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CASE REPORT

Radiological Findings of Michel Aplasia

Ayse Umul, Hakan Demirtas, and Ahmet Orhan Celik

Faculty of Medicine, Radiology Department, Suleyman Demirel University, Isparta, Turkey

Corresponding author: Ayse Umul, MD. Suleyman Demirel University, Faculty of Medicine, Radiology Department, Isparta, Turkey (ayseumul@gmail.com)

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ABSTRACT

Introduction: Congenital abnormalities of the inner ear is the most common cause of neurosensory hearing loss. Michel inner ear deformity is a rare developmental anomaly refers to the total aplasia of the inner ear. It is caused by developmental arrest of otic placode early during the third week of gestational age. **Case report:** We have discussed here that three year old girl diagnosed Michel aplasia with temporal bone computed tomography (CT) and temporal magnetic resonance imaging (MRI) findings.

Key words: neurosensory, aplasia, otic placode, imaging findings.

1. INTRODUCTION

Congenital inner ear abnormality is the most common cause of neurosensory hearing loss (1). Inner ear aplasia, also referred to as Michel aplasia, is due to failure of development of the otic placode occurring before the third week of gestation (2, 3). As the name suggests, this anomaly was first reported by Michel in the autopsy report of a 12-year-old boy with a history of congenital deafness (4). Multi-detector, thin slices CT is an effective diagnostic method for scanning inner ear abnormalities. Temporal MRI is useful in diagnosis with a high soft tissue contrast. Our aim is to reveal cross-sectional imaging findings of a three-year old female patient with Michel aplasia.

2. CASE REPORT

A three-year old female patient with the pre-diagnosis of neurosensory hearing loss in the left ear admitted to our hospital for CT scan of the temporal bone. Cochlea, vestibule and semi-circular canals of the inner ear structures in the left ear were not monitored, while the internal auditory canal was monitored markedly narrowed compared to the right ear on the multi-detector thin-section CT (Figure 1). The seventh cranial (facial) nerve was observed to have an abnormal course. The seventh and eighth nerve complexes in the left ear were found to be hypoplastic and the 8th cranial nerve could not be monitored on the temporal MRI scan (Figure 2). A hypoplasia was found on the petrous bone. The Welcher basal angle was measured as 152 degrees and found to have increased in line with the platybasia. We considered Michel deformity radiologically due to the defined findings. The patient whom was not implemented a cochlear implant was given recommendations.

3. DISCUSSION

During the embryonic development of the inner ear, otic placodes arise from the surface of ectoderm on either side of the rhombencephalon at approximately the third gestational week and subsequently, otocysts develop. At the fifth week, a diverticulum buds from the otocysts forming the endolymphatic sacs, followed by the cochlea and vestibule. The membranous cochlea completes 1 to 1.5 turns at the end of the sixth week and 2.5 turns at the end of the seventh week. The semi-circular canals start to develop from the utricle segments of the otocysts at the seventh to the eighth gestational week. The superior canals form first, followed by the posterior and then the lateral canals. The inner ear structures reach their adult forms by the end of the eighth week (2).

It is the Jacker classification that is the most commonly used classification of

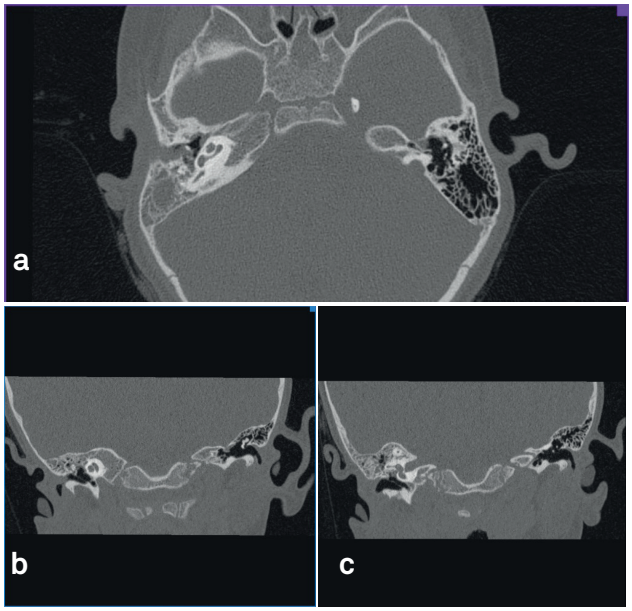

Figure 1. Cochlea, vestibule and semi-circular canals are not monitored in the left ear on the axial (a) and coronal reformatted MPR (b and c) images in CT scan of the temporal bone.

inner ear anomalies (2).This classification was made based on the time of developmental arrest during embryogenesis and anatomical development. Accordingly, inner ear anomalies are examined in five (5) groups: 1) Michel aplasia (complete labyrinth aplasia); 2) Mondini anomaly (incomplete division); 3) Cochlear aplasia; 4) Cochlear hypoplasia; 5) Common cavity.

Sennaroğlu et al. (5) modified this classification and grouped the inner ear anomalies by their decreasing severity as Michel deformity (complete labyrinth aplasia), cochlear aplasia, common cavity, IP-1 (cystic cochleovestibular malformation), cochlear hypoplasia and IP-2 (classic Mondini deformity).

