

SEVERE NEUROLOGICAL COMPLICATIONS OF CHICKENPOX

Report of four cases

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Neurological complications caused by chickenpox are estimated as approximately 0.01%-0.03%. Frequent complications related to central nerve system involvement are cerebellar ataxia and encephalitis, and rare complications are transverse myelitis, aseptic meningitis, Guillian-Barré syndrome, meningoencephalitis, ventriculitis, optic neuritis, post-hepatic neuralgia, herpes zoster ophthalmicus, delayed contralateral hemiparesis, peripheral motor neuropathy, cerebral angitis, Reye syndrome and facial paralysis. In present study, additional four cases were presented who diagnosed as chickenpox within one year and developed neurological complications. Cerebellar ataxia developed in two of our cases while cerebellar ataxia plus encephalitis was present in one case and peripheral type facial paralysis in the other.

Key words: Chickenpox; Neurological Complications; children

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INTRODUCTION

Chicken pox is mostly seen in pre-school and school-age children. The primary infection is characterized by generalized vesicular dermal exanthemas which are extremely contagious. After the primary infection period, the virus may stay latent in the spinal and cranial ganglia neurons, and then become reactive after the latent term resulting in a herpes zoster infection.

Neurological complications caused by chicken pox is estimated approximately 0.01%-0.03%. Complications related to the central nerve system are frequently seen in Chickenpox. Cerebellar ataxia and encephalitis are seen frequently; while transverse myelitis, aseptic meningitis, Guillian-Barré syndrome, meningoencephalitis, ventriculitis, optic neuritis, post-hepatic neuralgia, herpes zoster ophthalmicus, delayed contralateral hemiparesis, peripheral motor neuropathy, cerebral angitis, Reye syndrome and facial paralysis rarely can be observed (1-3).

In the present study, four rare cases are presented who received diagnosis at our clinic within one year and developed neurological

complications due to chicken pox. Cerebellar ataxia was present in two of the cases, while cerebellar ataxia together encephalitis was seen in one case and peripheral type facial paralysis was present in the other.

CASE 1

A 9.5 years old girl was referred to our clinic with exanthema, failure to stand and tremor. We learned that a week ago itchy, liquid lesions appeared in her body accompanied with symptoms including failure in sitting, unable to walk and tremors. Physical examination of the skin revealed dried chickenpox lesions spread throughout the dermal layer. During neurological examination we observe that the girl was unable to sit and stand and had tremor in her hands. Deep tendon reflexes (DTRs) were vivid. Other organ system examinations gave no abnormality. Serum Varicella IgM was positive. Cerebrospinal fluid (CSF) study demonstrated leucocytes ($55/\text{mm}^3$) with normal protein and glucose levels. No microorganism growth in the CSF culture. Brain magnetic resonance imaging (MRI) and computerized brain tomography (CT) studies were found to be normal.

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A diffused slow wave activity was detected in the patient's electroencephalography (EEG). The patient was hospitalized with a diagnosis of encephalitis and cerebellar ataxia and treatment with acyclovir was initiated. The patient's general condition totally improved within 10 days and was discharged from the hospital with a recommendation to come for control visits.

CASE 2

A 8 years old boy referred to our clinic due to complaints (unable to walk and vomiting). We were informed that, he had chicken pox 15 days before referral of him to our hospital and was experiencing difficulty in walking and had vomiting attacks for 5 days. Neurological examination displayed an ataxia gait accompanied with dysarthric speech. Cerebellar tests gave bilaterally abnormal results. CSF study revealed no abnormality. The patient was diagnosed as cerebellar ataxia secondary to chicken pox and a supportive therapy was initiated. The patient improved and was discharged from the hospital 6 days after hospitalization.

CASE 3

A 4 years old girl was referred to our clinic due to complaints such as fever, exanthema and unable to walk. We were informed that, a week ago the patient was referred to a physician because of symptoms of itchy, fluid lesions which began from her trunk and then covering her whole body. The doctor prescribed aspirin and another drug which they did not know its brand name, and two days later the patient suffered from severe headaches and was unable to walk. Neurological examination displayed an ataxia gait while cerebellar ataxia tests were bilaterally abnormal. Serum varicella IgM was positive. CSF study was normal. Brain MRI was within normal limits. The patient was diagnosed as cerebellar ataxia secondary to chicken pox and supportive therapy was initiated. The patient significantly improved and was discharged from the hospital 6 days after hospitalization.

CASE 4

A 7.5 years old boy applied to our clinic due to symptoms of left peripheral type facial paralysis. The medical history of the patient demonstrated that, the boy experienced chicken pox a month ago and a peripheral type facial paralysis occurred 15 days ago. Physical examination revealed findings

related to peripheral type facial paralysis while the child's body was totally covered with hyperpigmented maculae resulted from an ex-chicken pox. Brain MRI revealed no abnormality. Facial paralysis was accepted as a complication of chicken pox and acyclovir treatment was initiated. During follow-up period facial paralysis completely healed.

