Computed tomography of external auditory canal atresia

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Patient: 10 years, female

Clinical History:

We present the case of a 10 year old patient with bilateral congenital external auditory canal atresia (EACA).

Imaging Findings:

The patient had congenital EACA. No concomitant disorders were present. A positive family history was present. A bone conducting hearing aid had been used, and overall speech and perception skills were good. However, hearing loss was progressive and there were difficulties with language courses in school. Clinical examination showed bilateral conductive hearing loss and EACA. To evaluate if reconstruction surgery was possible, high-resolution computed tomography (HRCT) of the temporal bones was performed. Thin (0.3 mm) sections were acquired. HRCT showed bilateral complete atresia of the external auditory canals (Fig. 1). At the normal sites of the tympanic membranes, osseous segments (the atresia plates) were seen. Laterally, the external auditory canals were closed by soft tissue. Bilaterally, the middle and inner ear structures were normal (Fig. 2-5). Also the remaining parts of temporal bones and their relations were normal. The auricles were not completely included in the field of view, but the included parts had normal configuration (Fig. 6). Based on the HRCT findings, the patient was found to be eligible for surgical reconstruction.

Discussion:

EACA is rare with a reported incidence of 1:10.000. Bilateral involvement is present in only 1/3 of the cases. Anomaly of the external ear and conductive hearing loss are the most common clinical findings of EACA. Inner ear anomalies are rarely seen, whereas auricle, middle ear and mastoid anomalies are common. In the present case however, the auricle, middle ear and mastoid were normal on both sides.

EACA may be an isolated malformation or part of an underlying malformation syndrome. Fourteen percent of patients have a positive family history, as was seen in the present case.

Surgical reconstruction of EACA is feasible. HRCT is the method of choice for evaluating temporal bone anatomy in the EACA patient. Temporal bone HRCT findings are used to select patients that are candidates for surgery, as well as to plan the operation, and to predict the postoperative hearing outcome.

A systematical approach in evaluating temporal bone HRCT in the EACA patient is to use the grading scale introduced by Jahrsdoerfer et al. According to this scale the presence or absence of 9 anatomical structures yields a score from 0-10. The higher the Jahrsdoerfer score, the higher is the probability of a good postoperative hearing outcome. In general, patients with a Jahrsdoerfer score < 6 are not considered surgical candidates. The 9 structures included in the Jahrsdoerfer grading scale are listed below together with a brief explanation on how to perform the grading. An important perquisite for using the grading scale is that cochlear function and inner ear structures must be normal. This is because patients with anomalies in the inner ear are not surgical candidates.

The Jahrsdoerfer grading scale:
1) Stapes: 2 points are assigned if the stapes is present, 0 if it is absent.
2) Oval window: The oval window diameter should be ? 2 mm where the oval window enters the vestibule (1 point).
A diameter < 2 mm gives 0 points.
3) Middle ear cavity: In the coronal plane, width of the middle ear cavity is measured from the promontory to the atretic bone. A measure ≥ 3 mm receives 1 point, whereas measures < 3 mm receive 0 points.
4) Facial nerve: If the course of the facial nerve is unfavorable for surgical approach it receives 0 points. Otherwise 1 point is assigned.
5) Incudomallear articulation: Unless severe deformity is present 1 point is assigned.
6) Mastoid pneumatization: A sclerotic, poorly or non-aerated mastoid receives 0 points. Otherwise 1 point is assigned.
7) Incudostapedial articulation: If the joint is well-defined, 1 point is assigned. In case of joint discontinuity, rotation or fusion to adjacent bony structures, 0 points are assigned.
8) Round window: If the round window diameter is < 1 mm in diameter, it is considered stenotic and assigned 0 points. Otherwise 1 point is assigned.
9) External ear: A normal and fairly developed auricle is assigned 1 point, but when configuration is poorer (microtia) 0 points are assigned.

**Differential Diagnosis List:** Bilateral external auditory canal atresia

**Final Diagnosis:** Bilateral external auditory canal atresia

**References:**

Description: Axial thin section showing bilateral external auditory canal atresia (arrows). Origin:
Description: Coronal thin section of the right middle ear and external auditory canal showing the bony atresia plate (arrow). Origin:
Description: Coronal thin section of the left middle ear and external auditory canal showing the bony atresia plate (arrow). Origin:
Figure 2

Description: Right auricle. Origin:
Description: Left auricle. Origin:
**Description:** Axial image showing normal incudomallear articulation in both middle ears. **Origin:**
Description: Coronal image showing normal Incudomallear articulation in both middle ears. Origin:
Figure 4

Description: Normal right stapes (arrow). Origin:
Description: Normal left stapes (arrow). Origin:
Figure 5

Description: Normal formation of the right bony labyrinth and internal acoustic meatus. Origin:
Description: Normal formation of the left bony labyrinth and internal acoustic meatus. Origin:
Figure 6

Description: Normal course of the descending portion of the right facial nerve (arrow). Origin:
Description: Normal course of the descending portion of the left facial nerve (arrow). Origin: