Management of the Temporal Bone Fibrous Dysplasia With External Auditory Canal Stenosis and Secondary Cholesteatoma in an Asian Population: A 11-Case Series

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Abstract

Objective: This article summarizes the experience of diagnosis and treatment of temporal bone fibrous dysplasia (FD) with external auditory canal (EAC) stenosis and secondary cholesteatoma in the Chinese population, in order to improve the quality of life of patients in the future.

Methods: Eleven patients with FD of the temporal bone who underwent surgery were retrospectively reviewed.

Results: All lesions originated from the temporal bone, and all involved of the EAC. There were 11 cases of cholesteatoma in the EAC, 4 cases of cholesteatoma in the middle ear. The most common symptoms were hearing loss (100%), tinnitus (36.4%), and otorrhea (36.4%). Two patients were severe-profound sensorineural hearing loss, and one patient was complicated with subperiosteal abscesses. All 11 patients underwent surgery. There were no perioperative complications in this series and median follow-up time was 4.2 years.

Conclusion: Temporal bone FD remains a rare diagnosis, especially in the Asian population. The lesions mainly lead to stenosis of the EAC, especially at the osteochondral junction. Cholesteatoma is the main complication of this disease, which is secondary to occlusion of the EAC with the growth of the lesion. Canaloplasty of EAC combined with wide meatoplasty can provide excellent prognosis in most cases.

Keywords
fibrous dysplasia, temporal bone, diagnosis, treatment

Introduction

Fibrous dysplasia (FD) is an uncommon benign but progressive skeletal disorder that destroys and replaces the normal bone with fibrous bone tissue. Formation of pathological tissues can lead to deformities, fractures, pain, and functional impairment.¹,² The incidence of FD is approximately 1 to 2 per 30 000 people, and both sexes are equally affected.³ Fibrous dysplasia accounts for 2.5% of all osseous neoplasms and 7.0% of all benign bone tumors.⁴,⁵ Fibrous dysplasia may occur at any age and most commonly affects patients younger than 30 years. The etiology of FD is controversial. de Reynal B⁶ first described FD in 1938 and believed that it was caused by the defect in the differentiation of the mesenchymal bone precursor. In 1957, George⁷ found that fibroblast hyperplasia also played an important role in the pathogenesis of FD. In general, there are 3 variant types classified by the clinical presentation: monostotic, nonsyndromic polystotic, and McCune-Albright syndrome (MAS).⁸

Fibrous dysplasia shows a racial predilection in that Caucasians account for more than 80% of affected patients while

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Asians account for only 1%.9 At present, there are few reports on the characteristics of patients in Asia. Fibrous dysplasia can invade the bones of the whole body, but it mostly appears in the bones of the trunk, limbs, and craniofacial region. Of these, craniofacial lesions usually occur in the maxilla. The temporal bone is rarely affected.

Although FD of the temporal bone generally grows slowly, it tends to cause some complications with disease progression, such as stenosis of the external auditory canal (EAC), conductive hearing loss, secondary cholesteatoma, otorrhea, and so on. The optimal management of patients with temporal bone FD remains a topic of debate and controversy.10 This article reviewed and analyzed 11 patients with temporal bone FD who were treated in our center in recent 10 years, and we summarized the experience of diagnosis and treatment in the Chinese population in order to improve the life quality of those patients in the future.

### Materials and Methods

All patients with confirmed diagnosis of temporal bone FD by pathology results from August 2009 to August 2019 were included in this study. The patient’s medical records were reviewed for demographic data, clinical presentations, diagnostic findings, histopathology, serum indicators, tumor classification, treatment strategies, length of clinical follow-up, and prognosis. In addition, the involvement of individual anomalies was also analyzed. Ethical approval was obtained from the Medical Ethical Commission of the Eye and ENT Hospital affiliated to Fudan University. The characteristics of these 11 patients are presented in Table 1.

