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Case report - Olgu sunumu

Two unusual case of congenital conductive hearing loss related to anomaly of stapes

Konjenital stapes anomalisine bağlı iletim tipi işitme kayıplı nadir iki olgunun sunumu

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Abstract

Congenital abnormalities may occur in an isolated form of congenital ossicular or combined with external ear. Isolated ossicular anomalies are rarely seen in clinical practice only, a few reports include series of patients and vast majority of publications are consisted of case reports. Treatment of congenital ossicular anomalies depends on the condition of ossicular and oval window anomalies. In presented article two case reports with severe oval window anomalies and absence of suprastructure of stapes with mobile footplate were published. Management of these cases was discussed with the review of the literature.

Key words: Congenital conductive hearing loss; congenital absence of the oval window; absence of the stapes.

Özet

Konjenital anomaliler izole kemikçik anomalisi halinde veya dış kulakla kombine anomali olarak görülebilir. İzole kemikçik anomalisi klinik pratikte çok ender olarak rastlanır. Bu anomalilerle ilgili literatürün çoğunluğunu vaka takdimleri oluşturmaktadır ve vaka serisi halindeki bildiriler oldukça sınırlıdır. Konjenital ossiküler anomalinin tedavisi kemikçiklerdeki ve oval penceredeki anomalinin varlığına ve biçimine bağlı olarak değişiklik göstermektedir. Bu makalede ileri derecede oval pencere anomalisi ve hareketli tabanla birlikte stapes suprastriktürünün yokluğu ile karakterize iki ender olgunu ve tedavisi literatür taraması ışığında sunulmuştur.

Anahtar sözcükler: Konjenital iletim tipi işitme kaybı; oval pencerenin konjenital yokluğu; stapesin yokluğu.

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Introduction

Congenital ossicular anomalies could either be isolated ossicular anomalies or they could be combined with other ear anomalies. The external ear and eardrum are found to be normal in isolated ossicular anomalies. Diagnosis of isolated ossicular anomalies is not readily apparent and otoscopy is normal. The associated conductive hearing loss may be mild or severe. A detailed assessment of patients with high resolution computed tomography (HRCT) usually provides the feature of the anomaly [1]. The definitive diagnosis can be made by exploratory tympanotomy. Teunissen and Cremers [2] classified congenital ossicular anomalies into four groups; isolated stapes ankylosis, stapes ankylosis with other ossicular anomalies, ossicular anomalies with mobile stapes footplate and aplasia or dysplasia of oval or round window (Table 1). The most common

isolated ossicular anomalies are stapes ankylosis and inkudostapedial discontinuity [3]. We report two cases with unusual congenital conductive hearing losses and management of these conditions are discussed by review of the literature.

Table 1: Cremers' classificaiton of congenital anomalies of middle ear

Class 1	Ears with congenital isolated stapes ankylosis
Class 2	Ears with stapes ankylosis in combination with a congenital anomaly of the ossicular chain
Class 3	Ears with congenital anomaly of the ossicular chain and at least a mobile stapes footplate
Class 4	Ears with aplasia or severe dysplasia of the oval window or round window

Cases

Case 1

Twenty-nine-year-old male patient admitted to our hospital with complaint of hearing loss. His medical history revealed no head trauma or ear infection. He had realized his complaint 15 years ago. After that time he had not had any medication or treatment. The external ear and eardrum on both sides were normal. The audiogram showed 47 dB conductive type hearing loss on his left side. There was 40 dB air-bone gap. Hearing level on his right side was normal (Figure 1). There was no acoustic reflex on the left side and AD type tympanometric curve was obtained. On the right side the tmypanometric assessment was normal. There was no obvious abnormal finding in the temporal bone computed tomography (HRCT) which was obtained in 1 mm sliced on axial and coronal plane (Figure 2). We decided to perform an explorative tympanotomy ossicular dislocation or otosclerosis.

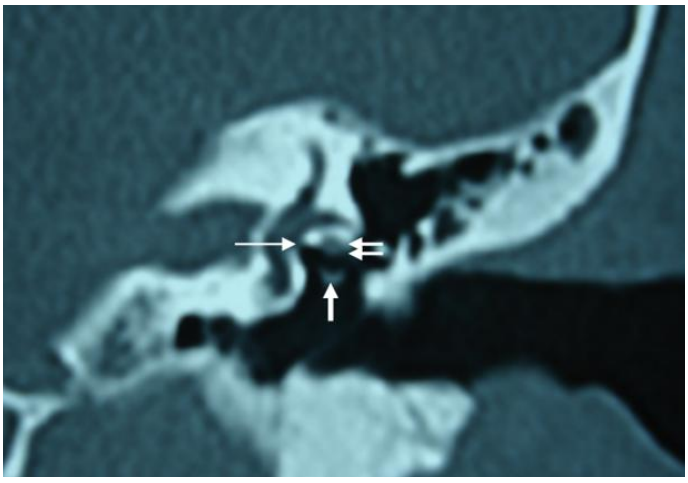

Figure 2: Severe anomaly of the oval window in coronal section of right temporal bone computed tomography of the case 1. Long white arrow indicates hypoplasic oval window; double white arrow indicates facial nerve; short white arrow indicates suprastructure of stapes.

