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RESEARCH ARTICLE

ACUTE DISSEMINATED ENCEPHALOMYELITIS FOLLOWING VARICELLA INFECTION

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Abstract

Inflammatory demyelinating disease affecting the central nervous system(CNS). Early treatment is the key to neurological recovery. We reportthe case of a 4-year-old girl who developed cerebellar ataxia one week after a varicella rash. Cerebral MRI revealed bilateral lesions of the white matter in favor of ADEM. Corticosteroid boluses associated with antiviral treatment were administered and resulted in a remarkable clinical improvement. Long-term follow-up is essential for assessing prognosis.

Observation:-

A 4-year-old child YA, with no significant medical history, was admitted to the pediatric emergency department due to ataxia.

Sixteen days before the onset of neurological symptoms, generalized vesicular-papular lesions were diagnosed as chicken pox. The progression was characterized by the development of walking and balance disorder.

Upon clinical examination, the patient was conscious, afebrile at 36.7 degrees Celsius.

Neurological assessments revealed static ataxia, drunken walking, negative Romberg with oscillations in all directions, axial hypotonia and segmental muscular strength reduced to 3/5 in both upper and lower extremities. Bilateral osteotendinous reflexes were brisk and symmetrical.

Dysmetria was noted on finger-to-nose tests on both sides of the body. The patient exhibited slow speech and behavioral disturbances, with no signs of meningeal irritation.

Skin examination disclosed diffuse crusty scarring throughout the body.

Cerebral magnetic resonance imaging (MRI) revealed bilateral and symmetrical lesions in the deep and peripheral supratentorial white matter, as well as in the cerebellar peduncles (Figure1) in favor of acute disseminatedencephalomyelitis (ADEM).

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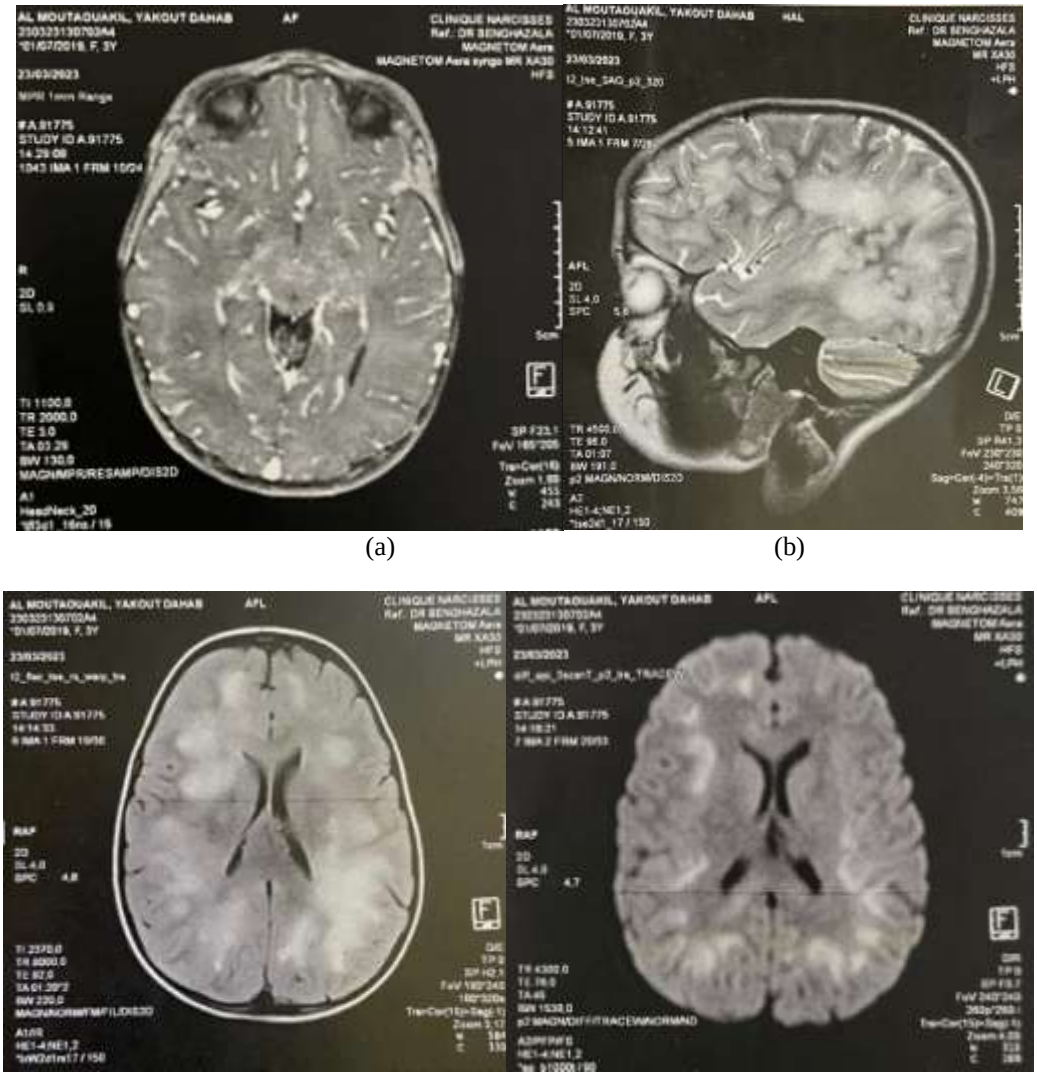

