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Case Report
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Mastoid Mucocele: An Uncommon Alert of Fibrous Dysplasia Onset: Case Report and Literature Review

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Abstract

The Problem: Fibrous dysplasia affects bone metabolism determining nonmalignant osseous change. Craniofacial skeleton is usually affected and temporal bone involvement is uncommon. Furthermore, when a concomitant rare mastoid mucocele is already affecting the same region, the related diagnosis can be even more challenging.

The Methodology: Here we report a radiological and surgical case, with CT, MRI and surgical imaging of a patient affected by a mastoid mucocele to be considered as a first sign of fibrous dysplasia onset.

Conclusions: Mastoid mucocele is a rare finding. To date, in medical literature only three cases are depicted. Etiology is still unknown even if multiple have been the hypothesis put forward. Although fibrous dysplasia temporal bone localization is well known, to our knowledge, this is the very first case in medical literature of a mastoid mucocele as an alert of fibrous dysplasia onset.

Keywords: Mucocele; Mastoid; Temporal bone; Head CT; MRI

Introduction

Mucocele is an expansile collection of sterile mucus which, lined by epithelium or covered by granulation tissue, could be frequently found in oral mucosa. Though only sporadically observable in the air bones cavities, among them paranasal sinuses are the most predominately involved areas while temporal bone is hardly affected. Pathology uncommon in adults and rare in childhood, mucocele is supposed to arise from the obstruction of the normal drainage of mucus in normal air cells [1]. As a matter of fact, mucosal thickening, bone overgrowth or fibrosis might lead to progressive inspissation of mucus and bone remodeling with local inflammation, thus determining the compression of the neighboring structures [2].

Clinical diagnosis, mostly relied on neuroradiological imaging (especially CT and MRI), can be challenging. Surgery is the gold standard for treatment whilst pathologic confirmation could be needed in atypical cases. Nowadays, medical literature provides for five cases of temporal bone mucoceles. Among them only three refer to the mastoid region [2-5].

Craniofacial localization of fibrous dysplasia, especially concerning paranasal sinuses and midface involvement, is well documented in medical literature as these regions are the most frequently affected. A recent article by Frisch has extensively presented which the clinical features of the fibrous dysplasia are and how temporal bone affection can be managed as the pathology is less common in this bone [6].

Also in consideration of the shortage of articles referring to temporal bone mucoceles available in medical literature, a case study of a mastoid mucocele to be considered an alert of fibrous dysplasia onset in a young patient treated with surgical drainage will follow.

Case Report

An 8-years-old boy, with medical history of right purulent otitis media occurred 3 years before, presented with subtle pain and swelling behind the right ear. For diagnostic reasons the patient underwent head CT and head MRI. Consequently a mastoidectomy was performed in order to drain the cyst. After the surgery the patient underwent a follow up head CT.

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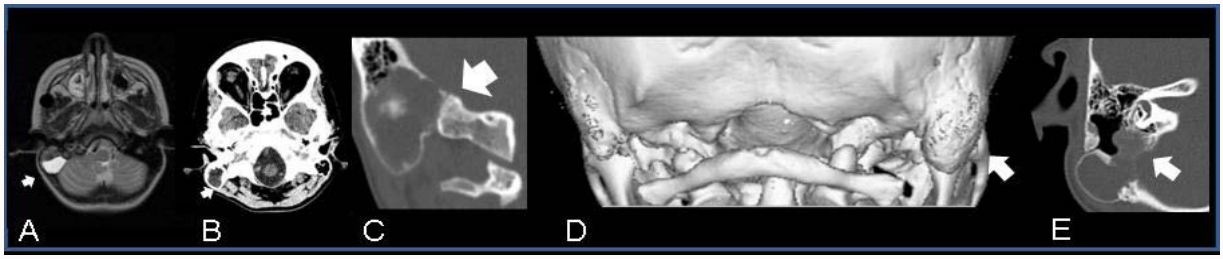

Figure 1: Neuroimaging of mastoid mucocele.
A) Brain MRI, axial T2 image showing a hyperintense mass in the right mastoid (arrow) dislocating the sigmoid sinus, consistent with a mastoid mucocele. B) Head CT axial image showing isodense tissue and loss of bone septa in the right mastoid (arrow). C) Head CT coronal image showing a loculated cavity (arrow) in the inferior right mastoid clearly divided by the healthy aerated mastoid. D) Head CT 3D reconstruction showing the enlargement of the right mastoid (arrow) compared to the contralateral one due to bone remodeling. E) Head CT coronal image showing a new cystic cavity (arrow) posterior to the middle ear.

