CASE REPORT

Inferior pontine segmentation abnormality in a child with sensorineural deafness: DTI analysis of fiber tracts

Summary MRI/DTI data are presented in a child with sensorineural hearing loss and swallowing disorder. MRI/DTI revealed hypoplastic 8th cranial nerves and an inferior pontine segmentation abnormality. Color-coded FA-maps revealed diminished/absent fiber tracts within the affected brainstem segment. This report may add another small puzzle piece to the ongoing research on brainstem malformations.

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Introduction

Malformations of the brainstem and cerebellum have been described in various conditions including chromosomal disorders [1], pontocerebellar hypoplasias types 1-5 [2], familial horizontal gaze palsy and scoliosis [3], Möbius syndrome [4] and more recently, pontine tegmental cap dysplasia [5]. Recent advances in neuroimaging and genetics have resulted in a more accurate classification of posterior malformations into disorders involving a) abnormal brainstem segmentation, b) segmental hypoplasia, c) postsegmentation abnormalities and d) abnormal cortical organization [6]. In this report, we present the magnetic resonance imaging (MRI) and diffusion tensor imaging (DTI) findings in a 12-month-old child referred because of sensorineural hearing loss (SNHL). MRI revealed hypoplastic 8th cranial nerves in combination with an inferior pontine segmentation abnormality.

Case report

A 12-month-old Caucasian female presented to our department for MR imaging of the brain and brainstem because of profound bilateral SNHL. She was born at 41 weeks gestation via spontaneous delivery. Pregnancy was uncomplicated. Apgar scores were 5/7/9. At birth she had respiratory dis-
Inferior pontine segmentation abnormality

Figure 1  
T1- and T2-weighted (CISS) sagittal MRI scans of the brain shows truncation of the inferior pons (arrow). Corpus callosum and remainder of the midline structures appear unremarkable.

Figure 2  
A,B: coronal (A) and axial (B) high resolution heavily T2-weighted CISS imaging of the cochlea and vestibulum. The internal auditory canals (arrows) and 8th cranial nerves are severely hypoplastic, right > left. The inferior pons is hypoplastic.
This case report would like to add an additional small piece to the complex puzzle of brainstem malformations by presenting and discussing a novel case of an inferior pontine segmentation abnormality accompanied by hypoplastic/absent vestibulocochlear nerves bilaterally.

The distinct brainstem abnormality found in this patient is characterized by the apparent truncation of the inferior portion of the pons. Our findings are best understood by a review of the normal pontine anatomy. The dorsal pons primarily contains cranial nerves V, VI and VII as well as their nuclei, the medial longitudinal fasciculus, the tectospinal tract, the medial leminiscus, the lateral leminiscus, the central tegmental tract, and parts of the superior cerebellar peduncle. The ventral part of the pons, which is the area of abnormality in our case, consists of the descending axons of the CST, CPT, pontine nuclei as well as transverse pontocerebellar axons, which are known as the PCT [6]. DTI analysis of our patient revealed a marked diminishment of the CPT, CST, PCT in the inferior portion of the pons. In addition, the ICP, MCP were diminished at the inferior level of the pons. This observation can be explained by the fact that during development, the PCT normally connect the pontine nuclei to the contralateral cerebellar cortex via the MCPs [8] and therefore abnormalities in the PCT would lead to an abnormality of the MCP.

In addition, conventional MR imaging also demonstrated absence of the 8th nerves and diminutive internal auditory canals bilaterally. We speculate that this may be attributed to an absence/malformation of the cochlear nuclei and/or course of the cochlear tracts within the brainstem. The cochlear nuclei are located lateral and dorsal to the restiform body in the floor of the lateral recess of the fourth ventricle, while the cochlear tract lies along the inferior border of the pons as cochlear nerves. In our case this inferior part of the pons appeared absent/hypoplastic.

Combining the findings of conventional MRI/DTI, the malformation observed in our case could have resulted from an early abnormal pontine segmentation as well as an impairment of cell proliferation of the pons and brainstem [6,8].

While our report may add another small piece of the puzzle to the ongoing research on brainstem malformations, it suffers from a number of limitations. It is a single case report, no neuropathological or detailed neurophysiological information is available, only those fiber tracts were studied that have been described previously. Our case report shows that several of these tracts were not identified in their usual location. It is however unclear if these tracts are absent, hypoplastic or possibly follow a different anatomical course. Future prospective studies using multi-tensor DTI should explore if these tracts are truly absent, follow a different pathway or if other neuronal networks have been recruited.

Conflicts of interest statement

The authors declare to have no conflicts of interest.

References

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Inferior pontine segmentation abnormality


