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IS IT REALLY TRANSVERSE MYELITIS? ACUTE FLACCID MYELITIS A RARE ENTITY

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ABSTRACT

Acute Flaccid Myelitis is a relatively new and rare disease entity which was recognized for the first time in United States in 20121. Usually children below 18 years of age present with sudden onset rapidly progressive flaccid paralysis sometimes following upper respiratory tract infection. To date no cases have been reported from Afghanistan and Pakistan. We are reporting case of a young girl who presented with sudden onset flaccid paralysis and respiratory distress. Her MRI Cervical spine showed longitudinally extensive transverse myelitis. One year after the onset of disease flaccid paraparesis continued despite various treatments. It is important to report cases of acute flaccid myelitis as it is a new disease with poor prognosis. People including clinicians are not aware of it.

Mesh: Myelitis, acute flaccid paralysis, flaccid

INTRODUCTION: Acute Flaccid Myelitis is a rare but extremely disabling condition, for which no treatment is available so far. Mostly children present with rapidly progressive flaccid paralysis. Sometimes swallowing and breathing difficulties accompany. The initial spinal MRI scans show longitudinally extensive lesion like Transverse Myelitis. Different treatments including steroids and IVIG have been tried without any success.

Case Description:

A 14-year-old girl from Afghanistan presented to a local Hospital in Peshawar in March 2016. Her symptoms started in Afghanistan. She had a 10 days history of flu like illness followed by lower backache and sudden onset lower limb paralysis. Two days later upper limb weakness started with respiratory distress. Speech, swallowing, bladder and bowels were normal. There was no history of exposure to sick individuals, toxins, insect bites, rash, joint pains, headache, vomiting or visual disturbance. Family history was negative for any familial diseases, vasculitis, childhood paralysis/malignancies. She was not taking any regular medications before this. On arrival in hospital she was intubated secondary to respiratory distress. She was then transferred to Shifa International Hospital, Islamabad for further management. On examination she was afebrile, blood pressure was 120/82 mm/Hg. Her higher mental functions and cranial nerves including fundoscopy were normal. There were no signs of meningism. She had reduced tone in all 4 limbs. Power was 1/5 in both upper limbs and 0/5 in both lower limbs with absent reflexes and mute plantars. Superficial abdominal reflexes were also absent. However, she had normal pin prick, position and vibration sensations. Rest of systemic examination was unremarkable.Her Complete Blood Count (CBC), Electrolytes, renal functions and liver function tests were all in normal range. Her Cerebrospinal Fluid (CSF) showed White Cell Count 980/ul, Lymphocytes 93%, Proteins 104mg/dl, Culture: Negative. Viral serology was not done due to non-availability. MRI cervical spine showed longitudinally extensive hyperintense lesion on T2/FLAIR extending from C3-C7 (See Figure A).

Diagnosis of Transverse Myelitis of uncertain etiology was made. She was pulsed with high dose Methylprednisolone 1g/day for 5 days but without any improvement. Subsequently, Intravenous Immunoglobulins (IVIG) 0.4g/kg/day for 5 days was given. Repeat CSF showed WCC 144/ul, Lymphocytes 90%, Proteins 23mg/dL. Serum Aquaporin 4 and ENA profile were negative. Despite treatment her clinical condition did not improve. She received Rituximab 500mg without any benefit. The family took her to India for a second opinion in September 2016, where shehad repeat cervical spine imaging. It showed complete resolution of the hyperintense lesion previously seen on MRI (See Figure D). She received a second dose of Rituximab 500mg. She was again reviewed by Neurology team in Shifa International Hospital in May 2017. During this time, she had

remained bed bound, fully dependant on her family for all activities of daily life. As before there were no bladder or bowel complaints. She continued to have flaccid paralysis with marked muscle wasting and occasional fasciculations. Power had improved to 2/5 in both upper limbs. But remained 0/5 in both lower limbs. Reflexes were absent and plantars were mute bilaterally. Sensations were normal bilaterally. Nerve Conduction Studies and Electromyography (NCS/EMG) was done which showed severe neuropathy/neuronopathy with active denervation. There were normal motor units in First Dorsal Interosseous and absent in lower limbs.
(See Table 1 below)

