wjpmr, 2020,6(2), 01-03

SJIF Impact Factor: 5.922

WORLD JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.wjpmr.com

Case Report

ISSN 2455-3301

WJPMR

RARE CASE OF GOODHILL SYNDROME

Dr. A. Mennouni*, H. Ngham, H. Ardhaoui, S. Rouadi, R. Abada, M. Roubal and M. Mahtar

Azemmour Casablanca.

***Corresponding Author: Dr. A. Mennouni**

Azemmour Casablanca.

Article Received on 25/11/2019 **Article Revised on 15/12/2019** **Article Accepted on 05/01/2020**

ABSTRACT

The association of otosclerosis with a Malleus ankylosis called goodhill syndrome is very rare, accounting for less than 1% of congenital malformations of the middle ear.[1] We report the case of a 46-year-old woman with no family history of deafness who had been consulting for bilateral deafness, evolving for 10 years, with no associated signs such as vertigo or tinnitus. Otoscopic examination show normal eardrums, The audiogram includes a conductive hearing loss of 60 dB d. Computed tomography is the main examination for the visualization of the ossicular chain and the search for other anomalies. The diagnosis is confirmed during the surgery. We have reported a case of bilateral Malleus ankylosis associated with otosclerosis. This makes our study interesting because it is rarely described in the literature. The objective of this study was to analyze the findings of middle ear exploration and the frequency of ossicular in patients with suspected otosclerosis. bone bridge (Figure 1). The surgery consists of an ossiculoplasty, by section of Malleus's neck with interposition with Incus. We obtained a good result by gain of 20 dB in the main frequency. Our patient is satisfied. The other side will be programmed a year later.

KEYWORDS: Goodwill syndrome, bilateral Ankylosis; otosclerosis, syndrome of the house; The otosclerosis; Malleus interposition; incus.

INTRODUCTION

Conductive hearing loss is a common cause of ENT consultation. Otosclerosis dominates etiologies by frequency, and house syndrome is a very rare cause.

The goodwill syndrome discovered during an intervention on a suspicious ear which causes a real problem for the surgeon.

The hypothesis of congenital etiopathogeny is the most probable. Tomodensitometry (CT), thanks to its Multi planar reconstructions, offers a good analysis of the ear and thus allows to exclude or confirm other differential diagnoses.

OBSERVATION

A 46-year-old patient with no history of the disease consults for unilateral right-sided deafness that has been evolving for 10 years, associated with bilateral tinnitus gradually worsening without recurrent ear infections or trauma.

On examination, the eardrums are normal. external ears normally formed (external auditory canal pavilion).

Audiometry shows right unilateral conductive hearing loss with a loss of 60 db and a rinne of 40 db.

The volume CT shows abnormal ossification of the anterior ligament of the right hammer producing a bone bridge connecting the head of the hammer to the anterior wall of the attic associated with bilateral otospongiosis foci, with flattening on the left and focus on the right. figure 1.

The patient is operated on, and the diagnosis was confirmed intraoperatively by showing attical bone synostosis with a fixed calliper.

The gesture took place in two stages; first stage consists of performing a transposition of the anvil with hammer neck section, the second stage consists of the realization of a platinotomy with interposition of a Teflon piston.

Hearing acuity improved, which was confirmed by audiometry with a 20 db control.

The patient is followed regularly to detect a recurrence. Our patient is satisfied. The other side will be programmed a year later.

