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Facial palsy and Valsalva-induced vertigo in a patient with temporal bone cystic fibrous dysplasia

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ABSTRACT

A 29-year-old woman, known with polyostotic fibrous dysplasia, presented with facial palsy, vertigo, and ipsilateral neck pain. Radiology revealed a large temporal bone cyst, extruding intracranially, eroding the labyrinth and Fallopian canal. Histopathology of the cyst wall retrieved through transmastoidal biopsy showed a mutation in the Gs-α gene, confirming cystic degeneration of fibrous dysplasia, making this a unique case in which both the inner ear and facial canal are affected by this condition.

1. Introduction

Fibrous dysplasia (FD) is a benign entity in which normal bone is replaced by immature bone and fibrous tissue, caused by mutations in the G_s-α gene [1–4]. It is not commonly seen in the temporal bone [1,4]. Patients with temporal bone FD often present with painless swelling or hearing loss, usually secondary to ear canal deformities or ossicular involvement [1,2]. Different types of FD are known, differentiated by extent of disease, such as monostotic, polyostotic and McCune-Albright syndrome, or by radiological presentation, such as pagetoid, sclerotic and cystic [2]. FD affecting the labyrinth or fallopian canal is rare, with only a few cases reported in literature of each [1,2]. We present the unique case of a patient with both facial palsy and labyrinthine fistula caused by polyostotic cystic temporal bone FD.

2. Case report

The patient is a 29-year-old woman with a history of polyostotic fibrous dysplasia and fibromyalgia, who presented to our clinic with right-sided neck pain, dizziness and right-sided facial weakness. The neck pain had been present for two weeks and was induced by an ordinary neck movement. One week later, the patient noticed dizziness after lifting her child. Ever since, she became progressively dizzy with every head movement, which initially led to nausea and vomiting. After her GP started with Betahistine a few days later, her vertigo subsided slightly. At that time, her GP also noticed that the right corner of her mouth was palsied.

On examination, both ears looked normal and apart from minimally uneven corners of her mouth the patient's face was symmetrical. She showed a spontaneous grade 1 nystagmus to her right and a positive right-sided Hennebert sign. When performing Valsalva and the Hennebert test, an upbeat counter-clockwise rotary nystagmus was seen (*see Video 1, which demonstrates nystagmus during Valsalva*). Furthermore, she was found to have hypoesthesia on the upper, middle and lower part of the right side of her face. All other cranial nerves were intact. Pure tone audiometry is shown in Fig. 1. CT and MRI imaging showed extensive ground glass deformities throughout the skull and a large lesion of the right petrous bone (*see* Figs. 2 & 3). The compilation of findings led to a presumptive diagnosis of an aneurysmal bone cyst, causing bony destruction of the labyrinth and fallopian canal with compression of the facial nerve.

Right cortical mastoidectomy revealed spongeous bone, of which a sample was sent for histopathology (*see* Fig. 4). A cystic lesion was encountered, which encroached the lateral semi-circular canal, the facial nerve and posterior dura. Incision caused clear yellowish fluid to leak out. A biopsy was taken from the wall of the cyst. The facial nerve was identified in the middle ear and followed towards the mastoid, where it was not possible to divide the cyst wall from the facial nerve. Part of the cyst wall was removed, resulting in an opening in the cyst of 20 × 8 mm. The dehiscent part of the posterior semi-circular canal could

