Klarov L.A., Barashkov N.A., Teryutin F.M., Popov M.M., Soloviev A.V., Pshennikova V.G., Fedotova E.E., Romanov G.P., Luginov N.V., Fedorova S.A.

The Results of Computer Tomography (CT) and Magnetic Resonance Imaging (MRI) of a rare case of Bilateral Inner Ear Aplazia

ABSTRACT

The results of computer tomography (CT) and magnetic resonance imaging (MRI) of a rare case the analysis of the images is performed and a detailed description of changes on CT and MRI studies is given. This case of aplasia is characterized by symmetrical agenesia of the of bilateral aplasia of cochlea and semicircular canals of the inner ear are presented. In this article cochlea and semicircular canals, and bilateral abnormality of the facial nerve canal, and abnormality of the internal auditory foramen on both sides, as well as unilateral agenesis of the abducent nerve and bilateral agenesis vestibulocochlear nerve. Results of the study indicate a high informative method of CT in the diagnosis of the inner ear abnormalities, as well as the possibility of using MRI in visualizing the cerebellopontine angle for the entire description of its clinical picture.

Key words

Michel aplasia, sensorineural deafness, CT of temporal bones, MRI of cerebellopontine angle and inner ear.

INTRODUCTION

Congenital hearing loss, registered at a frequency of an average of 1 per 1000 live births [1]. The etiology of this disease is heterogeneous. However, according to various authors, congenital anomalies of the inner ear in the average recorded in 20% of individuals with congenital hearing loss. In the structure of the inner ear malformations more than 90% of cases occur in Mondini anomaly - common cavity and hypoplasia of the cochlea, and about 1% cases is Michel aplasia [3].

Michel aplasia is a rare anomaly of embryonic development of the structures of the inner ear. For the first time this anomaly was described by P. Michel in 1863. (Subsequently became known as the "Michel aplasia") on autopsy material 11-year-old deaf boy who died at Children's Hospital of Strasbourg in France [2]. In the study of sectioned material P. Michel said symmetrical bilateral absence of the cochlear and vestibular structures of the inner ear (cochlea and complete

agenesis of the semicircular canals), which is characterized by abnormalities of the skull base and facial nerve canal dystopia and the jugular vein. [2]

Currently, for in vivo diagnosis of anomalies of the inner ear is used radiological methods of investigation, according to the standard classification on the presence / absence and / or resizing of certain structures proposed by R.K. Jackler in 1987 [3].

In this paper first summarizes the results of a computer (CT) and magnetic resonance imaging (MRI) of the rare cases of Michel aplasia, identified patient from Yakutia with congenital bilateral deafness and neuropathy abducens right.

MATERIAL AND METHODS

Patient

The presented case of Michel aplasia was detected in a patient with congenital bilateral sensorineural deafness, which was followed up by Audiology-Logopaedic Center of the Republican Hospital №1 - National Center for Medicine, Ministry of Health of the Republic of Sakha (Yakutia). The patient was examined by an audiologist, speech therapist, psychoneurologist. Study of hearing was conducted with the threshold tonal audiometry on the unit «Clinical Tonal Audiometer - GSI® 60» (Grason Stadler, USA) in a soundproof chamber calibrated Republican Audiology-Logopaedic center. The patient underwent functional studies: reflexometer, tympanometry (Amplaid, Italy).

From anamnesis we know that the patient is born in 1995 (at the time of the study 15 full years), sex, female, Russian, was born from the X pregnancy (birth in time, the weight of 3 kg) III childbirth. The impact of negative factors (ionizing radiation, medication, infectious disease) during pregnancy parents deny. Consists in the dispensary with 3 years Audiology-Logopaedic center of the Republican Hospital №1 (Yakutsk) with a diagnosis of congenital bilateral sensorineural deafness. Enrolled in a correctional boarding school type I for deaf children (Yakutsk). Consists followed up by a neurologist (psychomotor retardation, neuropathy abducens right vestibular ataxia in stage subcompensation) and ophthalmologist (hyperopia I st. OU, retinal angiopathy OU, exotropia). Psychosomatic status corresponds to the age. Abnormalities of the cardiovascular, endocrine systems have been identified. When audiological study, examine the patient observed systemic underdevelopment of speech, speech develops through learning, perception on the basis of visual, spoken and whispered speech are not perceived around the ear. Preferred type of communication based on gestural vocabulary. When the threshold tonal audiometry on both sides recorded remnants of hearing at frequencies of 125,

250, 500, 1000 Hz on air at levels of 75, 90, 100, 100 dB, respectively, according to the International Classification of degrees of hearing loss, deafness corresponds to sensorineural type.