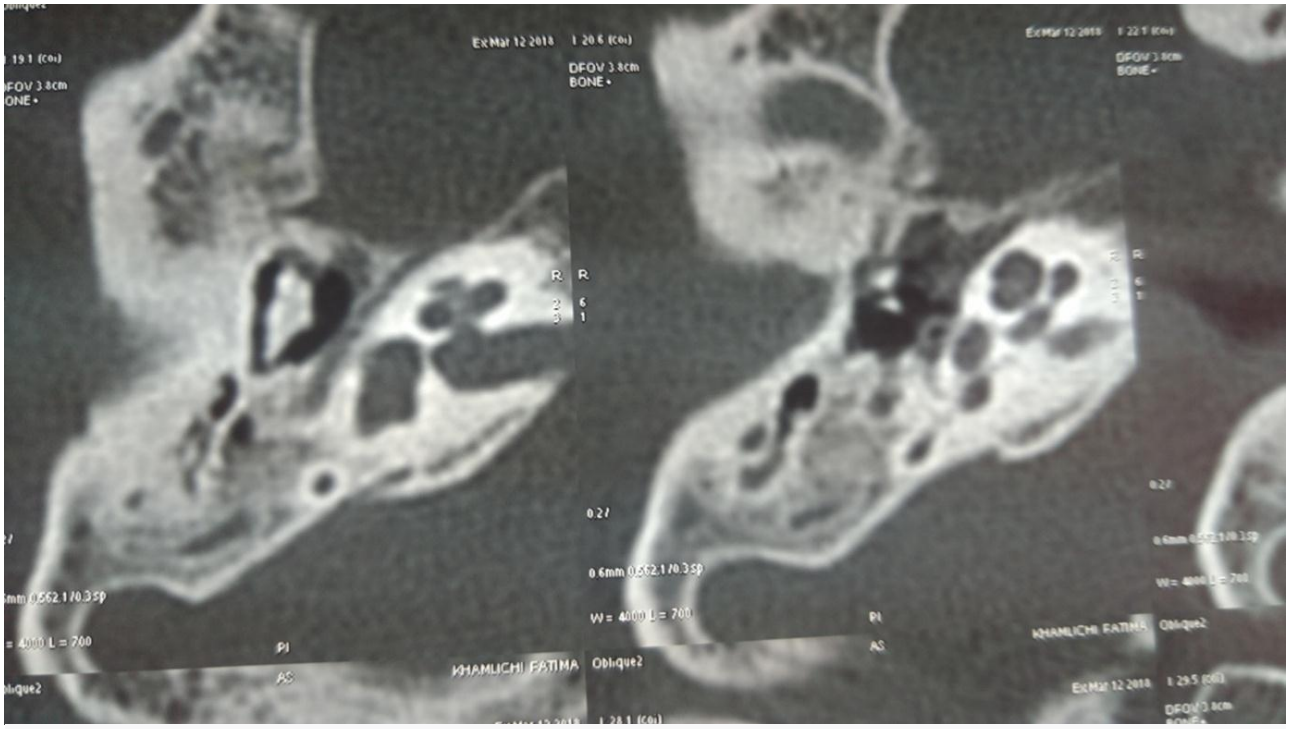

Figure 1: CT examination of a right bone in a bone window showing a bone bridge connecting the head of the hammer to the anterior wall of the attic, associated with a platinum thickening.

DISCUSSION

Otosclerosis is a primary osteodystrophy of the otic capsule, specific to the human species, responsible, when it manifests itself clinically, for conductive hearing loss.

Congenital etiopathogeny seems the most likely and is based on several findings healthy ear which a normal pneumatization and in the absence of inflammatory changes or otological history.

The frequency of associations with other congenital anomalies and the existence of bilateral forms is even rare.

The association of otosclerosis with fixed hammer head syndrome is an entity first described by Goodhill in 1966, and corresponds to all clinico-radiological manifestations resulting from ossification of anterior ligaments or hammer making a bone bridge connecting it to the roof of the attic. This synostosis can be uni- or bilateral. Its frequency is rarely estimated at 1% of transmission deafness.

This association further aggravates the conductive hearing loss and makes the surgery more complex with less satisfactory postoperative results.

The first-line imaging is CT scan, it is the key examination, that can detect a fixation of the hammer head to the attic wall, and signs of otosclerosis associated. CT can also be used to study the other elements of the ossicular chain, the path of the facial nerve and the detection of certain abnormalities of the inner ear that can modify the surgical conduct.

Volume CT, thanks to its high spatial resolution and the quality of its reconstructions, makes it possible to show the foci of otosclerosis in the form of limited circumscribed hypodensities at one or more points of the otic capsule whose seat of predilection is fissula antéfenestrum.

The classification proposed by Charachon et al. takes into account the description of the ossicular anomalies and their embryological origin:

Type 0: the ossicular chain is normal but with an atresia plate: the chain is fixed by adhesion between the atresia plate and the hammer handle.

Type I: ankylosis of the ossicles at the attic level, malformation of the hammer head and / or anvil body, incudo-malleal block. The facial path is normal.

