Trakia Journal of Sciences, Vol. 10, Suppl. 2, 143-146, 2012

Available online at:
http://www.uni-sz.bg

ISSN 1313-7050 (print)
ISSN 1313-3551 (online)

ACUTE DISSEMINATED ENCEPHALOMYELITIS IN 10-YEARS OLD CHILD. A CASE REPORT

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ABSTRACT

Acute disseminated encephalomyelitis (ADEM) is a monophasic, demyelinating disease of the central nervous system that predominately affects prepubertal children. It is typically characterized by an abrupt onset of neurologic symptoms preceded by an infection or recent immunization. The purpose of our study is to describe a case of a 10 years old girl, who fell ill without former predisposition with a temperature, neurologic deficit with a paralysis, visual disorders, and liquor alteration. It was only one episode, after which the patient improved. Clinical, laboratorial, microbiological, and instrumental investigation were used. The MMR changes established the diagnosis ADEM. The case was studied up to 5 years after the onset of the disease. The child is fully recovered. The clinical features of ADEM, differential diagnosis and therapy are discussed.

Key words: ADEM, neurological deficit, MRI, MS

INTRODUCTION

ADEM is a monophasic, nonvasculitic, inflammatory, demyelinating disease that can affect the entire central nervous system (CNS). It might be indistinguishable from many forms of encephalitis except for its patchy distribution of demyelinating lesions that may have a striking similarity to lesions caused by multiple sclerosis (MS). Unlike MS, ADEM is considered a disease of singular occurrence from which most people fully recover. ADEM appears to occur most frequently after childhood infections such as measles, smallpox, chickenpox, or their respective vaccinations. The exact mechanism of disease pathophysiology is unknown. It is thought that the CNS becomes inflamed with subsequent demyelination because of a primary autoimmune response or an immune response secondary to an infection. It is difficult to estimate the incidence and prevalence of ADEM. In the United States, postvaccination ADEM for the prevention of mumps, measles, and rubella and for tick-borne encephalitis occurs in 10-20 out of 100,000 individuals (1). ADEM occurs worldwide, although it is thought to affect more white people in higher latitudes, similar to MS. Mortality has been estimated as high as 10%—20% (2), and as low as less than 2% (3). Morbidity usually involves visual, motor, autonomic, or intellectual deficits, and epilepsy if gray matter is affected. Deficits may persist for several weeks; however, most deficits resolve within 1 year (3). Mental retardation is possible, especially in children younger than 2 years of age at disease onset. Males and females are equally affected. Female adolescents with ADEM are more likely than males to develop MS in the future (4), though there is no strong scientific theory to explain this occurrence.

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MATERIALS AND METHODS

This article described the course of a young female diagnosed with acute disseminated encephalomyelitis (ADEM) with a typical presentation. The patient was a 10-year-old female – L.V.H. Clinical, laboratorial, microbiological, virological and instrumental (LP, CT, MRI) investigations were performed. The patient was followed up to 5 year after the beginning of the illness.

L.V.H. was presented to the Department of Child Infection Diseases with sudden onset of fever up to 38°C, muscle and stomach aches, bilateral frontal headache, nausea, vomiting. She became drowsy, with unsteady gait, with numbness and tingling in all four extremities. She felt exhausted. Past medical history was not significant for a preceding infectious illness or recent immunizations. Approximately a week ago she complained of sore throat.

The girl was not in contact, but performed verbal demands. Her skin was sweated, pale and cold. Pathological deviation from lungs and heart were not determined. The stomach was not tender, liver was enlarged on the 1 cm under the right costal arc. Pelvic reservoirs were out of control. Her mental status when she entered in hospital was altered – she was soporous. On neurological examination her deep tendon reflexes were more extensive bilaterally. Hyperesthesia was demonstrated. She did stereotypical ballistic movements with left limbs. There was a spastic hemiparesis on the right side. Pathological reflexes as Babinski were provoked. On the 3-th day ballistic movements were disappeared. The temperature persisted up to 38,4°C during a week. The face was as a mask with bilateral midriasis. There was observed a mild central paresis of n. facialis. On the 7-th day the meningeal signs were manifested with neck stiffness, upper and lower Brudzinski, bilateral Kernik. Lumbar puncture showed a discreet disturbances.

Blood indexes (complete blood count and some biochemical parameters) were showed in **Table 1.**

Table 1. *Blood parameters*

Time\ Indexes	**1-st day**	**7-th day**	**15-th day**	**30-th day**	**3-th mo**	**6-th mo**	**5-th year**
ESR mm	**7**	**14**	**13**	**9**	**5**	**12**	**10**
Leuc . 106/l	**13,6**	**20,2**	**16**	**12,8**	**7,2**	**5,9**	**6,4**
Hb g/l	**124**	**86**	**96**	**102**	**118**	**129**	**134**
ASAT UI/l	**11**	**71**	**67**	**47**	**22**	**13**	**17**
ALAT UI/l	**20**	**158**	**157**	**66**	**40**	**23**	**28**
Glucosa mmol/l	**11,5**	**8,4**	**9,1**	**5,6**	**4,3**		

CSF examination revealed slightly elevated albumin level and cell counts with a high percentage of lymphocytes. **Table 2.** The liquor had negative serologic, molecular and/or rapid antigen tests for accessible infectious agents (bacteria, cytomegalovirus, herpes simplex viruses 1, influenza A, Coxackie B) and for oligoclonal bands.