DISCUSSION

Neurological complications secondary to chicken pox are very scarce; commonly they may occur during or after exanthemas. Neurological findings are less frequent prior the exanthemas occurs. Cerebellar ataxia, encephalitis and rarely, facial paralysis can be seen as neurological complications of chickenpox. The most serious complication that involves the central nerve system secondary to chicken pox is encephalitis. Encephalitis, the most serious CNS complications of varicella, has an incidence of 1-2 episodes per 10,000 varicella cases, with the highest incidence in adults and infants (1). The role of active varicella-zoster virus (VZV) replications in the pathogenesis of chicken pox encephalitis is not yet defined. However, it is reported that post-infectious demyelination or direct viral cytopathology may play a significant role. Cerebellar ataxia may also accompany the disease picture but in the majority of cases, this condition is seen alone. Fever, headache, vomiting, and change in general condition and convulsions may present a week after the exanthema appearance. Neurological examination generally reveals ataxia, hypertonia or hypotonia, hyporeflexia or hyperreflexia, positive plantar response, hemiparesis and sensorial disturbances. Pressure is increased in CSF findings with a mild to moderate degree of lymphocytosis, a slight degree of increased protein content and normal level of glucose. The EEG of the patients displays a diffused slow wave activity. Cranial imaging may display edema. Mortality is approximately 5-10% while the ratio of convulsion between long-term sequels is 10-20% (1,3-5).

In the present study, among four of our patients who developed neurological complications due to chicken pox, only two patients suffered from cerebellar ataxia, one patient from encephalitis + cerebellar ataxia and the other experienced left facial paralysis. In our first patient neurological symptoms occurred at the 4th day of the eruptions. Serum Varicella IgM was positive and CSF study contained 55 leucocytes. A

positive response was received from acyclovir treatment in first patient who had encephalitis plus cerebellar ataxia. In our second and third cases cerebellar ataxia alone developed.

Cerebellar ataxia occurs in about 1 in 4000 varicella cases (1). Cerebellar ataxia is more frequent in nine and six years old children. Prognosis is better if encephalitis is absent. However the pathogenesis of the condition is not completely well known, but a direct viral invasion of the cerebellum or an immunological status is considered to play a role in the pathogenesis of ataxia. The presence of specific antibodies against VZV indicates a replication of VZV in CNS. Findings related to ataxia are rare; however findings may occur prior to exanthemas or two weeks after eruptions. Headache, vomiting, lethargy, stiffness in the neck and nystagmus may occur. CSF findings are usually normal, but a mild degree of lymphocyte pleocytosis and increased content of protein may accompany the condition. The majority of the patients improve within 1-3 weeks without any sequel (1,3,4).

Facial paralysis that develops as a neurological complication of chicken pox is reported as a rare condition in the literature. In our 4th case, peripheral type left facial paralysis developed 15 days after exanthemas of chickenpox. However, due to insufficient facilities our laboratory we did not have the opportunity to investigate the existence of Varicella Zoster Virus IgG and IgM. Oral acyclovir treatment (20 mg/kg/day, once at per 6 hours) accompanied with a supportive therapy was administered for a period of five days. Facial paralysis in chicken pox can be seen prior or after exanthemas. It may also occur five days before chicken pox exanthemas appears and during the 16 day period after chicken pox is diagnosed. Hematogenesis which is observed during the pre-exanthema term follows a neural pathway after exanthema occurs.

In a study where 60 cases with neurological complications due to chicken pox were assessed the ratios of encephalitis/encephalopathy was determined as 23.3%, cerebellar ataxia 21.7%, meningitis as 8.3%, cerebral infarcts as 13.3% and facial paralysis as 8.3%. The frequency of facial paralysis is reported as 7-10% among all causes of non-traumatic paralyzes. The symptoms in patients who developed facial paralysis due to chicken pox totally responded to the therapy at the first three days while it was unable to obtain a total improvement in cases

who received therapy three days after the condition occurred (3,6,7).

Consequently, as use of vaccine against Chickenpox in our region is not a routine procedure of childhood vaccination practice, we would like to emphasize that neurological complications of chickenpox will continue to be seen.

REFERENCES

1. Gnann JW. Varicella-Zoster virus: atypical presentations and unusual complications. *J Infect Dis* 2002; 186: 91-98.
2. LaGuardia JJ, Gilden DH. Varicella-zoster virus: a re-emerging infection. *J Invest Dermatol Symp Proc* 2001;6:183-7
3. Shiihara H. Neurological complications of varicella-zoster virus (VZV) infection. *No To Hattatsu* 1993;25:128-34
4. Gershon AA, Larussa P. Varicella-Zoster virus infections. In: Samuel L.Katz, Anne A. Gershon, Peter J. Hotez (eds). *Infectious Diseases of Children*. St. Louis: Missouri, 1998, pp 620-49
5. van der Flier M, van Koppenhagen C, Disch FJ, et al. Bilateral sequential facial palsy during chickenpox. *Eur J Pediatr* 1999, 158; 807-8
6. Albeck H, Ninn- Pedersen K. Acyclovir in the treatment of facial palsy due to zoster virus. *Ugeskr Laeger* 1989; 151: 90-2
7. Deda G, Çaksen H, İçağasıoğlu D, İnce E. A case of chickenpox associated with facial nerve palsy. *Pediatr Dermatol* 2002; 19:95-6