Type II: The chain is continuous but fixed at the descending leg of the anvil or at the caliper. The most common is isolated stapedobuccal ankylosis. The facial path can be changed.

Type III: discontinuity and breakage of the chain at the level of the long anvil apophysis or caliper. The facial path is often abnormal.

Type IV: complex malformation.

The main differential diagnosis with conductive hearing loss with normal eardrum are :

L otospongiosis: is characterized by limited circumscribed hypodensity at one or more points of the otic capsule. The preferential localisation is the fissula antefenestram, the most constant embryo fibro-cartilaginous residue located on the anterior bank of the oval window.

Minor malformations of the chain: the diagnosis of minor aplasia of the chain can be discussed with the presence of a fix non evolutionary congenital hearing loss. The notion of family history, lack of otitis history, flag malformation, congenital stenosis of the external auditory canal, congenital facial palsy also point to this diagnosis.

Traumatic lesions of the anterior ossicular chain of the anvil and the incudo-stapedial and incudomalleal joints.

The treatment of Goodhill's syndrome is based on the restoration of normal ossicular mobility. It consists of making a classic anvil transposition with a section of the hammer's neck; or, more simply, if the fixation is isolated, to directly suppress attical synostosis while respecting ossicular continuity.

CONCLUSION

Goodhill syndrome is a rare and poorly understood cause of normal tympanic transmission deafness.

The co existence with other abnormalities such as otosclerosis is possible. CT remains the exam of choice.

REFERENCES BIBLIOGRAPHIQUES

1. FeeG A. ArchivesofOtolaryngology 88, 477 Freeman P & Edmonds C., 1968.
2. Archives of Otolaryngology 95, 556 V.Goodhill, 1972.
3. M. Lahkim, F.Z. Laamrani, H. En-Nouali, H. Boumdin, T. Amil. Goodhill syndrome: a rare cause of transmission deafness. La Lettre d'ORL et de chirurgie cervico-faciale, 2015; 15(9): 342.
4. Seidman MD, Babu S. A new approach for malleus/incus fixation: no prosthesis necessary. Otol Neurotol, 2004; 25(5): 669-73.
5. William H. Lippy, MD; Arnold G. Schuring, MD; Moshe Ziv, MD. Stapedectomy for Otosclerosis with Malleus fixation. Arch otolaryngol_vol, july, 1978.
6. Bruzzo M, Braccini F, Cacès F, Vallicioni JM, Chays A, Magnan J. Le syndrome de la tête du marteau fixée. Ann O_tolaryngol Chir Cervicofac, 1998; 115(5): 279-83.
7. Teunissen EB, Cremers WR Classification of middle ear anomalies. Report on 144 ears. Ann Otol Rhinol Laryngol, 1993; 102: 606-12. 2. Goodhill V. Pseudo-Otosclerosis. Laryngoscope, 1960; 70: 722-57.
8. Swartz JD, Faerber EN Congenital malformations of the external and middle ear: high-resolution CT findings of surgical import. AJR Am J Roentgenol, 1985; 144: 501-6.
9. Cremers CW, Teunissen E The impact of a syndromal diagnosis on surgery for congenital minor ear anomalies. Int J Pediatr Otorhinolaryngol, 1991; 22: 59-74.
10. Esteves SD, Silva AP, Coutinho MB, Abrunhosa JM, Almeida e Sousa C Congenital defects of the middle ear--uncommon cause of pediatric hearing loss. Braz J Otorhinolaryngol, 2014; 80: 251-6.
11. House HP Early and late complications of stapes surgery. Arch Otolaryngol, 1963; 78: 606-13.
12. De Souza C, Goycoolea MV, Sperling NM Otosclerosis: diagnosis, evaluation, pathology, surgical techniques, and outcomes. Plural Publishing, USA, 2014.
13. Bales P Scott-Brown's Otolaryngology (5th Edn) Published by London Boston: Butterworths, London, 1997.
14. Baily BJ Head and Neck Surgery: Otolaryngology (3rd Edn) Lippincott Williams & Wilkins Publisher, USA, 2001.
15. Flint PW, Haughey BH, Lund VJ, Niparko JK, Richardson MA, et al. Cummings Otolaryngology - Head and Neck Surgery (5th Edn) Philadelphia: Mosby Elsevier, USA, 2010.