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CASE REPORT

Life-saving decompressive craniectomy for acute disseminated encephalomyelitis in a child: a case report

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Introduction

Acute disseminated encephalomyelitis (ADEM) is an inflammatory demyelinating immune-mediated disorder which is more common in the pediatric population. It is often associated with a viral or bacterial infection or as a complication after vaccination. The pathophysiology involves transient autoimmune response directed at myelin or other self-antigens, possibly by molecular mimicry or by nonspecific activation of autoreactive T cell clones [2, 10]. Acute clinical presentation is characterized by a rapid onset of encephalopathy, multifocal neurological features, convulsions, and impaired consciousness with fever. There are no biological markers of the disease. Brain and spinal MRI is the imaging modality of choice to point out white matter lesion in ADEM. Mortality is actually lower than 5 % in children [2, 14]. Prognosis is more serious in patients hospitalized in the intensive care unit: mortality can reach 25 % [13]. Intracranial hypertension resulting from cerebral edema in ADEM can quickly lead to secondary brain damage, herniation, or death. Usual treatment options of ADEM includes: high-dose intravenous methyl prednisolone, intravenous immunoglobin, and plasma paresis [2]. The indication of surgical treatment is still under debate.

Herein, we report the first case of a pediatric patient presenting with an acute course of ADEM who needed intracranial monitoring, steroids, and emergency hemicraniectomy, with a favorable outcome.

Case report

An 18-month-old girl was hospitalized in our institution for left hemibody seizures with fluctuating consciousness, a few hours after a febrile rash had appeared on her face and anterior chest. On day 2, due to the persistence of neurological symptoms with worsening consciousness, she was treated with acyclovir and underwent a brain MRI. This showed a widespread hyperintense FLAIR/T2 white matter lesion revealing an acute demyelinating disease (Fig. 1). Electroencephalogram showed diffuse bilateral slowdown. Cerebrospinal fluid showed normal protein and glucose levels and negative HSV and enterovirus PCR. At that time, aciclovir was stopped, and corticotherapy was continued (20 mg/kg/day). On day 5, a fundoscopy exhibited bilateral papilledema with punctiform petechiae. Within hours, she became increasingly confused and presented bradycardia. She was transferred to the intensive care unit (ICU) and was intubated and ventilated. Immunoglobulin therapy was given for 2 days. A CT scan revealed cerebral edema with reduction of the space of the peri-pontine cisterns and on the skull base. After discussion with the intensivists, we decided to monitor intracranial pressure through a ventricular drainage (Fig. 2). Intracranial hypertension oscillated between 40

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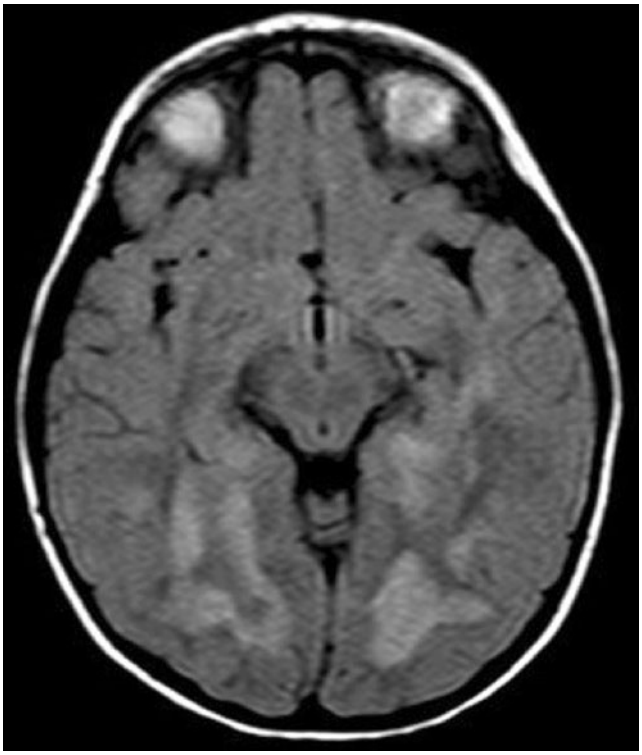

Fig. 1 Widespread posterior white matter hyperintensity on MRI (FLAIR)

