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CASE REPORT

Pyramidal Bony Bar Fixating the Lenticular Process: An Unusual Congenital Middle Ear Anomaly

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We report the case of a male child with a bilateral pyramidal bony bar connecting and fixating the lenticular process of the incus to the bony posterior wall of the retrotympanum, thus obstructing ossicular chain movement and causing a bilateral conductive hearing loss. Removal of the pyramidal bony bar was performed while sparing the stapedial muscle's tendon. The ossicular chain regained its mobility and the round window membrane reflex could be elicited. The air-bone gap pure-tone average of 0.5, 1 and 2 kHz improved from 45 dB to 9.7 dB at 2 weeks and 6.7 dB at 4 months after surgery. The difference between the ossified stapedial muscle's tendon, the elongated pyramidal eminence and our case will be discussed.

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Introduction

Congenital anomalies of the ossicular chain are rare and may present in various ways. Because they generally occur behind an intact tympanic membrane, computed tomography can be very useful in determining the type of deformity preoperatively. By means of a case report we will demonstrate the subtle differences in ossicular chain anomalies and how this specific case of ossicular chain anomaly was treated surgically.

Case Report

We report the case of a bilateral pyramidal bony bar connecting and fixating the lenticular process of the incus, obstructing ossicular chain movement and causing a bilateral conductive hearing loss.

A 5-year-old boy presented at the out-patient clinic with hearing loss and delayed speech and language development. No problems had occurred during pregnancy, but the patient was admitted to the neonatal intensive care unit for one night due to mild respiratory distress during the C-section. No abnormalities were detected by the paediatrician in the neonatal period. The universal new-born hearing screening was normal. The patient did not experience recurrent upper respiratory infections. There was no history of trauma. Family history for hearing loss was negative. It was unclear to the parents at which age the hearing loss had occurred, nor could they confirm a rapid or progressive onset. He had never experienced vertigo before.

During otoscopy a normal-appearing tympanic membrane was found bilaterally. Liminal audiometry revealed a bilateral conductive hearing loss with an air-bone gap (ABG) of 45 dB on the right side and 38.3 dB on the left side (pure-tone average of 0.5, 1 and 2 kHz).

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Tympanometry demonstrated a type C curve on the right side (-90 daPa) and a type A tympanogram on the left side (-50 daPa).

Several conditions were considered in the differential diagnosis including tympanosclerosis, otosclerosis, congenital ossicular chain fixation, absence of the oval or round window and a persistent stapedial artery.

A temporal bone CT scan was performed and revealed a pyramidal bony bar connecting the lenticular process of the incus to the bony posterior wall of the retrotympanum (Figure 1). This abnormal structure seemed to be in continuity or in close proximity with the pyramidal eminence and the tendon of the stapedial muscle's tendon. Additionally, the radiologists reported a high-riding bulb on the right side. No other causes of conductive hearing loss of middle or inner ear origin were found. A MRI scan was performed to evaluate the posterior fossa and cerebellopontine angle, but this examination did not reveal any abnormalities.

We decided to perform an exploratory tympanotomy on the right side (the worst hearing ear) to confirm the diagnosis suggested by imaging and to improve the hearing if possible. A transmeatal approach was used to explore the middle ear. Palpation of the ossicular chain revealed a mobile malleus, but a fixation of the incus. The stapes superstructure was not visible. No round window membrane reflex could be elicited. Just lateral and in conjunction with the lateral face of the pyramidal eminence, a bony bar originated at the bony wall of the retrotympanum and connected with the lenticular process of the incus, thus causing fixation.

The pyramidal bony bar was removed using a microdrill and curette, while taking care to avoid injury to the chorda tympani. The osseous prominence was fixed to the lenticular process while leaving the stapes superstructure mobile and the long process of the incus. The stapedial muscle's tendon was intact and not ossified. The stapes superstructure and footplate were normal. The surgeon's drawing and photographs taken during surgery can be found in Figures 2 and 3.

Afterwards, the ossicular chain regained its mobility and the round window reflex could be elicited by palpating the malleus handle.

Figure 1. High-resolution CT images of the right temporal bone: a case of an elongated pyramidal eminence. Axial image. (1) Lenticular process; (2) stapes capitulum; (3) pyramidal bony bar or elongated pyramidal eminence.

The ABG (pure-tone average of 0.5, 1 and 2 kHz) improved from 45 dB preoperatively to 9.7 dB at 2 weeks and 6.7 dB at 4 months after surgery.

