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A clinical case of cortical deafness in a patient with symmetrical cystic transformation of cerebral hemispheres

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Cortical deafness is a complete absence of the function providing a person with an ability to perceive audible signal caused by defects of cortical centers of hearing of both cerebral hemispheres. This rare pathology is caused by peculiarities of bilateral organization of hearing in cerebral cortex. The article presents a case of diagnosing cortical deafness confirmed with data of a clinical trial and magnetic resonance imaging (MRI) of the brain. It ought to be observed that accurate diagnosis was provided by prescription of the child's brain MRI despite the absence of specific indications for such an examination.

Keywords: *cortical deafness, symmetrical cerebral cysts, MRI diagnosis, viral encephalites, children.*

INTRODUCTION

True cortical deafness confirmed by visual MRI images of the cortical defect zone is rarely encountered in clinical practice. We deem necessary to publish this case, as it has the demonstrative potential, particularly, for educational purposes, and illustrates effectiveness of the multidisciplinary approach to detection of this rare pathology in children.

CLINICAL CASE DESCRIPTION

Boy D. aged 14. The child's parents, who are permanent residents of one of the Middle Asian countries sought outpatient neurological care in order to specify the adolescent's condition and determine the further treatment tactics. Anamnestic data obtained from parents and the child's medical record are scarce. The normally physically and mentally developing child had idiopathic meningoencephalitis at the age of 1 year 3 months. It is known that the pathology started with hyperthermia and convulsions; coma developed within 2 days. At admission to the inpatient hospital, the child was immediately taken to the resuscitation unit, where he remained for 40 days. No information on the treatment; the boy was discharged from the inpatient hospital with diagnosis of meningoencephalitis. At discharge from the inpatient hospital, the child was unable to turn, crawl, walk and speak, was almost unable to understand speech when addressed, i.e. he lost all the previously acquired (in the process of ontogenesis) psychomotor skills. The boy was later subjected to rehabilitation courses (drug treatment, massage, exercise therapy), in the setting whereof motor functions and speech understanding when addressed partially recovered; however, the child has remained unable to speak.

Neurological examination data

Positive reaction to the examination, difficulty in establishing contact due to the child's unfamiliarity with Russian. No meningeal or non-focal cerebral symptoms. Undisturbed sleep.

Cranial nerves. Right mouth angle downturn, pseudobulbar palsy (dysarthria, dysphagia).

Locomotion. Awkward gross motor skills; the child is able to walk unassistedly, pulls the right leg under; residual (pyramidal) changes; slightly heightened muscle tone, especially on the right-hand side; reinforced tendon reflexes; diminished muscle strength in hands; awkward completion of a finger-to-nose test; impaired fine motor skills.

Psychoemotional sphere. The child communicates with gestures, is unable to speak; difficulty understanding complex constructions translated by parents.

Anamnestic and examination data doubtlessly indicated a classic residual neurological pathology characterized by:

- acute organic affection of the brain in the onset (in this case – at an early age);
- no disease progression in the rehabilitation period and later; slow varying recovery of the impaired functions.

In such a situation, practical value of the neurological consultation consists in determination of rehabilitation potential and correction of the rehabilitation pattern. The more time has passed since the development of acute condition, the more obvious is the prognosis: in this case, twelve years of recovery is quite enough for accurate conclusions.

Therefore, the need in an additional expensive laboratory-instrumental study, such as brain magnetic resonance imaging (MRI) was not immediately obvious, as prognosis and rehabilitation are almost never determined by neuroimaging data in such situations. However, as the patient had never completed brain MRI (including the acute phase), we decided to additionally examine brain using this technology.

MRI data

Symmetrical brain substance cystic transformation zones with a perifocal glial component are visualized in the postfrontal-superotemporal-anteroparietal segments of the large hemispheres. The lesional zones spread from deep white matter to cortex and affect the primary sensorimotor (rolandic) area and the auditory cortex. In the left hemisphere, the cystic transformation affects a slightly larger area, including the hippocampal area and the anterior mediobasal areas of the temporal lobe characterized by secondary enlargement of the left lateral ventricle and the corresponding convexital spaces. Hypoplasia of the cerebellar vermis characterized by enlarged large occipital cistern and development of an extracerebellar cyst.

Pic. 1 shows large symmetrical bilateral cysts affecting the adjacent segments of the superotemporal, anteroparietal and postfrontal areas. Pic. 2 shows how cystic transformation resulted in almost complete absence of the superotemporal matter, where cortical auditory centers and auditory speech centers ought to be localized.

