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Article

Pulsatile Tinnitus Caused by Highly Vascularized Fibrous Dysplasia of Temporal Bone with Centrally Located Fibrous Cyst

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Abstract: To date, the management and mechanism of pulsatile tinnitus (PT) caused by fibrous dysplasia (FD) of the temporal bone have not been discussed in detail. Therefore, this study is carried out to introduce PT secondary to monostotic temporal bone FD with cystic and lytic development in simultaneity complicated by the expansile bone lesion compressing over the sigmoid sinus. A 46-year-old female patient diagnosed with FD presented with severe persistent PT seeking medical assistance. Radiologic modalities displayed a 74.3 cm^3 monostotic FD with a presumably high-vascularity lytic change of the bone lesion located in the posterior portion of the mastoid cavity. The patient's PT was non-pulse-synchronous and was irrelevant to the intracranial blood flow revealed by ultrasonographic examination. A subtotal petrosectomy was performed after a meticulous preoperative assessment. During surgery, an unprecedented elastic cystic structure with three pedicles filled with vascular fluid was appreciated at the center of the FD lesion. PT was resolved after the removal of 90.8% of the total volume of the highly vascularized low-bone mineral density fibro-osseous tissues and the fibrous cyst. There were no PT reoccurrence and no postoperative complications during a two-month follow-up. Albeit rarely encountered, caution is required as PT secondary to FD is highly associated with the vascularity of the bone lesion.

Keywords: Pulsatile Tinnitus, Fibrous Dysplasia, Cyst, Mastoid, Temporal Bone

1. Introduction

Fibrous dysplasia (FD) is a congenital disorder characterized by the abnormal proliferation of fibrous tissue which affects any bone in the body [1]. It is estimated to prevail in 1 to 2 per 30,000 people with equal distribution in both sexes [2]. The pathological characteristic of FD is the expansion of cortical bone with gradual replacement by fibrous tissue, i.e., weak immature bone interspersed with fibrous stroma and disorganized bone trabeculae likened to Chinese characters [3]. In contrast to the polyostotic FD and McCune-Albright syndrome which multisite lesions, excessive autonomous hormone production, and/or unilateral café au lait spots are involved, the monostotic FD is the most common form of the three subtypes (approximately 70% of cases) whereby the pathologic bone production is limited to a single site [1,2].

Temporal bone FD is a rare diagnosis. It occurs in 25 to 70% of patients with craniofacial skeleton involvement, most exhibiting the polyostotic pattern [2,4,5]. Findings of physical/radiological examination may reveal cosmetic deformity, severe or complete stenosis of the external ear canal, otitis media, and cholesteatoma. Since multisite involvement and slow progression are common in FD, symptoms at the time of presentation vary among patients [1,2,6]. About 27−40% of patients maintain asymptomatic due to slow progression of temporal bone FD. Headache or conductive hearing loss, or in a combination thereof, are the common presenting symptoms [1,2,6]. The bone lesion is complicated by the cystic development, sclerotic change, and/or engraftment of an aneurysmal bone cyst filled with amble or vascular fluids [1,7]. These types of settings are infrequent and are often found with an accelerating progression [1]. In an extremely rare occasion when the osseous structure near the inner ear is eroded, vertigo or pulsatile tinnitus (PT) may occur [8]. Thus, surgical intervention is often required in patients with FD progression [1,9].

PT is the abnormal perception of pulse-synchronous vascular somatosound [10,11]. Unlike the abnormal vasculature and/or osseous structures such as sigmoid sinus wall anomalies and/or vascular stenosis from where the vascular bruits are commonly generated [10−13], PT caused by mass effect is less common [12]. Because of a relative paucity of data in otologic literature, PT is estimated to occur in 6−25% of patients with temporal bone FD according to a systematic review and a 66-case two-tertiary otologic referral center study published in 2014 [2,6].

