Post-traumatic Acute Disseminated Encephalomyelitis: Case Report

Abstract

We report a case of a child who developed acute disseminated encephalomyelitis after a trauma at home. This is a 25-month-old child, born of a non-consanguineous marriage with no perinatal history or any recent viral infections, with good psychomotor and intellectual development.
Diagnosis of encephalopathy can be difficult in children, with nonspecific, subtle, and transient symptoms, such as drowsiness or irritability that can be easily overlooked, especially in the emergency department. The best available diagnostic tool is based on clinical and radiological findings. MRI could be useful for a precise and early diagnosis necessary to solve some unanswered questions.

Keywords: psychomotor, acute disseminated encephalomyelitis, radiological findings, monophasic immune-mediated inflammatory disease

I-Introduction

The term acute disseminated encephalomyelitis (ADEM) was first used in 1950 as a rare monophasic immune-mediated inflammatory disease of the CNS that is potentially, functionally, and vitally serious (1). It is a curable from of encephalomyelitis with rapid onset and combination of multifocal neurologic symptoms (2). ADEM diagnosis requires two essential elements: typical clinical presentation and magnetic resonance imaging (MRI) findings (3).
Its association with various infections is well described, less often with vaccination such as polio and rarely with trauma but can also occur without any cause (4-6).
We report a case of a child who developed ADEM after a trauma at home.

II – Case report:

We report a case of 25-month-old child, born of a non-consanguineous marriage with no perinatal history or any recent viral infections, with good psychomotor and intellectual development. He presented with gait disorders 48 hours after a fall from a staircase with a cranial impact point.
On examination, he had cerebellar ataxia, bidirectional horizontal nystagmus, divergent strabismus of the right eye, abnormal head movements, and hypersialorrhea. Both blood test and cerebrospinal fluid analysis were normal.
Brain MRI showed (Fig.1) inflammatory demyelinating lesions of the supra- and subtentorial white matter, located in the juxtaventricular and juxtacortical regions. Large areas of hyper intense signal in axial sections, and there is an associated right thalamic lesion. No gadolinium enhancement of lesion was observed. The associated involvement of the basal ganglia points toward an acute disseminated encephalomyelitis.
He was treated with a bolus of intravenous methylprednisolone (30mg/kg/d for 3 days) followed by tapered oral corticosteroids for one month with complete recovery of the motor and neurological deficit.

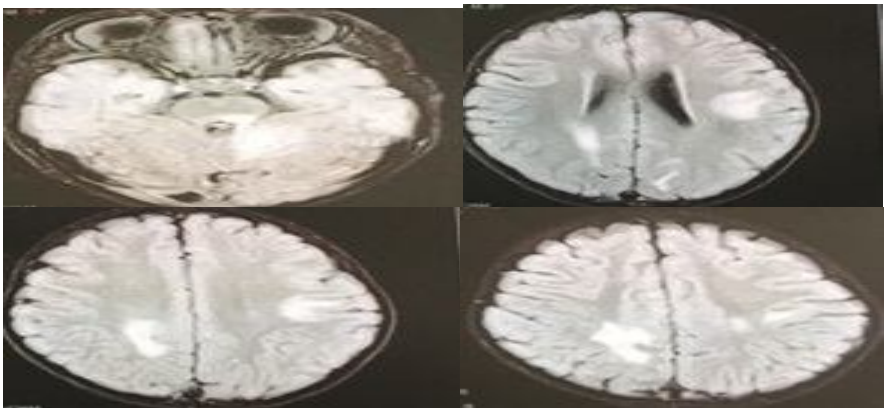

Fig 1: cerebral MRI showed hyperintense large, patchy lesion multifocal affecting cerebellar, pontine, and cerebral area.

III-DISCUSSION

We described an atypical case of ADEM that initially manifested after head trauma. This case report suggests a possible association, which should be considered in the management of patients with any kind of head trauma; whether minimal or severe, especially in the presence of neurological or neuropsychiatric symptoms. It can affect both adults and children (7). This frequency in children was explained by the greater frequency of vaccinations and exposure to infectious antigens (8,9).

In the definition and pathogenesis of ADEM, infections and vaccination seem to be the exclusive triggers of this condition. This case report is a call to investigate whether ADEM could indeed occur "as a result of trauma". It could contribute to early diagnosis and management of this condition, which is not always easy to treat and can sometimes be fatal.

