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Case Report

Monostotic fibrous dysplasia of temporal bone: a case series and literature review

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ABSTRACT

Fibrous dysplasia is a benign, progressive fibro-osseus disease. Typically, there are three variants monostotic, polyostotic and McCune Albright syndrome. Monostotic variant being the commonest of all, affects mainly the craniofacial bones and ribs. Fibrous dysplasia affecting the temporal bone may be challenging at times. Here, we present our case series of three temporal bone fibrous dysplasias along with the literature review.

Keywords: Fibrous dysplasia, Temporal bone, Monostotic

INTRODUCTION

Fibrous dysplasia is a benign condition primarily affecting the bone. Pathologically, there is fibrosseous proliferation where normal bone gets replaced with fibrous tissue and immature bone. There are three distinct types of this pathology mentioned so far; monostotic, polyostotic and McCune Albright syndrome. Monostotic variant accounts for 70% of the cases and represents the milder spectrum of the disease. Typically, craniofacial bones and ribs are involved.[1] Temporal bone involvement, although rare accounting for <10% of cases, is one of the challenging situations in otorhinolaryngology.[2] Here, we present a case series of temporal bone fibrous dysplasia focusing on the variability of clinical presentations, diagnosis and the management.

CASE REPORT

Case 1

A 23 years male presented with complaint of aural fullness in right ear for last 2 years. The fullness was insidious in onset, persistent and gradually progressive. It had no intermittent exacerbations or remissions. It was not associated with pain, discharge and swellings in and around the ear and the neck region. There were no other aural, nasal and throat symptoms relating to the chief complaints. His past medical history was insignificant. There was no surgical intervention done in the past. Family history of the patient revealed no similar complaints in the family members and absence of any ear disease running in the family. He doesn't smoke and doesn't consume alcohol. There was no significant history of drug allergy. General examination of the patient didn't reveal any abnormal findings. Complete ENT and head and neck examination revealed a hard-swelling arising from posterior superior canal wall in the bony part occluding the whole bony EAC. Cartilaginous EAC was normal. While probing the swelling, it was bony hard in consistency. It was arising from the posterior superior canal wall and completely occluding the EAC. Rinne was negative on the right side and positive on the left. Weber was lateralized to right ear. Rest of the ENT examination was normal. An initial diagnosis of canal exostosis was made and the patient was advised for HRCT temporal bone and pure tone audiometry. HRCT temporal bone revealed sclerotic bony expansion with ground glass appearance involving

the right zygoma, petrous temporal and mastoid bone. The bony expansion was centered in the medullary bone and had abrupt transition zone (Figure 1). Pure tone audiometry showed a 63 dB conductive hearing loss in right ear. Diagnosis of right temporal bone fibrous dysplasia was made. Since, the affected regions were surgically in assessable, only biopsy from the EAC was planned. Biopsy was carried out through post aural approach. Patient was followed up after 4 weeks, where histopathological report showed features of fibrous dysplasia. There were fragments of irregular bone without osteoblastic rimming and intervening stroma showing proliferation of bland spindle cells (Figure 2). The patient was advised to remain in follow up 6 months.

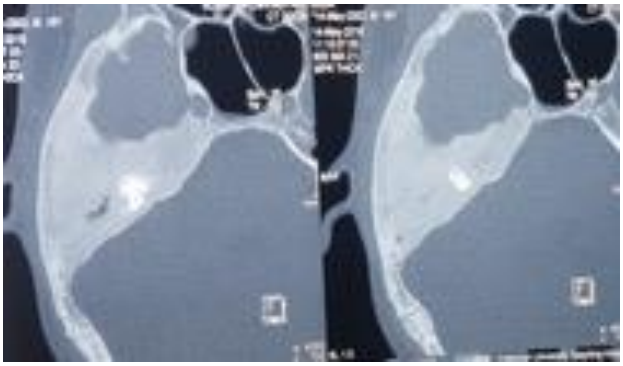

Figure 1: HRCT temporal bone showing ground glass appearance.

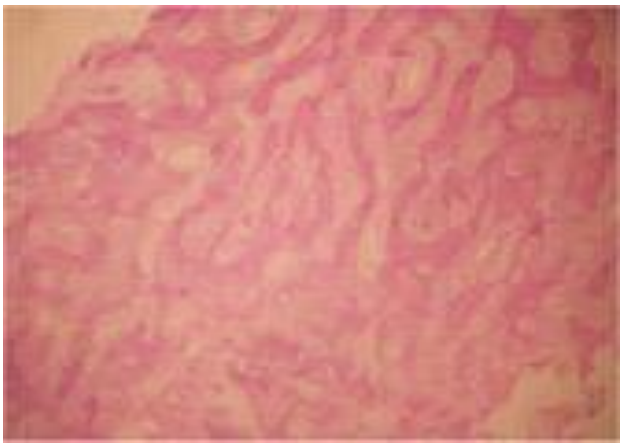

Figure 2: Histopathological slide showing fragments of bone without osteoblastic rimming with intervening stroma showing proliferation of bland spindle cells.