Table 2. *CSF indexes during the hospital period*

Time – day / CSF indexes	**1**	**7**	**10**	**21**
Albumin g/l	**0,16**	**0,64**	**0,12**	**0,15**
Cells – Leuc.106/l	**20**	**28**	**18**	**2**
Sediment % Ly	**62**	**65**		
Glucosa mmol/l	**4,6**	**3,7**	**3,4**	**3,6**

With recovering of consciousness, improvement of the neurological status was reported, spasticity was slowly decreased, muscle strength was improved. There was visual disturbances – absence of vision, achromatopsy which lasted 10 days. On the 15-th day the status was more improved – child was smiling and talking with desire. Meningeal signs were disappeared.

There were observed some complaints of diencephalon type such as hot waves, periodic face redness, temperature peaks up to 39°C and following sweetness. In the hospital an infection of urinary tract caused by P. aeruginosa was proved – most probably catheter-associated.

S. aureus from nasal swab was isolated.

CSF electrophoresis was not proved oligoclonal IgG chains.

Therapy was performed with antimicrobial medicine, osmotic diuretic, high doses corticoids and immunovenin and others**. Table 3.**

Table 3. *Medicines used in case and its duration*

DRUGS	DOSE	DURATION
Ceftriaxone	**2 x 1g IV**	**14 days**
Sol. Mannitoli 10%	**4 x 250 ml IV**	**5 days**
Immunovenin	**x 400 mg/kg IV**	**2 - five days courses**
Urbason	**x 30 mg/kg IV**	**3 - five days courses**
Dexamethason	**4 x 4 mg IV**	**5 days**
Nootropil	**2 x 400 mg PO**	**38 days**
Milgamma	**3 x 1 caps.**	**24 days**

Individual cynesitherapy was started immediately.

At the discharging the patient felt good, on neurological examination her deep tendon reflexes were still a little bit more extensive on the right side. The muscle tonus of the four limbs was lightly increased, the gate was slightly unstable without coordination disturbances.

On the 4-th day after the illness beginning MRI – native and with contrast (Magnevist) – was performed. It revealed supratentorial hiperintense lesions of multifocal distribution of T2 bilateral and more at frontal, parietal and occipital lobe on the right side. After a contrast enhancement a hardly perceptible capitation of Magnevist was observed in subcortical wite matter localized frontal and parietal lesions. These date were typical for ADEM.

On the 1-th month MRI showed the same, but less intense.

On the 6-th month there were no already pathological findings.

Cathamnestic following showed enhanced irritability, fast tiredness, complaints described as "leg heaviness", which persisted approximately 3 months. Last medical examine done 5 years after the acute phase of the disease demonstrated a teenager on good physical and mental condition. Complete blood count test presented no deviation. The patient attended school and spared not phisical efforts

DISCUSSION

Our patient was 10-year old. ADEM is more common in children, with a mean age of 7.1 years and a peak range of 3—10 years (5). She was female. As is known Female adolescents with ADEM are more likely than males to develop MS in the future (5), though there is no strong scientific theory to explain this occurrence. The disease began at December. There is a seasonal risk with a peak in the winter and spring; the lowest incidence occurs in July and August (6).

The definition of ADEM had not been well defined yet. Leading researchers provided criteria to facilitate diagnosis including:

1. History of recent infection, although it may have been clinically silent.
2. Monophasic disease course.
3. Disseminated CNS disease with neurologic findings.
4. Absence of metabolic or infectious disorders. (7)

Evidently our patient reached these criteria. She had not a history with predisposing conditions except a probably mild angina. The disease had a monophasic course with disseminated feature of neurological signs. The results of lumbar puncture showed clear liquor under increased pressure. In regard to ADEM, the CSF can be normal or, more likely, show nonspecific changes including an elevated protein and pleocytosis with lymphocytic predominance, a normal glucose, and negative cultures (8). In case of LVH findings were with a mild proteinorachia and slight pleocytosis. Laboratorial findings were on **table 1**.

Microbiological investigations did not show patogenetic microbes.

T2-weighted images showed ADEM lesions that are more pronounced with poorly defined margins in deep white matter and periventricular sparing.

MRI defined such a lesions in our patients since 4-th day, then they gradually decreased up to the 3-th month. On the 6-th month there was no pathological sighns.

ADEM is a diagnosis of exclusion. In differential diagnosis we discussed:

1. Viral encephalitis (Entero 71, Coxackie B, CMV)
2. Hyperkinetic encephalitis
3. MS

ADEM tends to have neurologic signs and symptoms not typically present in MS, such as he adache, nausea, vomiting, drowsiness, and meningismus. It is also important to differentiate between ADEM and viral encephalitis. Like MS, this distinction is not always apparent. ADEM may also be associated with visual loss in one or both eyes and spinal cord involvement these problems are also uncommon in encephalitis (8).

The therapy was provided as modern science attitude required.

The usual prognosis is excellent for most patients diagnosed with ADEM (1,8). Relapses after ADEM are uncommon. Despite of that mental retardation is possible, especially in children younger than 2 years of age at disease onset (3). L.W.H. was fully recovered without relapses and sequels.

Degree of recovery was not found to be related to the severity of the illness; even children who developed quadriparesis or blindness or went into a coma were able to recover fully (3). Garg found that all the patients in his study that developed MS did so within 1 year of the initial ADEM episode [9].

CONCLUSION

ADEM is a potentially severe demyelinating disorder likely to be increasingly diagnosed as more magnetic resonance imaging studies are performed on patients with acute encephalopathy. Further characterization of the central nervous system inflammatory response will be needed to understand ADEM pathogenesis, to improve diagnostic and treatment strategies and to distinguish ADEM from MS.

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