Alternatively, the patient was examined by norms born in 1997, female, diagnosed with congenital Yakut bilateral sensorineural deafness type not established etiology, with preserved bodies of the outer, middle and inner ear, without comorbidity with other organs and / or systems.

Computed tomography of the temporal bone pyramid

Analysis of the petrous held on 4 slice CT scanner Somatom Sensation 4 (Siemens, Germany) in axial projection with thick tomographic layer 1 mm step promotion table 1mm increment reconstruction of 1 mm (program InnerEarSpi), voltage 120 kV, current 70 mA.

When visualizing structures petrous used 2D images as a native axial plane, and in the mode MPR reformation, using the "bone" filter with a window width of 4000 HU, window level +700 HU.

Magnetic resonance imaging area cerebellopontine angle

The study was conducted on magnetic resonance imaging Magnetom Espree (Siemens, Germany) with a magnetic field strength of 1.5 T. We used the isotropic sequence T2 ci3d with slice thickness of 0.6 cm, with an isometric voxel 0.6 x 0.6 x 0.6 cm. With the parameters of the sequence TE (TimeEcho) - 2.81 and TR (TimeRepetition) - 6.25, with a resolution of the image matrix of 384 x 512, with FOV (fieldofview) - 135 x 180.

When rendering of the facial and abducens image used as a native axial plane, and in the mode MPR reformation.

Ethical control

This work was approved by the local ethics committee in biomedical ethics at FGBU "YSC ILC" SB RAMS, Yakutsk, Protocol №16 from April 16, 2009 CT and MRI - research conducted with the informed written consent of the parents of the patient.

RESULTS AND DISCUSSION

For the first time the results of computed tomography and magnetic resonance imaging of the temporal bone and cerebellopontine angle gives a detailed description of a rare case of congenital anomalies of the cochlear and vestibular structures of the inner ear - Michel aplasia.

CT imaging petrous examined patients is shown in Figure 1 (A and B). In a series of CT tomograms visualized ear canal size and shape is not changed. It should be noted that the study oto- abnormalities of external ear was also not detected. On CT images ossicular chain is not changed. The patient has the right (Fig. 1A) changes from the mastoid not detected, but the structure of the left mastoid diploetic type cells observed in the posterior slight thickening of the mucosa, suggesting previously recovered otitis media with low persistence. Internal auditory canal is visualized, but deformed, band-shaped, narrowed to 0.25 to 0.16 and the right from the left (Fig. 2.A). Channel deformed facial nerve on both sides, but is rendered throughout, narrowed in the mastoid section to the rear knee, and expanded in the drum (Fig. 2.B). The patient on the CT images snail, water snails and vestibule, and semicircular canals are not defined on both sides.

On the basis of complaints, anamnesis, clinical trial data, the CT examination the patient diagnosed with «bilateral congenital abnormality of the inner ear (Michel aplasia). Bilateral sensorineural deafness». However, the results do not fully explain the neurologic symptoms in patients (psychomotor retardation, neuropathy abducens right vestibular ataxia in stage subcompensation) except vestibular ataxia, which is likely due to the absence of the semicircular canals. In this regard, we have carried out an additional magnetic resonance imaging, as principle of obtaining CT images does not allow a detailed assessment of the pathology of cranial nerves in the cerebellopontine angle.

MR imaging of the cerebellopontine angle is shown in Figure 3. Similarly, the CT study on MRI images indicated the absence of structures of the inner ear (cochlea and the semicircular canals) and symmetric narrowing of the internal auditory canal on both sides.

