Now you see the drum, now you don't: a rare case of conductive hearing loss in a child

N. Seymour*, R. Tranter, K. Argiris and N. Saunders

Department of Otolaryngology, Brighton & Sussex University Hospital Trust, UK

Abstract: *Introduction*: Fibrous dysplasia is a benign disease of the bones, which affects the differentiation and maturation of osteoblasts. Fibrous dysplasia can affect the craniofacial bones, which can be asymptomatic or present with symptoms commonly found in an ENT outpatient clinic.

Method: We present an unusual case of temporal bone fibrous dysplasia. This is used as a platform to discuss craniofacial fibrous dysplasia further and the investigations and management.

Results: Other cases have been reported on temporal bone fibrous dysplasia, but our case is unique in the presentation. Gold-standard diagnosis for craniofacial fibrous dysplasia is by histopathological sampling, although computerised-tomography imaging can be used to diagnose fibrous dysplasia if a biopsy cannot be sought.

Conclusion: Fibrous dysplasia of the temporal bones is uncommon. Many cases from other series are found incidentally from radiology reports.

Keywords: Temporal bone, fibrous dysplasia, craniofacial, auditory canal, hearing loss.

INTRODUCTION

Fibrous dysplasia is a slowly progressive and benign disease of the bones. It affects bone development and replaces normal bone with fibrous tissue. It specifically affects differentiation and maturation of osteoblasts (cells which synthesis bone) [1].

Fibrous dysplasia was first described by Von Recklinghausen at the end of the 19th century, although specimens showing characteristic lesions were found in prehistoric Anglo-Saxon remains by both Albright and McCune & Brunch in 1937 [2, 3].

There are three subtypes of the disease: monostotic, polyostotic, and McCune-Albright syndrome.

Monostotic is the most common, yet least aggressive, comprising 70% of cases, although this may be higher as lesions are often asymptomatic. As its name suggests it affects one bone. Lesions most commonly involve the long bones or skull, and it is usually diagnosed at 20 – 30 years of age [4].

The polyostotic form (approximately 30% of cases) usually has a childhood onset and affects multiple bones [4].

Lastly, McCune-Albright syndrome occurs in 3% of cases and is characterised by polyostotic fibrous dysplasia, endocrinopathies and skin pigmentation [3, 4].

So, why is fibrous dysplasia of interest to ENT surgeons? Fibrous dysplasia can involve craniofacial bones. In fact, craniofacial dysplasia is now commonly thought of as a subgroup of its own, and it is perhaps more common than we think.

We present an unusual case of craniofacial fibrous dysplasia and use this as a platform to discuss the investigations and management of craniofacial fibrous dysplasia, with some learning points from the case.

CASE REPORT

Our patient, a twelve-year-old boy, had been under routine review in the ENT outpatient clinic for 7 years with otitis media with effusion causing a bilateral conductive hearing loss, successfully treated with grommets.

He re-presented to clinic with a short history of a few weeks, with worsening hearing in his right ear. He had no tinnitus, otalgia or cranial nerve defects. There was also no notable past medical or family history. He is a twin, but his brother, who has Prader-Willi syndrome does not have any hearing deficit.

Otoscopy of the right ear showed a narrowed external auditory canal, but a normal tympanic membrane. During the next month, his ear canal significantly narrowed further, to the point where the

*Address correspondence to this author at the ENT Department, Brighton & Sussex University Hospital Trust, Eastern Rd, Brighton, East Sussex, BN2 5BE, UK; E-mail: nickyseymour@doctors.org.uk