Discussion

To categorize this disorder according to Cremers' classification, we would suggest a class 3 congenital anomaly of the ossicular chain with a mobile stapes footplate, caused by a pyramidal bony bar (without ossification of the stapedial muscle's tendon) and consequent lenticular process fixation. [1]

In our opinion, the pathology presented differs from the elongated pyramidal eminence (which includes a prolonged circumferential ossification) and the ossified stapedial muscle's tendon (which affects the tendon itself), although it might share a common pathophysiological pathway with the former. Ossification of the tendon is probably caused by interhyale failing to differentiate into fibrous tissue, while elongation of the pyramidal eminence and the pyramidal bony bar are probably caused by prolonged condensation of mesenchymal tissue around the belly of the stapedial muscle. This hypothesis is based upon the work of Rodriguez-Vazquez (personal communication). [2]

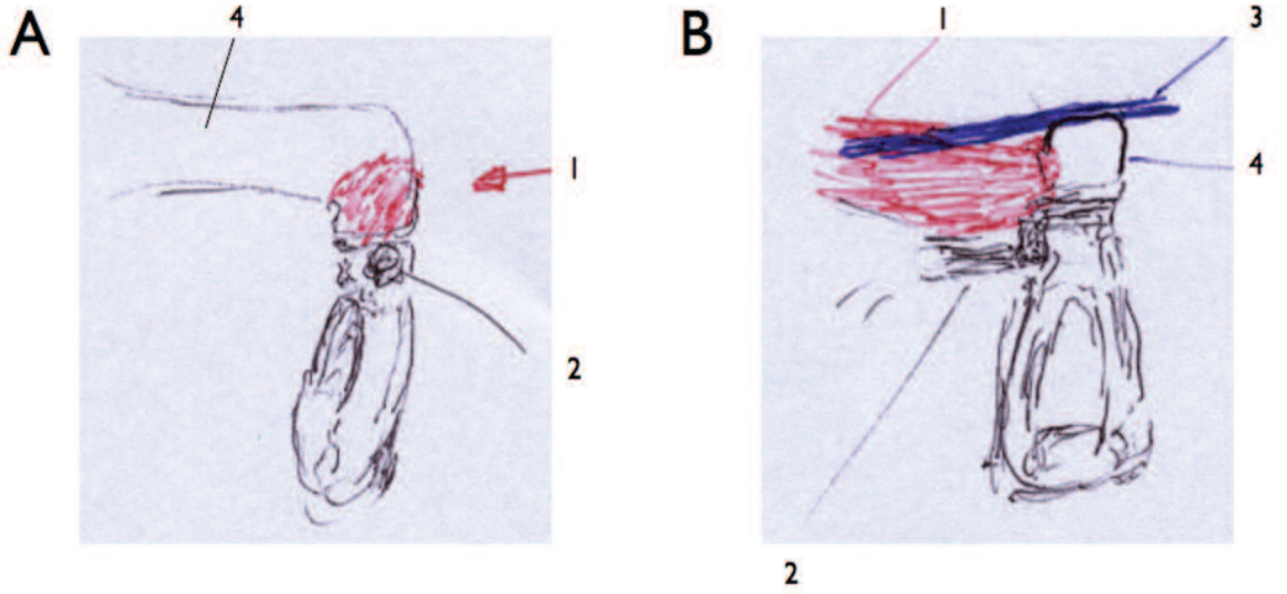

Figure 2. Surgical drawing of the incus-stapes interface. (1) Pyramidal bony bar; (2) stapedial muscle's tendon; (3) chorda tympani; (4) long process of the incus.
A. The incus-stapes interface from the pyramidal eminence's perspective.
B. The incus-stapes interface from a different point of view to demonstrate the distinction between the stapedial muscle's tendon and the pyramidal bony bar.

