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Acute Flaccid Myelitis in A 9- year- old Girl: A Case Report

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Abstract

Acute Flaccid Myelitis (AFM), a polio-like paralysis in children no older than 21, has recently reemerged and increasing numbers of such cases have been reported worldwide since 2012. Accurate and early diagnosis of this condition could help with better management of the disease. A 9- year- old girl with chief complaint of headache, fever and vomiting was subsequently affected by an acute paralysis in upper left limb followed by lower limb paralysis. An enhanced signal in T2 parallel to C2-C7, involving the central cord section in Magnetic resonance imaging (MRI) of the spinal cord and an asymmetrical neuropathy pattern reported in Electromyography (EMG), and nerve conduction velocity (NCV) tests led to diagnosis of AFM. Lack of efficacy of administered Intravenous immunoglobulin and no evidence of spastic paralysis in the three months follow up of the patient helped with definite elimination of Guillain-Barre´ syndrome (GBS) as the possible cause, and confirmed AFM diagnosis. Unfortunately, there is no specific treatment currently available for AFM.

Key Words: Acute Flaccid Myelitis, Acute Flaccid Paralysis, Children, Neurologic condition.

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1- INTRODUCTION

Since poliomyelitis has been eradicated, acute flaccid myelitis (AFM), initially referred to as "polio-like disease", has rarely been observed. The term "acute flaccid myelitis" was then adopted to refer to this condition so that it can be distinguished from classical poliomyelitis. Increasing numbers of severe cases with flaccid paralysis with very similar clinical manifestations to those of poliomyelitis have been reported worldwide since 2012; however, the causes were different and mostly unidentified. These cases were unusual and concerning in terms of disease manifestation and its numbers (1).

Increased incidence of AFM in California and Colorado in 2014 drew epidemiologists' attention and had them propose a case definition to better track the number of patients with this clinical condition. The definition included "patients not older than 21 years of age, with acute onset of focal limb weakness and an MRI study showing a spinal cord lesion largely restricted to the gray matter" (2, 3). Acute flaccid myelitis was also declared a reportable disease by the Center for Disease Control and Prevention (CDC) in August 2014 so that the sudden increase of the cases could be further understood (4). A detailed medical history, MRI imaging and ruling out similar conditions such as transverse myelitis and Guillain Barré syndrome (GBS) as potential causes are of great importance in differential diagnosis of AFM (5).

Unlike transverse myelitis, only motor function is affected in AFM in which the second motor neuron is damaged without involving the first motor neuron or sensory afferent nerves. No signs of demyelination are observed in AFM in contrast to Guillain Barré syndrome. In this paper, we present a case of AFM, a recently reemerging and alarming illness.

2- CASE REPORTS

A 9-year-old girl complaining of pulsing headache for two days following a 1-day history of fever and vomiting was presented to the hospital. The patient was assessed for meningitis signs and showed positive signs of meningeal irritation at the time of referral to the hospital. She was admitted in Abolfazl hospital, Kashmar, Iran, with primary diagnosis of meningitis and necessary treatment was started. Three days after the onset of the treatment, disability in holding her head and neck was observed which was followed by an acute paralysis in upper left limb and a subsequent paralysis in lower limb. The patient was then transferred to the Intensive Care Unit (ICU). A brain and neck computed tomography (CT) scan was carried out which revealed no abnormality. She was transferred to the neurology center for further investigation.

A lumbar puncture (LP) was performed. Cerebral spinal fluid (CSF) data was normal except for slightly elevated protein level (white blood cell =zero, red blood cells =150, sugar=73, PR=69%). CSF culture was also normal and no growth was observed. Assessment of serum immunoglobulin and other tests such as HIV, brucellosis, VDRL, blood and stool culture were carried out, all of which were normal. Electroencephalography (EEG) test also showed normal results. Magnetic resonance imaging (MRI) of the brain and neck showed an enhanced signal in T2, in long segment and parallel to C2-C7, involving the central cord section and leading to a slight expansion of the cord (**Figure1**). The patient was examined in terms of viral Antigen (Ag) in stool culture two times which showed negative results in both tests. An asymmetrical neuropathy pattern was reported in Electromyography (EMG), and nerve conduction velocity (NCV) tests, proposing sequel of polio-like infection and rare variants of Guillain-Barre´

syndrome (GBS) as possible diagnoses. Intravenous immunoglobulin (IVIG) was administered in two consecutive days. However, it was not effective, which weakened the probability of GBS being the diagnosis of the condition. Ultimately, acute flaccid myelitis was presented as the final diagnosis considering all observations. The patient was prescribed prednisolone and was discharged from hospital. However, she was followed up for three months; no spastic paralysis was observed, persistent flaccid paralysis remained in the upper left limb while other parts of the body showed improvement. These final observations ruled out GBS and confirmed AFM as the final diagnosis.