Post-traumatic ADEM is rarely reported in the literature. Neuroinflammation is well established as a key secondary injury mechanism after traumatic brain injury, and it has long been considered a contributor to the damage following brain injury (10)

Association with polio vaccine has been reported (11). Other explanation of its occurrence post infectious through the involvement of T cells cross-reacting with Myelin basic protein antigens include HHV-6 (12), coronavirus (13), influenza virus hemagglutinin (14) and EBV (15) Viral or bacterial epitopes ex urinary (16) resembling myelin antigens with ability to activate myelin-reactive T-cell clones by molecular mimicry (17).

It has also been described in association with the use of certain drugs such as sulfonamides para-amino-salicylic acid and streptomycin.

The age of presentation is usually between 5 and 8 years in children described in association with rabies and smallpox vaccines.

Updated consensus criteria for ADEM (which remains a diagnosis of exclusion) requires all the following criteria:

- First event of encephalopathy plus multifocal neurologic deficits.
- Presumed inflammatory demyelinating cause.
- Encephalopathy (altered consciousness or behavior unexplained by fever, systemic illness, or post-critical symptoms).
- Brain MRI abnormalities consistent with acute phase (3 months) demyelination.
- Typical brain MRI lesions:
    1. Diffuse, poorly delineated, large (>1 to 2 cm) lesions.
    2. Involving mostly cerebral white matter.
    3. T1 hypointense lesions should not be observed.
    4. Deep lesions of the gray matter (e.g., thalamus or basal ganglia) may be present, especially in children

MRI is extremely important in establishing the diagnosis of ADEM. Lesions are usually large and patchy, but different sizes can be identified in the same patient (from a few millimeters to several

centimeters). In large lesions with a tumor-like appearance, mass effect is usually mild or absent. Ovoid lesions (Dawson's fingers) are much less frequent. (18,19)
Clinical symptoms (convulsions, headache, fever, focal neurological deficits, cerebellar syndrome, consciousness disorders, transverse myelitis) (20) usually appear three weeks after vaccination (21,22).
The initial symptoms of ADEM may include lethargy, fever, headache, vomiting, meningeal signs, and seizures, including status epilepticus. Encephalopathy ranging from behavioral changes and persistent irritability to coma. Focal neurologic deficits may be difficult to determine in the obnoxious or very young child, but common neurologic signs of ADEM include visual loss, cranial neuropathies, ataxia, and motor and sensory deficits, as well as bowel, and bladder dysfunction with concomitant demyelination of the spinal cord. The clinical course is usually rapidly progressive over days. Intensive care unit admission may be required, especially for patients with brainstem dysfunction or elevated intracranial pressure. Diagnosis of encephalopathy can be difficult in children, with nonspecific, subtle, and transient symptoms, such as drowsiness or irritability, that can be easily overlooked, especially in the emergency department. ADEM lacks a specific biological marker, and diagnosis depends upon retaining a high index of clinical suspicion and always considering the condition in differential diagnoses (23). Although ADEM has been stated to occur in post-traumatic brain injury (16), this association is rarely reported in the literature (24). Brain CT may be normal or show hypodense areas. MRI visualizes multiple hyperintense patches of proton density in T2 and FLAIR in the supratentorial white matter, brainstem and cerebellum, often associated with localizations in the brain. The association with lesions of the basal ganglia, especially the thalamus, is an argument in favor of ADEM, but the preservation of the periventricular white matter rather points to ADEM (25). MRI abnormalities sometimes appear delayed compared to the first clinical signs and these same abnormalities may still increase, while the clinical picture is regressing (26).
In case of a single lesion with mass effect, the problem of differential diagnosis with a tumor process or an abscess arises (27,28). There is increasing evidence of reduced mortality and improved outcome in patients treated with high-dose intravenous corticosteroids, and Intravenous Immunoglobulin (IVIG) (29).
Therapeutic approaches for ADEM include steroids, IVIG or plasmapheresis. High-dose steroid therapy is recommended in the form of IV methylprednisolone (10-30 mg/kg/day) or dexamethasone (1 mg/kg) for 3 to 5 days followed by tapered the oral steroid over 4 to 6 weeks.
Recovery is reported in 50-80% of patients if treatment is continued with tapered oral steroids. IVIG should be used at a dose of 1-2 g/kg as a single dose or over 3 to 5 days. IVIG is recommended if steroid therapy fails or if demyelination recurs. In a small number of case series, the use of plasma exchange has been reported for children diagnosed with ADEM.

IV- Conclusion:

There appears to be a common pathway by which the brain responds to aggression, whether it is physical trauma or infection. Whether this can lead to ADEM is a question to be resolved by further study. A single case report can only introduce the subject, especially in the face of the near absence of diagnostic tools in developing countries and the absence of studies.
The best available diagnostic tool are clinical and radiological findings. MRI could be useful for a precise and early diagnosis necessary to solve some unanswered questions

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