Nerve Conduction Studies (Table 1)

Nerve Tested	Site	Amp (mv)	Conduction Velocity	F wave
Median APB	Wrist	8.6	54	22.7
	Elbow	7.3		
Ulnar ADM	Wrist	12.7	58	22.3
	Below Elbow	12.3	63	
	Above Elbow	11.2		
Tibial	Ankle	Not Recordable	-	-
	Knee	NR	-	-
	Ankle	NR	-	-
Peroneal EDB	Ankle	NR	-	-
	Below Knee	NR	-	-
	Above Knee	NR	-	-
H Reflex		NR	-	-
Nerve Tested		Latency (m)	Amp (mv)	Conduction Velocity
Median		2.8	111	62
Ulnar		2.5	96	61
Radial		1.8	97	91
Sural		3.7	37	47
Superficial Peroneal		3.0	31	64

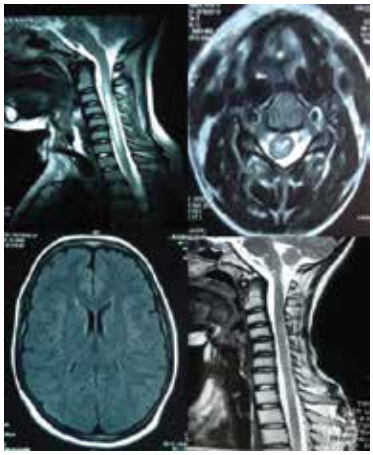
She did not come back for any further follow up.
These findings and clinical course are like other cases of acute flaccid myelitis where patients present with rapidly progressive flaccid paralysis without any improvement despite aggressive treatment with high dose steroids, IVIG and plasmaphresis.

Discussion and Conclusion:

Acute Flaccid Myelitis is a relatively new and rare disease entity, effecting children ranging from 22 months to 15 years. The illness usually begins with a prodromal phase of febrile illness with flu like symptoms, usually followed by headache, neck stiffness and backache. The patients then develop rapidly progressive lower motor neuron type limb weakness reaching nadir within hours to days[2,3,4,5]. Cranial nerves can be involved in some cases.Respiratory distress is the most severe symptom which requires ventilatory support. Bowel bladder involvement can also occur occasionally. Most patients have mild-to-moderate CSF pleocytosis (WCC Range 0-888) on initial lumbar puncture, with lymphocytic predominance in most cases [2,3,4,5]. CSF protein is mildly elevated with normal glucose[2,3,4]. Oligoclonal bands and Aquaporin 4 antibodies are negative. MRI Spine shows longitudinally extensive lesions in gray matter which are non-enhancing with gadolinium contrast6. Gradually the signal abnormality becomes more well defined within the anterior horn cell region6. After weeks to months, most spinal cord lesions resolve and nerve root enhancement not initially present on early imaging become apparent in some cases[4,5,6].
Electrodiagnostic studies demonstrate a motor neuropathy or neuronopathy with no sensory abnormalities[2,3,7].To date its etiology remains unknown3. It is a notifiable disease in USA. Antivirals and immunomodulatory drugs like high dose steroids, IVIG and plasmaphresis have been tried without success. Gradually many patients develop muscle wasting. Most of the patients do not show any significant recovery. Supportive care remains the mainstay of treatment.

Conclusion:

Acute Flaccid Myelitis should be considered in the differential diagnosis of patients presenting with acute flaccid weakness. Most clinicians are unaware of this newly described entity. Proper diagnosis is important in counselling and prognostication.



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Hina Yusuf; data collection, data analysis, manuscript writing, manuscript review
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