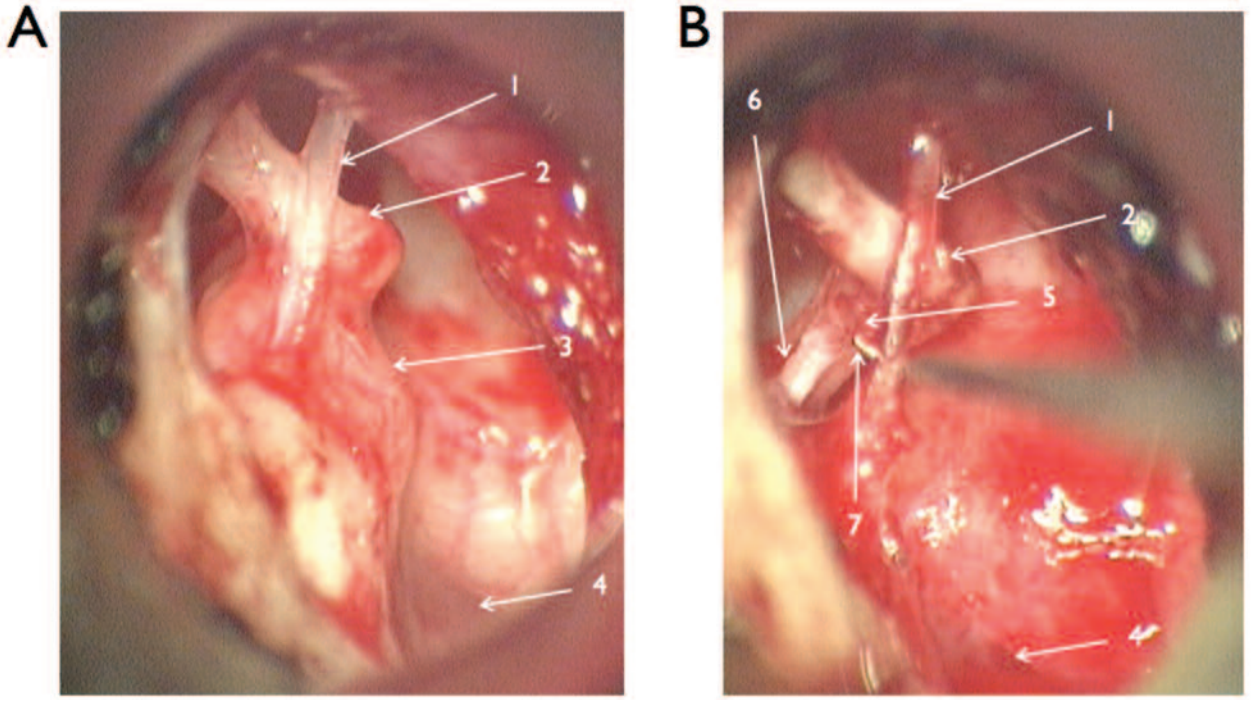

Figure 3. Intraoperative findings. Transmeatal view during exploratory tympanotomy. (1) Chorda tympani; (2) long process of the incus; (3) pyramidal bony bar; (4) round window niche (5) stapes superstructure (6) stapedial muscle's tendon.
A. Intraoperative view before removal of the pyramidal bony bar.
B. Intraoperative view after removal of the pyramidal bony bar. The instrument (7) is pointed at the insertion of the stapedial muscle's tendon in the stapes superstructure. Note that the tendon is intact.

Nandapalan and Tos have reported on isolated congenital stapes superstructure fixation including a pathology called *stapes - pyramidal bony bar with normal stapedial muscle's tendon*, categorized as a class 2 congenital middle ear anomaly according to Cremers (i.e. a stapes ankylosis associated with other congenital ossicular chain anomalies).[3] We think the case presented in our report has similarities to the stapes – pyramidal bony bar, but instead of fixating the stapes superstructure, it fixates the lenticular process (i.e. a *lenticular process – pyramidal bony bar with normal stapedial muscle's tendon*). Because it is the lenticular process which is fixated by the bony bar, we think it should be defined as a class 3 anomaly since the stapes footplate and superstructure *per se* are mobile.

Preoperative temporal bone CT scan can distinguish between ossification of the stapedial muscle's tendon and osseous malformations of the pyramidal eminence. In case of an elongated pyramidal eminence, the triangular conus of the pyramidal eminence is touching the incudostapedial joint; in case of an ossified stapedial muscle's tendon, the pyramidal eminence is in the correct position while a linear calcified structure (the ossified stapedial muscle's tendon) can be observed (example in Figure 4). However, preoperative CT scans do not allow differentiation between the elongated pyramidal eminence and the pyramidal bony bar (touching the stapes superstructure or lenticular process).

Until now, nineteen cases of stapedial muscle's tendon ossification were reported in literature, seven cases have been reported on elongation of the pyramidal eminence and six cases have been reported on a stapes - pyramidal bony bar without ossification of the stapedial muscle's tendon.[4-19] In our opinion, this is the first report of a pyramidal bony bar fixating the lenticular process (instead of the stapes superstructure).

Conclusion

In this congenital malformation case a pyramidal bony bar is connecting the lenticular process of the incus to the bony posterior wall of the retrotympanum. By obstructing ossicular chain movement, it caused a conductive hearing loss in early childhood.

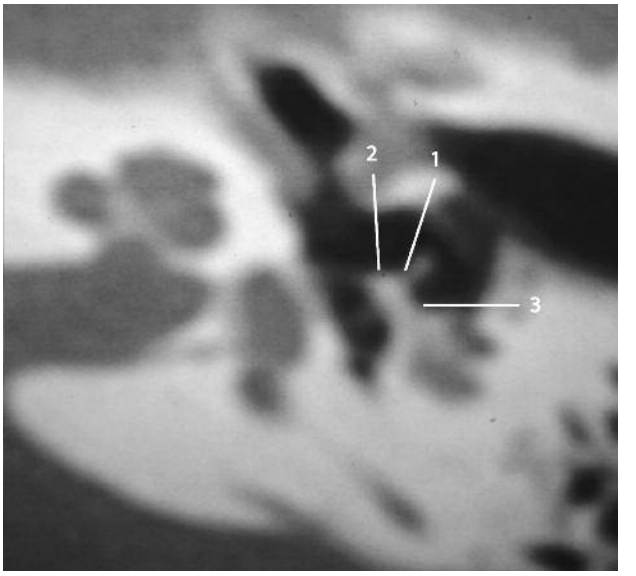

Figure 4. High-resolution CT images of the left temporal bone in another patient: a case of an ossified stapedial muscle's tendon. Axial image. (1) Lenticular process; (2) stapes capitulum; (3) ossified stapedial muscle's tendon.

Notwithstanding the dramatic improvement of the resolution of CT scans, the final diagnosis is made my middle ear inspection. Removal of the pyramidal bony bar can restore ossicular chain mobility resulting in an excellent hearing outcome.

Conflict of interest: None.

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