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Case Report

Fibrous Dysplasia of the Temporal Bone with External Auditory Canal Stenosis and Secondary Cholesteatoma

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Fibrous dysplasia is a slowly progressive benign fibro-osseous disease, rarely occurring in temporal bones. In these cases, most bony lesions developed from the bony part of the external auditory canals, causing otalgia, hearing impairment, otorrhea, and ear hygiene blockade and probably leading to secondary cholesteatoma. We presented the medical history of a 24-year-old woman with temporal monostotic fibrous dysplasia with secondary cholesteatoma. The initial presentation was unilateral conductive hearing loss. A hard external canal tumor contributing to canal stenosis and a near-absent tympanic membrane were found. Canaloplasty and type I tympanoplasty were performed, but the symptoms recurred after 5 years. She received canal wall down tympanomastoidectomy with ossciculoplasty at the second time, and secondary cholesteatoma in the middle ear was diagnosed. Fifteen years later, left otorrhea recurred again and transcanal endoscopic surgery was performed for middle ear clearance. Currently, revision surgeries provide a stable auditory condition, but her monostotic temporal fibrous dysplasia is still in place.

KEYWORDS: Fibrous dysplasia, temporal bone, external auditory canal, cholesteatoma

INTRODUCTION

Fibrous dysplasia is a slowly progressive benign fibro-osseous disease. The etiology of fibrous dysplasia is regarded as a correlation with the congenital mutation of the G-protein subunit α gene in 20q13.2 [1]. Histologically, normal bones are slowly replaced with abnormal irregular mixtures of fibrous tissue and mature trabecular bones in the affected areas. Three subtypes are classified by the clinical presentation: monostotic (70%), polystotic, and McCune-Albright syndrome (with metabolic abnormalities and ipsilateral Café-au-lait spots) [2, 3].

There may be complaints of localized symptoms, such as swelling, tenderness, or even pathological fractures, in the affected area. Fibrous dysplasia mostly appears in the bones of the trunk and limbs. Craniofacial involvement is found in 10%-30% of monostotic cases and in more than 50% of polyostotic cases [1, 4]. Fibrous dysplasia of the skull mainly affects the maxilla and mandible bones. Craniofacial diseases arising from temporal bones account for approximately 18% of cases. It usually originates from the bony parts of the external auditory canals, approximately 80%-85% [5], and causes symptoms such as conductive hearing loss, otalgia, fullness, and otorrhea. Obstruction-related secondary cholesteatoma often develops in patients with obvious canal stenosis.

CASE PRESENTATION

A female patient came to our clinic at the age of 24 years because of progressive left hearing loss for 2 years. There was no otorrhea, tinnitus, or vertigo, but there was a complaint of persistent mild left temporal tenderness. A hard protruding lesion with complete lumen narrowing was found from the upper wall of her left external ear canal. We checked the pure tone audiometry, and there was an approximately 40 dB air-bone gap over the left ear, which was compatible with unilateral conductive hearing loss.

A series of temporal HRCT (High-resolution computed tomography) (Siemens Somatom HiQ; Erlangen, Germany) showed an extensive bone lesion manifesting as expansile appearance, mixed with an osteolytic and a ground-glass appearance of the left temporal and parietal bones, including mastoid and petrous part noted, compatible with monostotic fibrous dysplasia (Figure 1a, b). Canalplasty and type I tympanoplasty were performed due to the almost absent left eardrum. The left ossicular chain was confirmed to be intact during surgery. Pathological reports of the protruding canal mass revealed fibrous dysplasia. Her hearing ability gradually improved after surgery.

