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Case Report

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External auditory canal stenosis due to isolated fibrous dysplasia of temporal bone: a case report

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ABSTRACT

Benign disease of bone marrow in which marrow is reorganize into fibrous tissue and immature woven bone, is known as fibrous dysplasia. Involvement of temporal bone is rare and can be isolated or in monostotic or polystotic form with various otologic manifestations. We are here reporting a case of fibrous dysplasia of the unilateral temporal bone who presented with hearing loss and stenosis of the external auditory canal. External auditory canal stenosis due to fibrous dysplasia was managed with canalplasty and patient was free of recurrence till last follow up. Fibrous dysplasia of isolated temporal bone is a rare entity which requires high grade of suspicion in a patient who presents with unusual bony swellings in the ear. Its management usually includes resection of the most affected part of temporal bone in order to achieve auditory canal patency and restoration of hearing. After surgery, a close follow up is warranted due to its propensity of recurrence.

Keywords: Benign, Canalplasty, External auditory canal, Mastoidectomy, Radiotherapy

INTRODUCTION

Benign disease of bone marrow in which marrow is reorganize into fibrous tissue and immature woven bone, is known as fibrous dysplasia. The temporal bone sporadically may be involved in disease process, approximately 100 cases described until now.[1-3] Schlumberger presented the first temporal bone fibrous dysplasia case in 1946.[4] It may occur as part of Albright syndrome characterized by multiple bony lesions, abnormal pigmentation, endocrine dysfunction, and precocious puberty in girls or it may exist alone in the monostotic or polyostotic form. The monostotic form is more common and usually occurs in the skull, ribs, proximal femur, or tibia. In the polyostotic form, skull lesions are seen in more than 50% of patients.

CASE REPORT

Informed written consent of the patient had been taken for reporting and publication of all types of data related to this case. A 21-year-old boy presented to department of ENT with chief complaint of progressive hearing loss in right ear. On otoscopy only small part of tympanic membrane was visible. Otoendoscopy (Figure 2) through the stenotic segment showed normal tympanic membrane in most of the parts. Pure tone audiometry revealed moderate conductive hearing loss in the right ear. High resolution computed tomography (CT) of temporal bone showed expansion of diploic space involving right temporal bone extending in mastoid process and petrous part. There was ground glass appearance of stroma with partial obliteration of right mastoid air cells suggestive of fibrous dysplasia (Figure 1). Mild narrowing of internal auditory canal was also evident. Right external auditory canal was also narrow in its middle part. Cochlea and semicircular canal appeared normal in CT scan. On left side all structures appeared to be normal (Figure 1). These findings were suggestive of fibrous dysplasia of right side of temporal bone (Figure 1). Patient was counseled and planned for canalplasty. Post auricular approach was used for canalplasty. Nerve monitor was

used to check the integrity of facial nerve. After elevating postauricular musculoperiosteal flap, tympanomeatal flap was elevated. Bony stenosis was drilled with diamond burr and canalplasty done to achieve a normal diameter of external auditory canal. External auditory canal was then packed with antibiotic soaked ear pack. Repeated canal packing was done in order to maintain canal patency. Biopsy of the drilled bone came out as fibrous dysplasia.

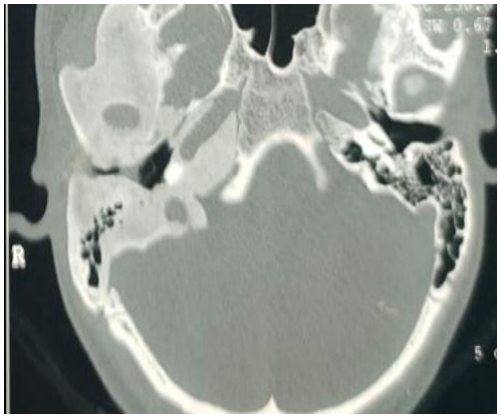

Figure 1: HRCT temporal bone showing expansion of diploic space involving right temporal bone extending in mastoid process and petrous part. There is ground glass appearance of stroma with partial obliteration of right mastoid air cells suggestive of fibrous dysplasia. There is mild narrowing of internal auditory canal. Right external auditory canal is narrow in mid part.

