



Fibrous Dysplasia of the Temporal Bone Presenting with Vertigo

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Fibrous dysplasia (FD) is a slowly progressive, benign, chronic fibro-osseous tissue disease, which is characterized by abnormal proliferation of fibrous tissue interspersed with normal or immature bone.

In this report we describe a patient with monostotic fibrous dysplasia that involved temporal bone presenting with vertigo, and discuss diagnostic work-up, histopathology, and treatment options of the disease, reviewing the literature.

Keywords: fibrous dysplasia, vertigo,

Introduction

Fibrous dysplasia (FD) is a slowly progressive, benign, chronic fibro-osseous tissue disease, which is characterized by abnormal proliferation of fibrous tissue interspersed with normal or immature bone.

Von Recklinghausen first described the term fibrous dysplasia, although Lichtenstein first suggested the terms fibrous dysplasia and polyostotic fibrous dysplasia in 1938. Ten to 25% of the monostotic form of FD involves craniofacial region [1]. Anatomically, the most commonly involved area in the skull is the ethmoid bone (71%) [1]. The next most common site of skull involvement is the sphenoid bone (43%), followed by the frontal bone (33%) and maxilla (29%) [1]. 24% of the disease involves temporal bone [2]. In this report we describe a patient with monostotic fibrous dysplasia that involved temporal bone presenting with vertigo, and discuss diagnostic work-up, histopathology, and treatment options of the disease, reviewing the literature.

Case Report

A 35-year-old woman attended the outpatient clinic of Azerbaijan Medical University,

Otorhinolaryngology Department with the complaints of a vertigo, headache, and dizziness for 6 months. Previously she had applied to a public hospital with the same complaints and medical treatment was administered with the diagnosis of vestibular neuritis but as the complaints did not regress, she was referred to our clinic. The previous medical and family history was non-specific. In her physical examination, the external auditory canal and tympanic membrane were normal. Nasal, nasopharyngeal, oral and oropharyngeal, hypopharyngeal examinations with rigid and fiberoptic endoscopy were otherwise normal. Audiologic tests, including pure-tone and impedance audiometry, were normal and other laboratory investigations were all in normal limits. Neurological and cranial nerve exams were within normal range except for vestibular nerve dysfunction demonstrated by caloric testing and electronystagmography that showed reduced vestibular function on the left side. Temporal bone computed tomography (CT) demonstrated bone growth with temporal bone sclerosis and narrowing of internal auditory canal on the left, compatible with picture of temporal bone fibrous dysplasia on the left and normal right temporal bone. As the other system and neuro-

logical examinations were normal the vestibular symptoms were related to temporal bone fibrous dysplasia due to irritation or compression of vestibular nerve. High dose corticosteroids were administered (1mg/kg metilprednisolon) and the symptoms regressed one week after the treatment.

Discussion

Fibrous dysplasia (FD) is a slowly progressive, benign, chronic fibro-osseous tissue disease, which is characterized by abnormal proliferation of fibrous tissue interspersed with normal or immature bone. The precise etiology of fibrous dysplasia is currently unknown. The disease tends to develop in the pre-adolescence years, and is predominant in male patients (2:1).

Fibrous dysplasia can be classified as unifocal (the monostotic form of the disease, MFD involving only one bone) 70%, multifocal (the polyostotic form of the disease, PFD involving multiple bones) 30% or part of McCune Albright Syndrome (bone involvement associated with skin lesions and endocrinopathies) 3%. Unifocal type typically involves the ribs and femur. The lesions grow slowly and growth usually stops after puberty. Because many patients are asymptomatic and are often diagnosed incidentally, the incidence of the monostotic form is considered to be highest. 10 to 25% of the monostotic form of the disease involves craniofacial region. Ethmoid bone is the most commonly involved region in the skull followed by sphenoid and frontal bone. Involvement of temporal bone is seen %24 of the cases. Determining the true incidence of fibrous dysplasia, particularly for the more prevalent monostotic form, is difficult because many patients are asymptomatic and are often diagnosed incidentally after radiographic evaluation for other reasons [4].

Temporal bone involvement usually presents with clinical symptoms because narrowing of external auditory canal leads to progressive conductive hearing loss [3]. Other symptoms include retroauricular bulging, otalgia tinnitus and otorrhea. When the ear capsule is involved then sensorineural hearing loss may occur. In about 40% of the cases cholesteatoma may develop, and it is the most frequent complication of the disease [5]. Also the involvement of the facial nerve is seen in 10% of the patients. In our case, the fibrous lesions of the temporal bone led to narrowing of the left internal auditory canal causing vestibular nerve dysfunction. The reduced activity of the vestibular nerve on the left side was demonstrated by caloric ENG and balance tests. We believe that the reduced diameter of the internal auditory canal compressed the vestibular nerve causing irritation and dysfunction of the nerve.

The differential diagnoses of temporal bone fibrous dysplasia include Paget's disease, hyperparathyroidism, local reaction to meningioma, osteoma, eosinophilic granuloma, osteochondroma, and sarcomatous neoplasm [6].

It is not always possible presently, to control fibrous dysplasia since there is no conservative treatment. The simple presence of the lesion does not justify surgical intervention. The bone invasion of external auditory canal (enough to produce conductive hearing loss), recurrent infections and secondary cholesteatoma in the external auditory canal are the indications for surgery of temporal bone fibrous dysplasia. The secondary complications

of fibrous dysplasia may be secondary external cholesteatoma behind the canal stenosis or obliteration (16-40 %), erosion of the middle ear ossicles inner ear capsule and fallopian canal leading to a labyrinthitis and facial palsy. Involvement of the middle and posterior cranial fossa dura, lateral sinus, jugular bulb and carotid artery may also be seen. If the hearing is affected or recurrent infection or secondary canal cholesteatoma is detected, surgical intervention must be performed. The surgeon must be aware that surgery of the dysplastic temporal bone can be hazardous because landmarks are often obliterated and intra-operative bleeding can be vigorous [7].

As hearing of our patient was within normal limits and the only symptom was vertigo we started medical treatment with steroids and the symptoms regressed.

In conclusion, we report a rare lesion effecting internal auditory canal and interestingly affecting only vestibular nerve in same side. the symptom regressed via steroid. Otherwise we need operated and enlarged the canal.

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