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Asymptomatic fibrous dysplasia of the temporal bone

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Abstract

Introduction: Fibrous dysplasia is a bone disorder of unknown origin in which normal bone is replaced with fibrotic tissue and disorganised bone trabeculae. The temporal bone is rarely affected. Because of the slowly progressive course of the disease, many mild cases may never be recognised and are found incidentally. We present a patient with fibrous dysplasia of the right temporal bone, who had few complaints.

Objective: A 62-year-old man was incidentally found to have fibrous dysplasia of the temporal bone on routine computed tomography scan.

Method: One case report.

Results: Computed tomography showed a thickening of the right temporal bone with a ground-glass appearance. The 62-year-old man opted for watchful waiting.

Conclusion: We have presented an asymptomatic fibrous dysplasia of the temporal bone. Mild cases may never be recognised and are found incidentally because of their slow progression.

Key words: Fibrous Dysplasia; Temporal Bone; CT Scan

Introduction

Fibrous dysplasia is a slowly progressive bone disorder of unknown origin in which normal bone is replaced with fibrotic tissue and disorganised bone trabeculae. It was first described by McCune and Bruch in 1937.[1] The disease is classified into three variants: monostotic (involving one bone), polyostotic (involving more than one bone) and McCune–Albright syndrome (associated with endocrinopathies and skin hyperpigmentation).[2] Fibrous dysplasia of the temporal bone is very rare. Most involvement of the temporal bone is unilateral and associated with conductive hearing loss attributable to compression of the external auditory canal. Sensorineural hearing loss (SNHL) is usually due to otic capsule involvement by fibrous dysplasia or secondary cholesteatoma. However, the progression of the disease is so slow that it takes years before patients become symptomatic.

This report presents a patient with fibrous dysplasia of the temporal bone incidentally found on a routine computed tomography (CT) scan.

Case report

A 62-year-old man consulted the Department of Neurosurgery after a transient episode of syncope and a slight headache. He was referred to the ENT department with an abnormal shadow on the right temporal bone.

There was no external deformity of the contour of the patient's head and no skin lesions. Otoscopic evaluation did not demonstrate any stenosis of the right external auditory canal and the tympanic membrane was normal. Cranial nerve function and cerebellar function were intact. Hearing in both ears was within normal limits by pure tone audiometry examination. Caloric responses did not demonstrate canal paresis on either side. There was no family history of bone disorders, early puberty, or other endocrine or genetic abnormalities.

A CT scan of the right temporal bone demonstrated extensive thickening of the diploic and medullary spaces with a ground-glass appearance, moderately dense new bone and fibrous tissue (Figure 1). There was considerable expansion of the temporomandibular joint, but the patient did not have any functional complaints regarding mastication. The external auditory canal (Figure 2), middle ear, ossicular chain, cochlear capsule and internal auditory canal (Figure 3) appeared normal. Both the inner and outer cortices of the involved bones were intact, an important characteristic of fibrous dysplasia. Magnetic resonance imaging (MRI) showed a low-to-intermediate signal on T1-

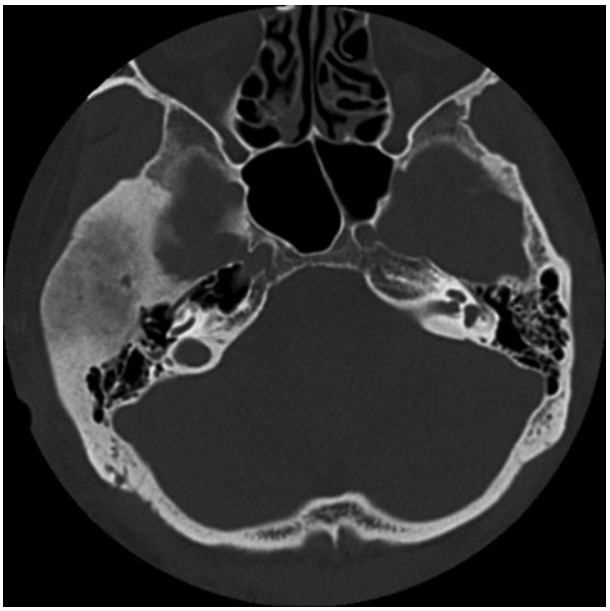

FIG. 1

Axial computed tomography scan demonstrating a massive fibrous dysplasia in the right temporal bone. The overlying cortical bone remained intact.

weighted images and a heterogeneous signal on T2-weighted images (Figure 4).

