INTRODUCTION

Benign fibro-osseous lesions (BFOLs) of the maxillofacial bones comprise a diverse group of pathologic conditions ranging from developmental lesions to neoplasms [1]. Regardless of subtype, all BFOLs demonstrate replacement of normal bone by fibrous connective tissue with an admixture of mineralized product, including osteoid, mature bone, and/or cementum-like calcifications. Clinically they may be asymptomatic or may cause significant cosmetic and functional disturbances [1-3]. Radiographically, BFOLs may manifest as solitary, multifocal, or multi quadrant disease. They may be defined ill or well; they may have a radiolucent, mixed radiolucent-radio paque, predominantly radio paque, or ground glass appearance; they may be monostotic or polyostotic lesions rarely cross the midline [4]. Fibrous dysplasia usually arises within the first or second decades of life, manifesting clinically as a slow-growing, painless expansion of the involved bone [5]. Facial asymmetry may be apparent. In rare cases, the expansion may be more rapid or begin to accelerate after a period of slow growth, resulting in marked facial deformity and potential nerve entrapment [6]. Active growth typically slows or ceases around the time of puberty or after skeletal maturation; however, sporadic periods of re growth may occur in adulthood [7]. Temporal bone dysplasia is rare [8-11] and usually asymptomatic. This paper presented three cases.

CASE PRESENTATION

Case 1

A 10 years old male presented a painless swelling of the left mastoid region of two years duration. At that time there was no hearing loss, ear discharge, pain, facial nerve affection or vertigo. Five months prior to presentation, the parents noticed progressive hearing loss on the side of the swelling and sought medical advice. The child was healthy without obvious morbidity. There was a painless smooth diffuse swelling of the left mastoid region blending with the rest of the skull. The auricle was slightly pushed outwards. The external auditory canal is obliterated by firm swellings of the anterior and posterior walls with intact skin. There were no other swellings all over the body, no sites of abnormal pigmentation. There was no evidence of any endocrinological abnormalities.

Audiological evaluation revealed left-sided conductive hearing loss of 45 dB. The right ear has a normal audiogram.

Diffuse affection of all parts of the temporal bone by a homogenous appearance of ground glass appearance. The middle ear space is preserved but restricted. Ossicles appear intact but entrapped by the restricted space. The lateral two-thirds of the external auditory meatus were obliterated by swellings of all the walls. The drum is obscured by the swellings. The otic capsule is spared but looks as a hyperdense island within the ground glass bone. The internal auditory meatus looks normal with a comparable diameter as the opposite side although in some cuts it looks narrower at its porus. There is apparently no neural compression with a normally appearing fundus and facial canal (Figures 1, 2).

Due to the absence of signs of nerve entrapment, and the high risk on the facial nerve in the event of exploration, the patient and his parents were counseled to wait and observe. They were also instructed to report immediately whenever there were signs of hearing deterioration, facial twitchs of facial weakness. A repeat scan is scheduled 6 months after the initial presentation.
Case 2

This was an 18 years old boy presenting with an attack of acute suppurative otitis media with fever, otalgia and otorrhea. He was treated medically at another facility by intravenous ceftriaxone, local ear drops and analgesics. His symptoms abated but there was still some tenderness over the mastoid and hearing loss. A CT scan was ordered and showed cloudiness of the middle ear and mastoid cells. There was also a suspicious cavity in the mastoid (Figure 3,4). A congenital cholesteatoma was suspected and an MRI was also performed. It showed a rounded mass with a hyperintense signal on T1 and a hypo intense signal on T2 (Figure 5,6). Consequently, an exploratory mastoidectomy was scheduled. It showed a smooth cavity full of fibrous tissue and bony spicules. The mastoid was otherwise healthy and the middle ear full of seromucous fluid. Biopsy revealed a fibro-osseous lesion.

DISCUSSION

Fibrous dysplasia is a rare benign intra medullary fibro-osseous lesion, which may present in either monostotic or polyostotic forms [1,12-14]. It is a genetic non inherited condition caused by missense mutation in the GNAS1 gene on chromosome 20 [15,16]. It is characterized by abnormal proliferation of fibrous tissue interspersed with normal or immature bone. It occurs in equal proportions in males and females, most often during the first two decades of life. In 3-5% of cases it may be...
CONCLUSION

Fibro-osseous lesions of the temporal bone are uncommon. They usually present with minor symptoms and signs and in most cases do not warrant any management. However follow up is mandatory and surgical treatment may be necessary if there are any functional or cosmetic effects.

REFERENCES