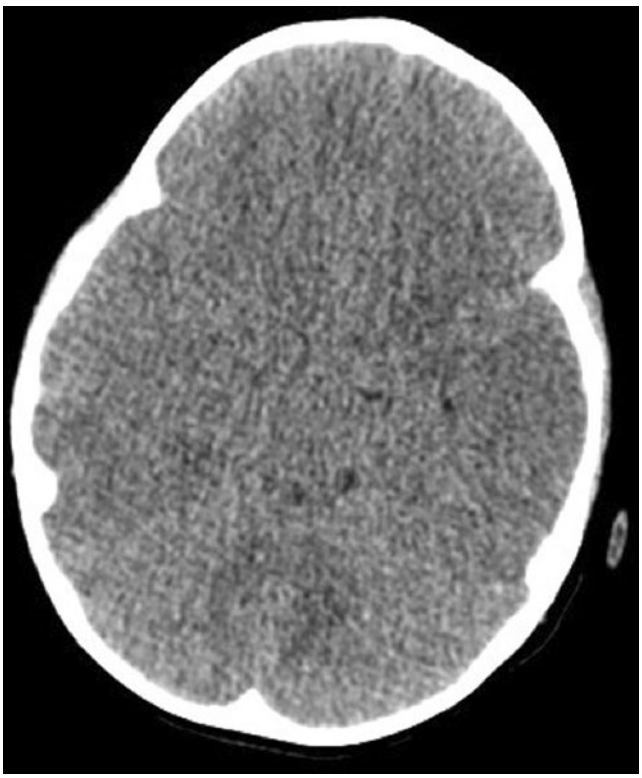

Fig. 2 Diffuse cerebral edema. Disappearance of the basal cisterns and diffuse brain swelling

and 70 mmHg, which was first treated with mannitol and barbiturates. Yet, intracranial pressure oscillated between 20 and 30 mmHg. Twelve hours later, intracranial pressure had again increased, associated with bilateral mydriasis and bradycardia. We decided to perform a life-saving large decompressive left hemicraniectomy. The dura mater was noted to be tense and opened in an X shape. The brain appeared edematous. Dural plasty enlargement was performed with Gore-tex (Fig. 3). Following this, she was transferred again to the ICU. Postoperative intracranial pressure was 14 mmHg.

The ventricular drain was in place for 4 days. Intracranial pressure was 10 mmHg during 2 days, and then the drain was removed. Postoperative CT scan revealed the cerebral edema had disappeared and showed evidence of hypointensities in the white matter.

Five days after the hemicraniectomy, sedation was stopped; a right hemiparesis appeared, with absence of eye tracking and eye deviation to the right. The child was extubated on day 10.

Eleven days after the decompressive hemicraniectomy, the child moved from the intensive care unit to the Department of Pediatric Neurology, and then she spent 1 month in a rehabilitation center.

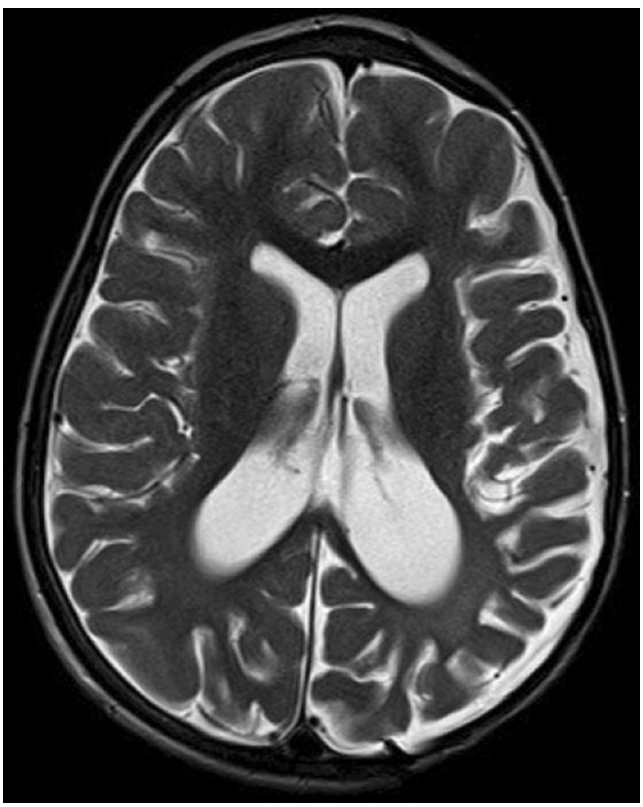

Fig. 3 T2 brain MRI 14 months after decompressive craniectomy: mild atrophy on the left side with slight enlargement of the ventricular cavities

Two months later, the child was readmitted for an autologous cranioplasty. Eye tracking was normal. Her motility had improved with a slight motor weakness of the right upper limb.

Two years later, the child had a discrete right hemiparesis; she can walk and use her upper limb. She speaks fluently. MRI shows only mild left hemisphere atrophy with slight enlargement of the ventricular system (Fig. 3).

Discussion

Acute disseminated encephalomyelitis is an inflammatory immune-mediated disorder which is more frequent in children. In 2007, the International Pediatric Multiple Sclerosis Study Group defined ADEM as "the first clinical event with a polysymptomatic encephalopathy, with acute or subacute onset, showing focal or multifocal hyperintense lesions predominantly affecting the Central Nervous System (CNS) white matter." This definition points out the difficulty to differentiate ADEM from a multiple sclerosis at the beginning of its history [22]. The pathophysiology is still unclear and probably involves a transient autoimmune response directed at myelin or other self-antigens, possibly by molecular mimicry or by nonspecific activation of autoreactive T cell clones [2, 10]. Brain biopsies are performed in difficult cases [7, 8]. Histologically, ADEM is characterized by perivenous demyelination and infiltration of the vessel wall and perivascular spaces by lymphocytes, plasma cells, and monocytes [4].