Case 2

A 48 years old lady presented with decreased hearing in right ear for 4 years. It was gradual in onset and progressive with no fluctuations. There were no other complaints of ear discharge, tinnitus and pain in and around the ear. She had no other nasal and throat complaints that could be related to her chief complaints. Her past medial history, personal history and family history were all insignificant. Her menopause started one and half years ago and her previous menstrual cycles were regular. She had no history of drug allergy. General examination of the patient was normal. On examining her right ear, there was narrowing of right EAC in the bony part with bulge from posterior, inferior and superior walls. On probing the swelling was bony hard however, nontender. Rinne test was negative on the right side and positive on the left with Weber test lateralized to the right ear. Nose, oral cavity, oropharynx, hypopharynx and neck examination yielded normal findings. With a working diagnosis of right EAC exostosis, HRCT temporal bone and PTA was planned. PTA revealed 42 dB conductive hearing loss in right ear and normal hearing in left ear. In the HRCT scan of temporal bone there was sclerotic bony expansion involving the mastoid process and tympanic potion of the temporal bone narrowing the bony EAC (Figure 3).

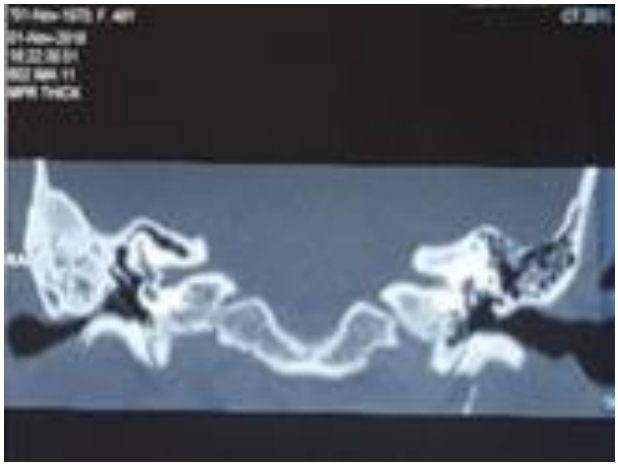

Figure 3: HRCT temporal bone showing sclerotic bony expansion involving mastoid and tympanic portion on right side.

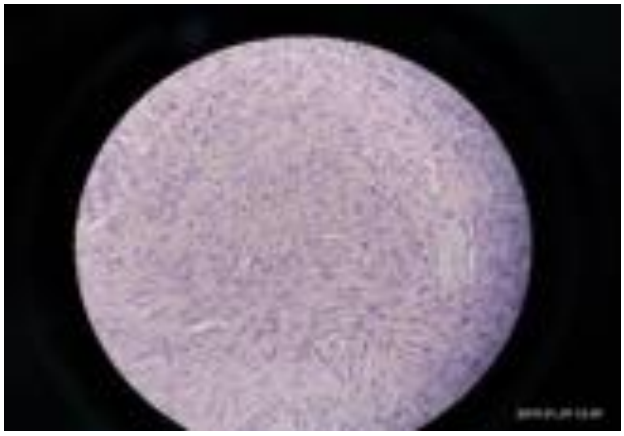

Figure 4: Histopathological slide showing fragments of bone with osteoblastic rimming.

Middle ear and the ossicles were normal. Based on the HRCT temporal bone findings a canal wall down mastoidectomy was planned as the disease involved the mastoid and posterior wall of EAC. Through post-aural

route, a complete canal wall down mastoidectomy was done and the disease was removed as a whole. There was soft fibrosseous bone present in the mastoid process and posterior wall of EAC. Middle ear was normal and the ossicles were intact and mobile. Patient was discharged on 4th post-operative day, with pack and sutures removed on 6th post-operative day. On follow up histopathological report of the bone fragment showed features of irregular bone showing osteoblastic rimming. It was compatible with fibrous dysplasia (Figure 4).