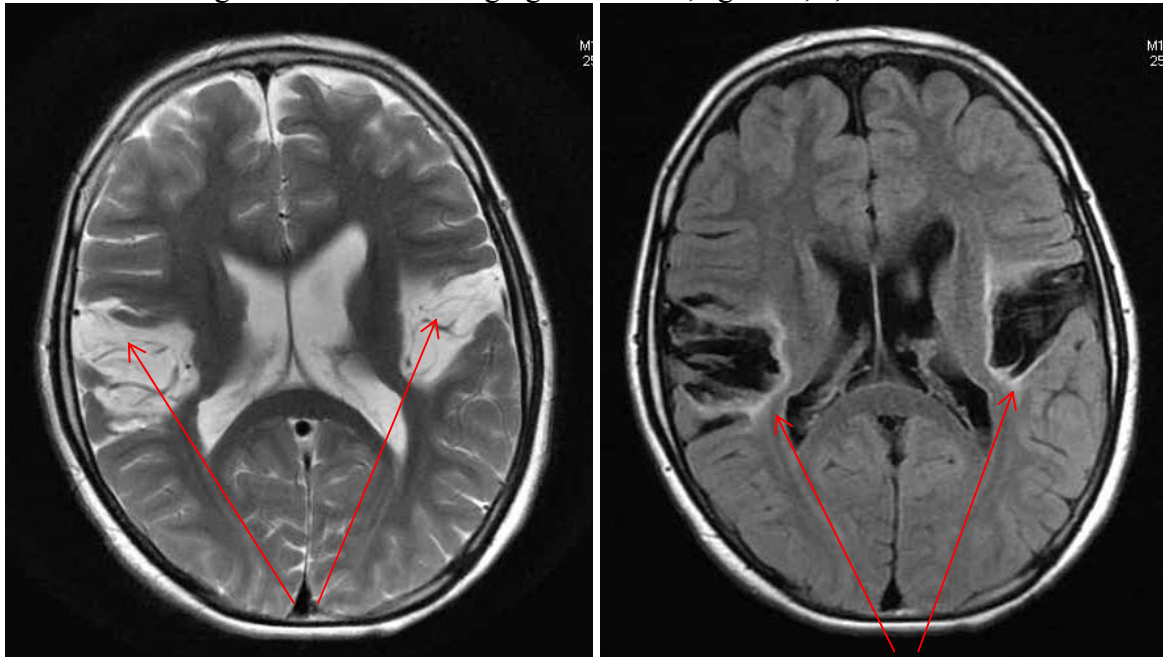
The child was appointed to audiological examination on the grounds of the detected affection of cortical auditory centers. Tympanometry (measurement of tympanic membrane mobility parameters) demonstrated type A tympanogram (norm). Ipsilateral acoustic reflex was registered in both ears. Delayed evoked otoacoustic emission was registered in both ears. These examinations ruled out pathology of the peripheral ear structures and of the cochlear outer hair cells.

The performed tonal threshold audiometry revealed deafness (pic. 3). The examination was based on the child's behavioral response, as he did not understand the task.

The detected deafness is cortical (due to cystic transformation of the higher brain centers of temporal lobes).

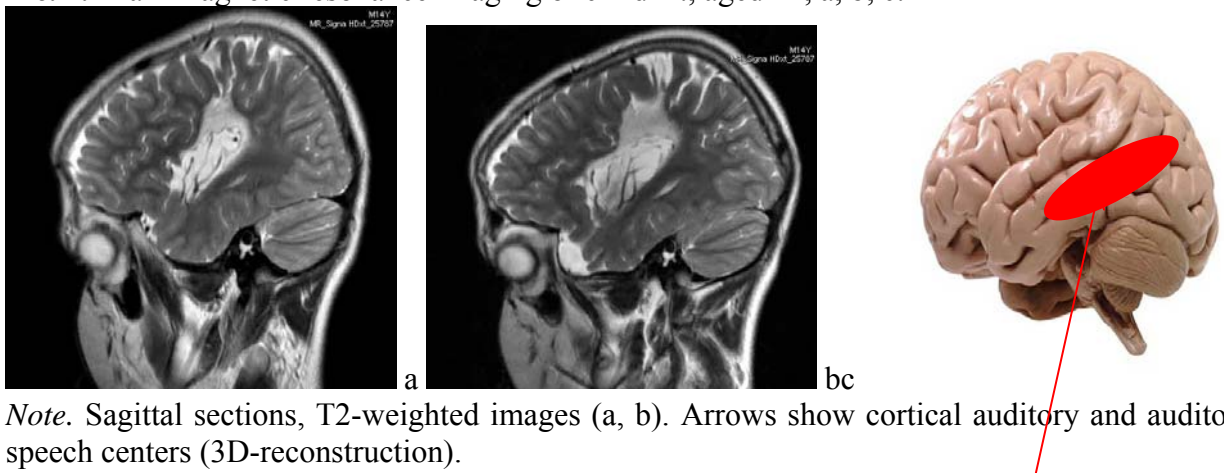
Cortical deafness plays a significant role in the development of intellectual incapacity in this patient: it results in gradual failure to understand oral speech, which, together with affection of adjacent speech centers of temporal, frontal and parietal lobes, predetermines failure of verbal logical thinking and inability to speak.