Michel aplasia that is the most severe and rare type in these classifications occurs due to the non-formation of otic placodes in the beginning of the development. Cochlea, vestibule and semi-circular canals do not exist. Michel aplasia can be seen in girls or boys and it does not have any significant gender difference (6-8). Although most of the cases reported are bilateral, cases that are unilateral as in our case are also seen. Exposure to thalidomide (9), congenital CMV infection (10) and genetic diseases (11) are predisposing factors.

Vestibulocochlear nerve, which is the eighth cranial nerve, and its ganglion are formed by otic vesicle cells during the fourth gestational week (12). Otic vesicle also ensures continuity and growth of the eighth nerve (13). This nerve is aplastic in patients with Michel aplasia because they have no otic vesicle development. Absence of the eighth nerve causes decrease in size and calibration of the internal auditory canal containing only the seventh nerve, as in our case (14).

Facial nerve, which is the cranial nerve, develops normally, but exhibits an abnormal course (15).

Michel aplasia may be associated also with skull base and vascular anomalies. Accompanying skull base anomalies are petrous bone hypoplasia, platybasia and aberrant jugular bulb veins (4). Our case had platybasia, too. The incidence of posterior fossa anomalies also increased.

Accompanying tracheoesophageal, cardiac or extremity malformations and facial palsy were reported in some cases with Michel aplasia (7, 16).

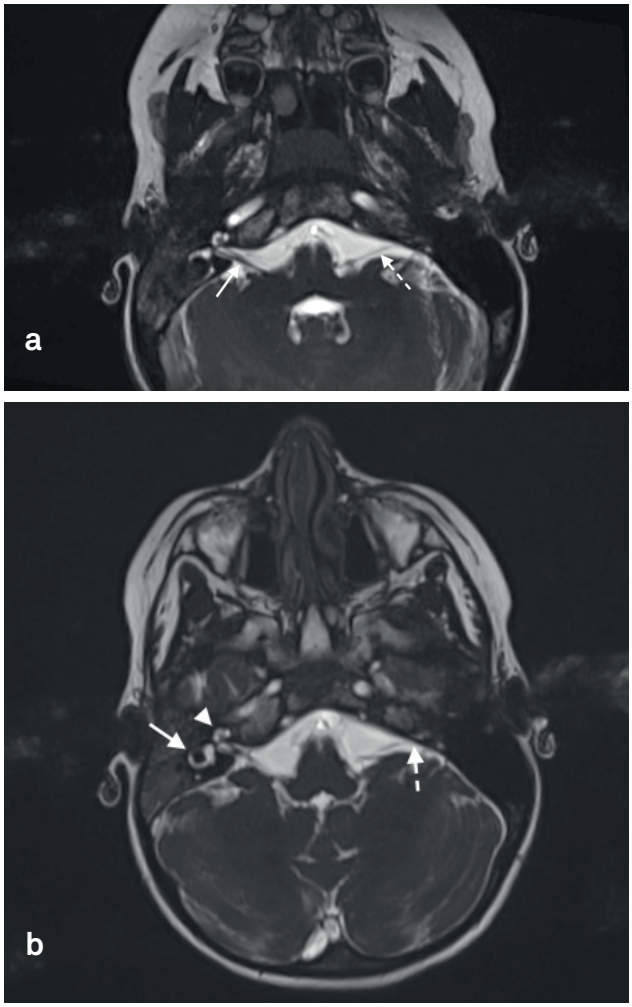

Figure 2. a) The right arrow on the axial T1-weighted MR image shows the seventh and eighth nerve complexes and the left dashed arrow shows the seventh nerve. The eighth nerve is not monitored in the left ear. b) Semi-circular canals and cochlea on the left are shown with the arrows on the axial T1-weighted MR image. The left dashed arrow shows the hypoplastic internal auditory canal.

CT ensures scanning of the bony labyrinth and otic capsule in patients with congenital neurosensory hearing loss (17). Multi-detector CT plays an important role in diagnosis due to its ability to obtain very thin sections and features of anatomical detailing and high resolution. CT findings in Michel aplasia are diagnostic. It manifests with absence of the cochlea, vestibule and semi-circular canals, which are the inner ear structures, narrowing of the internal auditory canal and flattening of the middle ear medial wall.

MRI is a very useful diagnostic tool that ensures distinguishing lesions forming a high contrast soft tissue. That it does not contain any ionizing radiation is an important advantage especially in pediatric age group (18). Stenosis of the internal auditory canal, absence of the seventh and eighth cranial nerves and abnormal course of the seventh cranial nerve in MRI allows for establishing the diagnosis.

Recognizing Michel aplasia is important in planning cochlear implantation because there is no place for implantation in Michel aplasia where the internal auditory canal is markedly narrow and the vestibulocochlear nerve does not exist.

4. CONCLUSION

Congenital inner ear abnormality is the most common cause of neurosensory hearing loss and Michel aplasia is characterized by a total absence of inner ear. Diagnosis is established based on cross-sectional imaging findings. There is no place for cochlear implantation in treatment.

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