To date, there has been no in-depth description of the PT mechanism associated with FD. Thus, this study is conducted to introduce a rare case of monostotic highly vascularized temporal bone FD compressing over the sigmoid sinus, in which PT presents as an initial and the only symptom. After prudent physical and radiologic examinations, the surgical removal of the FD to resolve PT was performed. The validity to adopt surgical intervention to eliminate PT and the mechanism of PT associated with FD were scrutinized in this case report.

2. Materials and Methods

A 46-year-old female with left-sided persistent PT with a duration of ten months was consulted in April 2020. She described a low-frequency non-pulse-synchronous drum-like percussive sound. The amplitude of her PT had aggravated 4 months after the initial onset but did not progress afterward. The patient stated that her PT was distressful during nighttime and became intolerable over time. Her quality of life and ability to focus at work or sleep at night were severely impaired. She rated the annoyance of her PT a level of 10 out of 10 grades (severity grade 1 to 10: 1–3 representing mild tinnitus, 4–7 moderate tinnitus, and 8–10 severe tinnitus), and her tinnitus handicap inventory score was 62. There was no headache, hearing loss, vertigo, and other associated symptoms of FD reported by the patient. Family history of bone disorders and/or other endocrine/genetic abnormalities was uneventful.

On physical examination, there was no external deformity of the facial structure or skin lesions. Otoscopic examination excluded the stenosis of the external auditory canal, and the tympanic membrane was normal. Pure tone audiometry showed that her hearing in either ear was within normal range.