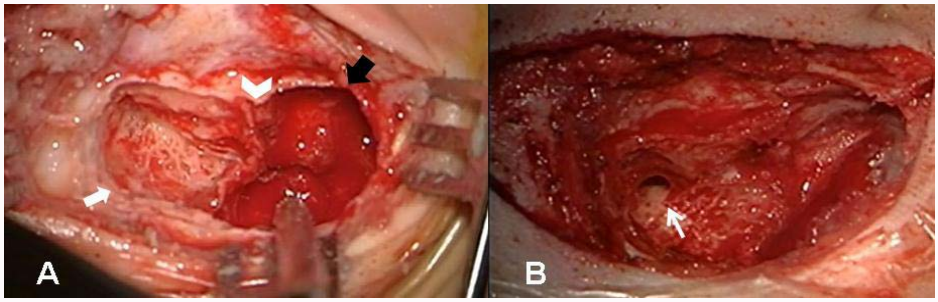

Figure 2: Surgical images during right mastoidectomy for mucocele.
A) The open right mastoid shows the healthy bone (white arrow), the cavity of the mucocele (black arrow) with a highly vascularized wall and a bone septum dividing the two regions (arrowhead). B) The surgical field after the exposure and drainage of the mucocele with decompression of the sigmoid sinus. The wide aditus ad antrum (small white arrow) provides a proper aeration of the cavity.

Results

After the clinical evaluation, the patient underwent head CT that showed a cyst-like lesion (Figure 1B) clearly divided from the healthy aerated mastoid cells by a bone septum (Figure 1C); the mastoid proved to be enlarged (Figure 1D). At MRI, the lesion appeared hyperintense on T2 (Figure 1A) and iso-hypointense on T1 and diffusion weighted imaging revealed an intense peripheral contrast enhancement (not shown in figure). The surgical inspection disclosed a mucus collection with a highly vascularized cyst wall (Figures 2A and B).

After surgery the patient underwent a follow up CT (Figure 1E), which show a growing cystic lesion, similar to the one that drove to mastoidectomy in a more anterior area, in the posterior part of the middle ear.

Discussion

Fibrous dysplasia is a rare idiopathic, progressive bone remodeling disorder. Etiology is still unknown. Craniofacial involvement is frequent, particularly with regards to membranous bones such as maxilla, mandible, frontal ethmoid, and sphenoidal. Temporal bone affection is reported to range from 25% to 70% of the cases [6]. The bone remodeling pattern can be monostotic, affecting a single cranial bone (70% of cases), or polyostotic thus involving multiple cranial bones (30% of cases) [7]. Typically, first symptoms debut during the first or the second decade of life, males are less frequently affected and clinical features depend on both the site and growth of the lesion. Prepubertal adolescents can show rampant bone remodeling slowing down in adulthood [8].

Whenever mastoid is involved, recurrent mastoiditis and conductive hearing loss can be symptomatic indicators.

To our knowledge, no case report in medical literature even suggests that fibrous dysplasia might appear concomitantly to a mastoid mucocele.

Only five cases of temporal bone mucoceles are covered by medical literature [2-5], the latter one reporting lesions found three times in the mastoid [2-5] and only once in the middle ear [5] or in the petrous apex [3].

According to the most recent literature, CT and MRI neuroimaging techniques are crucial for diagnosis and surgical planning [5]. In fact, neuroimaging findings provide with sufficient information to differentiate mucocele from other osteolytic lesions of the mastoid (e.g. cholesteatoma, cartilaginous tumors, schwannoma, meningioma, petrositis, glomus tumors, metastatic tumors, giant cell tumors, large air cell, cholesterol granuloma, histiocytosis and lymphoma) [3] while, at the same time, disclosing data about both the involvement of the neighboring structures and the best surgical approach to the lesion.

Although surgical intervention can be delayed in case of asymptomatic lesion, in light of the fact that mastoid mucocele, likewise paranasal sinuses mucocele, is expansile [5], drainage of the cyst must be performed whenever adjacent structures are involved [2,9]. For such a purpose, mastoidectomy should be considered a valid approach to drain the lesion and provide for an adequate aeration.

Conclusions

The etiology of temporal bone mucocele is still unknown although some authors reported history of chronic otitis media in patients affected by a temporal bone mucocele [4,5,10], noteworthy,

our patient suffered of right purulent otitis media 3 years before the exordium of the retroauricular swelling. The very same authors suggest that recurrent local inflammation is supposed to potentially determine mucosal or bone thickening affecting the ostium of a normal air cell thus causing a local mucus collection.

In our specific patient, bone remodeling typical of the fibrous dysplasia might have been started proximal to a mastoid cell ostium, therefore determining its occlusion causing the collection of mucus and such atypical bone remodeling.

Postsurgical neuroradiological CT examination showed typical fibrous dysplasia cystic bone remodeling. Tissue samples are currently under genetic evaluation to exclude McCune-Albright syndrome.

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