Explorative tympanotomy was performed via transmeatal approach under general anesthesia. Posteriosuperior bony annulus was drilled with a diamond burr to expose incudostapedial joint. The suprastructure of the stapes was hypermobile. Both cruses of the stapes were deficent. There was a dimplike depression on oval window and it was

positioned medial to tympanic wall. The oval window was retracted and there was no real footplate and annular ligament. Then we realized that there was a severe hypoplasia of the oval window. The facial canal was normal. A vestibulotomy was performed by a pick and a teflon prothesis was emplaced. There was no complication during the operation. Postoperative hearing gain was 22 dB and air conducting hearing level was 15 dB on the left side (Figure 1).

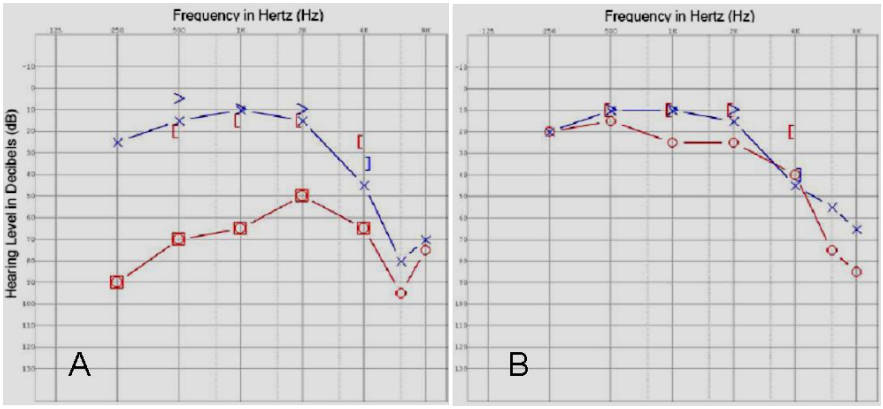

Figure 1:Audiogram of the case 1. A; Preoperative, B; Postoperative.

Case 2

Sixteen-year-old-female patient with hearing loss was admitted to our outpatient clinic. The examination of the external ear and eardrum were normal. The audiogram showed 62 dB conductive type hearing loss on her right side, there was 52 dB air-bone gap. Her hearing level was 8 dB at left ear (Figure 3). Impedance audiometry showed normal curves on both sides. There was no acoustic reflex on the right side.

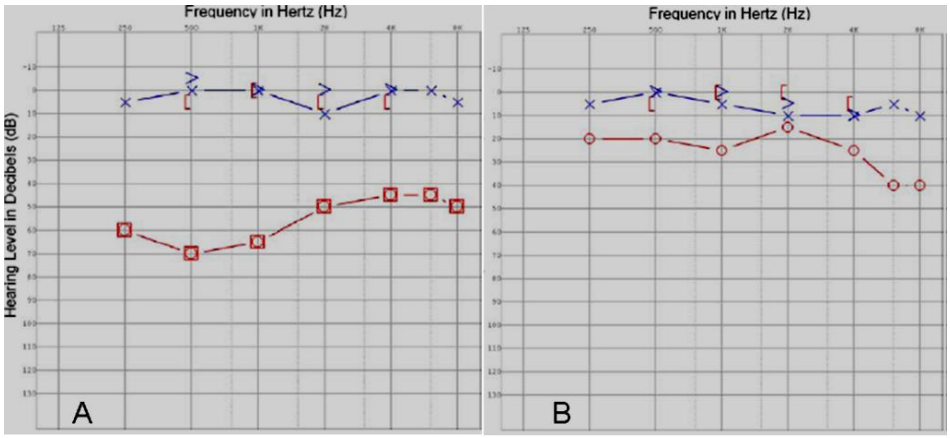

Figure 3: Audiogram of the case 2. A; Preoperative, B; Postoperative.