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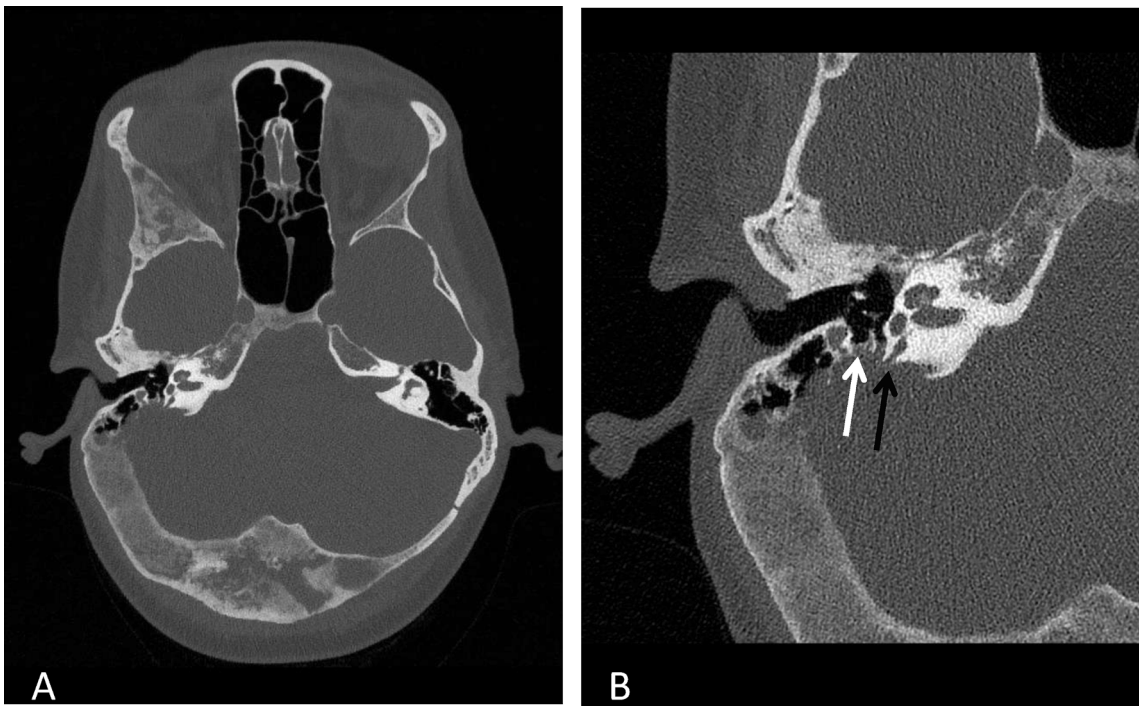

Fig. 1. A) Transversal CT image. A mostly right-sided thickening of the skull is seen with ground glass aspect of the sphenoid wing, petrosal apex and occipital bone. Some cystic areas can be identified. A classic aspect of fibrous dysplasia. **B)** Detailed image of the right petrosal bone. Posterior osseous erosions are seen, direcly adjacent to the ground glass appereance of the occipital bone. The white arrow shows erosion of the facial canal and the black arrow erosion of the posterior semicircular canal.