(a) (b)

(c) (d)

Figure 1:- Brain MRI image showing areas of hyperintensity involving subcortical white matter of both cerebral hemispheres, and pedunculus cerebellaris Medius bilaterally in Axial T1 (a),Sagittal T2 (b), Axial T2-FLAIR (c), and Axial DWI (diffusion-weighted imaging).

Lumbar puncture showed clear, acellular cerebrospinal fluid with normal glucorachia and proteinorachia. Culture resulted was sterile, and meningeal polymerase chain reaction (PCR) was negative.

The diagnosis of ADEM following varicella was retained.

The patient received treatment with a 1g/1.73m2/d intravenous methylprednisolone bolus for three days. Acyclovir was administered at 20 mg/kg every eight hours for a total of 14 days.

The resolution of symptoms was spectacular; marked by the recovery of muscular strength,normalized walking, and improved voice.

Discussion:-

Acute disseminated encephalomyelitis (ADEM), also called post-infectious encephalitis, is an acute inflammatory demyelinating disease of the central nervous system,presenting with altered consciousness, neurological deficits, and white matter lesions on MRI [1].

The occurrence of CNS complications following varicella is well-documented, with cerebellar ataxia and hypotonia being the most common neurological manifestations [2.3].

The time interval between the onset of the varicella rash and neurological symptomsvaries from several days before to two weeks after the eruption [4].

In our case, neurological signs appeared one week after the initiation of the varicella rash.

Acute disseminated encephalomyelitis was the most likely diagnosis in our report, due to the rapid development of neurological dysfunction after the rash and subsequent deterioration.[4] It has been attributed to immune-mediated post-infectious demyelination or direct viral invasion. [5]

Cerebral MRI in our case revealed multifocal lesions predominantly affecting the white matter, consistent with ADEM.

Acellular clear cerebrospinal fluid with sterile culture and negative meningeal multiplex PCR was observed in this patient.

Although the presence of varicella zoster virus (VZV) DNA and anti-VZV antibodies in CSF typically supports the diagnosis, their absence in our case suggests an indirect role of VZV in ADEM [5].

Thefirst-line treatment forADEMtypically involves intravenous methylprednisolone (1g/1.73m2/day), for 3 to 5 days, alongside antiviral therapy using intravenous acyclovir at 20 mg/kg/8 hours. However, the efficacy of antiviral treatment remains unclear. Some authors have reported that acyclovir is indicated due to disease severity, [3,6] while others have not recommended it, as it may promote additional viral replication and progressive damage [6 ,7]

Acute disseminated encephalomyelitis is usually considereda monophasic disease, but recurrent cases have been reported in the literature.[8]

Our patient’s acute onset with a single clinical episode classifies the condition as monophasic.Recurrence in subsequent follow-ups would prompt consideration of alternative diagnoses such as multiphasic ADEM or recurrent ADEM [5].

Predicting treatment response and disease progression has led to the development of scoring systems, emphasizing the importance of long-term follow-up for better management [9].

Conclusion:-

A positive treatment response has been observed in post-varicella acute disseminated encephalomyelitis (ADEM) with the administration of high-dose steroids along with antiviral therapy, leading to significant neurological recovery. The importance of long-term follow-up cannot be overstated in determining the prognosis of such cases [9].

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