CASE REPORT

Bilateral Ramsay Hunt Syndrome in a Pediatric Patient

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Bilateral facial paralysis is an unusual clinical entity that occurs in less than 1% of patients with facial paralysis. In children, bilateral facial paralysis is even rarer. The etiological assessment can be difficult. Herpes Zoster can cause serious infections, bilateral facial paralysis and Ramsay Hunt syndrome in pediatric patients. In this paper we report the first case in the literature according to our knowledge, with bilateral facial paralysis due to Ramsay Hunt syndrome in a pediatric patient.

Pediatric bilateral facial paralysis is a rare clinical entity with an exhaustive differential diagnosis. In both adults and children, Lyme disease, trauma, and idiopathic causes are most commonly reported; however, in children, acute otitis media and congenital facial diplegia are also important in the differential diagnosis [1]. Herpes Zoster can cause severe infections in pediatric patients and should be mentioned in the etiology of bilateral facial paralysis (BFP) in children.

Varicella-zoster virus, member of Herpes viridae family has structural characteristics like a lipid envelope surrounding a nucleocapsid with icosahedral symmetry, a total diameter of 180-200 nm and centrally located double-stranded DNA. Varicella-zoster virus lies latent in sensory root ganglion for years in a patient who had chickenpox earlier. Some precipitating factors may reactivate it especially when immunity of patient wanes leading to Herpes zoster, a sporadic disease. Involvement of geniculate ganglion of sensory branch of facial nerve leads to Herpes zoster oticus also known as Ramsay Hunt syndrome [2].

In this paper we present bilateral facial paralysis due to Ramsay Hunt syndrome in a 7-year-old male patient who was treated with systemic steroid and antiviral medications and in whom an excellent result was achieved.

CASE REPORT

A 7-year-old boy with an unremarkable medical history attended to our clinic with left sided immobility in his face, pain in both ears and generalized vesicular eruptions in his lower and upper lips. The symptoms were progressive and had begun two days prior the admission to the hospital. On the day of admission to the hospital the patient's mother noted that his face was bilaterally immobile. On physical examination he had bilateral painful and sensitive auricle and external canals but normally appearing tympanic membranes. Vesicular lesions were widespread on lower and upper lips with adherent crusts and forming groups. Bilateral House Brackmann Grade V facial paralysis was observed. On further questioning we've revealed that, a brother who was born a month ago and this was assumed to be a stress factor. Tuning fork tests and pure tone audiometry were normal. Acoustic reflexes were absent on both sides. The remainder of the neurological and physical examinations was noncontributory.

Computerized tomography findings of brain stem, cerebellopontine angle, temporal bone and skull base were normal. However in magnetic resonance imaging (MRI) with gadolinium contrast we observed an abnormal enhancement in facial nerve within distal internal auditory canal, geniculate ganglion and tympanic segment. A smear from floor of vesicle stained with Giemsa stain showed degenerating cells with multiple nuclei. This favored the clinical diagnosis of Herpes Zoster oticus. This diagnosis was confirmed by detection of IgM antibodies to Varicella-Zoster virus by ELISA test.

Acyclovir was given in dose of 10 mg/kg every 8 hr for 7 days and prednisolon acetate was administrated in daily doses of 1/mg/kg for 3 days and tapered 10 mg in every three days from intravenous route. Progressive recovery had been observed after the fifth day of treatment and at the eleventh day, facial nerve functions on both sides were Grade II on House Brackmann scale. Vesicular eruptions were recovered well with daily dressings with topical rifampicine.

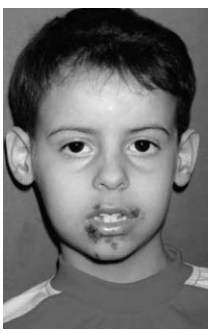

Figure-1: Patient at time of presentation, bilateral Ramsay-Hunt syndrome with presence of Bell's phenomenon and vesicular lesions with adherent crusts.

DISCUSSION

In the evaluation of a child with bilateral facial nerve paralysis, history plays an important role in the diagnosis. Also the otologic symptoms, including ear pain, drainage, hearing loss and mastoid tenderness are of primary importance [2]. The differential diagnosis of BFP is difficult. A standardized diagnostic work-up designed to investigate every possible etiology has been advocated in the past but this approach is no longer practical as there are too many diseases to test and this costs too much [3,4]. Fortunately, as in the case we presented above, usually there are specific findings in history or examination which make diagnosis easier. Detection of IgM antibodies to Varicella-Zoster virus by ELISA test confirmed our preliminary diagnosis. The patient was also under severe psychological stress since one month and psychological stress is identified as a potential risk factor for zoster that might suppress cell-mediated immunity. Psychological stress was the sole risk factor present in this patient, making him prone for severe and recurrent infection of Herpes Zoster.

The first and the most important part of the diagnostic work-up for BFP is therefore a thorough history and physical examination. A neurologist should be consulted as soon as possible to assist in this evaluation. The history should be directed toward eliciting symptoms of the diseases in the differential diagnosis list. As may be seen in our case, the history should lead the clinician to an individually designed work-up, directed to the diagnoses of highest clinical suspicion thereby eliminating low yield evaluations.

MRI images in patients with Ramsay Hunt syndrome usually have been reported in literature as contrast enhancement of the seventh and eighth nerve trunks within the distal internal auditory canal and along the labyrinthine segment as well as enhancement of the cochlea, vestibule, and parts of the semicircular canals [5]. Also intense enhancement of the geniculate ganglion; the tympanic and mastoid facial nerve segments, and the blister lesions of the external auditory canal are evident [6]. No correlation was found between intensity, extension, and duration of the enhancement and the clinical and intraoperative findings, as well as with electroneuronography in facial palsy but it is still useful in differential diagnosis [7]. In our case, we observed similar MRI findings showing abnormal enhancement in facial nerve within the distal internal auditory canal, geniculate ganglion and tympanic segment.

Recent reports suggest that treatment with intravenous acyclovir in addition to systemic steroids decreases the risk of permanent facial nerve palsy in Ramsay Hunt Syndrome. Results of relevant control trials have not yet been published (2). On follow up of this patient, facial nerve recovered on both sides, and it was more apparent on the right side. This was documented by electroneuronography which showed greater amplitude of muscle compound action potentials on right side as compared to left. This might have been due to the onset of acyclovir therapy and attack of zoster oticus on the right side. The time interval between the onset of acyclovir therapy and attack of zoster oticus is the most relevant prognostic factor in recovery of these patients and this hypothesis is supported by findings in studies of Mcgrath N. (1997) [8], Raschilas F (2002) [9] and Strupp M. et al (2004) [10].

CONCLUSIONS

Herpes zoster can cause serious infections like bilateral Ramsay Hunt syndrome in pediatric patients. It should be treated as promptly as possible with systemic steroids and intravenous acyclovir. Intravenous acyclovir therapy in cases of herpes zoster oticus is effective in control of disease and prevents the risk of permanent facial palsy. Treatment should be started early in the course of disease preferably within 72 hours from the start of disease. However we have to keep in mind that in Ramsay-Hunt syndrome the recurrence can be seen as the viruses are generally located in geniculate ganglion.

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