# Congenital hearing loss explained in adulthood. Computed tomography of the temporal bone in hemifacial microsomia. A case report.

Dietrich Beitzke[1*], Ulrike Wiesspeiner[1], Peter Brader[1], Markus Beitzke[2], Josef Simbrunner[1]

*1. Department of Radiology, Medical University Graz, Graz, Austria*

*2. Department of Neurology, Medical University Graz, Graz, Austria*

* **Correspondence** Dietrich Beitzke, MD., Department of Radiology, Section of Neuroradiology, Medical University Graz, Auenbruggerplatz 9, 8036 - Graz, Austria
(dietrich.beitzke@aon.at)

Radiology Case. 2009 Oct; 3(10):1-4 :: DOI: 10.3941/jrcr.v3i10.302



ABSTRACT

We present a case of complex hemifacial microsomia (HFM) which was diagnosed at the age of 46 years. Imaging findings of a complex deformity of the temporal bone are presented and connected to a broad range of clinical symptoms. Computed tomography (CT) imaging indications are discussed briefly.



## CASE REPORT

### CASE REPORT

A 46 year old male patient was referred to our department of otolaryngology because of symptoms of sudden left sided hearing loss in combination with tinnitus. He had been noticed to have a right-side congenital hearing loss but never underwent clinical or diagnostic exploration.

The clinical evaluation of the left ear showed a thickening of the tympanic membrane (TM) and a perforation in the anterior lower section. An audiogram of the left ear showed signs of mild conductive hearing loss. Clinical evaluation of the right ear revealed microtia grade II according to the classification of Weerda with a low set right auricle and an atretic external auditory canal (EAC). TM was not seen in the endoscopic exploration. An audiogram of the right ear revealed severe conductive hearing loss. Additional neurological evaluation revealed a light paresis of the peripheral part of the right facial nerve.

Because of conductive hearing loss on both sides and right sided microtia standard two-plane multislice computed tomography of the temporal bone was performed.

Computed tomography of the right temporal bone confirmed stenosis and low position of the EAC (Fig.1A). Behind the EAC stenosis, in front of the anterior part of the TM, a soft tissue mass was detected (Fig1B, 2, 3A-C) but there were no signs of bony erosion indicating a cholesteatoma. Evaluation of the right middle ear revealed reduced width of the middle ear cavity with dysplasia and partial fusion of the malleolus and incus (Fig.3, A-C). Right mastoid bone was hypoplastic and its pneumatization was completely absent (Fig.1, 2). The right internal carotid artery canal showed a reduced diameter in the petrous segment (Fig.2). The facial nerve canal showed anterior displacement in the mastoid segment (Fig. 1C). No anatomical abnormalities were found at the right inner ear.

Imaging results were completely compatible with otolaryngology findings of the patient and led to the diagnosis of hemifacial microsomia (HFM). Additional conventional X-ray of the cervical spine revealed missing posterior fusion of the vertebral arch C6 and C7 (Fig. 4).

## DISCUSSION

HFM is a congenital disease affecting the development of aural, oral, mandibular and vertebral structures resulting in a broad range of otolaryngology symptoms with a marked variability. Due to the anatomical complexity of this region proper CT imaging is crucial for diagnostic and preoperative issues. Knowledge of the underlying embryology is demanded in image interpretation in developmental disease of the temporal bone.

HFM was initially described by Goldenhar in 1952(1). Gorlin et al. defined it as a condition affecting oral, aural, mandibular and vertebral structures. Due to this dispersion the name oculoauriculovertebral spectrum (OAVS) was also introduced. The incidence of HFM/OAVS is about 1/5600 and the male to female ratio is at least 3:2 (2). Symptoms are limited to one side only in most of the cases and are varying from mild to severe.

As the middle and external ear develops from the first and second branchial arch and the endodermal first pharyngeal pouch starting in the 4th gestational week, structures arising from these origins are involved in HFM. The first and the second branchial arch originate into the ossicular chain. The first endodermal pharyngeal pouch develops into the future auditory tube, the middle ear, the antrum mastoid and the inner layer of the TM.