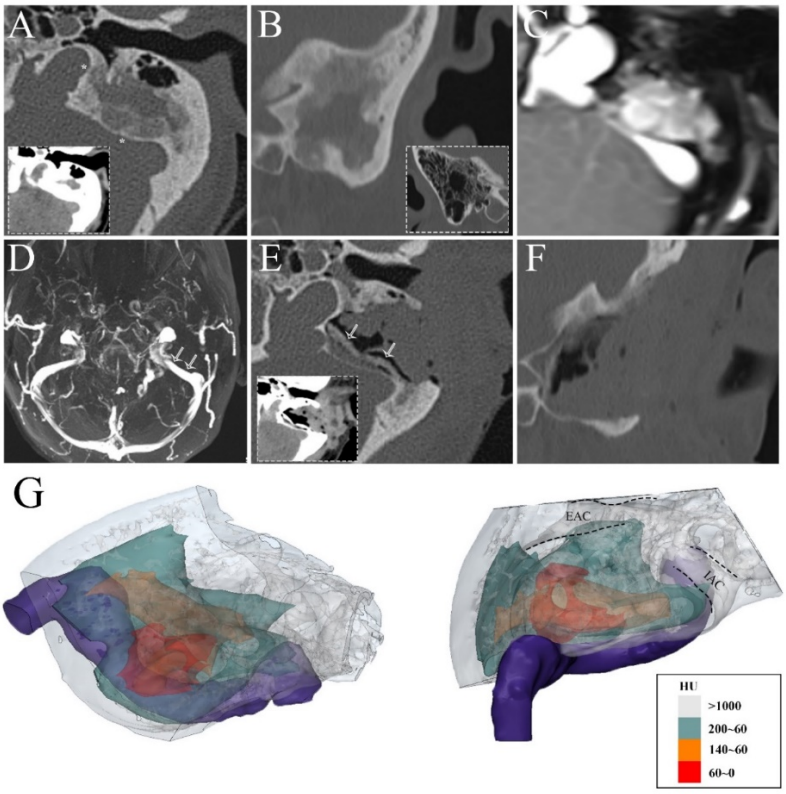

Fig. 1. (**A**) The high-resolution temporal bone computed tomography (CT) showcasing a diffusive sign of radiolucent ground-glass matrix confined to the posterior portion of the mastoid cavity. The heterogenous lytic change of the bone lesion with a tendency of cystic development characterized by less-defined margins is detected. White asterisk indicates FD compression/involvement of sigmoid sinus plate/jugular bulb region. (**B**) The Sagittal CT plane displays FD possessing the entirety of the mastoid cavity and change of mastoid pneumatization. (**C**) Contrast-enhanced magnetic resonance (MR) demonstrates enhancement of mass lesion with vague signs of fluid-fluid levels compressing the upper curve of the sigmoid sinus. (**D**) 2D Time-Of-Flight MR showcasing a notch-like defect due to a roughly 15% diameter decrease of the sigmoid sinus caused by the withdrawal of the sigmoid plate secondary to FD compression (white arrows). (**E**) and (**F**) Postoperative CT scan took 5 hours after surgical intervention. The bone lesions within the mastoid cavity with were radically resected. The sigmoid sinus plate was reconstructed using the normal autologous cortical bone of the mastoid (white arrows). (**G**) The three-dimensional planar reconstruction of the high-resolution temporal bone computed tomography (CT). The characteristics of the bone mineral density of the lesion are displayed using a different range of the Hounsfield unit (HU). EAC indicates external auditory canal; IAC indicates internal acoustic canal.

The temporal bone CT scan revealed that a large bone lesion situated at the left temporal bone was limited to the posterior region of the mastoid bone. The diffusive "ground-glass" appearance was distributed in the middle-posterior portion of the mastoid cavity with a gradient osteolytic change of cystic structure in the center of the lesion (Fig. 1). There was no sign of lesion invading posteriorly into the posterior cranial fossa or anteriorly compressing the posterior wall of the external ear canal. Horizontal CT slices demonstrated that the petrous and lateral surface of the sigmoid wall remained intact. However, the sigmoid plate was thinned and backwardly compressed due to the expansion of FD. Further analysis utilizing the planar three-dimensional reconstruction of the left temporal bone using MIMICS 13.0 (Materialise, Belgium) demonstrated that the lesion had a comparatively lower bone mineral density, lower Hounsfield unit (HU), of the bone lesion in the range of 200−0 HU. The preoperative lesion size was 74.1 cm^3. The contrast-enhanced MR revealed that the mass at the posterior mastoid region extended to the jugular bulb. FD compression over nearly 15% of sigmoid sinus diameter at the upper curve was observed.

The ultrasonographic examination was performed using a color-coded Doppler ultrasonographic system (Esoate My Lab Class C, Italy). The subject was requested to lie supinely on the examination bed with a neutral head position. PT persisted by completely collapsing the lumen of the upper jugular vein and/or internal carotid artery under the observation of ultrasonography. The subject reflected that the displaying sound of the jugular vein/internal carotid artery was completely dissimilar in both rhythm and pitch of her PT. After excluding the sigmoid sinus/internal carotid flow as a source of PT, the vascularity of the bone lesion was considered a potential source of PT. Thereafter, the patient demanded to receive surgical intervention to resolve her devastating PT with urgency.