Case 3

A 52 years old male presented with gradual swelling in left supra-auricular area for past 7 years. Swelling initially was of small size around 2x2 cm. There were no known initiating factors for the swelling. It then gradually increased in size causing progressive disfigurement of the skull. There were no intermittent fluctuations in the size of swelling. It was hard and painless. The swelling was not associated with other symptoms such as headache, earache, aural fullness, decreased hearing, vertigo, vomiting and other focal neurological deficits. He had no significant past history of chronic medical diseases such as HTN, DM, TB, etc. He is a non-smoker and non-alcoholic. There was no history of drug allergy. General examination was normal. Local ENT, head and neck examination revealed there was a generalized swelling in left temporal region measuring about 7x8x1 cm. The swelling extended anteriorly upto zygoma, posteriorly 5 cm anterior to occipital protuberance, superiorly 4 cm from superior attachment of the pinna and inferiorly upto tip of mastoid (Figure 5). It was non tender and hard on palpation with no mobility. Surface was smooth and the margins were well defined. Left external auditory canal was narrow compared to the right due to bulge in posterior bony EAC wall which was hard on probing. Tympanic membrane couldn't be visualized. Rinne was negative on the left with weber test lateralized to the same ear. Rest of the examination of nose, oral cavity, oropharynx and neck were normal. Benign bony tumor was suspected and the patient was advised for HRCT temporal bone. CT scan showed expansile bony lesion affecting the squamous, petrous, and tympanic portion of the temporal bone with heterogeneous density (Figure 6). Pure tone audiometry showed mild conductive hearing loss of 28 dB in the left ear. Diagnosis of fibrous dysplasia of left temporal bone was made. Shaving and recontouring squamous portion of left temporal bone was done with cortical mastoidectomy by extended post aural approach (Figure 7). Temporalis muscle was divided to expose the squamous portion. There was soft fibro-osseous tissue present between outer and inner table of squamous potion of the temporal bone, root of zygoma and superior and posterior wall of external auditory canal. Following the procedure, temporalis muscle was sutured back after filling the cavity with absorbable gel pack. Wound was closed in 3 layers. Patient was discharged on 6th post-operative day after suture removal. However, he developed wound gape on the 10th POD for which he was readmitted and resuturing was done. The sutures were removed after 6 days. Histopathological report on follow up showed osseous tissue composed of irregularly shaped trabeculae of immature bone without osteoblastic rimming. These features were suggestive of fibrous dysplasia (Figure 8).

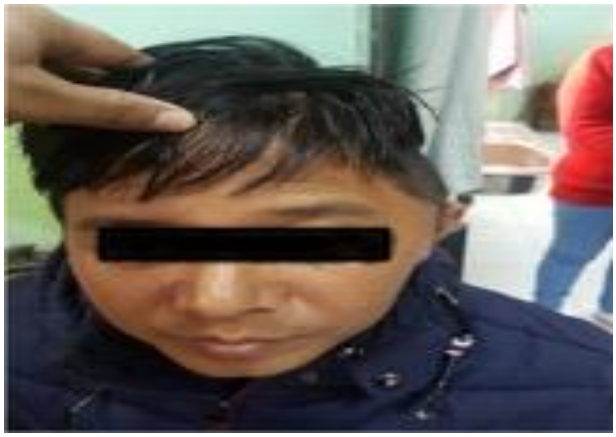

Figure 5: Case with fibrous dysplasia of left temporal bone.

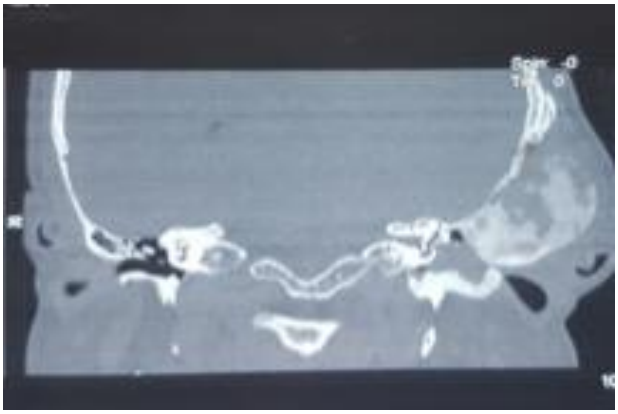

Figure 6: HRCT temporal bone showing expansile bony lesion with heterogenous density of left temporal bone.