His fainting and headache disappeared the following day and there have not been any further complaints. Therefore, surgical exploration has not been performed.

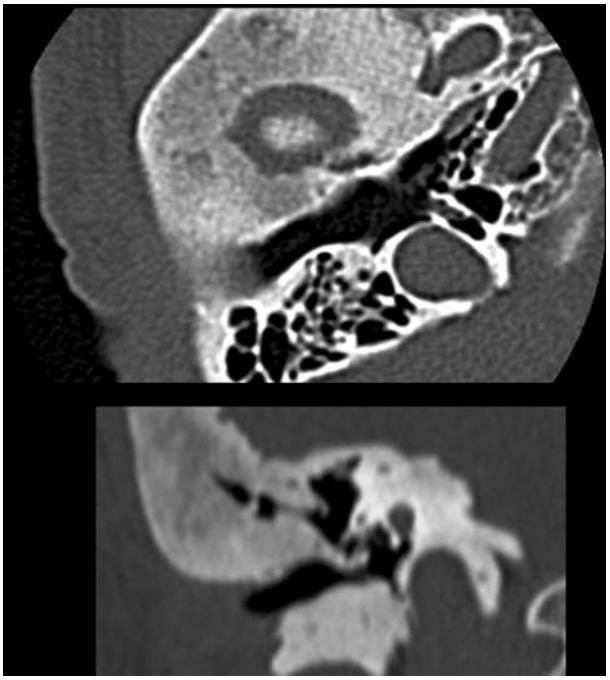

FIG. 2

Computed tomography scan of the right temporal bone demonstrating no narrowing of the external auditory canal. There is expansion of the disease into the temporomandibular joint. Upper = axial image; Lower = coronal image

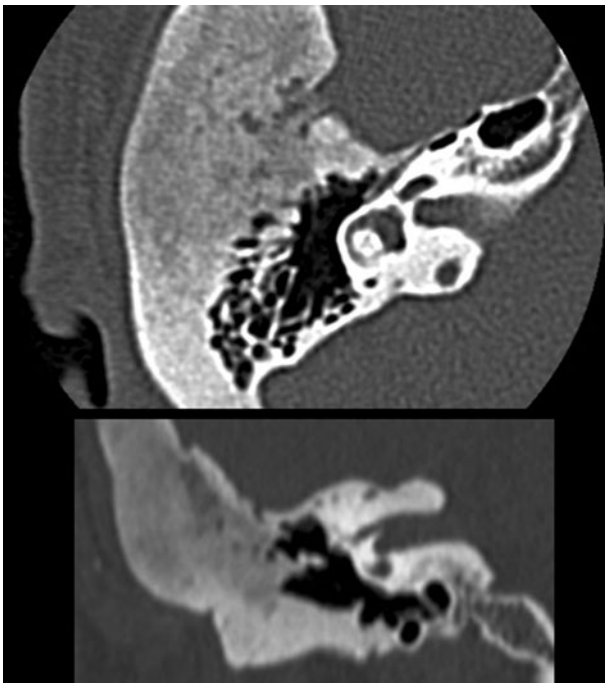

FIG. 3

Computed tomography scan of the right temporal bone demonstrating no narrowing of the internal auditory canal. Upper = axial image; Lower = coronal image

Discussion

Fibrous dysplasia is a relatively uncommon condition of the bone and rarely affects the temporal bone. It is classified into three subgroups: (1) monostotic fibrous dysplasia – involvement of a single bone (70 per cent); (2) polyostotic fibrous dysplasia – involvement of multiple bones (27 per cent); and (3) McCune–Albright syndrome – bony involvement associated with endocrinopathies and skin hyperpigmentation (3 per cent).[2] Craniomaxillofacial bony involvement occurs in only 10 per cent of monostotic fibrous dysplasia cases.[3] The maxilla and mandible are the most frequently affected craniomaxillofacial bones. The temporal bone is affected in only 18 per cent of those with skull involvement.[4,5]

