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CASE REPORT OPEN ACCESS

Cryptic Temporal Bone Fibrous Dysplasia with Predominant Vestibular Symptoms: A Case Report

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Abstract
Background: Temporal bone fibrous dysplasia is relatively uncommon and typically manifests as a monostotic form of the disease. It often presents with noticeable temporal bone deformity, clinically appearing as a hard bony swelling behind or around the ear, external auditory canal stenosis, and conduction deafness. While symptoms such as aural fullness, tinnitus, and hearing loss are commonly reported, specific vestibular symptoms like vertigo and nystagmus are considerably less frequent.
Case Presentation: In this report, we describe the diagnostic features, management, and outcome of a 37-year-old Nigerian patient with fibrous dysplasia of the right temporal bone, who presented with recurrent debilitating vertigo, along with tinnitus and aural fullness, but without any obvious temporal bone deformity, which posed an initial diagnostic dilemma. She underwent an exploratory mastoidectomy and received a postoperative stat dose of IV zoledronate, along with oral dexamethasone tapered over 10 days, resulting in significant symptom resolution.
Conclusion: This unusual presentation underscores the importance of considering temporal bone dysplasia as a potential differential diagnosis for unilateral tinnitus, regardless of the presence of clear bony deformity.

Keywords: Fibrous dysplasia, Temporal bone, Vestibular symptoms

Background

Fibrous dysplasia (FD) is an uncommon, histopathologically benign disease characterized by the progressive replacement of normal bone marrow by proliferating fibro-osseous tissue that expands and thins the overlying cortex. FD accounts for about 2% to 3% of bone-derived tumours and usually occurs throughout the skeleton. Although it has a predilection for the craniomaxillofacial bones, temporal bone involvement is uncommon (1, 2, 3).

Fibrous dysplasia is presently classified into three subtypes based on the distribution of lesions and associated symptoms. The most common subtype is monostotic fibrous dysplasia, which involves a single bone, while the polyostotic form involves multiple bones, including the ribs, craniofacial or long bones. McCune-Albright syndrome is the third

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form and is associated with endocrine abnormalities and café-au-lait spots. Involvement of the temporal bone in this disease process was first reported in 1946 (4, 5), and accounts for less than 10% of all reported cases (6).

Monostotic fibrous dysplasia of the temporal bone most commonly presents with bony expansion and overt deformity of the site (6, 7, 8). This usually is the initial pointer to the diagnosis. Other associated symptoms include otalgia, hearing loss, tinnitus and less commonly, vertigo (8).

In this report, we describe a case of fibrous dysplasia affecting the left temporal bone, presenting mainly with debilitating vertigo but no obvious cosmetic deformity or significant loss of hearing.

Case presentation

A 37-year-old Nigerian female presented to our clinic with a 3-year history of insidious-onset, recurrent vertigo and a 1-month history of persistent tinnitus and fullness in the left ear.

Vertigo was described as a recurrent non-positional spinning sensation lasting about 10-20 minutes per episode with an average of 3-4 episodes per day, occasionally associated with nausea and vomiting, and significantly affecting her daily activities. Tinnitus was characterized as a high-pitched subjective continuous sound in the ipsilateral ear, louder in quiet settings. However, despite the tinnitus and sensation of ipsilateral ear fullness, she reported no reduced hearing, ear pain, discharge, or overt swellings around the ear. There was no prior history of head or ear trauma.

Clinical examination revealed normal-looking pinnae and mastoids bilaterally. Otoscopy showed a dull but intact left tympanic membrane. The facial nerve function was intact.

Pure tone audiometry showed slight conductive left-sided hearing loss with normal hearing on the right. Tympanometry revealed a Type A Tympanogram on the left and a Type A on the right. Due to the prolonged duration of the non-positional vertigo, and its debilitating nature, we requested a cranial CT-Scan which showed increased thickness of the left temporal and zygomatic bones with expansion by a mixed density lesion and irregular margins. There was also partial opacification of the left mastoid air cells, more marked along the tegmen tympanum, the petromastoid canal and anterior petrous air cells, which was reminiscent of osteomyelitis of the left petrous apex and temporal bones. (Figure 1).

No intracranial nor Cerebello-pontine mass lesion was noted.