Pic. 1. Brain magnetic resonance imaging of child D., aged 14, a, b.



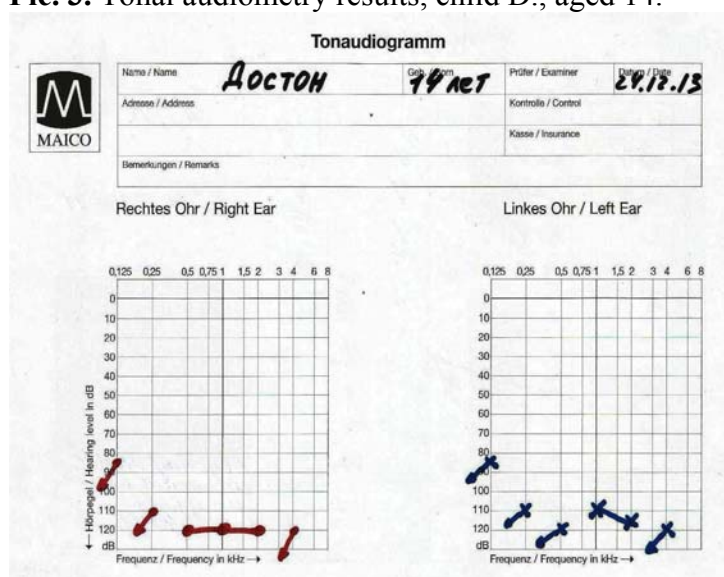
Note. Axial sections, T2- and FLAIR-weighted images. Arrows show cystic transformation of the brain matter (a), glial component (b).

Pic. 2. Brain magnetic resonance imaging of child D., aged 14, a, b, c.



Note. Sagittal sections, T2-weighted images (a, b). Arrows show cortical auditory and auditory speech centers (3D-reconstruction).

Pic. 3. Tonal audiometry results, child D., aged 14.



Neuropsychological examination

The boy understands almost nothing when addressed; is able to execute simple instructions explained with gestures. Expressive speech – individual sounds. Visual active thinking, kinesthetic praxis and imitation tests are infeasible. Gross and fine motor skills are only insufficiently developed. Programming, regulation and control of one's actions are underdeveloped. No interest in tasks and aptitude for learning. No interest in results of one's actions. Noticeable intellectual deterioration.

As is evident from the foregoing, alteration of higher cortical functions is not restricted to failure to understand oral speech and impaired verbal logical thinking caused by affection of the upper temporal segments; it is also manifested with consequences of cystic transformation of the adjacent anteroparietal, postfrontal and mediobasal segments of the left temporal lobe and hippocampus – impaired visual active thinking, kinesthetic praxis, programming, regulation and control of one's actions.

Laboratory diagnosis of the neuroinfectious condition's etiology was not performed, as it is inconclusive when performed long after the exacerbation. Analysis of hemostatic parameters performed to rule out high risk of acute cerebrovascular disease did not reveal any deviations. Taking into consideration analysis of the anamnestic data, available medical documentation and examination results, there is no reason to revise diagnosis of encephalitis as a causative agent of brain damage and functional impairment of the cortex. However, etiology of encephalitis remains unknown.

