생명을 위협하는 뇌간 압박과 급성 수두증을 유발한 급성 소뇌염: 사망 증례보고

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Acute Cerebellitis Causing Life-threatening Brain Stem Compression and Acute Hydrocephalus: A Fatal Case Report

Acute cerebellitis is a rare inflammatory disorder that occurs most frequently in children. The typical clinical course of acute cerebellitis is benign. However, in some cases of acute cerebellitis, fulminant cerebellar swelling with obliteration of the fourth ventricle causes brain stem compression and acute obstructive hydrocephalus, which can be life-threatening and require emergent neurosurgical procedures. We describe the case of a 4-year-old girl whose acute cerebellitis caused brain stem compression, acute obstructive hydrocephalus, and death. The patient was admitted to the emergency department because of a severe headache that had persisted for 5 days. Neurological examinations revealed no specific abnormality. Brain magnetic resonance imaging (MRI) demonstrated diffuse swelling and high signal intensity lesions in the cerebellar hemispheres and vermis, obliteration of the fourth ventricle, and brain stem compression on T2-weighted images in conjunction with mild hydrocephalus. Cerebrospinal fluid (CSF) analysis revealed white blood cell and red blood cell counts of 180/mm³ and 0/mm³, respectively; protein and glucose concentrations of 263.6 mg/dL and 37 mg/dL, respectively; and negative culture results. Despite aggressive treatment, the patient developed sudden cardiorespiratory arrest on day 2. Although emergency neurosurgery was performed after cardiopulmonary resuscitation, her condition progressed to brain death, and she died on day 29. This case suggests that timely and appropriate neurosurgery should be actively considered to relieve increased intracranial pressure in the early phases of acute cerebellitis.

Key Words: Intracranial hypertension, Hydrocephalus, Neurosurgical procedures, Death, Child

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Introduction

Acute cerebellitis is a rare inflammatory syndrome of the cerebellum, which can occur as a primary infectious, postinfectious, or postvaccination disorder¹⁾. Although no infectious pathogen is isolated from the cerebrospinal fluid (CSF) in most cases, the pathogenic agents that have been reported to be associated with acute cerebellitis include the varicella virus, measles virus, mumps virus, rubella virus, Epstein-Barr virus, cytomegalovirus, herpes simplex virus, influenza virus,

parainfluenza virus, poliovirus, coxsackie virus, *Salmonella typhi*, *Bordetella pertussis*, and *Mycoplasma pneumoniae*²⁾.

The clinical course of acute cerebellitis is typically benign and self-limiting. However, life-threatening complications such as brain stem compression and acute obstructive hydrocephalus resulting from severe cerebellar swelling can occur in the acute phases of the disease^{1,2)}. We describe a fatal case of acute cerebellitis with life-threatening brain stem compression and acute hydrocephalus.

Case Report

A 4-year-old girl was admitted to the emergency department of our hospital; she presented with severe headache, which began 5 days earlier. Her headache was initially accompanied by fever, dizziness, and dysarthria. Fever subsided one day prior to admis-

sion but the remaining symptoms were aggravated. She was previously healthy with no history of medication, head trauma, or recent immunization. Her vaccinations were up to date. On arrival at the emergency department, the patient intermittently screamed owing to the severe headache. Neurological examination revealed that she was fully alert and well-oriented. Pupils were isocoric with prompt light reflex. Other cranial nerves were also intact. The patient showed normal motor power in the extremities, and the Babinski sign was absent. Cerebellar function tests, such as the finger-to-nose test, rapid alternating movement test, and tandem gait test were not performed due to the patient's poor cooperation. Magnetic resonance imaging (MRI) of the brain demonstrated diffuse swelling and high signal intensity lesions in the cerebellar hemispheres and vermis, obliteration of the fourth ventricle, and brain stem compression on T2-weighted images in conjunction with mild hydrocephalus (Fig. 1). CSF analysis showed the following measurements: white blood cells,

Fig. 1. Brain magnetic resonance imaging findings. (A) Axial T2-weighted magnetic resonance imaging (MRI) shows diffuse areas of high signal intensity in the cerebellar hemispheres and the vermis. (B) Contrast-enhanced axial T1-weighted MRI shows strong signal enhancement of both cerebellar hemispheres and the vermis. (C) Axial fluid attenuated inversion recovery MRI shows obliteration of the fourth ventricle and brain stem compression resulting from diffuse cerebellar swelling (arrow). (D) Axial fluid attenuated inversion recovery MRI shows dilatation of the lateral and third ventricles with periventricular interstitial edema.

