

ACUTE CEREBELLAR ATAXIA AS A COMPLICATION OF VARICELLA IN FOUR CHILDREN*

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Varicella is usually a mild and self-limiting disease associated with few complications. Although it has been reported that encephalitis is seen in 0.10% to 0.26% of the cases of varicella, acute cerebellar ataxia is much more rare. The pathogenesis of acute cerebellar ataxia caused by varicella has not as yet been explained¹⁻³. The purpose of this article is to present four cases of acute cerebellar ataxia caused by varicella, which as far as we know is the largest sampling taken from our region.

Case Reports

Four patients with cerebellar ataxia and varicella were examined at Hacettepe University Children's Hospital. Three of the children were five-years-old and one was nine-years-old. There were two males and three females. The children were admitted to the hospital with symptoms of vomiting, tremor and ataxia of one or two days duration. They had all been in good health one week prior to admission when they developed chickenpox with typical skin lesions. On the fifth or sixth day of the eruption, they began to complain of vomiting, muscular weakness and incoordination of the lower extremities. The symptoms rapidly progressed and on the day of admission they were unable to sit, stand or walk.

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All the systemic and neurologic findings of the patients were normal except for abnormal cerebellar findings. Hyperactive deep tendon reflexes were determined in two patients.

In all the patients routine complete blood count, urinalysis and cerebrospinal fluid findings were within normal limits. Skull x-rays revealed no abnormalities. EEG examination which was performed on only one patient revealed 5-6 cycl/sec theta and 7-8 cycl/sec alpha activity. Computerized tomograms of the brain were normal in three subjects and slight brain edema was found in one.

All the clinical findings of the patients gradually disappeared and they were discharged in good health without any neurologic sequelae on the seventh to twelfth day after their admission.

Some of the clinical and laboratory findings of the patients are summarized in Table I.

Discussion

Varicella usually runs its normal course and recovery is usually without complication. However, in five percent of the cases, local bacterial infections such as otitis media, skin abscess, suppurative lymphadenitis, pyoderma, cellulitis and erysipelas occur⁴.

The incidence of neurologic complications recorded at 0.2 percent is much more rare^{1,2,4,5}. These include cerebellar ataxia, meningoencephalitis, disseminated or transverse myelitis, acute ascending spinal paralysis, polyneuritis, optic neuritis, and paralysis of the cranial nerves. The average latent period for encephalitic symptoms to occur is six days after the exanthem appears, however in some cases, it is three to four days before the exanthem. The symptoms which may be gradual or sudden in onset frequently include fever, headache, vomiting, meningismus and drowsiness. Other symptoms occasionally noted during the illness include tremor, chorea, urinary retention or incontinence, and pupillary changes⁴⁻⁸. The majority of the neurologic complications subsides within one or two weeks. Encephalitis has a mortality rate of 5-10 percent^{5,8} and some 15 percent of the cases are left with permanent neuropsychiatric disabilities such as mental retardation, palsies and blindness⁸. There is a correlation between the severity of the encephalitis, cerebrospinal fluid findings and the ultimate prognosis, but convulsions are unfavorable signs¹.

In some cases of varicella with unassociated signs of encephalitis, an acute ataxic syndrome occurs five to ten days following the appearance of the eruption or rarely, even before the rash is noted¹⁻³. Truncal ataxia resulting in a markedly incoordinated gait is often the only sign of neurologic dysfunction. In others, the

TABLE I: Some Clinical and Laboratory Findings of Patients with Acute Cerebellar Ataxia and Varicella

Age	Sex	Presenting symptoms	Appearance of presenting symptoms	Appearance of skin lesions of varicella	Main neurologic findings	Duration of neurologic findings	Cerebrospinal fluid Appearance	Pressure	Protein	CSF/Blood glucose	Cells	EEG	Cranial X-ray	Computerized tomography of brain
5	F	Ataxia Tremor Vomiting Lethargy Headache	2 days ago	7 days ago	Cerebellar, Hyperactive DTR	11 days	Clear	90 mmH ₂ O	36 mg/dl	69/135 mg/dl	11 lymphocytes/mm ³	5-6 cycl/sec theta and 7-8 cycl/sec alpha activity	Normal	Normal
5	M	Ataxia Tremor Vomiting Lethargy	1 day ago	7 days ago	Cerebellar, Hyperactive DTR	12 days	Clear	70 mmH ₂ O	72 mg/dl	80/75 mg/dl	none	-	Normal	Normal
										85/90 mg/dl	11 lymphocytes/mm ³	-	Normal	Normal
										59/106 mg/dl	none	-	Normal	Normal
5	M	Ataxia Tremor	1 day ago	7 days ago	Cerebellar	8 days	Clear	160 mmH ₂ O	24 mg/dl					
9	F	Ataxia Vomiting Headache	2 days ago	8 days ago	Cerebellar	7 days	Clear	110 mmH ₂ O	36 mg/dl					

cerebellar signs are preceded by headache or vomiting and are combined with lethargy, irritability and evidence of increased intracranial pressure. The spinal fluid in these patients is usually clear. Although the pressure is generally normal, it is sometimes moderately increased, and the cell count is slightly increased⁷.

Acute ataxic syndrome is transient in most cases with complete recovery after a week or so¹⁻³. The four cases presented presumably had acute cerebellar ataxic syndrome caused by varicella as demonstrated by their clinical and laboratory findings. It is important to make a definite differential diagnosis from other causes of acute cerebellar ataxia, for example nystagmus is quite rare in acute ataxic syndrome caused by varicella.

Since complete recovery from acute cerebellar ataxia caused by varicella occurs within a few days or weeks, differentiation must be made between acute ataxic syndrome and meningoencephalitis resulting from varicella and other acute cerebellar disorders in order to predict the ultimate diagnosis of these disorders.

Summary

Four cases of acute cerebellar ataxia caused by varicella are presented and the related literature is discussed. Three of the subjects were five years of age and one was nine years of age. There were three females and two males. The patients presented with symptoms of vomiting, tremor and ataxia approximately five or six days after the appearance of typical varicella lesions. Cerebrospinal fluid, EEG, cranial x-ray and computerized tomography findings were all within normal limits. All four patients recovered completely within seven to twelve days after the appearance of neurologic findings without any sequelae.

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