Using MRI reconstructions was visualized area cerebellopontine angle. MRI images in the projection of the seventh and eighth cranial nerves on both sides determined by one nerve that makes it impossible to clearly differentiate them from each other. However, given that the vestibular-cochlear nerve (eighth pair) consist of a 3-ex fibers (lower and upper vestibular and cochlear nerves), and the fact that there are no organs of the inner ear, it is likely to MRI images are defined facial nerves (seventh pair). In the left cerebellopontine angle, posterior to the internal auditory canal is determined small arachnoid cysts with a clear thin capsule (size 17 x 7 mm). On the right is defined similarly arachnoid cysts, slightly invaginated into the internal auditory canal (7 x 7 mm). Also on MR images there is a lack abducens right. Left abducens the ordinary course, not changed. The absence of MRI images of the right abducens fully explain the reasons for divergent strabismus in patients surveyed on the right side.

Thus, the results of the study indicate a high informative method of CT in the diagnosis of abnormalities of the inner ear [4], as well as the possibility of using MRI to visualize the cerebellopontine angle, for the most complete explanation of the clinical picture.

ACKNOWLEDGEMENTS

This work was supported by grants of RFBR (14-04-01741_a) (15-44-05106-r_vostok_a) project of the Ministry of Education and Science of the Russian Federation Civil Code №6.656.2014/K, SB RAS integration project number 92, as well as the Grant of Head of the Republic Sakha (Yakutia) for young scientists, professionals and students in 2015 (#79 from 06.02.2015).

REFERENCES

1. Mehl A.L. The Colorado newborn hearing screening project, 1992-1999: on the threshold of effective population-based universal newborn hearing screening / A.L. Mehl, V. Thompson // Pediatrics. – 2002. – 109. – 1:E7.

2. Michel P. Me´moire sur les anomalies conge´nitales de l'oreilleinterne / P. Michel // Gazette Me´d de Strasbourg. - 1863. – Vol. 23. – P. 55–58.

3. Jackler R.K. Congenital malformations of the inner ear: a classification based on embryogenesis / R.K. Jackler, W.M. Luxford, W.F. House // Laryngoscope. - 1987. – Vol. 97, № 40. – P. 2–14.

4. Zelikovich E.I. CT of the temporal bone in the study of the inner ear structures in detection of the causes of neurosensory hypoacusis / E.I. Zelikovich // Vestn. otorinolaringol. - 2004. - № 6. - P. 25-31.

AUTHORS

1. Klarov Leonid Alexandrovich - radiotheropist, CBI Republican Hospital №2 – Emergency Medical Centre, eizonix@mail.ru
2. Popov Mikhail Mikhailovich - radiotheropist, CBI Republican Hospital №2 – Emergency Medical Centre, quentus@bk.ru
3. Luginov Nikolay Vasilievich - cand. med. sciences, head of Radiotherapy
4. Fedotova Elvira Egorovna – cand. med. sciences, audiologist, chief non-staff surdologist of MH RS(Y), head of the Republican surdologic centre RH №2 – NCM, FSBCE YCC KMP sakhasurdo@mail.ru

5. Barashkov Nikolay Alekseevich – cand. biol. sciences, research associate, barashkov2004@mail.ru
6. Teryutin Fedor Mikhailovich – junior research associate, rest26@mail.ru
7. Pshennikova Vera Gennadievna – research assistant, FSAEE Northeast Federal University psennikovavera@mail.ru
8. Soloviev Aysen Vasilievich - engineer, nelloann@mail.ru
9. Romanov Georgiy Prokopievich – research assistant, gpromanov@gmail.com,
10. Fedorova Sardana Arkadievna – doct. biol. sciences, head of the Scientific laboratory of the Institute of natural Sciences

1 - Department of Radiology, Republican Hospital № 2 – Center of Emergency Medicine, (Yakutsk)

2 - Laboratory of Molecular Genetics, Department of Molecular Genetics, Yakut Scientific Center of complex medical problems, (Yakutsk)

3 - Laboratory of Molecular Biology, Institute of Natural Science, M.K. Ammosov North-Eastern Federal University (Yakutsk)

4 – Audiology-logopaedic center, Republican Hospital № 1 - National Centre of Medicine (Yakutsk)