The etiology of HFM is heterogeneous and is associated with vasoactive drug intake during the first weeks of gestation especially in conjunction with smoking. It may also be associated to different chromosome aberrations.

Indication for primary CT evaluation of the temporal bone include conductive hearing loss, deformed auricle, external auditory canal abnormalities, chronic otitis media/cholesteatoma, postoperative conditions, trauma and peripheral facial nerve paralysis if there is evidence for a cholesteatoma or a middle ear mass (3). Chronic otitis media/cholesteatoma and postoperative conditions additionally require contrast enhanced images. Our case nicely demonstrates the complex connection between the pathologic development during the gestation and a broad range of otolaryngology symptoms that could nicely be explained by multislice CT of the temporal bone. Besides diagnostic issues, imaging is necessary if surgical intervention is planned. With recent technical advances in multirow detector CT scanners imaging is nowadays be performed in axial plane acquiring a volumetric CT data set with 0.5 to 0.7 mm slice thickness for multiplanar reconstruction.

Congenital pathologies of the EAC include atresia and stenosis. Atresia can be bony, membranous and mixed. EAC stenosis like in our case may cause retention of cerumen and debris but may also induce a cholesteatoma of the EAC, normally a rare entity. CT imaging findings of a cholesteatoma include signs of bony erosion, missing in our study so the soft tissue mass was more likely consisting of cerumen or debris (4). Our patient also showed pathologic findings of the middle ear cavity and the ossicular chains congruent with the findings of Mayer et al. (5). The authors proved in a retrospective analysis of 92 cases that the grade of aural dyplasia is linked to the serverity of EAC stenosis or atresia, the lack of pneumatization of the mastoid, the size of the middle ear space and the grade of aberration of the ossicular chain. Slight aberrations of the carotid canal like in our case were also described in this study. An abnormal course of the facial canal is also often observed in aural dyplasia and EAC stenosis. The exact course of the facial canal is very important in preoperative imaging of the temporal bone as the posterior genu of the facial nerve is often placed anteriorly in HFM. Pneumatization of the mastoid is also linked to the grade of aural dysplasia. Inner ear deformities are usually not observed in hemifacial microsomia (5). Cervical vertebral malformations in HFM include fusion of vertebral bodies and missing fusion of vertebral arches.

Several syndromes are also associated with aural dysplasia and therefore should be considered as differential diagnosis (3). Treacher-Collins syndrome has additional features of micrognathia and typically includes bilateral downward slanting eyes from facial bone underdevelopment. Crouzon disease also occurs bilaterally and, besides EAC stenosis or atresia, patients are usually suffering from brachycephaly because of cranial synostosis. In Nager syndrome there is underdevelopment of the cheek and jaw area, down sloping of the opening of the eyes and shortened forearms.

## TEACHING POINT

Thin cut multislice CT is the imaging modality of choice in evaluation the temporal bone in HFM and other conditions associated with aural dysplasia. Indications for primary CT of the temporal bone include auricular dysplasia, EAC stenosis, conductive hearing loss, and facial nerve paresis in case of detected mass in the middle ear.

## REFERENCES

1. Goldenhar M. Associations malformatives do I'oeil et de I'oreille, en particulier le syndrome dermo¨ide epidbulbaire - appendices auricularies - fistula auris congenita et ses relations avec la dysostose mandibulo-faciale. J Genet Hum 1952; 1:243-82.

2. Hartsfield JK. Review of the etiologic heterogeneity of the oculo-auriculo-vertebral spectrum (Hemifacial Microsomia). Orthod Craniofacial Res 10, 2007; 121-128.

3. Imaging of the temporal bone; Schwartz JD, Harnsberger R. 3rd ed. New York: Thieme; 1997.

4. Heilbrun ME, Salzman KL, Glastonbury CM, Harnsberger HR, Kennedy RJ, Shelton C. External auditory canal cholesteatoma: clinical and imaging spectrum. AJNR Am J Neuroradiol. 2003; 24:751-6.

5. Mayer TE, Brueckmann H, Siegert R, Witt A, Weerda H. High-resolution CT of the temporal bone in dysplasia of the auricle and external auditory canal. AJNR Am J Neuroradiol. 1997; 18:53-65.