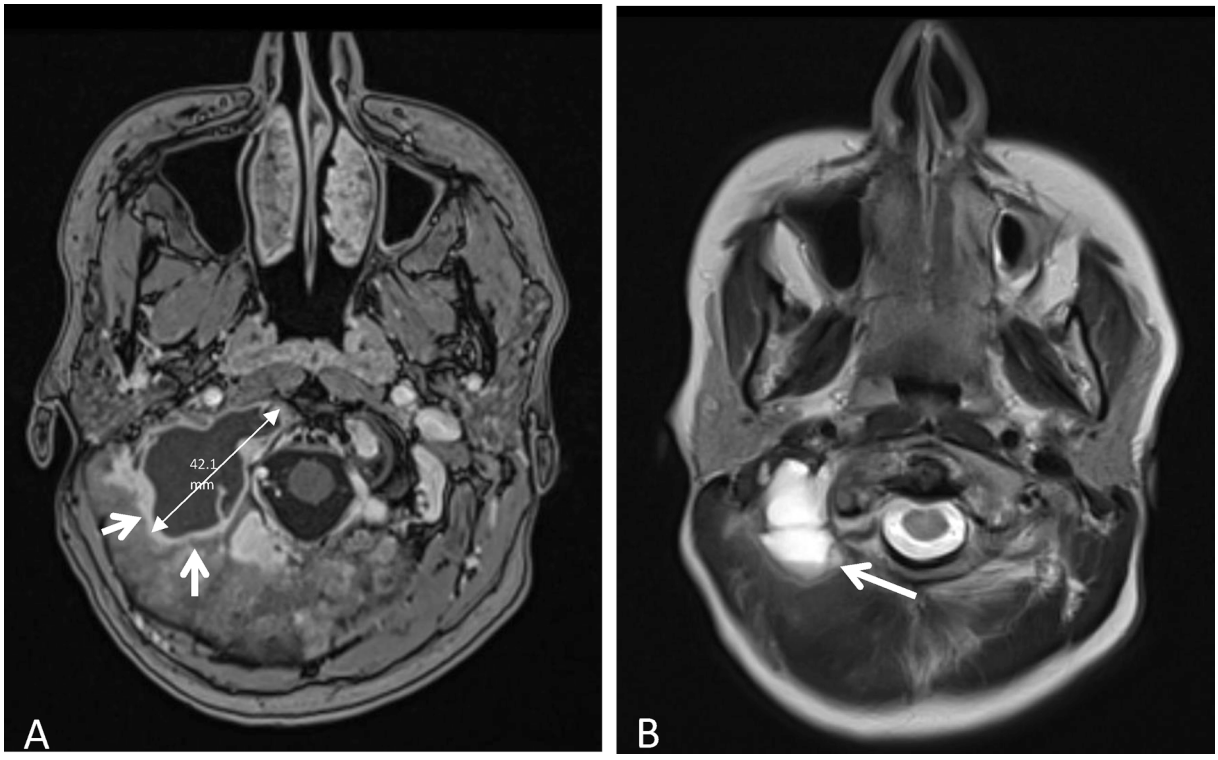

Fig. 2. A) Axial T1-weighted image post gadolinium. Expansion and heterogenous signal intensity of the occipital bone is shown, congruent with fibrous dysplasia. A cystic lesion is seen at the dorsal side of the right os petrosum with enhancement of the cyst wall (white arrows). **B)** Axial T2-weighted image shows the multi-loculated cystic lesion. The white arrow indicates a fluid–fluid level inside the cyst.