Five years after the first operation, there was a complaint of progressive left hearing impairment again. Further, some discharge with debris from the left ear was noticed. Repeat HRCT Aquilion-64, Toshiba Medical; Tokyo, Japan showed the remaining fibrous dysplasia of her left temporal and parietal bones, invading her external auditory canal. Additionally, a large amount of soft tissue filled the external and middle ear. The left ossicular chain was eroded to an extent where only the partial footplate of the

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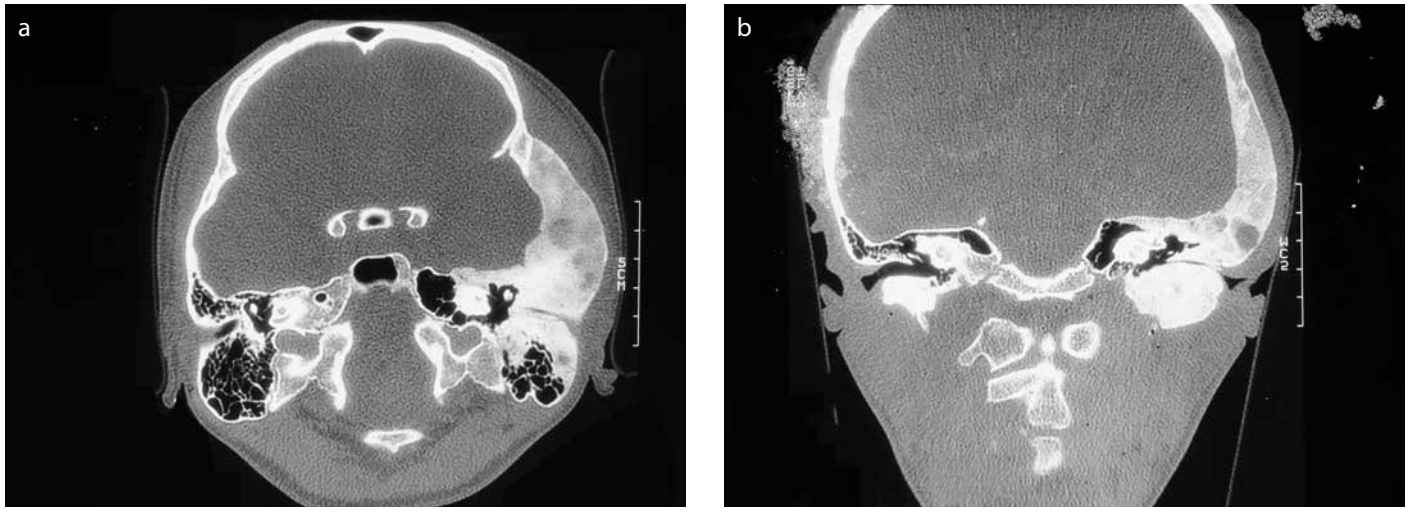

Figure 1. a, b. Axial (a) and coronal (b) views of the non-contrast temporal HRCT showed fibrous dysplasia of the left temporal bone with severe external auditory canal stenosis

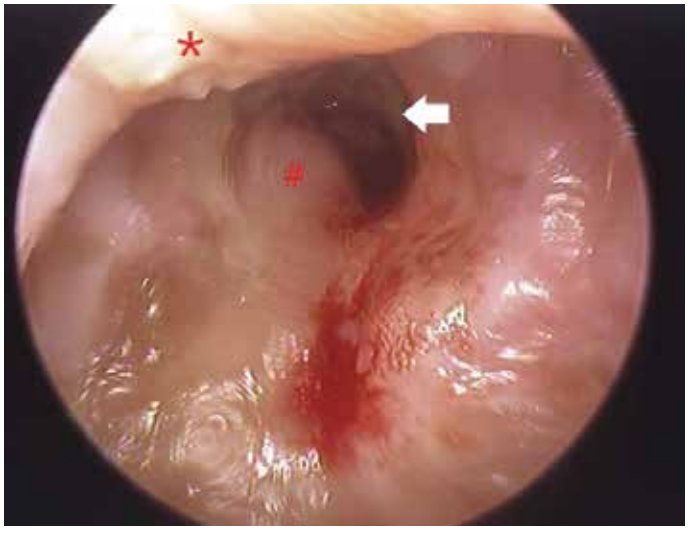

Figure 2. Otoscopy showed left meatal stenosis (*), residual canal fibrous dysplasia (#), and a bloody discharge with soft tissue filled the left middle ear cavity (white arrow)

stapes remained. Cholesteatoma involving the left external auditory canal and the middle ear was highly suspected. Revision canal wall down tympanomastoidectomy with type IV tympanoplasty was performed. Ossicular chain reconstruction was performed with total ossicular replacement prosthesis (TORP). An autograft conchal cartilage was placed between TORP and the tympanic membrane. The pathological result revealed the middle ear tissue to be a cholesteatoma. Postoperatively, pure tone audiometry showed partial improvement of her left hearing ability. Her auditory condition remained stable for years.