### Surgical Technique

The aim of the surgery was to recover function and prevent complications. The choice of surgical approach was based on the extent of lesion involvement, physical examination, imaging studies, and pathology tests. In this study, the main operative technique consisted of canaloplasty of EAC and wide meatoplasty. A standardized approach was performed under assisted general anesthesia using an endaural incision as described in previous publications.11,12 After debridement of cholesteatoma keratin debris of the EAC, the membranous auditory canal was incised. The fibrous tissue was sharply detached from the bony ear canal down to the eardrum level. The bony ear canal was widened using diamond burrs of descending size. Subsequently, part of the concha, cartilaginous of EAC, and skin were removed, to create a widening of the cartilaginous portion of the EAC to adapt to the widened bony ear canal. The conchal flap is rotated toward the meatus to cover the part of the defect. Finally, an ear dressing with ribbon

### Table 1. Characteristics of 11 Patients of Temporal Bone Fibrous Dysplasia.

| Case | Sex/age/side | Main symptoms | Time from onset of symptoms to diagnosis (month) | Signs | Form | Cholesteatoma | Operation | Recrudescence | Follow-up (years) |
|------|--------------|---------------|-----------------------------------------------|-------|------|--------------|------------|---------------|-----------------|
| 1    | M/15/R       | HL, otorrhea  | 2                                             | EAC stenosis, TM granulation                 | Monostotic | Y, EAC | Y           | N               | 2               |
| 2    | M/17/B       | HL, tinnitus  | 6                                             | EAC stenosis, TM granulation                 | Polyostotic | Y, EAC | Reoperation Y |                 | 3               |
| 3    | M/38/L       | HL            | 240                                           | EAC stenosis, TM perforation                 | Polyostotic | Y, EAC, ME | Y           | N               | 2               |
| 4    | M/59/L       | HL, otorrhea, PFP | 60                     | EAC stenosis, TM perforation, HB: II        | Polyostotic | Y, EAC, ME | Y           | N               | 4               |
| 5    | M/36/L       | HL            | 6                                             | EAC stenosis, TM (--)                        | Monostotic | Y, EAC | Y           | N               | 4               |
| 6    | M/20/L       | HL            | 24                                            | EAC stenosis, TM (--)                        | Monostotic | Y, EAC | Reoperation Y |                 | 10              |
| 7    | F/46/L       | HL, otorrhea, vertigo | 72                  | EAC stenosis, TM (--)                        | Polyostotic | Y, EAC, ME | Y           | N               | 6               |
| 8    | M/26/L       | HL, tinnitus  | 84                                            | EAC stenosis, TM (--)                        | Polyostotic | Y, EAC | Y           | Loss            | 3               |
| 9    | M/51/L       | HL, tinnitus  | 6                                             | EAC stenosis, TM (--)                        | Monostotic | Y, EAC | Y           | N               | 6               |
| 10   | M/38/L       | HL, otalgia   | 96                                            | EAC stenosis, TM (--)                        | Monostotic | Y, EAC | Y           | Loss            | 3               |
| 11   | M/22/L       | HL, otorrhea  | 2                                             | EAC stenosis, TM perforation                 | Polyostotic | Y, EAC, ME | Y           | N               | 3               |

Abbreviations: EAC, external auditory canal; HB, House-Brackmann; HL, hearing loss; L, large; M, male; ME, middle ear; PFP, peripheral facial paralysis; TM, tympanic membrane.
Results

Demographics

Among the 11 (12 ears) patients with temporal bone FD managed at the authors’ centers, the mean age at time of diagnosis was 33.4 (15-59 years) years and 10 (90.9%) were men. Ages of onset in most patients were less than 30 years. One patient had bilateral disease. One patient reported a history of sudden sensorineural hearing loss (SSHL; Table 1). The tumors mainly lead to stenosis of the external auditory meatus, especially at the osteochondral junction. In patients with MFD, 100% accounted for 45.5% tumors involved the EAC, 40% involved the sphenoid bone, 50% involved the middle ear or mastoid, 20% involved the petrous bone. In patients with PFD, 100% of tumors involved the EAC, 20% involved the petrous bone. In patients with PFD, 100% of tumors involved the EAC, 50% involved the middle ear or mastoid, 83.3% involved the sphenoid bone, 50% involved zygomatic bone, and 16.7% for the ethmoid bone, frontal bone, maxilla, carotid canal, and petrous bone (Table 4). There were 11 cases of cholesteatoma in the EAC, 4 cases of cholesteatoma in the middle ear. One case of peripheral facial paralysis was caused by secondary cholesteatoma of middle ear.

Supplementary Examination

The level of alkaline phosphatase (ALP) and phosphorus in peripheral blood of the adolescent group (range from 15-17 years) was 192.0 ± 86.56 U/L and 1.27 ± 0.14 mmol/L, which was significantly higher than that of the adult group (range from 20-59 years; 125.87 ± 22.65 U/L and 1.02 ± 0.20 mmol/L, P < .05). The level of serum calcium of the adolescent group was slightly higher than the adult group (26.73 ± 12.49 dB vs 20.55 ± 9.58 dB), but the difference was not significant (P > .05; Table 2).