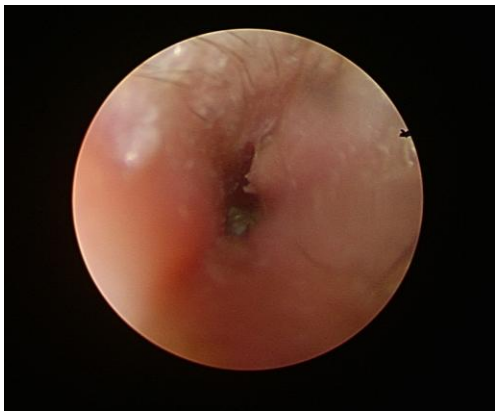

Figure 2: Endoscopic picture of right external auditory canal showing stenosis.

DISCUSSION

Benign disease of bone marrow in which marrow is reorganize into fibrous tissue and immature woven bone, is known as fibrous dysplasia. There is failure of the normal remodeling process.[5] Histopathological finding of disease is fibrous stroma arranged in a whorled pattern. The gene that encodes the α-subunit of stimulatory G protein is suffering from mutation in this disease.[6] Increased level of cyclic adenosine monophosphate modifies the transcription and expression of manifold downstream genes, which eventually result in pathologic lesion.[7] Clinical presentation of disease comprises bony deformity, pathologic fracture, and cranial nerve palsies. The disease starts early in life, the monostotic form may become latent at puberty, whereas the polyostotic form can continue to progress. Among all reported cases of fibrous dysplasia, the monostotic form is most common (70%), the polyostotic form in 23%, and Albright syndrome in 7%. In temporal bone the process generally begins as a painless, slowly progressive swelling that involves the mastoid or squama. Most common presentation of disease is narrowing of external auditory canal and occurs in about 80% of cases. Exostoses can be taken as diagnosis for the narrowing of canal, but fibrous dysplasia is detected during the second or third decade of life; in consistency soft, spongy, and gritty during surgery. Cholesteatoma can be formed in external auditory canal due to deception of keratin debris. Conductive hearing loss can occur due to entanglement of middle ear and ossicles or impediment of Eustachian tube. Facial nerve paralysis can be seen due to involvement of facial canal. Sensory neural hearing loss and vertigo is also seen sometimes when disease process encroaches otic capsule. Disease having a racial predilection, in Caucasians account for 80% of cases and Asians only 1%.[7] Fibrous dysplasia can be divided into three radiological patterns: pagetoid (56%) with a "ground glass" appearance due to a mixture of dense and radiolucent areas of fibrosis, sclerotic (23%) with homogeneous dense areas, and cystic (21%) with a radiolucent ring, oval or round, and a capsule of dense bone.[7] CT is useful in diagnosis and to know the extent of disease. Management of fibrous dysplasia is symptomatic. Operative procedures should be limited to biopsy and relief of functional deficits. Canalplasty and meatoplasty recommended for correction of the stenosis of external auditory canal. For cholesteatoma canal wall down mastoidectomy can be done. Restenosis as a result of regrowth of fibrous dysplasia occurs, and multiple procedures are sometimes needed. Due to risk of malignancy radiotherapy is contraindicated.[8] Long-term follow-up is indicated because of the potential risk of facial nerve involvement and for the progression of conductive hearing loss to profound sensory neural hearing loss.[9] Differential diagnosis of fibrous dysplasia are simple bone cysts, nonossifying fibromas, osteofibrous dysplasia, adamantinoma, low-grade intramedullary osteosarcoma, and Paget disease.

CONCLUSION

Fibrous dysplasia of temporal bone is a rare disease which can be isolated or associated with multiple bone involvement. The main feature of disease are hearing loss which is mainly conductive, can be sensorineural and vertigo when inner ear involvement is there. Treatment is mainly symptomatic and surgical procedure like canalplasty, mastoidectomy for stenosis and cholesteatoma.

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