6] Postauricular swelling is caused by the increase of fibrotic bone between the 2 cortices, which usually leads to cosmetic problems. In our patient, however, the inner cortex of the temporal bone was not intact because of the erosion of the mastoid cavity caused by the cholesteatoma. It is therefore likely that the identified fibrotic tissue extended into the mastoid cavity with the cholesteatoma. Because the inner cortex is devoid of resistance if the outer cortex is intact, the fibrotic tissue that extended into the mastoid cavity was lysed by the cholesteatoma. As a result of that lysis and despite the presence of massive FD in the mastoid portion of the right temporal bone, our patient did not exhibit the swelling that usually causes a cosmetic abnormality in the postauricular region. He had been wearing his eyeglasses without difficulty and had not adjusted the right temple bar for 20 years.

The only treatment for FD is surgical intervention. Conservative surgery or complete removal of the dysplastic lesion is indicated,[11] and the extent of the surgical procedure should be determined by the degree of the functional and cosmetic deformity. The primary objectives of surgical therapy are the restoration of hearing and the prevention of complications such as chronic external otitis, the effects of a hidden cholesteatoma, the destruction of ossicles, or facial palsy.[12] A patient with stage 1 or stage 2 FD should undergo a total resection of fibrous dysplasia. If that is not an option, then when complications such as conductive hearing loss, stenosis of the EAC a cholesteatoma, or facial palsy develop, appropriate conservative surgical treatment should be performed in patients with stage 3 FD.[7] We performed a Bondy-type modified mastoidectomy with a wide meatoplasty in our patient.

CONCLUSIONS

In this report, we describe a patient with bilateral FD of the temporal bones and a concomitant cholesteatoma who had experienced progressive unilateral hearing loss of 32 years' duration. This case demonstrates that FD is an active process that slowly progresses throughout the patient's lifetime. We suggest that what has been considered the monostotic form of FD may instead be an early stage of the polyostotic form. In patients with a cholesteatoma and concomitant FD of the temporal bone, a radical mastoid operation is indicated. In our patient, the cholesteatoma prevented the swelling of the temporal bone that usually leads to cosmetic deformity. Periodic CT scanning is effective in assessing the progression and recurrence of FD and in detecting the development of complications.

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