

Temporary Consciousness Disturbance in Van der Knaap Disease after Minor Head Trauma

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INTRODUCTION

Van der Knaap disease, also known as megalencephalic leukoencephalopathy with subcortical cysts (MLC), is a slowly progressive neurodegenerative disease. The disease is characterized by macrocephaly, mild gross motor developmental delay, gradual onset of ataxia, spasticity, dysarthria, dystonia, and sometimes extrapyramidal findings. A defining trait of this illness is macrocephaly, which commonly manifests at birth or throughout the first year. The disease is known to have a slow progression. During childhood, motor and intellectual functions remain mostly preserved despite diffuse white matter involvement

In this report, a 10-year-old girl with van der Knap's disease, which became comatose after minor head trauma and gradually improved, is presented.

CASE PRESENTATION

A 10-year-old patient, who was followed up with the diagnoses of epilepsy and Van der Knaap disease, presented with deterioration of consciousness after minor head trauma

She only had a response to painful stimuli and did not respond to verbal stimuli. She needed nasogastric tube for feeding but no respiratory support. After head trauma, the patient underwent a non-contrast CT scan that showed prominent diffuse white matter hypodensity (Figure 1A). Her cranial magnetic resonance imaging showed showed diffuse bilateral symmetrical white matter hyperintensities (Figure 1B). EEG showed slow background activity with epileptic charges from right temporal legion. The patient was using sodium valproate (30 mg/kg/day) and she had no seizures about two years. Her comatose status lasted for over two months, and then gradually resolved. Eight months after the trauma, the patient started walking independently for short periods of time. She has dysarthric speech.

DISCUSSION

Van der Knaap disease is a rare kind of leukoencephalopathy first reported in mid '90s characterized by macrocephaly from infancy, white matter involvement, clinically manifested as mild neurological symptoms.

The disease is known to have a slow progression. Mental function deteriorates slower than motor function and patients usually maintain their cognition despite severe motor deterioration. Almost all children have epileptic seizures in early childhood (range between 1,5 and 14 years) which are usually precipitated by a minor head trauma and sometimes seizures fascilitate the deterioration of clinical course

During clinical course of van der Knaap syndrome, slow progression period may precede a subsequent period with worsening of the neurological condition. After clinical worsening, some patients may have a stable period or even have tendency to improve. Head trauma or febrile diseases may also cause clinical worsening, Bugiani et al. presented a case of van der Knaap who developed onsciousness disturbance after minor head trauma and recovered after four months. Our patient also developed coma after minor head trauma. Over time, the patient's clinic recovered. Even after eight months from the trauma, a fully independent walking could not be achieved.

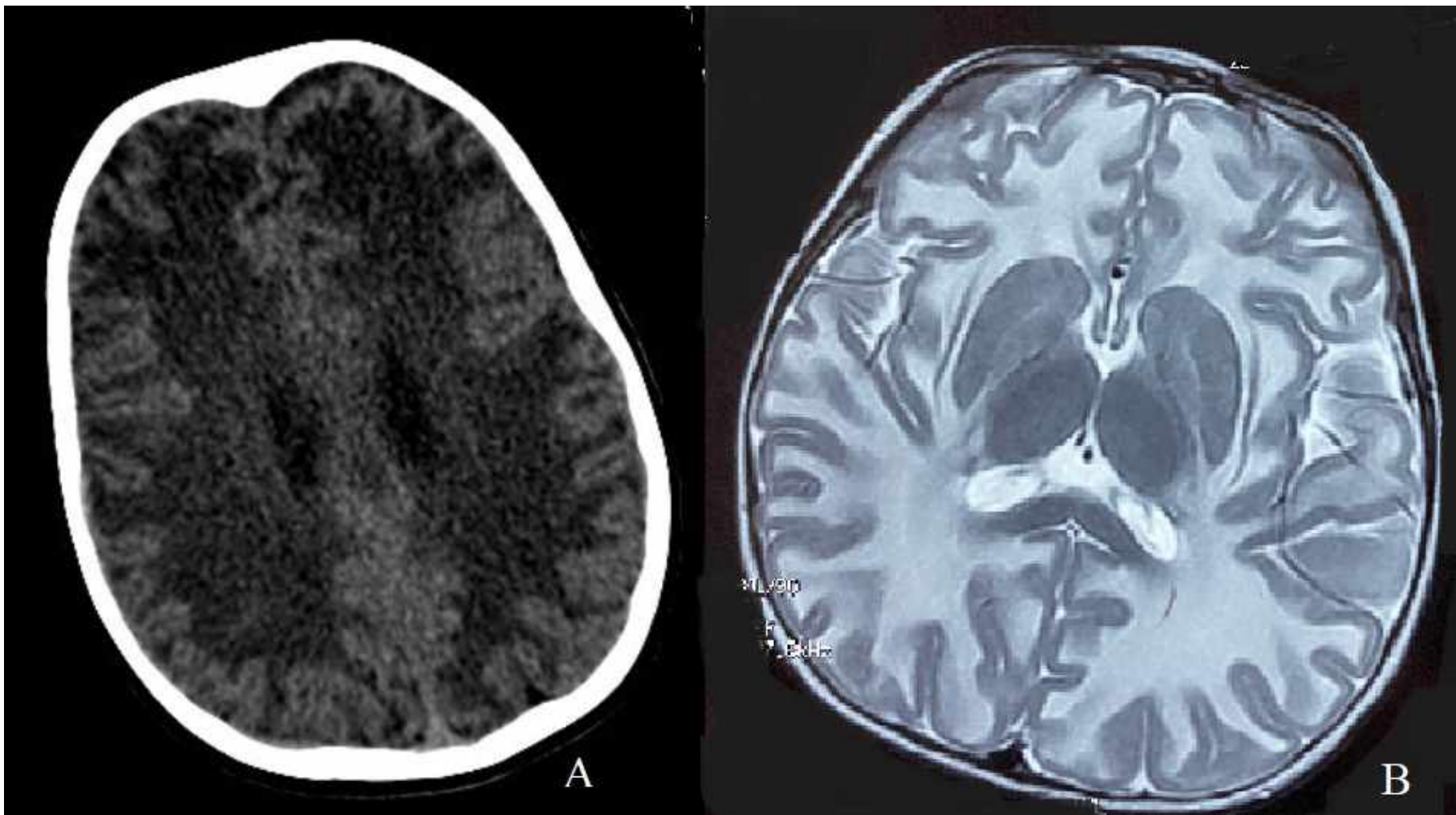


Figure 1: A) Axial CT scan that showed prominent diffuse white matter hypodensity, B) Axial : FLAIR image reveals diffuse bilateral symmetrical white matter hyperintensities

CONCLUSION

Neurodegenerative diseases can occur after a physical stress situation such as mild head trauma or febrile infection. And also minor head trauma may cause consciousness disturbance in van der Knaap disease.

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