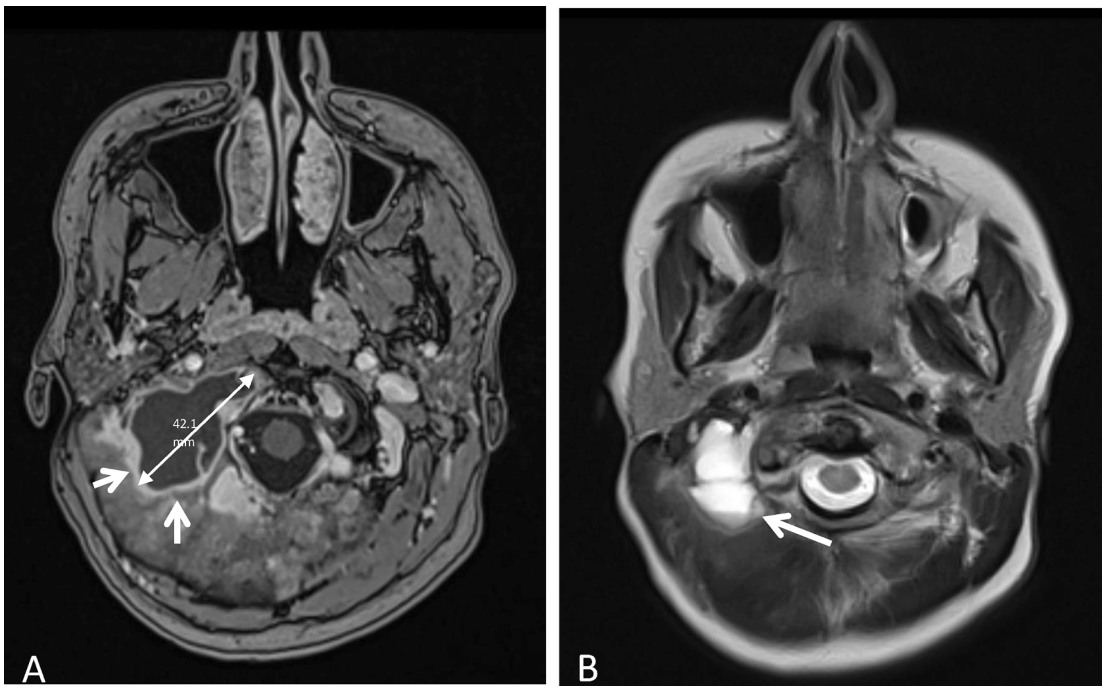

Fig. 3. A) Axial T1-weighted image post gadolinium. Expansion and heterogenous signal intensity of the occipital bone is shown, congruent with fibrous dysplasia. A cystic lesion is seen at the dorsal side of the right os petrosum with enhancement of the cyst wall (white arrows). **B)** Axial T2-weighted image shows the multi-loculated cystic lesion. The white arrow indicates a fluid–fluid level inside the cyst.

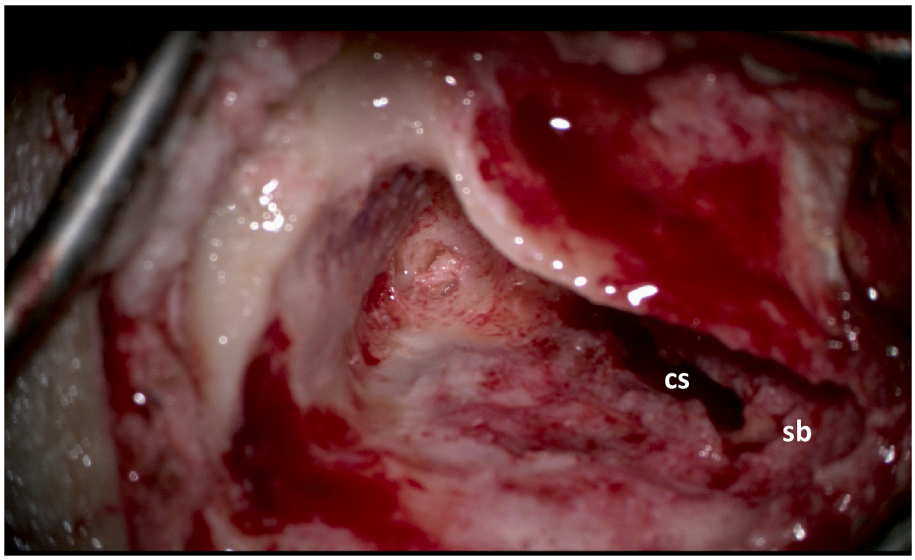

Fig. 4. Intraoperative photo of right mastoidectomy, illustrating pathological spongeous bone (sb) and the retrofacial infralabyrinthine space where the cyst was marsupialised (cs).

not be identified because of the cover by the cyst wall. Because it was deemed to hazardous for the inner ear to remove this last layer, it was decided perioperatively to apply bone dust and fibrin sealant on the expected site of dehiscence. This was further covered with temporal fascia.

Post-operatively, the patient's facial palsy subsided within a few days, she did not suffer from any more vertigo spells and her hearing was subjectively symmetrical. The pathological specimens of the mastoid bone and cyst wall sample both showed signs of discontinued trabeculae, surrounded by abundant fibrous tissue, in a typical pattern of FD, as described by Hartley et al., [5]. Genetic analysis followed, which found a mutation on exon 8 of G_s-α gene, confirming the diagnosis. Pure-tone audiometry several months post-operatively showed similar thresholds to her pre-operative examination. Post-operative CT is shown in Fig. 5.