E-ISSN: 2311-1917/17

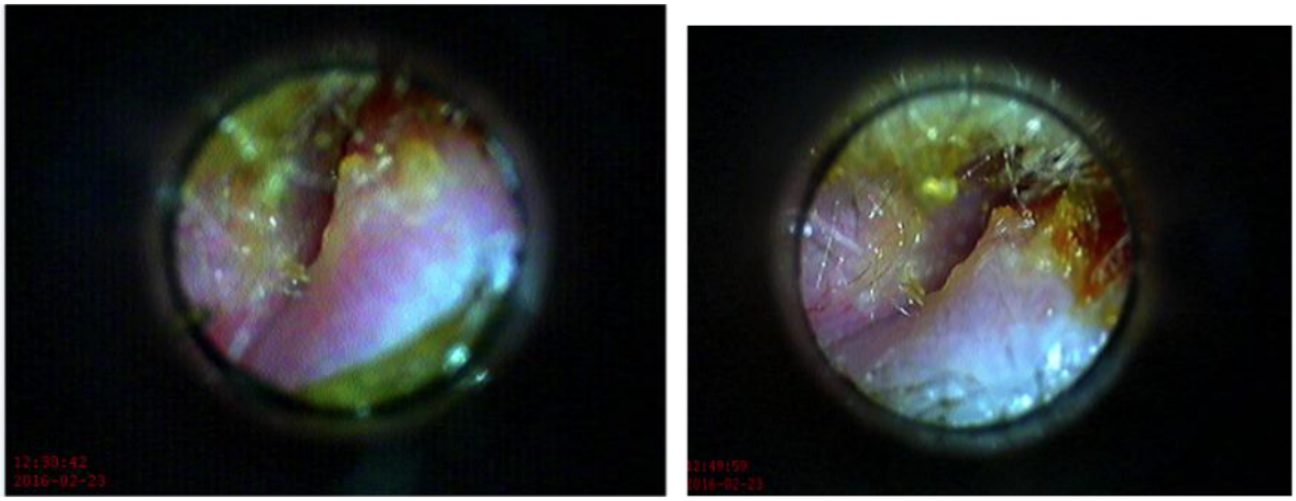

Figure 1: Right external auditory canal. Tympanic membrane cannot be visualised.

tympanic membrane could no longer be visualised (Figure **1**). Pure-tone audiometry showed a worsening moderate conductive hearing loss on the right side.

A high-resolution computerised tomography (CT) scan revealed a bony narrowing of the external auditory canal with expansion and ground-glass opacification of the petrous temporal bone, extending to involve the skull base at the junction of the middle and inner ear (Figure **2**). These findings are in keeping with fibrous dysplasia of the petrous temporal bone.

There is evidence that fibrous dysplasia in children becomes less active or even burns out after puberty [3]. As the patient is approaching this age we decided to monitor him with a further CT scan, with a view to possible surgery over the next couple of years. Reconstructing the ear canal is a significant undertaking in a child and it would be preferable to do this only once after the onset of puberty.

DISCUSSION

Fibrous dysplasia of the craniofacial bones can be asymptomatic or may cause deformity and dysfunction, depending on which structures it invades or compresses.

Craniofacial fibrous dysplasia causes a range of symptoms from those commonly seen in the ENT clinic such as hearing loss, nasal congestion and recurrent sinus infections to more unusual presentations like unilateral facial swelling or pain.