An explorative tympanotomy was performed under general anesthesia. Suprastructure of the stapes was absent and the long process of the incus was short. The facial nerve was open and hanging over the footplate. The footplate of the stapes was intact and mobile. Incus was removed and a cartilage fragment was harvested from tragal cartilage. Short process of the incus was shortened with diamond burr. A hole was created on the cartilage and long process of the incus was adapted in the hole and they were attached each other with bone cement (Figure 4). The prepared prosthetic bone and cartilage complex was placed over the footplate and tympanic membrane replaced over the prosthesis. Postoperative hearing gain was 42 dB and air conducting hearing level was 20 dB on the right side. The patient had no complaint postoperatively.

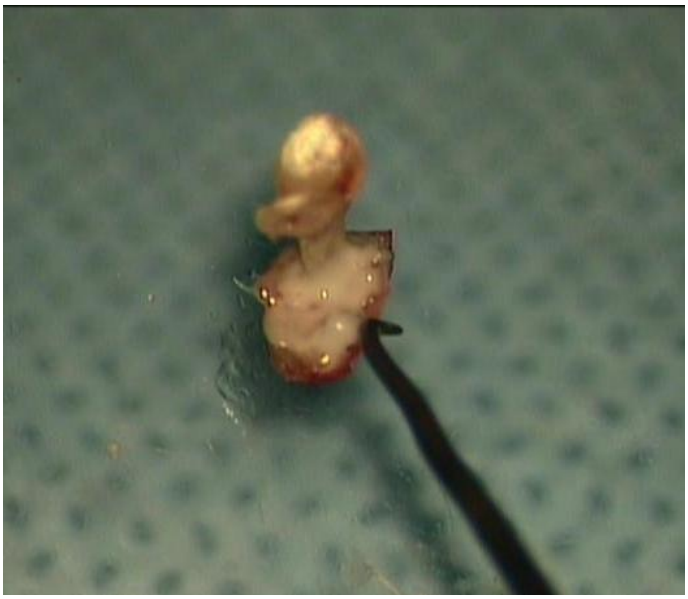

Figure 4. The prosthetic complex made from incus and tragal cartilage.

Discussion

The oval window is an anatomic and developmental junctional zone forming the interface between the middle and the inner ear. The embryology of oval-window atresia is related directly to the development of second branchial arch structures. When the primitive stapes fails to fuse with the primitive vestibule, the oval window can not develop, resulting in its congenital absence. Malformation of the stapes is an expected concomitant finding given the close relationship of the stapes to the oval window during this developmental sequence. Partial absence or hypoplasia of the oval window represents a malformation of intermediate severity.

Several authors have reported congenital absence of oval window (CAOW) [4-8]. In 1976, Jarsdoerfer [8] reviewed the literature and found 45 ears with CAOW. In his series, only 4% of the patients had a normal stapes, 43% of middle ears had no stapes, and 41% had a malformed stapes. In another surgical publication, Lambert [9] reported seven patients with CAOW and noted that the stapes was either absent or severely abnormal in all. The findings in our case were similar to the literatures discussed before. There was a depressed bony plate on the place where the oval window should had been. The stapedial cruras were short and had not connect to the footplate.

Cremers' class III ossicular anomaly consists of anomalies of ossicular chain with mobile footplate. Teunissen and Cremers reported 27 ossicular anomalies with mobile footplate [10]. Eight of the cases had discountinuity and 19 of the cases had fixation. There was only one case with missing suprastructure and aplastic long process of the incus. Cousins and Milton reported that one case had absence of stapes with deficent long process of the incus [11]. But they did not specify if the anomaly was isolated ossicular anomaly. Similar to our case, 2 cases reported by Isenberg and Tubergen [12] had absent stapes suprastructure with short long process of the incus. However, there is no previously reported case with absent suprastructure of the stapes with mobile plate and transposition of facial nerve over the footplate as seen in the second case. Such anomalies could be seen together with facial nerve course anomalies [13]. One of the theories that explain this condition is a fusion between the otic capsule and the second branchial arch which developed too late by preventing the facial nerve from moving anteriorly [14]. Most of these anomalies were presented with congenital major anomalies.