FIGURES

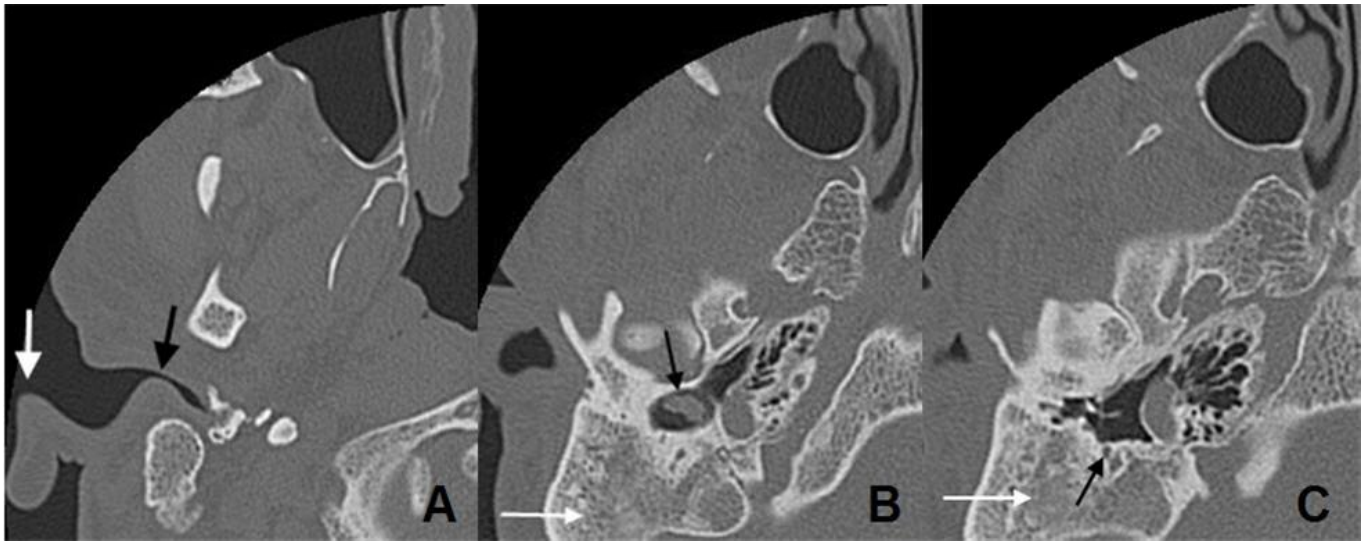


**Figure 1:** Axial computed tomography images in a 46 year-old man with hemifacial microsomia (native, 120kv/120mAs):
A: Stenosis of the external auditory canal (black arrow) and dysplasia of the auricle (white arrow).
B: Soft tissue mass in the external auditory canal next to the tympanic membrane (black arrow). Note the complete absence of pneumatization of the mastoid (white arrow).
C: Again absence of pneumatization of the mastoid bone (white arrow). Anterior displacement of the facial nerve canal in the mastoid segment (black arrow).