3. Discussion

FD is a benign skeletal disorder with an aetiology that is not yet fully understood [2,3]. It forms around 7% of all benign osseous tumors and when found in the skull it affects the temporal bone in approximately 24% [2]. Usually, it is diagnosed in patients in their first or second decade [2,3]. Three types of FD can be distinguished, namely monostotic (±70%), polyostotic (±30%) and McCune-Albright syndrome (±3%)

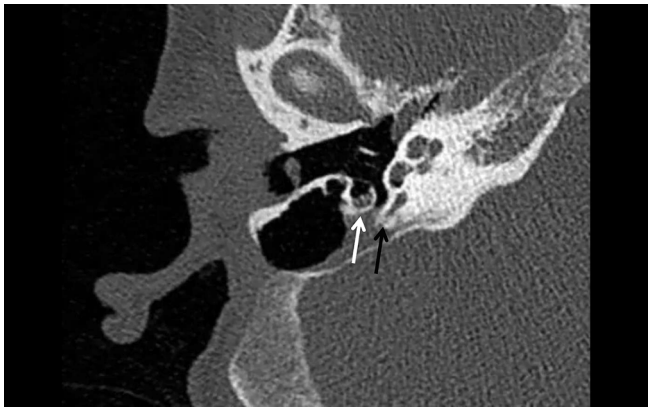

Fig. 5. Postoperative detailed image of the right petrosal bone showing the absence of the previously seen cyst. The white and black arrows indicate the re-ossification of the Fallopian and posterior semi-circular canal, respectively, after their coverage with bone dust, fibrin sealant and fascia.

[2,3]. When FD is found in the head and neck area, it is usually the monostotic type [2,3]. Monostotic and polyostotic FD are differentiated by severity, with the latter having an earlier onset, with more extensive skeletal and craniofacial involvement [3]. McCune-Albright syndrome, the most severe form of FD, is associated with endocrine abnormalities, pigmented cutaneous lesions and pre-mature closure of the epiphyses [3].

Most commonly, patients with FD of the temporal bone present with progressive hearing loss (80%), which is usually conductive, resulting from external auditory canal obliteration or ossicular chain erosion. Sensorineural hearing loss, however, has also been reported [2]. Recurrent external otitis, cholesteatoma, labyrinthitis, facial palsy and labyrinthine fistulas are other temporal bone FD manifestations [1,2,6]. The upbeat, anti-clockwise nystagmus of the current case during Valsalva is a textbook example of right-sided posterior semi-circular canal stimulation. To the authors' knowledge, no patient has yet been described who presented with both facial palsy and a labyrinthine fistula as a result of cystic FD.

Radiologically, a "ground-glass appearance" is typical for FD, along with sclerosis, erosion and cortical thinning [1]. FD on CT imaging can be further differentiated into pagetoid (56%), sclerotic (23%) and cystic (23%) types [2]. Of other cases reported with facial nerve or semi-circular canal involvement, most patients showed cystic FD, too [1,2,6]. In cystic FD, fluid–fluid levels may be seen within cysts, as was the case in the current patient [4]. Fluid-fluid levels become apparent on CT or MRI when substances of different density are allowed to settle within a cyst and when the imaging plane is perpendicular to the fluid level [4]. This finding can aid in the radiological diagnosis of the cyst, as it is seen more frequently in bone as opposed to soft tissue lesions [4]. Aneurysmal bone cysts and telangiectatic osteosarcoma are the most common lesions containing a fluid–fluid level [4]. Because of its association with FD and because of the found fluid–fluid level, an aneurysmal bone cyst was our working diagnosis after imaging. However, the gold standard in differentiating between these diagnoses remain CT findings supported by histopathology [1,2], and after the biopsy results from surgery, which did not show macroscopic or histological signs of an aneurysmal bone cyst, this diagnosis was discarded. Genetic analysis of tissue samples showed presence of a mutated G_s-α gene, which alters activity of the G_s protein signal transduction pathway and is a suggested etiologic factor of FD [3].

4. Conclusion

This case shows the extent of erosion that cystic FD can cause in the middle and inner ear. It is a unique presentation of facial palsy and vertigo, induced by Valsalva and the Hennebert test, owing to the destruction of labyrinth and Fallopian canal. In a patient with these symptoms and FD, cystic degeneration should be considered. Diagnosis should involve radiology and a biopsy of the cyst wall, and treatment should consist of removal of as much of the cyst as possible.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.inat.2022.101622.

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