180/mm³; red blood cells, 0/mm³; protein, 263.6 mg/dL; and glucose, 37 mg/dL, with a negative culture result. The intracranial pressure was not determined because the patient continuously moved during lumbar puncture despite the use of sedative hypnotics. Serological tests were negative. Serum immunoglobulin (Ig) M against *Mycoplasma pneumoniae* was positive (5.8; positive >1.0) but CSF IgM against it was negative (0.4). CSF polymerase chain reactions for enterovirus, herpes simplex viruses 1 and 2, varicella zoster virus, Epstein-Barr virus, *Haemophilus influenzae*, *Neisseria meningitidis*, *Group B Streptococcus*, and *Listeria monocytogenes* all tested negative. CSF oligoclonal IgG bands and myelin basic protein produced negative results. Blood and urine cultures were also negative. The patient was treated with empirical antimicrobial agents (vancomycin, cefotaxime, and acyclovir), intravenous methylprednisolone (30 mg/kg/day), and mannitol. However, she continued to complain of headache along with intermittent screaming despite no obvious signs of neurological deterioration. On day 2, the patient developed sudden cardiorespiratory arrest. Follow-up brain computed tomography obtained after resuscitation showed aggravated cerebellar swelling, hydrocephalus, and brain stem compression (Fig. 2). Despite emergency external ventricular drain, decompressive suboccipital craniectomy, and C1 laminectomy, patient condition evolved to brain death. The patient died on day 29.

Discussion

Most patients with acute cerebellitis show full clinical recovery, although some exhibited long-term neurological sequelae ranging from mild tremor to profound ataxia, and cognitive deficits³. In rare cases, during the acute phase of the disease, fulminant cerebellar swelling may lead to acute hydrocephalus and fatal brain

stem compression, presenting as unconsciousness and cardiorespiratory depression, cranial nerve dysfunction, and sudden death, as described in this case¹. Obliteration of the fourth ventricle by cerebellar swelling can obstruct CSF flow, which could eventually develop into acute hydrocephalus and tonsillar herniation, or sudden death. This case also showed that MRI-detected obstruction of the fourth ventricle by cerebellar swelling may predict an impending fatal condition.

The microscopic pathology of acute cerebellitis is characterized by edematous cerebellar changes, with the infiltration of lymphocytes, monocytes, macrophages, and eosinophils⁴. One case report concerning fatal acute cerebellitis showed that inflammatory cell infiltration was confined to the molecular layer, with the internal granular cell layer and deep white matter apparently being spared⁴. In this case, however, we did not perform a brain biopsy or pathologic examinations because of the fulminant and life-threatening deterioration of the patient's condition.

CSF analysis is important for diagnosing acute cerebellitis. Although it may reveal variable features, typical CSF findings in acute cerebellitis include mild pleocytosis and elevated protein levels⁵. MRI is also among the most important diagnostic tools for acute cerebellitis. Unlike acute cerebellar ataxia, acute cerebellitis can be diagnosed from cerebellar lesions detected by neuroimaging⁶. MRI findings in acute cerebellitis are characterized by hyperintense cerebellar lesions on T2-weighted images and diffusion-restricted cerebellar lesions on diffusion-weighted images⁷⁻⁹.

The clinical presentation of acute cerebellitis is variable. The common symptoms are truncal ataxia, dysarthria, dysmetria, tremor, and headache². However, a recent literature demonstrated that 50% (30/60) of the patients with acute cerebellitis did not present with cerebellar signs⁷. Cerebellar symptoms can be overlooked or masked if the neurological examination is limited

Fig. 2. Brain computed tomography findings. (A) Note the markedly dilated lateral ventricles on the computed tomography (CT) scan obtained soon after cardiorespiratory arrest. (B) Note the collapsed lateral ventricles and the tip of the ventricular catheter (arrow) on the CT scan obtained on the day of the emergency external ventricular drain and decompressive craniectomy. (C) Note the severe cerebellar swelling, cisternal space obliteration of the posterior fossa, brain stem compression, and the status of the suboccipital craniectomy and C1 laminectomy on the CT scan obtained on the day of surgery.

because of patient irritability or alteration in the state of consciousness.

The medical management of acute cerebellitis includes the use of high doses of corticosteroids, appropriate antibiotics and antiviral agents, and osmotherapy. Despite intensive medical treatment, development of obstructive hydrocephalus requires emergent neurosurgical interventions, such as external ventricular drainage, ventricular peritoneal shunt, and posterior fossa decompression^{1,2)}.