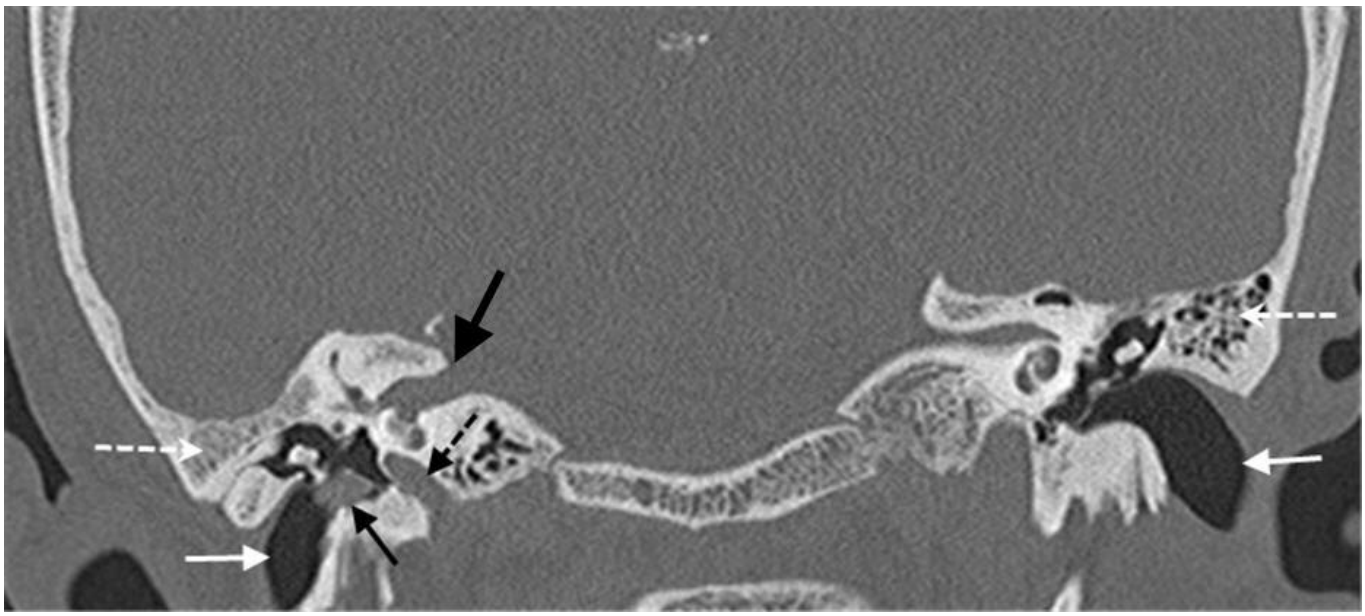

**Figure 2:** Coronal computed tomography (native, 120kv/120mAs) image in a 46 year-old man allows a good overview of the pathologic findings in hemifacial microsomia: Complete absence of pneumatization and hypoplastic appearances of the right mastoid bone (white dotted arrows). Right external auditory canal shows reduced diameter (white arrows) and inside a soft tissue mass in front of the right tympanic membrane (black arrow). Note the narrowing of the right internal carotid artery canal (black dotted arrow). Additional shortening of the right internal auditory canal (black spaced out arrow).