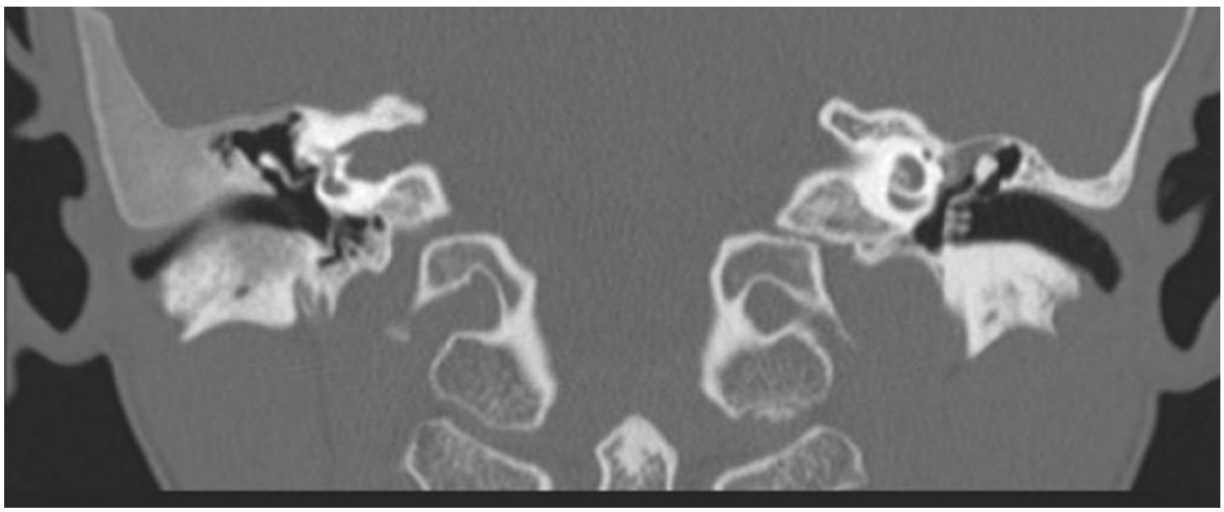

Figure 2: High-resolution CT scan image showing ground glass opacification and expansion of petrous temporal bone on the right.

Literature Search

An extensive *PubMed* literature search was carried out on craniofacial fibrous dysplasia, particularly of the temporal bone. Our main finding was a review of 66 cases over a 13-year period at two tertiary centres [5]. Our case is unique due to the rapid onset of symptoms over a short time period and also the age of our patient, with no other cases being reported in a paediatric patient.

Another case series of craniofacial fibrous dysplasia was carried out at The Johns Hopkins University over a 15-year period (1983-1998) [4]. They found 21 cases of craniofacial involvement, with only five patients having fibrous dysplasia of the temporal bones. They found that most cases were found incidentally on radiology reports [4].

Investigation and Management

The gold standard way to diagnose craniofacial fibrous dysplasia is by histolopathological diagnosis following biopsy. However, if the lesion cannot be biopsied due to poor access, or if it is well vascularised and biopsy may cause haemorrhage, then high-resolution CT imaging can provide a diagnosis. CT imaging shows expansion of the affected bone and loss the normal cortico-medullary differentiation, being replaced by a homogeneous ground-glass appearance, as in our case [5, 6].

The presence of the lesion alone does not usually justify surgery [4, 7]. Surgical intervention aims to restore function, prevent further complications from arising and improve the aesthetic appearance. Surgery therefore would be indicated with bone invasion of the external auditory canal, resulting in a significant conductive hearing loss, recurrent infections and secondary cholesteatoma in the external auditory canal and severe aesthetic deformity. A surgical decompression of the external auditory canal can be carried out to widen the canal and prevent further narrowing. Pathological bone is soft and spongy and therefore easily curetted; however, this cannot prevent recurrence [4, 8].

CONCLUSION

Fibrous dysplasia of the temporal bone is uncommon. Cases can be found incidentally from radiology reports. Therefore it is likely that it is more common than we think. Perhaps consider a diagnosis in atypical hearing loss, congestion or sinus infections and certainly in patients presenting with auditory canal stenosis.

Although craniofacial fibrous dysplasia is usually a monostotic form (affecting one bone only), consider that this may be part of a McCune Albright syndrome and take a thorough history and investigations into endocrinopathies.

Surgery is not the ‘go to’ management. It is best to avoid surgery prior to skeletal maturity due to recurrence; however, if the patient is symptomatic or rapid change in the lesion, this may warrant surgical management.

CONFLICT OF INTEREST

This case report presents no conflicts of interest

CONSENT

Consent for this case report has been given from the patient and his parents.

ACKNOWLEDGEMENTS

Thank you to Mr Robert Tranter for advice and support when writing this clinical report. Thank you to the patient and his parents for allowing this case report to be written.

FINANCIAL SUPPORT

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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Received on 21-11-2016 Accepted on 30-12-2016 Published on 09-01-2017

DOI: https://doi.org/10.12970/2311-1917.2017.05.01