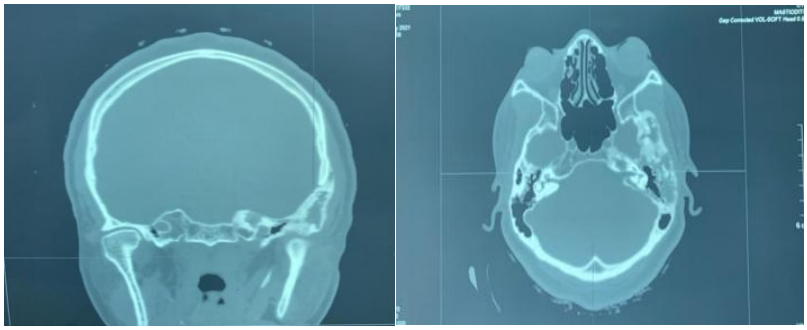

Figure 1: CT Temporal bone showing expansion of the left temporal bone expansion showing the classical "ground-glass" appearance, extending to the petrous apex.

Features were suggestive of fibrous dysplasia of the temporal bone with Ossifying fibroma as a close radiological differential. She subsequently had an exploratory mastoidectomy with the extirpation of the expanded mastoid cortex and trabecular-like bone that had almost completely occupied the mastoid. (Figure 2).

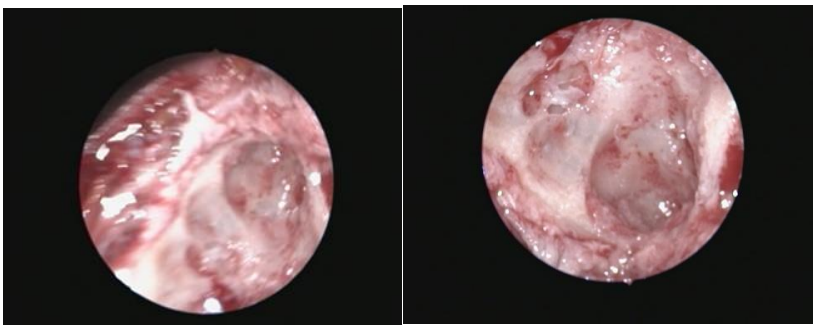

Fig. 2: Intra-operative images of cortical mastoidectomy and extirpation of the fibrous matrix from the mastoid

The histology revealed a benign fibro-osseous lesion composed of thin and irregularly curved trabeculae of immature woven bone surrounded by fibroblastic stroma comprising proliferating bland spindle-shaped cells, all in keeping with fibrous dysplasia. (Figure 3).

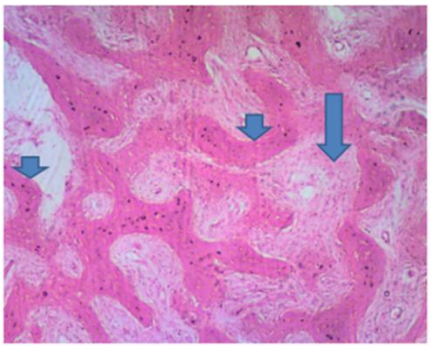

Figure 3: Photomicrograph (H&E x40) of the mastoid bony tissue showing irregularly curved immature woven bone (Arrowheads) lacking osteoblastic rimming and interspersed by benign stromal cells (Full arrows)

She received a stat dose of IV Zoledronate 4mg, as well as tabs Supplemental Vitamin D, Vitamin C, Vitamin B complex, and Dexamethasone tapered down from 4mg 8 hourly over a period of 10 days, with complete resolution of vertigo noticed from the 1st-week post-op.

She has now been followed for 18 months, with audiometry performed at 6 months and 1 year post-operatively. Despite improvements in middle ear compliance and a narrowing of the air-bone gap with improvement in hearing thresholds, she still has ipsilateral intermittent tinnitus. However, she reports a significant improvement in her quality of life following the resolution of vertigo and no recurrence after follow-up for 2 years.

Discussion

The precise etiology of fibrous dysplasia is currently unknown, and various theories have been proposed over the years. Most recently, it is thought to be caused by a non-heritable activating mutation in the α-subunit gene of the stimulatory G-protein coding gene. The disease tends to develop in the pre-adolescence years and is predominant in male patients (2:1) (9). The monostotic form of the disease has been reported to be found in as much as 70% of reported cases, such as in our index case. Fibrous dysplasia of the temporal bone is an uncommon entity, although its prevalence may be underestimated due to the frequent occurrence of cases only identified incidentally (9, 10).