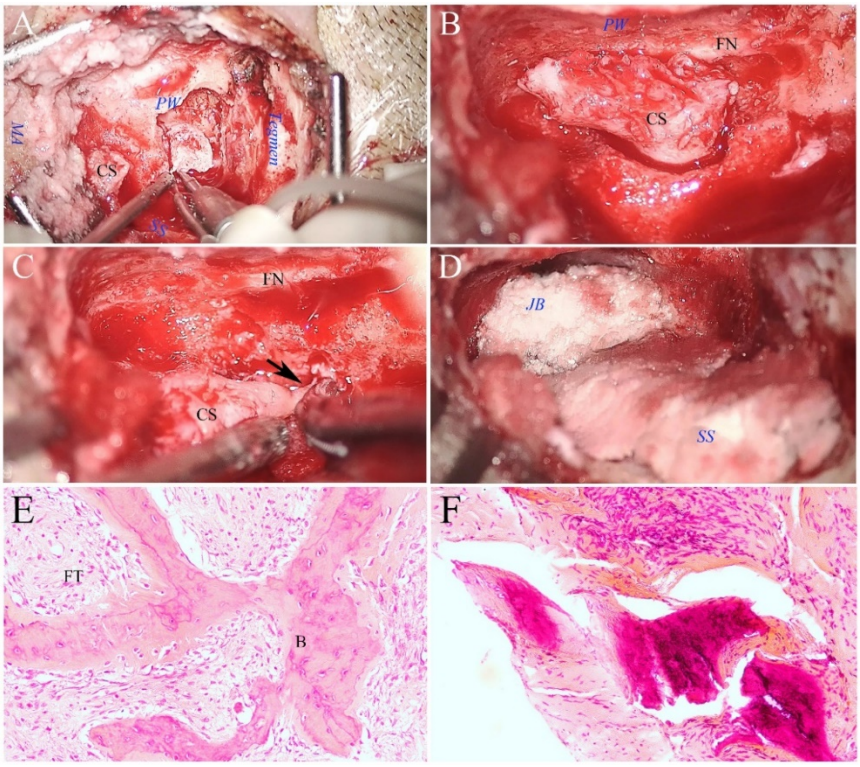

Fig. 2. Intraoperative graphs of FD and the aneurysmal bone cyst resection. PW indicates the posterior wall of the external auditory canal; SS indicates sigmoid sinus; MA indicates mastoid apex; CS indicates cyst lesion; FN indicates facial nerve and JB indicates jugular bulb. **(A)** The highly vascularized fibro-osseous tissues were easily removed using the otologic electrodrill. (B) Full exposure of an extraordinary elastic blood-rich cystic tissue after removing the surrounding fibro-osseous lesions. The cystic lesion was flattened after vascular fluids completely escaped. **(C)** Resection of the cystic tissue by dissecting the last of the three pedicles (black arrow). **(D)** Reconstruction of the sigmoid wall and jugular bulb using normal autologous cortical bone pate of the mastoid surface. **(E)** Classic Chinese-figure histologic pattern of fibrous dysplasia with spherical and ovoid cementum-like ossification interspersed in a proliferation of spindle cells throughout the lesion (hematoxylin-eosin, origin magnification ×40). FT indicates fibrous stroma; B indicates discontinuous trabeculae. **(F)** Histologic section of the cystic structure. Cystic development and fibrous boney elements were evidenced (hematoxylin-eosin, origin magnification ×40).

The subtotal petrosectomy was performed under general anesthesia with a standardized retroauricular approach (Fig. 2). During the skeletonization of the mastoid cavity, the removal of the highly-vascularized gritty and fragile boney structures was prone to hemorrhage. An unprecedented finding of an elastic cystic structure was gradually exposed during the removal of the vascularized bone lesion. After full exposure, the three-pedicle pouch-like fibrous structure was removed, and the remaining fibrotic osseous

tissues close to the sigmoid sinus and jugular bulb were resected. The autologous normal cortical bone of the mastoid surface was used to resurface the sigmoid sinus and jugular bulb. Postoperative CT demonstrated a subtotal dissection of 90.8% of the total volume of FD. Histopathological examination was reported as temporal bone FD. There have been no complications during the four-month postoperative follow-up. Remnant bone lesions with a total volume of 6.8 cm^3 located sporadically underneath the external auditory meatus remained quiescent and were carefully monitored. Her PT was significantly reduced after surgery.

3. Discussion and Conclusion

PT caused by secondary pathological effects of a growing mass is not commonly observed [12]. Studies of intracranial space-occupying lesions most often refer to the onset of PT as either a consequence of vascular compression owing to expansile mass effect or as a product of tumor vascularity abut cochlear, ossicular chain, and/or tympanic membrane [14]. While PT secondary to FD per se is an infrequent setting, it severely impacts the patient's quality of life so needs to be surgically eliminated once the surgical indication is clear and definite.

