Acute Fulminant Cerebellitis Managed by Posterior Fossa Decompression Surgery with Favourable Neurologic Outcome

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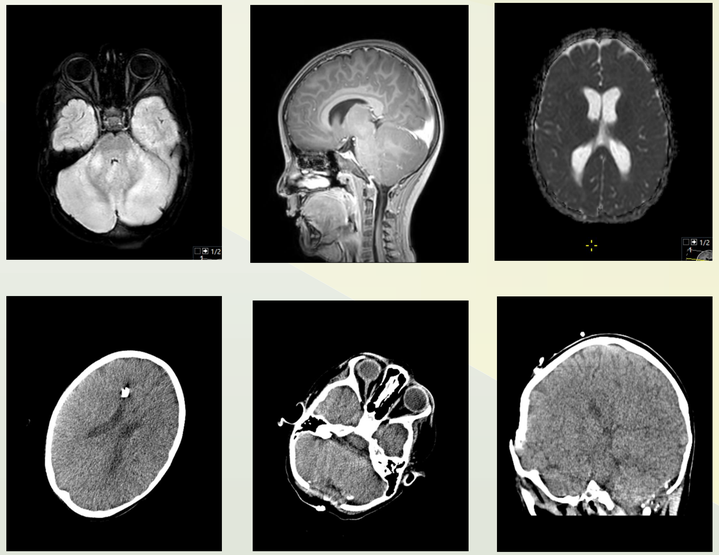
INTRODUCTION

Acute cerebellitis (AC) is a rare infectious-inflammatory syndrome with a variable clinical course, ranging from benign self-limiting cerebellar symptoms such as ataxia, nystagmus and dysmetria to a fulminant presentation requiring posterior fossa decompression surgery. (1,2) Although AC causes acute cerebellar dysfunction, often associated with fever, headache and nausea, in some cases cerebellar inflammation can lead to brainstem compression and cause altered level of consciousness, coma and even death. (3,4) Because of its infrequency and heterogeneous presentation, diagnosis requires a high level of suspicion and an emergent MRI scan of the brain. After diagnosis, treatment with antibiotics and high-dose corticosteroids should be started as soon as possible, and if posterior fossa compression occurs, extraventricular drainage and craniectomy should be performed to prevent morbidity and mortality. (5)

OBJECTIVES

We aimed to discuss the effectiveness of decompressive surgery due to fulminant acute cerebellitis.



BEFORE SURGERY

AFTER SURGERY

CASE

A 4-year-old previously healthy boy presented with vomiting, diarrhoea and altered mental status for 2 days. He was confused and his Glascow Coma Scale was 11 with stable vital signs. The rest of the physical examination was normal. Brain MRI showed widespread hyperintensity of both cerebellar hemispheres and the splenium of the corpus callosum, and marked cerebellar swelling. CSF analysis showed normal protein and glucose levels with mild pleocytosis. CSF PCR analysis was negative for common causes of viral meningoencephalitis. Serum anti-MOG was negative. Hypertonic saline was used to reduce posterior fossa oedema. Control brain MRI showed hydrocephalus and increased cerebellar oedema. Extraventricular drainage was placed, but the clinical findings gradually worsened. Apnoea and bradycardia developed, brainstem reflexes weakened, and the patient was intubated. Brain CT showed cerebellar herniation and brainstem compression. An urgent craniectomy and partial cerebellar resection was performed. The case improved rapidly in the postoperative period. During long-term follow-up, the neurological status of the case was normal for his age.

CONCLUSIONS

Acute cerebellitis is a "red flag" diagnosis that may require urgent pharmacological and surgical intervention. AC diagnosis requires a high level of suspicion and an emergency brain magnetic resonance imaging study. In this case diagnosed as acute cerebellitis with a fulminant course, the life-threatening volume and pressure increase in the posterior fossa could be controlled by resection of the cerebellar vermis with a favourable neurologic prognosis. Therefore, we emphasize that decompressive surgery may be life-saving in selected cases of cerebellitis with a fulminant course.

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