There was a significant difference between postoperative and preoperative mean AC thresholds (47.4 ± 20.08 dB vs 67.6 ± 16.89 dB, P < .05). The postoperative mean ABG was 18.8 ± 7.52 dB compared with 40.0 ± 6.07 dB in preoperative mean ABG and the difference was significant (P < .01; Table 3).

High-resolution computed tomography of temporal bone was used for initial evaluation and pretreatment planning (Figure 1). By virtue of inclusion criteria, all lesions originated from the temporal bone. Monostotic form of FD accounted for 45.5%, PFD 54.5%, and no MAS was found. The tumors mainly lead to stenosis of the external auditory meatus, especially at the osteochondral junction. In patients with MFD, 100% of tumors involved the EAC, 40% involved the middle ear or mastoid, 20% involved the Eustachian tube, 20% involved the carotid canal, and 20% involved the petrous bone. In patients with PFD, 100% of tumors involved the EAC, 50% involved the middle ear or mastoid, 83.3% involved the sphenoid bone, 50% involved zygomatic bone, and 16.7% for the ethmoid bone, frontal bone, maxilla, carotid canal, and petrous bone (Table 4).
Table 4. Scope of Temporal Bone Fibrous Dysplasia Damage in Imaging Examination.

| Scope            | Number (n = 11) | %      |
|------------------|-----------------|--------|
| MFD              | 5               | 45.5   |
| EAC              | 5               | 100    |
| ME               | 2               | 20     |
| Eustachian tube  | 1               | 20     |
| Carotid canal    | 1               | 20     |
| Petrous apex     | 2               | 20     |
| PFD              | 6               | 54.5   |
| EAC              | 6               | 100    |
| ME               | 3               | 50     |
| Sphenoid bone    | 5               | 83.3   |
| Ethmoid bone     | 3               | 50     |
| Frontal bone     | 1               | 16.7   |
| Maxilla          | 1               | 16.7   |

Abbreviations: EAC, external auditory canal; ME: middle ear; MFD, monostotic fibrous dysplasia; PFD, polyostotic fibrous dysplasia.

Management and Follow-Up Outcomes

All 11 patients underwent surgery. No patient was treated with radiotherapy. Canaloplasty of EAC and cholesteatoma resection were performed in all cases, including 10 with wide meatooplasty. Four patients underwent tympanoplasty and 5 patients underwent skin graft of EAC. One patient underwent decompression of facial nerve and 2 patients underwent reconstruction of ossicular chain.

There were no perioperative deaths in this series. Over a mean period of 4.2 years (range from 2-10 years), restenosis happened in 2 patients’ EAC in the third year after operation. So they underwent a secondary operation, and they were followed up for 2 years without recurrence. Two patients were lost to follow-up at 3 years (Table 5).

Discussion

Fibrous dysplasia is a rare benign lesion of bone disorder, characterized by a slow, progressive replacement of normal bone elements with proliferative fibrous tissue.\textsuperscript{13} Reports on cases with the temporal bone affected are uncommon, especially in Asians. Previous publications reported that there was no significant difference in gender, but 90\% of our patients were male.\textsuperscript{4,14} Temporal bone FD occurs in 25\% to 70\% of patients with craniofacial skeleton involvement, most frequently in the setting of PFD.\textsuperscript{3} In this article, 6 cases of temporal bone FD were PFD, accounting for 54.5\%. The most common clinical manifestations of the temporal bone FD were...
progressive stenosis of the EAC and conductive hearing loss, followed by tinnitus and otorrhea. There was no obvious clinical symptom in the early stage of the disease, and the median time delay between the first symptom onset and diagnosis was 54.2 months in this report. The lesion mainly leads to stenosis of the external auditory meatus, especially at the osteochondral junction. Cholesteatoma was the main complication of this disease, which was secondary to occlusion of the EAC with the growth of the lesion.9,12 All of the patients we reported have cholesteatoma, including 7 cases were limited to the EAC, and 4 cases invaded the middle ear. Two patients had SSHL, which may be due to cochlear destruction, inner auditory canal stenosis, or vestibular fistula. The prevalence of cranial neuropathy in temporal bone FD is rare. One of the patients in the current series had facial paresis caused by secondary cholesteatoma. For lesions beyond the temporal bone, the sphenoid and zygomatic bone were often invaded, accounting for 83.3% and 50%, respectively.