Clinical diagnosis

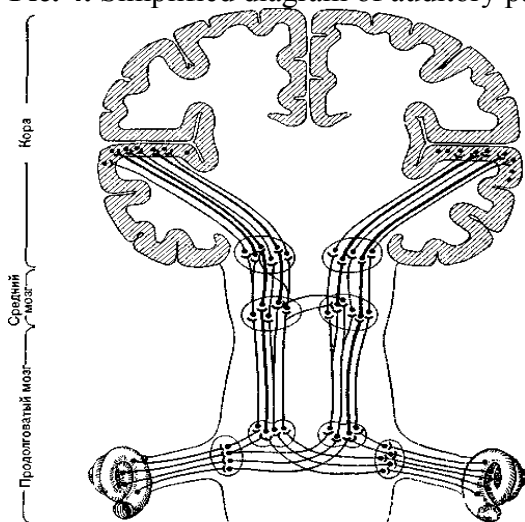
Consequences of meningoencephalitis: cortical deafness, dysarthria, dysphagia, bilateral upper motor neuron lesion, dyspraxia and intellectual incapacity.

Diagnosis of deafness will help to improve the child's quality of life and effectiveness of rehabilitation measures: education material presentation and instructing mechanisms will be revised; communication potential, ability to communicate and express oneself with gestures will improve. Overall, significance and intensity of the impaired cognitive functions by the age of 14 years indicate no chance of complete cognitive recovery of the adolescent. The following condition correction has been recommended to the child's parents on the basis of the examination results: sign language teaching at a specialized school for the deaf or a society of the deaf; neuropsychology and speech pathology sessions; exercise therapy; nootropic therapy course.

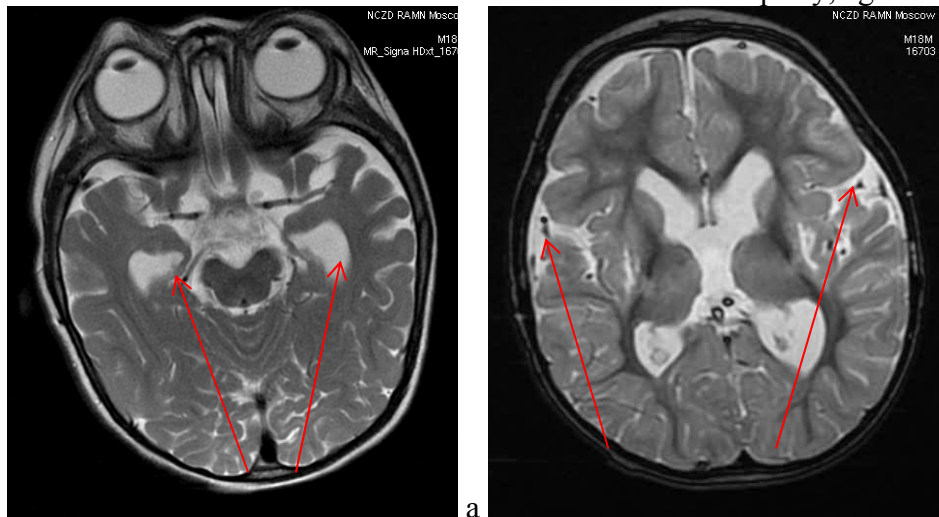
DISCUSSION

In this clinical case we encountered a rare case of classic cortical deafness diagnosis. It is well known that the primary auditory impulses simultaneously transmit from the periphery to cortical centers of both hemispheres (bilateral nature of the cortical auditory analyzer; pic. 4). That is why a relatively common unilateral affection of the cortical auditory center does not lead to deafness – only a partial loss of hearing may occur.

Pic. 4. Simplified diagram of auditory pathways from internal ear to auditory cortex [1].



Pic. 5. Selective neuronal necrosis in a child with cerebral palsy, aged 18 months, a, b.