Audiogram and tympanometric assessment can give us limited information. If there was a subtle anomaly, it would be recognized as a normal ossicular chain. Temporal bone HRCT is necessary if there is a suspicion of congenital anomaly and clinician should inform the radiologist if there was a suspicion of congenital ossicular anomaly [11]. HRCT could give us information about aplasia or severe dysplasia of stapes or round window and sometimes condition of the incudo-stapedial joint [15]. On the temporal bone HRCT, when an occluded oval window is seen in association with an anomalous horizontal facial nerve canal, the diagnosis of CAOW may be suggested [7]. Yet, most of the cases of CAOW were identified on surgical explorations. CT is also useful for detecting if there is another abnormal condition especially about facial nerve. CT in the 105-degree coronal and 30-degree axial plane can accurately track the facial nerve through the temporal bone [16]. In our case the stapes suprastructure was normal, the facial canal was not dehiscent and the bony plate was not so thick so the CT scan did not give us adequate information about the pathology. An exploratory tympantomy was required for definitive diagnosis.

Treatment of congenital ossicular anomalies depends on stapes and treatment of stapes anomalies depends on footplate fixation. If stapes footplate is mobile, stapedectomy is unnecessary. Discountinuity with mobile stapes footplate could be reconstructed by using autogenous or allogenic ossicles.

There is no definite answer to the question of which surgical technique is the best treatment for the CAOW. Hearing aid, vestibulotomy and ossicular reconstruction, and fenestration of the horizontal semicircular canal can be used to treat the patients with absence of stapes and oval window. Jahrsdoerfer [17] reported 13 patients with CAOW undergoing an attempt at corrective surgery, with 6 undergoing successful oval window drillout (OWD). Three of them achieved a closure in their air-bone gap between 10 and 15 dB. In two of them gap was closed between 20 to 25 dB. One of them had no improvement. Lambert [9] described a series of 7 patients with CAOW, with 6 patients undergoing OWD. Hearing results in his series demonstrated initial improvement followed by long-term diminish of hearing. In his series, he showed 2 patients had hearing improvement in pure tone audiogram (PTA) and 4 patients had minimal improvement of 0 dB to 10dB. Alarcon et al. [15] reported the largest series of CAOW treated with OWD procedure. 4 surgeries were aborted and 4 had revision surgery. Nine of patients had hearing improvement with OWD procedure. They found a statistically significant decline in hearing over long term period and they could not identify factors or comorbidities that could be predictive of success or failure.

Teunissen and Cremers [2] created neo-oval window with teflon piston bridge in 4 of 14 patients with CAOW and in 2 of them they were successful. They concluded that the opening of the vestibulum could lead to considerable damage to inner ear function. In our case, the oval window nish was visible. Thus, we could easily orientate where the vestibulotomy should be done. In our patient, the bony plate was not thick and we were able to perform vestibulotomy with a pick. In such a case, surgery would be less complicated and we could get more favorable hearing results.

Preoperative radiological assessment especially in suspected anomalies of the oval window and the facial nerve should be done in a special care. It is also important if there is an anomaly in the inner ear. Surgery should not be performed if there is an inner ear anomaly. Hearing level of the opposite ear must also be considered during decision for a surgery. The patient should be informed about the risks and long term results and hearing aids should always be kept in mind as an option.

Cousins and Milton [11] used teflon piston for isolated stapes anomalies with mobile foot plate in their two cases. In one case they obtained hearing improvement but the other resulted in dead ear. They also performed 6 reconstructive procedures for mobile footplate with disorders of the incus-stapes complex. They used interposition in 4 cases and malleus footplate piston in two cases. They obtained hearing gain in two cases. Teunissen and Cremers [10] reported four cases with malformed stapes, mobile footplate and normal malleus. They had reconstructed ossicles with allogenic incus. They had

performed type III tympanoplasty in one case which had mobile footplate with abnormal malleus and incus. The mean hearing gain in these 5 cases was 19 dB hearing levelDb. Jahrsdoerfer [16] reported facial nerve transposition technique for congenital ossicular chain abnormality treatment when facial nerve is passing over the oval window. After facial nerve transpositioning, TORP was used in 3 cases and TORP with a teflon system was modified to fit around the overhanging facial nerve. Hearing gain was observed in two of four cases. We used homogenous incus attached to a cartilage fragment for hearing reconstruction. This method is very safe because stapedectomy is unnecessary and there is almost no risk of dead ear. Furthermore, the risk of extrusion is minimal than the allogenic materials. We emplaced the thin cartilage over the oval window and under the overhanging facial nerve. The shortened incus was attached to the faraway side off the cartilage from the facial nerve. By this prosthetic complex, we set conduction between tympanic membrane and oval window with bypassing the overhanging facial nerve. This is a new technique which could be used for mobile footplate with overhanging facial nerve.

Isolated ossicular anomaly is a rare entity which mimicks otosclerosis and definitive diagnosis could be made by explaratory tympanotomy. Refinement surgical techniques and improvement prosthesis will raise efficiency of surgery and reduce surgical failure and complication rates.

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