This patient again returned again 15 years after the second surgery due to intermittent left otorrhea for weeks. Meatal narrowing due to canal fibrosis was observed, and there was mild bloody otorrhea from her left middle ear (Figure 2). The bacterial culture showed *Pseudomonas aeruginosa*. Ear drops of Ciprofloxacin were used twice a day for 2 weeks, and a repeat HRCT revealed a substantial soft tissue density in the middle ear, suspected recurrence of cholesteatoma (Figure 3a-d).

Extended temporal bone resection was suggested, but she refused it because of cosmetic reasons. Thus, transcanal endoscopic surgery was immediately performed after meatoplasty. During the operation, some granulation tissue was removed from TORP and the cartilage columella. No residual cholesteatoma could be found, so revision type III tympanoplasty was performed. After the third surgery, she had regular check-ups every 2 months, and currently, there is no further complaint of otorrhea.

Informed consent for reviewing and publishing the medical records was obtained from the patient during the follow-up period.

DISCUSSION

Fibrous dysplasia involving the temporal bones is rare. Mostly, there are complaints of unilateral headache, otorrhea, otalgia, fullness, temporal region mass, and trismus. In total, 80% of all patients have partial or total stenosis of the external auditory canal, corresponding to secondary conductive hearing loss or a cholesteatoma. Keratinized epithelial of the external auditory canal accumulates, and secondary cholesteatoma occurs in patients with canal stenosis [6].

Due to the destruction of cholesteatoma in the middle to the inner ear, 14%-17% of patients can be inflicted by sensorineural hearing loss. A few lesions cause facial nerve canal compression and facial paralysis. However, polyostotic lesions of the bilateral temporal bones are still being reported in the literature [7].

High-resolution computed tomography is the examination of choice if fibrous dysplasia is highly suspected. Homogenous ground-glass density, loss of the trabecular pattern, and asymmetrical thickening of the cortical wall are the typical manifestations. Three types of CT findings are classified: pagetoid (56%), sclerotic (23%), and cystic (21%) types. The pagetoid type represents the end stage of sclerotic and cystic radiographic patterns. Moreover, middle and inner ear cholesteatomas can be well evaluated on HRCT [4]. However, the definite diagnosis is based on the histopathological findings. Well-demarcated intramedullary lesions are grossly found, and a moderately cellular proliferation of fibroblasts embedded with the curvilinear