The mean age of occurrence of the three types is 14, 11 and 8 years old for the monostotic form, polyostotic form and McCune–Albright syndrome, respectively.[6] Yaunus and Haleem[7] reported an infant case in a nine months old with an absent kidney. Temporal bone involvement with fibrous dysplasia is twice as common in males as it is in females.[2] This disease has a racial predominance, in that Caucasians account for >80 per cent of all cases, African-Americans 2 per cent and Asians only 1 per cent.[4]

Pathologically, fibrous dysplasia appears as a poorly localised lesion replacing normal bone. The tissue contains disorganised bony trabeculae and spindle cells surrounded by a fibrous matrix.[4,7] Theories of 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 in

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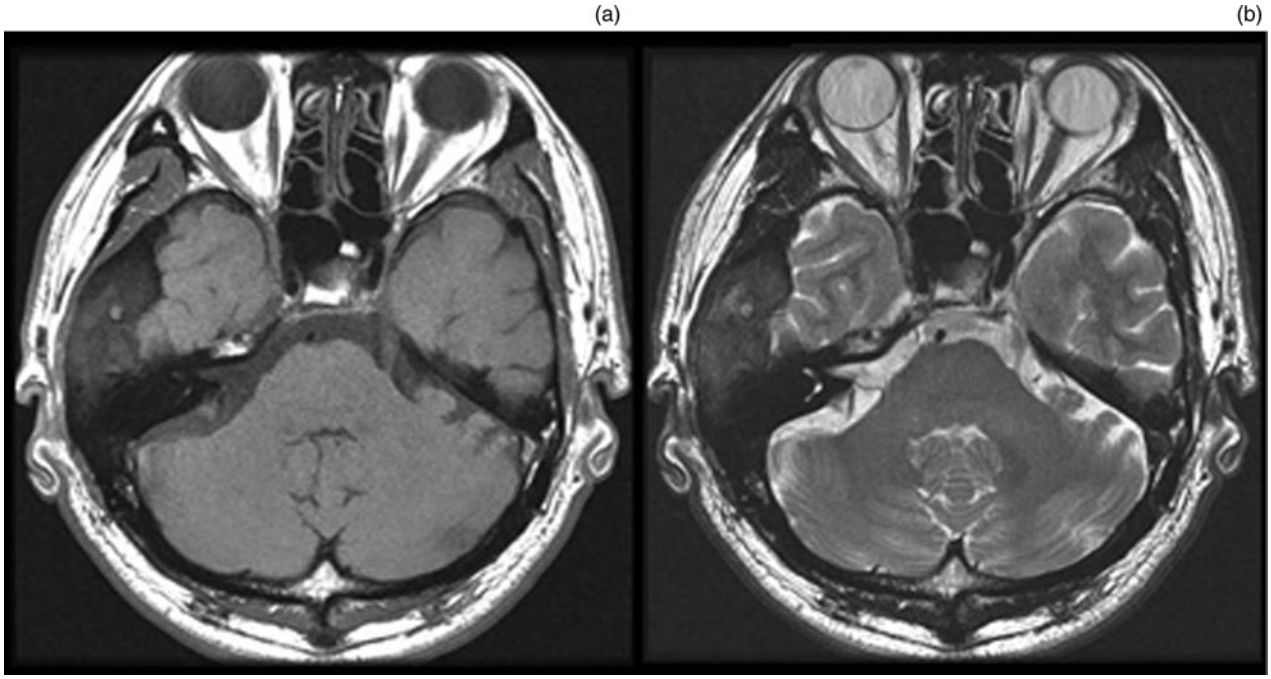

FIG. 4
Magnetic resonance imaging. (a) T1-weighted image. (b) T2-weighted image.

cancellous bone maintenance.[2] The bony cortex is expanded and thinned, but usually remains intact, maintaining a smooth contour. There is no true capsule, and the margin between normal and abnormal bone is indistinct.