High-resolution computed tomography is especially important in the diagnosis of the temporal bone FD. The typical manifestations are homogenous ground-glass density, loss of the trabecular pattern, and asymmetrical thickening of the cortical wall.15 However, the imaging findings are not always typical and may be initially misdiagnosed by radiologists. Three types of CT findings are classified: pagetoid, sclerotic, and cystic types, and the pagetoid type is the end stage of sclerotic and cystic types. Besides, EAC and middle ear cholesteatomas can be well evaluated on HRCT.16 The magnetic resonance imaging appearance of FD is less characteristic and the typical characteristics are often very heterogenous pattern of enhancement in T1- and T2-weighted sequences. Magnetic resonance imaging might be useful in cases of cystic FD and in assessing the involvement of soft tissues. It is difficult to distinguish the temporal bone FD and some skull-base cases such as low-grade chondrosarcoma, atypical chordoma, or intraosseous meningoima from imaging and the final diagnosis depends on pathological examination.17 Fibrous dysplasia lesions may be quite vascular and bleeding can be brisk. If the lesion is quiescent or asymptomatic, and/or in the cranial base, a biopsy of FD may not be necessary. History, clinical examination, and the classic radiographic presentation are often adequate to establish the diagnosis of the temporal bone FD.18

There is no unified guidance on treatment standards, and the treatments of temporal bone FD still remain diverse. Currently, there is as yet no effective drug treatment. Although radiotherapy can inhibit the progression of disease, it has been reported that radiation may increase the possibility of malignant transformation.19 Hence, radiotherapy is also not recommended as an optimal choice. If there is no complication, conservative treatment is preferred. However, surgical interventions are recommended in the following complications: secondary cholesteatoma, peripheral facial paralysis, recalcitrant infection, or biopsy for exclusion of malignancy. The aim of surgery is to restore function, prevent complications, and improve cosmetic demands. The selection of surgical indications in our case series was secondary cholesteatoma.

Our experience is that if the patient has no complications, he can receive surgery until adulthood. The narrowing of the EAC may result in significant cerumen and keratin debris build-up. Therefore, it is recommended that regular examinations are usually required by the otolaryngologist to maintain EAC patency. The timing of surgery is a key aspect of decision-making, and the abnormal skeletal tissue hyperplasia become active in childhood.20 It may be beneficial to wait until growth has slowed of the FD and the patient has progressed beyond adolescence.18 In this report, 2 patients underwent a secondary operation, and the first surgery was performed during adolescence. It is reported that FD was related to increased osteoclastic resorption and ALP, calcium, and phosphorus in serum had relation with FD progression.21,22 At the same time, we also found that serum phosphatase were significantly higher in adolescence than that in adulthood, suggesting that the FD was active bone growth before adulthood. Surgery may be performed before FD maturity if there are severe complications; however, patients must be aware of the risk of regrowth.

Canaloplasty of EAC and wide meatoplasty are first considered. The lesions of EAC, such as cholesteatoma, can be removed at the same time. Because the surgery itself is relatively not complicated, the complication will rarely occur and the results are satisfactory. A skin graft can be an optional procedure for canal coverage when the skin defect of EAC is large. Tympanoplasty should be performed if symptomatic middle or inner ear invasion. Temporal bone resection is only performed for extended bony lesions. Complete resection of all lesions is not necessary because function can be restored well and the residual lesions did not significantly progress postoperatively after adulthood. However, a relatively large sample investigation including treatment and long-term follow-up will be necessary to provide a more comprehensive overview of this disease.

### Conclusion
Cholesteatoma is the main complication of temporal bone FD, which is secondary to occlusion of the EAC with the growth of

### Table 5. Treatment Strategy for 11 Patients.

| Treatment modality              | Number (n = 11) | %   |
|---------------------------------|-----------------|-----|
| Surgery                         | 11              | 100 |
| Radiosurgery                    | 0               | 0   |
| Observation                     | 0               | 0   |
| Surgical approach               |                 |     |
| Canaloplasty of EAC             | 11              | 100 |
| Wide meatoplasty                | 10              | 90.9|
| Cholesteatoma resection         | 11              | 100 |
| Tymanoplasty                    | 4               | 36.4|
| Skin graft of EAC               | 5               | 45.5|
| Decompression of facial nerve   | 1               | 9.1 |
| Reconstruction of ossicular chain | 2              | 18.2|

Abbreviation: EAC, external auditory canal.
the lesion. If the patient has no complications, they can be observed with serial cleanings/debridement in the clinic. Canalooplasty of EAC with wide meatoplasty is first considered, and if surgery itself is relatively not complicated, the complication will rarely occur and the results are satisfactory. Also, a long-term follow-up will be necessary for patients after surgery and that adolescent patients may be at higher risk of restenosis.

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