Symptomatic cases of temporal bone involvement present, most commonly with cosmetic deformity of the temporal bone, headache, slowly progressive external auditory canal stenosis and consequent conductive hearing loss (11). Hearing loss could also occur when there is gross affectation of the middle ear and/or Eustachian tube, as well as when the otic capsule is involved with resultant sensorineural hearing loss (13). Interestingly, hearing loss is not as common as one might expect, accounting for only 29% of symptoms in a review of 66 cases of temporal bone fibrous dysplasia and 22.4% of cases in another study of 130 subjects with craniofacial fibrous dysplasia (9, 10, 11, 12). It has also been shown that conductive,

rather than sensorineural hearing loss, is the more common form of hearing impairment in this condition (12). Our Patient had a near-normal hearing, with tympanometry showing features of reduced middle ear compliance that improved after surgery. This is probably due to the expansion of the mastoid cortex and reduced middle ear space by the dysplastic bone, which was extirpated. Other associated symptoms that have been described include cosmetic deformity from retro-auricular bulging, otalgia, otorrhea, tinnitus, and facial nerve palsy following erosion of the fallopian canal or pressure on an exposed segment of the nerve (11). Vestibular symptoms are seldom encountered. Frisch et al in their case series, reported a prevalence rate of 24% among 66 patients (9). Weiss et al. recently reported a case of Meniere's syndrome attributable to erosion of the petrous apex and retro-labyrinthine area, as well as obliteration of the vestibular aqueduct (13). Other radiologic findings associated with vertigo in temporal bone fibrous dysplasia include posterior semicircular canal labyrinthine fistula, internal auditory canal narrowing, as well as endolymphatic sac erosion, all of which had petrous temporal bone involvement in common (11, 12, 13). Our Patient also had disease extension to the petrous temporal bone in addition to extensive mastoid involvement. However, it is our opinion that her vertigo resulted from pressure from the expanded thinned-out cortices of the mastoid and petrous bone on the vestibular system rather than actual erosion of the bony labyrinth.

Tinnitus is an inherently more complex symptom and could result from widely varied causes including narrowing of the external auditory canal, various manifestations of middle ear involvement-from epitympanic disease and cholesteatoma to ossicular erosion, otosclerosis and tympanic cavity occupation by fibrous tissue, in addition to erosion of the bony labyrinth/cochlear or compression of the cochlear nerve in the internal auditory canal or the inner ear. Considering that our patient did not have any disease in the external ear canal, nor any intra-op features of cholesteatoma and that there was improved middle ear compliance after surgery, we were disappointed at the persistence of this symptom. Other case reports and series we came across that identified tinnitus as an associated symptom did not state its response to treatment (4, 9, 12, 13).

Our Patient did not have any pre-operative bone pain. However, considering the intraoperative indistinct margins between fibrotic and normal bone and the uncertainty of complete excision, we chose to use this adjunct treatment. Bisphosphonates are useful in the management of fibrous dysplasia, resulting in pain reduction, slowing osteoclastic activity and consequent bone destruction, and even increasing bone mineral densities with evidence of a reduction in bone turnover markers (14, 15).

Conclusion

Though unusual, Fibrous dysplasia of the temporal bone could present chiefly with vertigo and without any external bony deformity. Surgery remains the mainstay of treatment for symptomatic disease, with bisphosphonates proving a useful management adjunct.

List of Abbreviations

CT: Computed Tomography
FD: Fibrous dysplasia

Declarations

Ethical approval and consent to participate

Written informed consent was obtained from the patient's parent for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor.

Consent for publication

All authors gave consent for publication of the work under the Creative Commons Attribution-Non-Commercial 4.0 license.

Availability of data and materials

The tissue blocks and all essential data supporting the findings of this case are available upon reasonable request from the corresponding author.

Competing interests

The authors declare no conflict of interest.

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Authors' contributions

AMA: Lead clinician, conceptualization, manuscript review. UKC: Literature review, Manuscript writing, manuscript review. NJI: Conceptualization, Case pathologist, manuscript review. OO: Case clinician, case conceptualization, manuscript review. BBA: Manuscript review.

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