Case Report

Fibrous Dysplasia of the Temporal Bone

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INTRODUCTION

Fibrous dysplasia is a relatively uncommon condition that presents in the adolescent age group. Rarely, it is isolated to temporal bone. We present a case report of fibrous dysplasia that has involved the entire temporal bone.

CASE REPORT

A 16 year-old schoolboy presented with a history of left postauricular swelling. He first noticed the swelling 2 years earlier and it had been progressively increasing in size. He did not complain of hearing loss, pain or any facial weakness. On examination, a smooth swelling was noted behind the left pinna, encompassing the mastoid prominence. It was non-tender, bony hard, smooth and diffuse with indistinct margins. The pinna itself was normal.

Otoscopic examination revealed narrowing of the ear canal posteriorly with normal tympanic membrane. Tuning fork examination and free-field voice test revealed clinically normal hearing. Other ear, nose and throat examinations were entirely normal. No abnormal skin or cutaneous pigmentation were noted. No cranial nerve palsies were noted. There was no other bony swelling elsewhere on his body. Pure ton audiogram revealed normal hearing.

Computed tomography (CT) revealed an opacity involving the entire left temporal bone, extending to the petrous apex and encompassing the labyrinth. It was radiolucent with ground glass or bubbly appearance (Fig. 1). In comparison, the right temporal bone was well pneumatized up to the petrous apex. A diagnosis of fibrous dysplasia was made on the basis of these findings. As the patient was asymptomatic and the swelling was not cosmetically obvious, he was managed conservatively. The natural progression of the disease is self-limiting. Hence the patient is being followed up with a view to possible surgical intervention if his hearing becomes impaired or if any obstructive symptoms appear.

DISCUSSION

Lichtenstein in 1938 coined the term fibrous dysplasia to describe a disorder characterized by the progressive replacement of normal bone elements by fibrous tissue. Fibrous dysplasia is a benign disease in which there is slowly progressive growth of abnormal fibro-osseous tissue which replaces the normal medullary bone. Histologically the tissue consists of disorganized bony trabeculae and spindle cells surrounded by a fibrous matrix. Theories about the aetiology of this disease include aberrant differentiation of the mesenchyme during bone formation, an arrest of bone at the immature woven stage, or a disturbance of cancellous bone maintenance [1,2].

Fibrous dysplasia can be monostotic, involving a single bone, or polyostotic, involving two or more bones. Albright’s syndrome describes a polyostotic disease associated with pigmented skin lesions, endocrine abnormalities and precocious puberty.

In the head and neck region, the skull and facial bones are involved in 10–25% of cases of monostotic fibrous dysplasia and in 50% of the polyostotic variety [3]. The ethmoids were the most commonly involved (71%), followed by the sphenoid (43%), frontal (33%), maxilla (29%), temporal (24%), parietal (14%), and occipital (5%) bones [4].

The most common presenting features of fibrous dysplasia involving the craniofacial region bones include atypical facial pain and headache, complaints referable to the sinuses, diplopia and proptosis, hearing loss, and facial numbness [4]. Hearing loss is usually conductive in nature but sensorineural hearing loss has been reported in 17% of patients [5]. Facial nerve involvement was seen in 10% and almost 40% had cholesteatoma.

In an extensive review of 69 cases by Nager et al. [1], the male-female ratio was 2:1 and the most common presenting symptoms were progressive hearing loss (56%), increasing size of the temporal bone (50%), and progressive occlusion of the external auditory canal (42%). Some of the patients even had multiple cranial nerve involvement.

A clinical staging system has been proposed for fibrous dysplasia. Stage 1 is for asymptomatic disease while symptomatic disease is stage 2, and stage 3 is for those with complications [6]. Management of stage 1 is basically conservative with regular follow-up, as in our patient. Stage 2 and 3 would need surgical decompression to alleviate symptoms.

The three major radiographic types of fibrous dysplasia are
pagetoid, sclerotic and cystic [3]. Any of these types may involve the temporal bone and related structures, including the external ear canal, middle ear, jugular foramen, or rarely, the otic capsule. The radiographic differential diagnosis includes Paget’s disease, ossifying fibroma, osteoma, meningioma, haemangioma and mucocoele [7]. Areas of cortical erosion and the characteristic coarse bony trabeculae of Paget’s disease differentiate it from fibrous dysplasia. The radiographic density and histological appearance of fibrous dysplasia and ossifying fibroma are similar. They can be differentiated on the basis of their growth pattern. Ossifying fibroma grows outwards from a finite centre producing mass effect on adjacent structures in contrast to the diffuse expansion in fibrous dysplasia. In meningioma there is always a soft tissue component adjacent to the bone while haemangiomas contain very coarse, ray-like heavy bony trabeculae. Mucocoeles have a well-defined bone margin and expansile growth pattern but can be confused with the purely radiolucent type of fibrous dysplasia that contains no bone spicules. In this instance, magnetic resonance imaging (MRI) would be useful as mucocoeles will show a lack of enhancement.

Definitive diagnosis and treatment hinges on surgical debulking and biopsy. In this patient no biopsy was taken as the patient was asymptomatic and the diagnosis was quite evident from the CT. Hearing loss and cholesteatoma are definitive indications for surgical intervention. Periodic CT can be used to follow the progression of the disease and assess the need for any surgical intervention [8].

REFERENCES