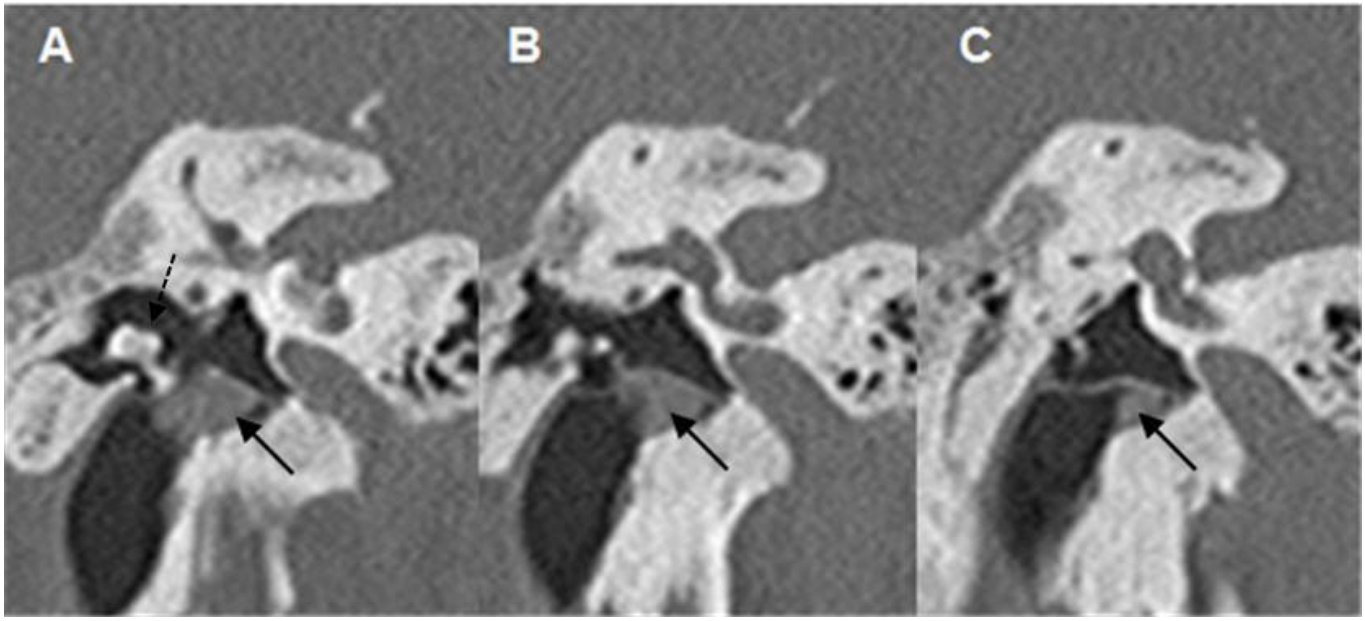


**Figure 3:** A-C: Serial coronal CT - images (slice thickness 1mm, native, 120kv/120mAs) of the right middle ear cavity in a 46 year-old man with hemifacial microsomia) shows dysplasia and partial fusion of the malleolus and incus within a small middle ear cavity (black dotted arrow in A). Soft tissue mass in front of the tympanic membrane (black arrows).

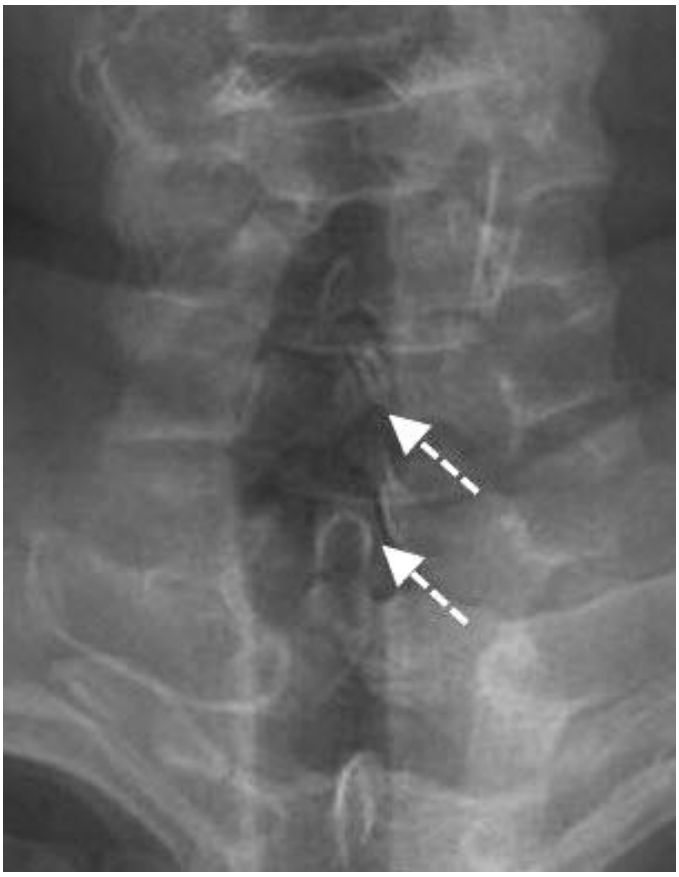

**Figure 4:** AP radiograph of the cervical spine in a 46 year-old man with hemifacial microsomia demonstrating missing posterior fusion of the vertebral arch C6 and C7 (white dotted arrows).

ABBREVIATIONS

HFM: hemifacial microsomia
CT: Computed tomography
TM: tympanic membrane
EAC: external auditory canal
OAVS: oculoauriculovertebral spectrum
AP: anterior posterior

KEYWORDS

Temporal bone, computed tomography, hemifacial microsomia

***Online access***

This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/302

***Interactivity***

This publication is available as an interactive article with scroll, window/level, magnify and more features.
Available online at www.RadiologyCases.com

Published by EduRad
www.EduRad.org