Barrionuevo *et al.*[8] classified fibrous dysplasia of the temporal bone into the following three stages according to the progression of the disease: stage 1, latent phase; stage 2, symptomatic phase; and stage 3, complication phase. Although our case was stage 1, almost all cases of fibrous dysplasia of the temporal bone in the 77 surveyed cases were in stage 3.[9] The most common presenting symptom is deafness. Other symptoms include post-aural or temporal swelling, otorrhoea, otalgia and pulsatile tinnitus. Abnormal physical findings are unilateral conductive or SNHL, stenosis of the ear canal including external auditory canal mass[10] and cholesteatoma. Cholesteatoma is seen in 40 per cent of cases and usually originates from the external canal wall.[5,11] Sensorineural hearing loss, resulting from involvement of the internal auditory canal, is seen in 17 per cent of patients.[4] McCall *et al.*[12] reported posterior semi-circular canal dehiscence arising from cystic degeneration of fibrous dysplasia. Facial nerve involvement is seen in 10 per cent of these patients.[5,13,14] Although not common, function of all cranial nerves can be affected.[2] The case presented here showed expansion of the disease into the temporomandibular joint, but did not show any symptoms. If displacement of the temporomandibular joint occurs, the patient may complain of dental malocclusion or temporomandibular joint dysfunction.[15]

The radiographic appearance varies with the ratio of fibrous and osseous tissue in the affected bone, ranging from radiolucency to radiopacity. Three radiographic patterns have been identified: pagetoid, sclerotic and cystic.[4,14,16] The pagetoid pattern seen in 56 per cent of cases is typified by a mixture of dense and radiolucent fibrous areas with bone expansion. Homogeneously dense areas with bone expansion are typical of the sclerotic pattern seen in 23 per cent of radiographic studies. The cystic pattern shows spheric or oval lucent regions with a dense boundary seen in 21 per cent of cases. Our case showed the most common pagetoid pattern, which is characterised by a ground-glass appearance on CT imaging. Magnetic resonance imaging, in general, does not give as much information as CT.[17] It exhibits a low-to-intermediate signal on T1-weighted images and a heterogeneous signal on T2-weighted images. Post-gadolinium T1- and T2-weighted imaging shows moderate to marked enhancement.[18] Signal intensity and the degree of enhancement depend on the relative proportions of bony trabeculae, cellularity, collagen and cystic changes.

- **This report presents a patient with asymptomatic fibrous dysplasia of the temporal bone**
- **Diagnosis was based on computed tomography and magnetic resonance imaging**
- **Surgical exploration and treatment was not performed due to the patient's lack of complaints**
- **Treatment should be based on patient symptoms and disease progression**

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Treatment and definitive diagnosis rely on surgical debulking and biopsy. The decision to treat depends on the degree of cosmetic or functional impairment. Fibrous dysplasia is a benign condition with a good prognosis. As fibrous dysplasia is likely to become quiescent, surgical intervention should be as conservative as possible.[5,7,14] The disease process usually stops at puberty.[7] However, when an aggressive or unpredictable progression occurs, including cases involving inner ear or skull base structures, appropriate surgical intervention should be considered. Despite some instances of recurrence, surgical treatment has revealed relatively good outcomes.[19,20] Radiation therapy should be avoided,[5,14,21] because past experience shows an increase in the incidence of malignant transformation to 44 per cent after radiation treatment.[22,23] Spontaneous malignant transformation is extremely rare (0.5 per cent);[16] however, malignant change is indicated by pain, aggressive growth and elevation of serum alkaline phosphatase.[24]

Conclusion

This report presents a patient with asymptomatic fibrous dysplasia of the temporal bone. Diagnosis was based on CT and MRI. Surgical exploration and treatment was not performed due to the patient having no complaints.