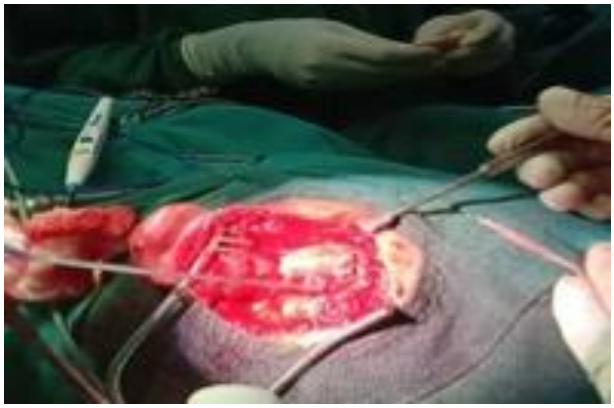

Figure 7: Per-operative picture of squamous portion of left temporal bone affected by fibrous dysplasia.

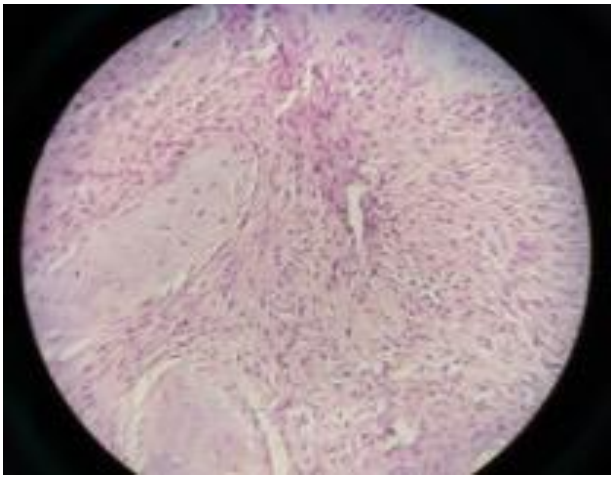

Figure 8: Histopathological slide showing osseous tissue composed of irregularly shaped trabeculae of immature bone without osteoblastic rimming.

DISCUSSION

Fibrous dysplasia is a benign but a slowly progressive disease. Regarding its etiology, several theories have been put forth with earlier theories suggesting disorders in mesenchymal differentiation and osteoblastic proliferation.[3,4] Some suggested arrest of bone at immature woven state led to development of fibrous dysplasia.[5] However, recent researches suggest a genetic basis with mutations involving proteins responsible for cellular signaling such as G_s-α which affects intrinsic GTPase activity.[6]

Male is to female ratio is 2:1, with prevalence being more common in first 2 decades of life.[7] In our series, two were male and one was female and the disease onset was on early third decade of the first case and 5th and 6th decades of the second and third case respectively. Temporal bone is not uncommon site to be involved. Various literatures have quoted various figures ranging from 10% to 24%.[2,8] Common presentations in a patient having temporal bone fibrous dysplasia are swelling in the mastoid region, aural fullness and decreased hearing from the obstruction of external auditory canal. On rare instances there might be sensorineural hearing loss due to diffusion of toxins in inner ear, pulsatile tinnitus, vertigo and facial nerve palsy.[7,9] Also, there is always a good chance for the development of cholesteatoma. In our study, two cases had progressive aural fullness while one had progressive swelling in the supra auricular area.

Investigation of choice is HRCT temporal bone. Various CT scan appearances of the disease have been described so far e.g. pagetoid, sclerotic, and cystic. Pagetoid pattern, being the most common, is characterized by a ground-glass appearance. Dense lesion is seen in the sclerotic pattern and cystic pattern has radiolucency in the center and hyperdense area in the surrounding.[4] These 3 different appearances can be seen in the CT scan of our three cases (Figure 1, 3 and 6).

Although imaging aids a lot in the diagnosis, histo-pathological examination is required for the confirmation. Microscopic appearance shows irregular trabeculae of woven bone intermixed with connective tissue stroma. A characteristic pattern 'cemmenticles' i.e., small bony spicules interspersed throughout the lesion is seen only in skull base lesions.[11]

Management depends on the symptomatic presentation. Lesions without any cosmetic deformity and absence of symptoms can be followed up conservatively with repeated scans. On the other hand, symptomatic lesions may require surgical intervention. However, complete surgical excision may not always be achieved. In our cases, indications for the surgery were progressive hearing loss and cosmetic deformity. Medical treatment is only reserved for the symptomatic relief. Literatures have shown results of bisphosphonates in reducing the incidence of fractures.[12]

CONCLUSION

Temporal bone involvement in fibrous dysplasia can present with a range of aural and extra aural symptoms. HRCT temporal bone is the investigation of choice. There may be variable CT scan findings which should be kept in mind. Definite surgical goal is not always achievable and the surgery should only be indicated in symptomatic cases. Progressive hearing loss, periauricular swellings and cholesteatoma are the commonest indications for the surgery in cases with temporal bone fibrous dysplasia.

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