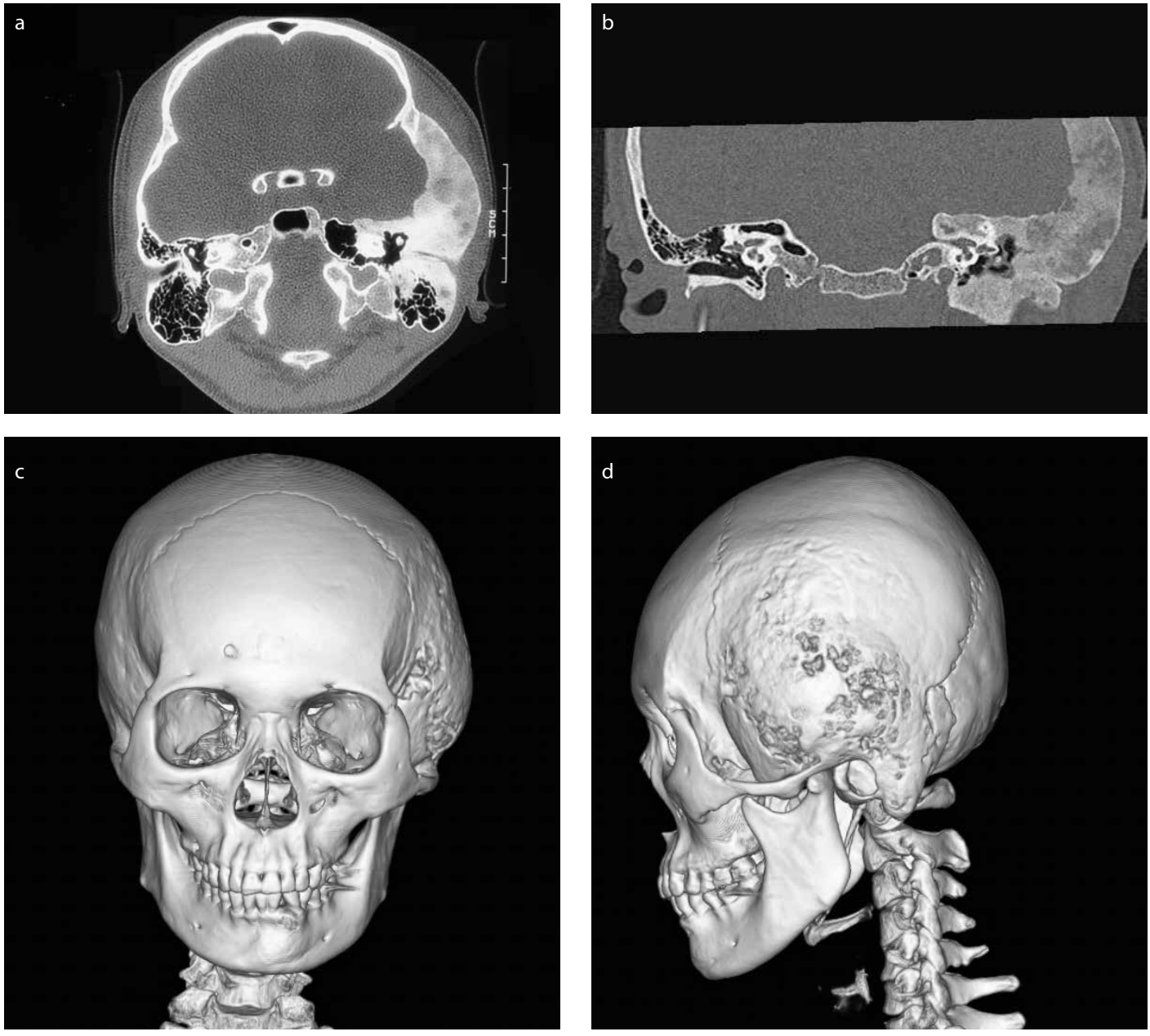

Figure 3. a-d. Repeated non-contrast temporal HRCT series: Axial (a) and coronal (b) views showed the progression of left temporal fibrous dysplasia with soft tissue opacities in the left middle ear. Three-dimensional reconstructed images showed anterior (c) and lateral (d) appearances of the expansile left temporal bone

trabeculae of the woven bone is noted under microscopy [8]. Malignant transformation in the temporal area is rare, with only approximately 1% of cases transforming into osteosarcomas [2, 9].

If asymptomatic, conservative treatment is first considered. However, surgical interventions are recommended in the following situations: cholesteatoma, external ear canal stenosis, recalcitrant infection, conductive hearing impairment, debris trapping, cosmetic deformity, cranial neuropathy with radiologic evidence of bony impingement, or biopsy for the diagnosis and exclusion of malignancy [2]. Canalplasty is first considered. A skin graft can be an optional procedure for canal coverage [1]. Tympanoplasty with or without mastoidectomy should be performed if symptomatic middle or inner ear invasion. Temporal bone resection is only performed for extended bony lesions. The aim of surgery is to restore function, prevent complications, and improve cosmetic demands [1].

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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