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Meningitis and Ramsay-Hunt syndrome in a 17-year old girl

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Abstract

INTRODUCTION: Ramsay Hunt syndrome (RHS) is a rare manifestation of varicella-zoster virus (VZV) reactivation in geniculate ganglion. It usually manifests with a characteristic triad of symptoms including ipsilateral ear pain, vesicles in the external auditory canal, and facial nerve palsy.
CASE: We present a case report showing RHS additionally manifested by meningitis and involvement of VIII cranial nerve. Clinical course was complicated by acute kidney injury induced by acyclovir therapy.
RESULTS: Despite the involvement of the geniculate ganglion and vestibulocochlear nerve in the course of herpes zoster, and the occurrence of acute kidney injury, the patient fully recovered.
CONCLUSIONS: A complete recovery of cranial nerves VII and VIII in the course of RHS can be achieved.

Abbreviations:

VZV	- varicella zoster virus
RHS	- Ramsay Hunt syndrome
CT	- computed tomography
MRI	- magnetic resonance imaging
CBC	- complete blood count
CRP	- C-reactive protein
CSF	- cerebrospinal fluid
CMV	- cytomegalovirus
HSV	- herpes simplex virus
DNA	- deoxyribonucleic acid
PCR	- polymerase chain reaction
mg/dl	- milligrams per deciliter
µmol/l	- micromoles per liter

INTRODUCTION

After primary infection with varicella zoster virus (VZV), manifesting as varicella (chickenpox), a latent asymptomatic infection establishes in sensory ganglia of cranial and spinal nerves. Reactivation and replication of VZV results in herpes zoster, manifesting with vesicular rash in the involved dermatome. Various neurological complications may occur, the most common is postherpetic neuralgia (Kennedy & Gershon, 2018). Reactivation of VZV in the geniculate ganglion presents as herpes zoster oticus, with vesicles in the ear or in the oral cavity – on the palate or on the anterior two-thirds of the tongue (Wagner

CASE REPORT

et al. 2012). Herpes zoster oticus associated with ipsilateral ear pain and peripheral facial palsy is diagnosed as Ramsay Hunt syndrome (RHS). The course of the disease may be complicated by the involvement of further cranial nerves and spinal ganglia, preferentially the trigeminal and vestibulocochlear nerves, spinal ganglia C2-C4. Herpes zoster oticus and RHS, similarly to herpes zoster in general, are mainly diagnosed in adult patients. The risk of VZV reactivation increases with age, the highest incidence is in adults older than 80 years. However, herpes zoster may occur in childhood, especially if varicella was experienced in first years of life or a patient is immunocompromised.

In this case report we present a complicated RHS in a pediatric patient with a successfully achieved complete recovery.

CASE REPORT

On the 8th of August 2017, a 17-year old immunocompetent girl was admitted to the Department of Neurology, Central Clinical Hospital of Medical University of Warsaw with a 4-day history of severe headache and nausea, without fever and emesis. She was in good general condition. Physical examination revealed horizontal nystagmus and dysmetria of upper limbs. Imaging tests: CT, MRI, MRI angiography and venography, transcranial ultrasound with Doppler showed no abnormalities. Laboratory results (CBC, CRP, electrolytes, glucose level, liver and kidney function) were normal. Two days later, meningeal signs appeared. Lumbar puncture was performed, and cerebrospinal fluid (CSF) examination revealed pleocytosis (377 cells/mm^3) with predominance of lymphocytes (82%), slightly elevated protein level 57 mg/dl [N: 15–45], normal glucose level 49 mg/dl [N: 45–80] and negative results of testing for anti-Borrelia burgdorferi antibodies. Viral meningitis was diagnosed and DNA PCR testing CSF for VZV, CMV, HSV1 and 2 was ordered. The girl was transferred to the Department of Children's Infectious Diseases, Medical University of Warsaw. She complained of headache, but her general condition was quite good and no abnormalities were found on physical examination. Supporting treatment was applied. Next day positive result of VZV DNA PCR testing in CSF was known and antiviral therapy with intravenous acyclovir (Acix™) was implemented. On the same day, the patient complained of pain and hypoacusis of the left ear. Simultaneously, vesicles in the left external auditory canal appeared and peripheral left facial nerve palsy occurred. She was diagnosed with RHS. Dexamethasone (Dexaven™) was administered intravenously. Symptoms of meningitis, vesicular rash and earache resolved in 9 days, hypoacusis and left facial palsy persisted and the patient was discharged home, oral prednisone (Encorton™) was recommended. Two days later, she returned to the hospital due to severe vertigo and vomiting. Physical examination revealed horizontal nystagmus and positive Romberg test. Left facial palsy and hypoacusis were still present. The patient was consulted by a neurologist, brain CT was performed (no abnormalities were found) and she was diagnosed with dysfunction of cranial nerve VIII in the course of RHS. Intravenous acyclovir and dexamethasone were administered again. In spite of treatment vertigo and vomiting persisted, but function of left facial nerve improved. Three days later the girl developed acute kidney injury (creatinine level was 168 μmol/l [N: 46–92]), which was interpretated as a side effect of acyclovir, so the antiviral therapy had to be withdrawn. Kidney injury manifested with hypertension that was treated with amlodipine (Amlozek™). Kidney function normalized in 3 days; then brain and temporal bone gadolinium-enhanced MRI was performed, it showed no abnormalities. During following days vertigo and vomiting subsided. After 17 days the girl was discharged home with normal kidney function and trace dysfunction of left cranial nerves VII and VIII. After a month she presented with a complete recovery.

DISCUSSION

The presented patient had typical symptoms of RHS with vesicular skin lesions in the left ear, ipsilateral earache, hypoacusis and facial palsy. Involvement of the vestibulocochlear (cranial VIII) nerve, manifesting with vertigo and vomiting, is also quite often observed. It may be explained by a close anatomic location of the vestibulocochlear nerve to the geniculate ganglion (Sweeney & Gilden, 2001). In our patient it occurred surprisingly after the antiviral treatment. Additionally, meningitis is not frequent complication of RHS and in our patient it was the first presentation of the disease. It could be a result of retrograde axonal spread of VZV after reactivation in the geniculate ganglion.

Damage of the involved cranial nerves VII and VIII can be shown by gadolinium-enhanced MRI (Chung *et al.* 2015). In our patient the result of imaging was normal, but it was performed rather late in the course of the disease, when clinical improvement was observed.

The facial nerve palsy in the course of RHS is described as more severe than Bell's palsy, the prognosis for the affected nerve is uncertain and complete recovery is uncommon (Monsanto *et al.* 2016). Early diagnosis and treatment with combination of antivirals and corticosteroids was reported as a factor influencing the prognosis. A combined therapy is recommended (Coulson *et al.* 2011). In the described case a complete recovery was observed, although the clinical course was complicated by acute kidney injury. The most probable cause was acyclovir therapy (Yildiz *et al.* 2012), therefore it was interrupted. The complete recovery may result from the young age of the patient, the absence of accompanying chronic diseases or lack of immunodeficiency, which promotes proper nerve regeneration during convalescence. During antiviral treatment it is

important to properly hydrate the patients, because acyclovir may crystalize in renal glomeruli and lead to acute kidney injury. It may also occur in patients with proper fluid supply, thus monitoring kidney function during the antiviral therapy is crucial.

CONCLUSION

Although prognosis for normal function of cranial nerves VII and VIII in the course of RHS is uncertain, complete recovery may be achieved.

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