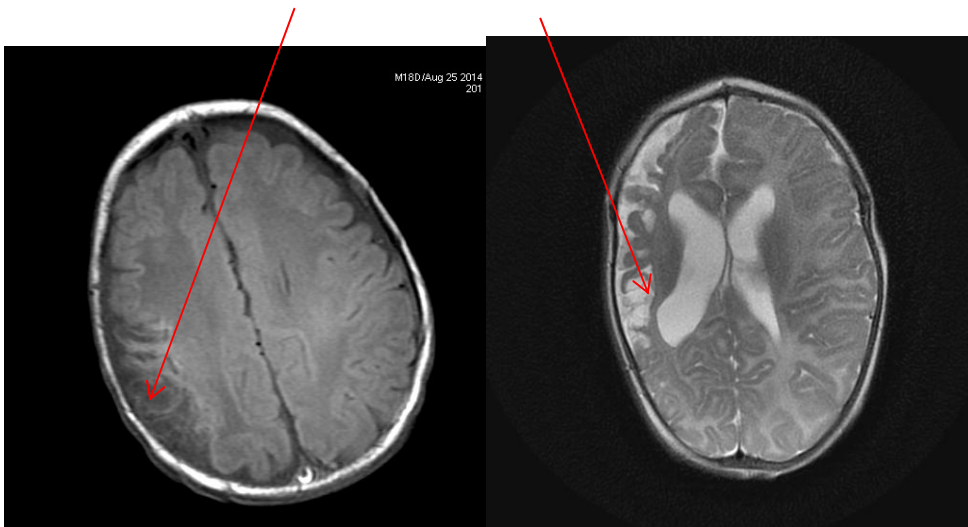
Note. Arrows show hippocampal areas (a), primary sensorimotor cortex (perirolandic areas; b).

Cortical deafness may develop only when both cortical auditory centers are simultaneously affected, which occurs rarely and determines exclusiveness of the given case. Literature review helped to reveal only one observation (by Canadian authors) of a patient with MRI-confirmed bilateral absence of temporal lobe segments, which resulted in cortical deafness [2].

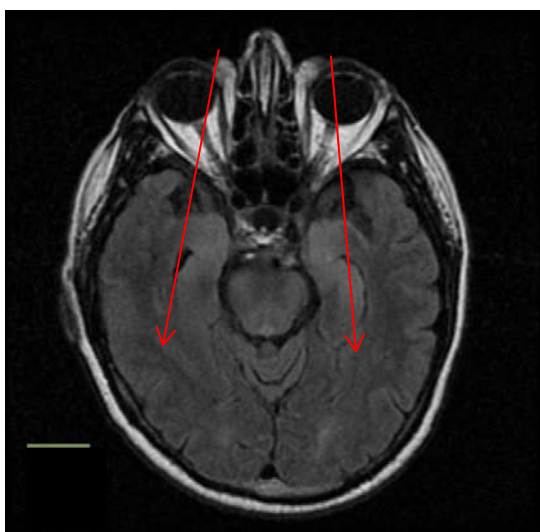
Cystic transformation of local symmetrical segments of brain hemispheres is indeed rare in radiodiagnosis. Symmetrical focal hemispheric damage may manifest selective neuronal necrosis in the zones of active myelinogenesis and synaptogenesis in the event of severe asphyxias in neonates; however, such changes are caused by a damaging factor in neonatality and restricted to the areas of hippocampus, basal nuclei and primary sensorimotor cortex (pic. 5).

Sharp-edged primary lesion focus is observed in older patients in the event of strokes or encephalites, which is why cystic transformation is local and asymmetrical (pic. 6). Dissemination of an acute process results in total brain lesion, not local symmetrical affection of certain areas. Symmetrical lesions are sometimes observed at viral (hepatic) encephalites [3, 4], but in such cases they are very limited in size (pic. 7) (unlike in the case under analysis). It is possible that this case was initially characterized by such a local symmetrical affection; however, ineffective therapeutic and resuscitation measures led to enlargement of the affected area (some anamnestic data suggest such a risk): thus, according to the parents, Haemodes was actively used for resuscitation; it is also known that the child remained at the resuscitation unit for more than 40 days.

Pic. 6. Focal cystic transformation in right hemispheres at ischemic stroke and meningoencephalitis (shown with arrows).



Pic. 7. Limbic encephalitis (shown with arrows) [F. Gaillard, Yu. Weerakkody et al. Herpes simplex encephalitis. Radiopaedia.org].



Conclusion

Thus, non-progressive course, slow recovery of the impaired functions and no suspicion of a neurosurgical pathology at a severe cerebropathy does not remove MRI from the diagnostic process if such a study has not been previously performed. In such a situation, MRI helps not only to specify the neuroanatomical affection substrate, but also (in some cases) to positively affect rehabilitation making it more effective. Presence of specialists in as many different fields as possible, availability of modern diagnostic equipment and a well-established pattern of specialists' interaction with various services at a medical center help to implement advantages of the multidisciplinary approach in practical pediatrics, one of which is the diagnostic range broadening for rare clinical cases.

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CONFLICT OF INTEREST

The authors have indicated they have no financial relationships relevant to this article to disclose.

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