Prior to the surgical intervention, there were originally three potential scenarios that expatiated the onset of PT: (1) hemodynamic alteration of sigmoid sinus due to FD compression that induces PT, (2) transduction of sigmoid sinus flow sound via holistic decrease of the bone mineral density of the mastoid bone as an intermedia that provokes audible PT, and (3) the highly vascularized lytic bone lesions that caused PT. Through cautious examinations and analyses, the first two scenarios were rejected. The subject did not respond to venous flow variations at the neck and the sound-proof sigmoid plate remained intact. This suggested that the sound of sigmoid flow was less likely to permeate through. Hence, PT originating from the sigmoid sinus is unlikely (factoring out scenario 1). Although it was suspected that the change of mastoid pneumatization rendered the entire mastoid bone an effective agent for flow-induced sound to transmit to the hearing apparatus [15], this notion contradicted the actual outcomes from the physical and ultrasonographic examination because PT persisted after deliberately reducing the sigmoid sinus outflow. In addition, the sonographic-detected rhythm and pitch of the subject's venous outflow did not match the patient's subjective perception of PT. As such, PT caused by the transduction of the sigmoid sinus flow sound is untenable (factoring out scenario 2). Instead, it was highly plausible that PT resulted from the vascularity of the lesion. As the mastoid air cells were progressively substituted with a massive volume of blood-rich immature bone structures, we extrapolated that the normal osseous structure near the hearing apparatus eventually failed to insulate the sound of FD vascularity from the inside out. It also explained why the patient's PT persisted even by interrupting both major in-and-out intracranial blood flow pathways and was not pulse-synchronous, suggesting her PT was not directly associated with extradural or the arterial system. Ultimately, the extrapolation was vindicated by the successful surgical resolvent of PT. Aside from complete removal of the tumor, PT secondary to tumor vascularity was effectively eliminated by stereotactic radiosurgery. The therapeutic effect was associated with progressive radiation-induced intimal fibrosis and stenosis of vessels of the mass [16]. However, radiation and chemotherapies were not effective and contraindicated owing to malignant transformations, e.g., osteosarcoma, in FD treatment [1]. This renders the excision of the tumor vascularity away from the hearing apparatus a last resort but effective method to eliminate PT.

The connection between FD vascularity and the cause of PT has been underappreciated. As PT devastatingly impairs the quality of life and even brings suicidal thoughts to those who suffer from PT, notwithstanding its rarity, this case study confidently demonstrates and reemphasizes the bonding between vascularity and PT. Cautions are required in cases presenting the lytic change of FD or the engraftment of cystic change filled with vascular fluids. There have been only two FD surgical case studies published in the last five decades which introduced the surgical resolvent of PT with a clear description of removing the vascular, gritty sponge-like bone [17,18]. Due to the lack of referencing information in the otologic literature, we cannot identify whether the fibrous cystic structure filled with vascular fluids or the vascularized fragile sponge-like bone lesion or both, causes PT. Through backward induction of PT elimination, it is concluded that the reduction of the vascularity of FD, as a whole, is positively related to surgical efficacy and can be a pivotal factor in surgical indication and pertinence.

PT presenting as an initial and only symptom induced by the osteolytic change of temporal bone FD with a centrally located cystic structure is extremely rare. Notwithstanding the rarity, this case study identifies that PT derives from the vascularity of FD rather than expansile compression of the extradural venous system. The surgical excision of the highly vascularized FD adjacent to the hearing apparatus ensures positive surgical efficacy of PT. The value of the information adds up to surgical pertinence and indications for surgery of temporal bone FD.

Author Contributions: Y-L H integrated all clinic and operative notes on the patient to create content for the case report and drafted the final manuscript. WQW is the lead surgeon who performed with Y-L H participated in the subtotal petrosectomy and edited the final manuscript. WQW and Y-L H followed up with the patients, provided clinical and operative notes, and supervised iterative drafting of the manuscript. All authors reviewed the manuscript prior to submission. The authors read and approved the final manuscript.

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