CLINICAL MANIFESTATIONS OF PATIENTS WITH BLOCH-SULZBERGER SYNDROME FOLLOWED UP AT SREBRNJAK CHILDREN’S HOSPITAL IN 5-YEAR PERIOD

Matilda Kovac Szigorić*, M Kovac Szigorić, A Miculinic, R Gjergja Juraski, L Tambic Bukovac, D Erco, D Plavec, B Nogalo, G Tesovic, M Turkalj. Srebrnjak Children’s Hospital

Bloch-Sulzberger syndrome is a rare genetically determined dermatosis, manifested by skin changes in combination with...
Abstracts

The aim of this case report is to describe rare form of migraine in pediatric patient and to point out difficulties of making the right diagnose and classification according to ICHD-III (International Headache Society 2018.)

Case Report A 14-year old girl driven by emergency team to pediatric emergency from school where she had severe unilateral headache with nausea and paresthesia in the right hand and leg about an hour before arrival. At arrival she was confused, unknowing the name of her school and dyphasic. Neurological exam showed dysphasia and right hemiparesis, in the next half an hour she was somnolent, only opening her eyes when shaken and started vomiting.

After two hours (three from the first symptoms) all clinical signs vanished, she was in her full consciousness, ambulatory with no neurological deficits!

Relevance Vestibular Schwannoma (VS) – is a benign encapsulated tumor, initially arises in the inner auditory canal from the neurolemma of the vestibular nerve with further growth towards the base of the skull. Basically, they are associated with neurofibromatosis type 2 (NF2). In children sporadic cases of VS are uncommon. We report a case of VS in teenager.

Methods A 15-year-old girl with a viral infection was diagnosed with unilateral acute sensorineural hearing loss (SNHL). There was no NF2 family history. General medical examination revealed no ENT-pathology. MRI showed no tumor and vascular malformations in the brain and temporal region. After the standard course of dexamethasone effect was not obtained. SNHL progressed over the next 9 months. Evaluation of advanced hearing tests showed: tympanometry (probe-ton 226 Hz) demonstrated Type A bilateral, TEOAE were documented bilateral, pure tone audiometry determined profound hearing loss on the right, Auditory Brainstem Response (ABR) was defined wave V latency on the right, ABR thresholds were detected with intensity stimulus level 85 dB SPL on the right and 30 dB SPL on the left. At the series X-ray CT images of temporal region were determined inner auditory canal deformation and extension. A second MRI scan revealed a bulky tumor coming from the inner auditory canal. The

Abstracts

First Episode of Sporadic Hemiplegic Migraine in Pediatric Patient: Diagnostic Phantom

Marija Minković*, Dubravka Duraković Fejdić, Zrinka Erčić Hrvaćanin, Ivana Daković, Carmen Kndža, general hospital ‘Dr. J. Bendević‘ Slavonski Brod

The 7-year-old girl, appealed to the department in satisfactory condition, with complaints of mental deficiency, reduced memory, behavior disorders, multiple hyperpigmented spots on the limbs and body. The child after birth had multiple vesicles on an erythematous background on the body, localized linearly, mainly on the flexor surface of the limbs, as well as on the skin of the chest and back. After birth, he was diagnosed with progressive erythema and vesicular rash. In connection with skin manifestations, she was examined for intrauterine infections – pathology wasn’t detected. The patient had no family history of skin diseases. Subsequently, the appearance of hyperkeratoses in the form of plaques, warts and lichenoid growths along the Blashko lines was noted in the affected areas of the body. The girl was under the supervision of a pediatrician. During the examination in the Department revealed: mental retardation, hyperpigmented spots of gray-brown color with light edges, on the extremities of the rash in the form of ‘mud splashes’, on the trunk – in the form of a spiral,‘ ring ‘or’ marble cake’, with the location of pigmentation along the lines of Blashko.

The course of Bloch-Sulzberger syndrome is suspected in a child with clinical manifestations of mental retardation, stages of the skin process with a debut in the newborn period. To confirm the diagnosis, a molecular genetic examination was performed: mutations of the IKBKG gene, which is located on the X-chromosome, were detected.

Typical skin manifestations allow you to clinically suspect the presence of Bloch-Sulzberger syndrome in newborns, to conduct timely molecular genetic diagnostics to make the correct diagnosis. In view, the importance of differential diagnosis in the neonatal period and the rare disease, a multidisciplinary team of neonatologists, pediatricians and neurologists to this disease is necessary.

Relevance Vestibular Schwannoma (VS) – is a benign encapsulated tumor, initially arises in the inner auditory canal from the neurolemma of the vestibular nerve with further growth towards the base of the skull. Basically, they are associated with neurofibromatosis type 2 (NF2). In children sporadic cases of VS are uncommon. We report a case of VS in teenager.

Methods A 15-year-old girl with a viral infection was diagnosed with unilateral acute sensorineural hearing loss (SNHL). There was no NF2 family history. General medical examination revealed no ENT-pathology. MRI showed no tumor and vascular malformations in the brain and temporal region. After the standard course of dexamethasone effect was not obtained. SNHL progressed over the next 9 months. Evaluation of advanced hearing tests showed: tympanometry (probe-ton 226 Hz) demonstrated Type A bilateral, TEOAE were documented as ‘Pass’ bilateral, pure tone audiometry determined profound hearing loss on the right, Auditory Brainstem Response (ABR) was defined wave V latency on the right, ABR thresholds were detected with intensity stimulus level 85 dB SPL on the right and 30 dB SPL on the left. At the series X-ray CT images of temporal region were determined inner auditory canal deformation and extension. A second MRI scan revealed a bulky tumor coming from the inner auditory canal. The