In this case, we failed to conduct timely neurosurgical interventions, because the patient showed no apparent signs of neurological deterioration except for severe headache and somnolence. Her pulse, blood pressure, and respiratory pattern were normal throughout the hospitalization period. Her pupils were light-responsive and not dilated before her sudden cardiorespiratory arrest. This suggests that MRI findings of obliteration of the fourth ventricle and acute hydrocephalus resulting from cerebellar swelling might be more critical than the clinical features of patients with acute cerebellitis when the time of neurosurgical intervention is determined.

In conclusion, since severe cerebellar swelling in acute cerebellitis can cause life-threatening obstructive hydrocephalus and brain stem compression, timely and appropriate life-saving surgical procedures should be actively considered to control the increased intracranial pressure in the early phases of the disease.

요약

급성 소뇌염은 주로 소아에서 발생하는 드문 염증성 질환이다. 전형적인 급성 소뇌염의 예후는 양호하나, 질병 급성기에 심한 소뇌 부종이 발생할 경우 제 4뇌실 폐쇄, 뇌간 압박 및 급성 폐쇄성 수두증을 유발하여 생명을 위협할 수 있으며, 응급 신경외과 수술이 필요할 수 있다. 저자들은 급성 소뇌염에 의해 뇌간 압박과 급성 수두증이 발생하여 사망에 이른 4세 여자 환자를 경험하였기에 보고하는 바이다. 환자는 5일간 지속되는 심한 두통을 주소로 응급실에 내원하였다. 내원 당시 환자의 신경학적 진찰에서 특별한 이상소견은 없었다. 뇌 자

기공명영상의 T2 강조영상에서 소뇌의 전반적 부종과 함께 소뇌 반구와 충부(vermis)에 고강도 신호가 관찰되었고, 제4뇌실의 폐쇄, 뇌간 압박, 경도의 수두증이 동반되었다. 뇌척수액 검사는 백혈구 180/mm³, 적혈구 0/mm³; 단백 263.6 mg/dL, 당 37 mg/dL, 및 세균 배양 검사 음성 소견을 보였다. 환자는 적극적인 약물치료에도 불구하고 입원 2일째 갑자기 심폐정지가 발생하였다. 심폐소생술 후, 응급 신경외과 수술이 시행되었으나 환자는 뇌사 상태로 진행하였고, 결국 입원 29일째 사망하였다. 본 증례는 급성 소뇌염의 초기에 생명을 위협할 수 있는 뇌압상승이 발생할 수 있으므로, 뇌압상승 조절을 위해 시의 적절한 신경외과적 수술이 적극적으로 고려되어야 함을 시사하였다.

References

- 1) Shkalim V, Amir J, Kornreich L, Scheuerman O, Straussberg R. Acute cerebellitis presenting as tonsillar herniation and hydrocephalus. *Pediatr Neurol* 2009;41:200-3.
- 2) Hacoen Y, Niotakis G, Aujla A, Siddiqui A, McCormick D, Bassi S, et al. Acute life threatening cerebellitis presenting with no apparent cerebellar signs. *Clin Neurol Neurosurg* 2011;113:928-30.
- 3) Hennes E, Zotter S, Dorninger L, Hartmann H, Häusler M, Huppke P, et al. Long-term outcome of children with acute cerebellitis. *Neuropediatrics* 2012;43:240-8.
- 4) Levy EI, Harris AE, Omalu BI, Hamilton RL, Branstetter BF 4th, Pollack IF. Sudden death from fulminant acute cerebellitis. *Pediatr Neurosurg* 2001;35:24-8.
- 5) Sawaisi Y, Takada G. Acute cerebellitis. *Cerebellum* 2002;1:223-8.
- 6) Desai J, Mitchell WG. Acute cerebellar ataxia, acute cerebellitis, and opsoclonus-myoclonus syndrome. *J Child Neurol* 2012;27:1482-8.
- 7) Poretti A, Benson JE, Huisman TA, Boltshauser E. Acute ataxia in children: approach to clinical presentation and role of additional investigations. *Neuropediatrics* 2013;44:127-41.
- 8) De Bruecker Y, Claus F, Demaerel P, Ballaux F, Sciort R, Lagae L, et al. MRI findings in acute cerebellitis. *Eur Radiol* 2004;14:1478-83.
- 9) Kamate M, Chetal V, Hattiholi V. Fulminant cerebellitis: a fatal, clinically isolated syndrome. *Pediatr Neurol* 2009;41:220-2.