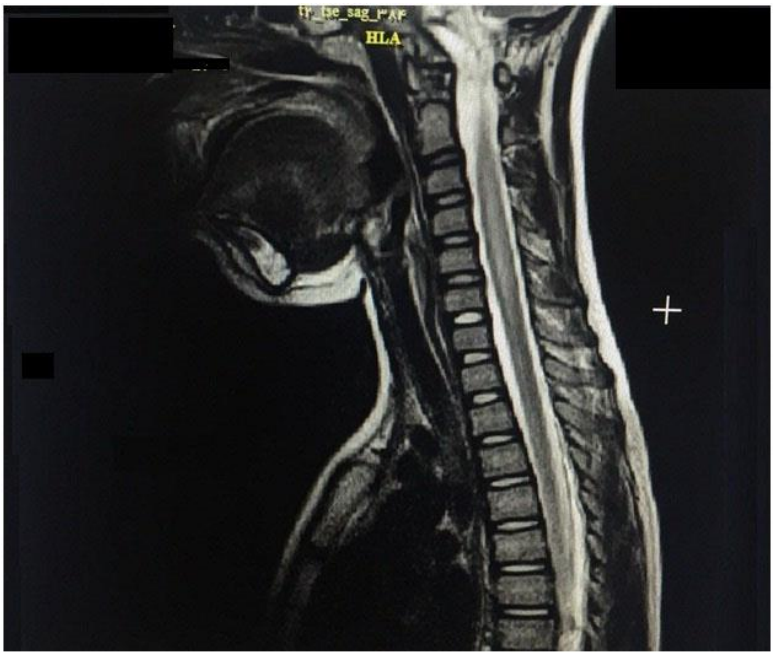

Fig.1: MRI of the spinal cord.

3- DISCUSSION

Alarming increase of AFM cases in recent years has made it necessary for emergency physicians to differentiate between AFM and other similar conditions. An accurate history, excellent neurological examination, and MRI findings help with the differential diagnosis of the underlying etiology of acute flaccid paralysis (3, 6). The present case was a 9- year- old girl with headache, fever, vomiting and positive signs of meningeal irritation at the time of referral to the hospital, which was finally diagnosed with AFM after thorough examinations and tests. Febrile respiratory illness in the two weeks prior to the onset of neurologic symptoms has been observed in many children diagnosed with AFM (7). Analysis of 120 cases of AFM occurred in 2014 in the United States showed that 56%, 25% and 9% of the patients had fever in combination with respiratory tract infection, only respiratory tract infection and only fever respectively in the days leading up to limb weakness (8). Sensory nerves are not typically affected in AFM and the condition generally appears as an acute focal spinal cord paralysis, in contrast to GBS, which often appears as an ascending paralysis, and transverse

myelitis that presents with motor and sensory loss, frequently with a sensory level (9, 10). In the present study, a progressive descending flaccid paralysis was started in three days with an acute paralysis in the upper left limb and a subsequent paralysis in the lower limb.

In the above mentioned study by CDC in the US in 2014, neurological symptoms were observed in 0 to 18 days after the respiratory tract infection or fever, with a median interval of 5 days. Of all patients, 34%, 23% and 43% were affected in the upper extremities, the lower extremities and both the upper and lower extremities respectively and 47% of the patients clearly showed a one-sided asymmetrical pattern (8). CT scan, Chest X-Ray, CSF analysis, blood culture, EEG and other bacterial and viral tests including HIV, brucellosis and viral tests in stool sample were all normal. CSF cytology of AFM cases often shows a pleocytosis, but may be normal (3); while more than 90% of recent cases have followed a mild viral infection, cause of most cases is unclear (11), since AFM is a clinical condition with several possible etiologies (6).

A variety of factors such as environmental toxins, genetic disorders and viruses including poliovirus, non-polio enteroviruses, West Nile Virus, and adenoviruses have been suggested as possible causes of AFM since 2018 (12). Although it is possible to detect pathogens in stool culture or respiratory secretions, this does not prove a pathogenic relationship (1). In our case, MRI showed an enhanced signal in T2, parallel to C2-C7 which involved the central cord section and led to a slight expansion of the cord. An asymmetrical neuropathy pattern was reported in Electromyography (EMG) and nerve conduction velocity (NCV) tests. Intravenous immunoglobulin (IVIG) was administered twice, eight and nine days after the onset of weakness; however, it did not work. The patient was discharged from hospital with prednisolone prescription and diagnosis of AFM. She was followed up for three months and except for the upper left limb, which still suffered from a flaccid paralysis; other parts of the body were improved. No evidence of spastic paralysis was observed during the follow up period, which in turn reconfirmed ruling out GBS. Intravenous immunoglobulins (IVIG), and plasmapheresis are the two main immunotherapy treatments for GBS but there is no clear indication for efficacy of IVIG, plasmapheresis, corticosteroids or antiviral medications in the treatment of AFM (1). Esposito et al., reported a significant reduction in the signs and symptoms of AFM in their case, a 4-year-old boy, after 4 weeks of treatment, which included 3 days of plasmapheresis and IVIG from the first day of weakness report, stopping Intravenous steroid therapy after 5 days and administrating prednisolone for 6 weeks (tapered in the final two weeks) (13).

In a study done by Chen et al., IVIG was used twice with pulse steroid therapy between the 2 IVIG administrations from the fifth day after reporting weakness; however, the weakness progressed (14). IVIG was also used in the present study like the two abovementioned cases, however different results were observed. Although the efficacy could not be attributed to a single treatment, since a combination of treatments were attempted in these studies, it seems that IVIG and pulse steroid therapy might be effective when used during the early stages of AFM.

4- CONCLUSION

Acute flaccid myelitis, a previously rare syndrome, is alarmingly reemerging worldwide. Case reports of this condition can both help the emergency physicians and pediatricians with more accurate and rapid differential diagnosis of the disease and help with further investigation of the

etiology of this sudden increase of AFM incidence, which in turn paves the way for developing effective treatments. Currently there is no gold standard treatment for AFM. Although steroids, intravenous immunoglobulin, or plasma exchange have been suggested based on the initial experience from the clinical events in 2014, further studies are required to reach a standard and widely accepted therapy.

5- CONFLICT OF INTEREST: None.

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