Brain MRI is necessary for diagnosis. ADEM is associated with characteristic lesions on FLAIR and T2-weighted images such as: large multifocal, hyperintense, bilateral, asymmetric lesions in the supratentorial or infratentorial white matter. The gray matter, basal ganglia, thalamus, and spinal cord may be involved. There is no evidence of previous destructive white matter changes [10, 21]. Periventricular lesions are reported currently (30–60 % of cases) as callosal locations are less frequent [15].

Management of ADEM is now well described although spontaneous improvement has been documented [20]. Medical treatment at the acute stage includes high-dose intravenous methylprednisolone, intravenous immunoglobulin, and plasma paresis [2]. Corticosteroid is the first-line treatment (10–30 mg/kg/day, up to a maximum of 1 g/day) [2]. After treatment, prognosis in ADEM is good with more than 50 % of the patients who achieved total functional recovery [18] while the others suffer only from mild paresis or trouble in cognition or behavior [2, 18]. However, evolution may be less favorable in some cases especially for very young children or children with hyperacute initial stage and hemorrhagic forms [1–3, 5, 17, 19, 23]. These children need to be managed in the ICU and may benefit from surgery. Mortality in ADEM varies from 1 to 12 % [1, 2, 12, 18]. Refractory intracranial hypertension is the cause of death in such cases.

In the majority of cases, the increasing intracranial pressure can be controlled by conservative treatment, which may include a combination of the following; 30° elevation of the head, mannitol therapy, hypertonic fluid infusion, brief hyperventilation, and mild hypothermia. When ICP levels remain refractory, the treatment options may also include barbiturate coma, and when all else fail, decompressive craniectomy.

Decompressive craniectomy in ADEM is reported rarely in medical literature [1, 17, 19, 23]. The only reported cases concern four adult patients and indicate that decompression in adult patients with ADEM is effective and associated with a favorable outcome. The four patients have mild residual neurological deficits soon after craniectomy [1, 17, 19, 23]. Our patient is the first reported child with ADEM who sustained a life-saving craniectomy after monitoring of the intracranial pressure (Fig. 4). Craniectomy was large, close to two thirds of the left hemicranium as performed in brain trauma patients [9, 11]. We feel that the large size of the craniectomy, in accordance with the recommended size for traumatic brain injury, contributed to the good recovery [9]. In our case, it was performed 4 h after evidence of refractory intracranial pressure. This short delay was possible due to the intracranial monitoring and may explain the very good

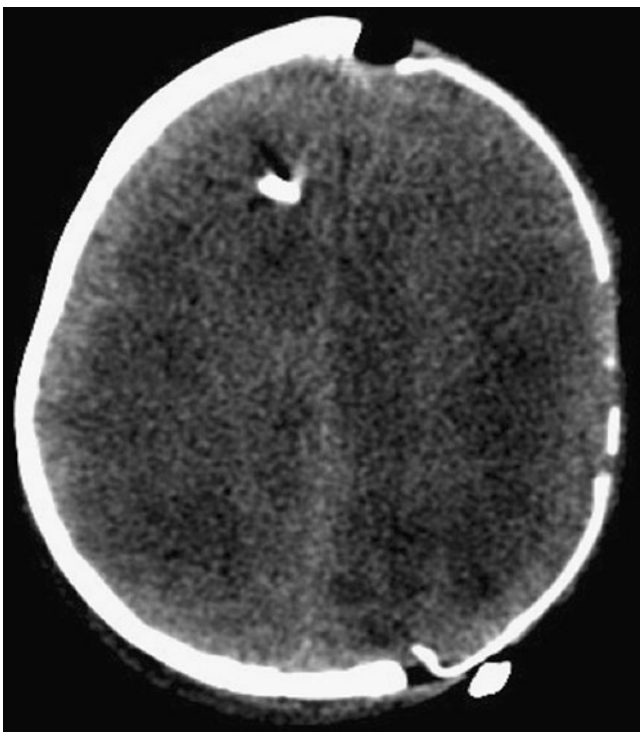

Fig. 4 Post hemicraniectomy CT scan with intracranial monitoring probe. Note the large craniectomy

outcome 2 years after the event. So, ICP monitoring looks to be a very good tool to manage children hospitalized in the ICU with high-grade ADEM. It permits a more rapid decision for hemicraniectomy once pressure has become out of control.

Conclusion

Although medical treatment alone is very effective to treat children suffering from ADEM, we must keep in mind that some cases are refractory to usual therapy. Yet throughout the medical literature there is much anecdotal evidence to suggest a larger role for decompressive craniectomy as an effective and safe treatment for cerebral hypertension secondary to multiple pathologies [6, 11, 16]. To our knowledge, this is the first reported case of a child treated successfully with intracranial monitoring and decompressive craniectomy for high-grade ADEM.

Conflict of interest We have no conflict of interest.

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