References

1 McCune DJ, Bruch H. Osteodystrophia fibrosa: report of a case in which the condition was combined with precocious puberty, pathologic pigmentation of the skin and hyperthyroidism, with a review of the literature. *Am J Dis Child* 1937;**54**:806–48
2 Nager GT, Kennedy DW. Kopstein E. Fibrous dysplasia: a review of the disease and its manifestations in the temporal bone. *Ann Otol Rhinol Laryngol* 1982;**91**(suppl 92):1–52
3 Windolz F. Cranial manifestation of fibrous dysplasia of bone. *Am J Roentgenol* 1947;**58**:51–63
4 Magerian CA, Sofferman RA, McKenna MJ, Eavey RD, Nadol JB Jr. Fibrous dysplasia of the temporal bone: ten new cases demonstrating the spectrum of otologic sequelae. *Am J Otol* 1995;**16**:408–19
5 Morrissey DD, Talbot JM, Schleuning AJ. Fibrous dysplasia of the temporal bone: reversal of sensorineural hearing loss after decompression of the internal auditory canal. *Laryngoscope* 1997;**107**:1336–40
6 Van Horn PE, Dahlin DC, Bickel WH. Fibrous dysplasia: a clinical pathologic study of orthopaedic surgical cases. *Mayo Clin Proc* 1963;**38**:175–89
7 Yaunus M, Haleem A. Monostotic fibrous dysplasia of the temporal bone. *J Laryngol Otol* 1987;**101**:1070–4
8 Barrionuevo CE, Marcallo FA, Coelho A, Cruz GA, Mocellin M, Patrocinio JA. Fibrous dysplasia and the temporal bone. *Arch Otolaryngol* 1980;**106**:298–301
9 Abe T, Suzuki M, Ueyama T, Ina K, Mogi G. A case of monostotic fibrous dysplasia of the temporal bone [in Japanese]. *Practica Oto-Rhino-Laryngologica* 1999;**92**:713–19
10 Wang YC, Chen YA. Fibrous dysplasia of the temporal bone presenting as an external auditory canal mass. *Otolaryngol Head Neck Surg* 2009;**141**:655–6
11 Tochino R, Sunami K, Yamane H. Fibrous dysplasia of the temporal bone with cholesteatoma. *Acta Otolaryngol Suppl* 2004;**554**:47–9
12 McCall AA, Curtin HD, McKenna MJ. Posterior semicircular canal dehiscence arising from temporal bone fibrous dysplasia. *Otol Neurotol* 2010;**31**:1516–17
13 Sakamoto M, Hayashida T, Sugasawa M. A case of fibrous dysplasia of the temporal bone: evaluation of treatment performed 23 years ago. *Otolaryngol Head Neck Surg* 2001;**125**:563–4
14 Reddy KTV, Vinayak BC, Jefferis AF, Grieve DV. Fibrous dysplasia of the temporal bone. *Ann Otol Rhinol Laryngol* 1994;**103**:74–6
15 Pouwels ABPM, Cremers CWRJ. Fibrous dysplasia of the temporal bone. *J Laryngol Otol* 1988;**102**:171–2
16 Lambert PR, Brackmann DE. Fibrous dysplasia of the temporal bone: the use of computed tomography. *Otolaryngol Head Neck Surg* 1984;**92**:461–7
17 Chee GH, Chen JM. Fibrous dysplasia of the temporal bone. *Otol Neurotol* 2002;**23**:405–6
18 Casselman JW, De Jonge I, Neyt I, De Clercq C, D'Hont G. MRI in craniofacial fibrous dysplasia. *Neuroradiol* 1993;**35**:234–7
19 Kim YH, Song JJ, Choi HG, Lee JH, Oh SH, Chang SO *et al*. Role of surgical management in temporal bone fibrous dysplasia. *Acta Otolaryngol* 2009;**129**:1374–9
20 Yang H, Chen S, Zheng Y, Xu Y, Zhang X, Xiong H *et al*. Surgical treatment of monostotic fibrous dysplasia of the temporal bone: a retrospective analysis. *Am J Otolaryngol* 2012;**33**:697–701
21 Yagoda MR, Selesnick SH. Temporal bone fibrous dysplasia and cholesteatoma leading to the development of a parapharyngeal abscess. *J Laryngol Otol* 1994;**108**:51–3
22 Slow IN, Stern D, Friedman EW. Osteogenic sarcoma arising in preexisting fibrous dysplasia: report of a case. *J Oral Surg* 1971;**29**:126–9
23 Kaushik S, Smoker WR, Frable WJ. Malignant transformation of fibrous dysplasia into chondroblastic osteosarcoma. *Skeletal Radiol* 2002;**31**:103–6
24 Schwartz DT, Alpert M. The malignant transformation of fibrous dysplasia. *Am J Med Sci* 1964;**247**:1–20

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