P 1-59

Rapid recovery in Miller-Fisher syndrome in a child with poor prognostic factors: A Case report

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Introduction

Guillain Barre Syndrome(GBS) is known to be a syndrome with several variant forms. Of those variant, Miller Fisher Syndrome(MFS) is not common in children and characterized by double vision, loss of balance and deep tendon reflexes. We present a child who had MFS with abrupt onset of profound weakness with multiple conduction blocks with good prognosis.

Case report

A 6-year-old male patient came to the emergency room. He complained of abrupt gait disturbance and left side weakness, difficulty in left lateral gaze, intermittent dysarthria and presented diplopia. Initial chest X-ray showed pneumonia and two days later, high fever up to 39.9℃was checked. Intravenous immunoglobulin therapy started, but his symptoms gradually worsened and five days later, the patient became tetraplegia and fell into respiratory failure, so ventilator care was started. Steroid pulse therapy was started just before the EMG, that is, 8 days after the onset. Muscle strength was generally trace grade according to the manual muscle test. Deep tendon reflex of upper limb and bilateral knee jerk were hypoactive, and ankle jerk was absent. Nerve conduction study (NCS) and electromyographic (EMG) examination were performed 9 days after symptom onset. Sensory NCS showed decreased amplitudes of sensory nerve action potential(SNAP) in bilateral sural and right ulnar nerves. Motor NCS presented drop in amplitudes of compound muscle action potential(CMAP) in right median, peroneal and bilateral tibial nerves, delayed conduction velocities in right median and bilateral tibial nerves(Table 1). Needle EMG and facial ENoG showed no definite abnormality. And there was no response of F wave in all sampled nerves. Above electrodiagnostic findings are compatible with inflammatory demyelinating polyneuropathy(AIDP). Comprehensive rehabilitation therapy including gait training, fine motor training and balance training was continued. After a month, the patient was able to walk independently. Still, mild impairment of balance and fine motor in upper extremities was remained, but he participated nearly all activities of daily living including running.

Conclusion

Early diagnosis of AIDP is crucial because it sometimes is life-threatening, but several treatments could lessen the disease severity and improve outcome. Generally, the prognosis of AIDP in children is better than adults. However, outcomes may be less favorable those with some risk factors. Such as child younger than 2 years, limb paralysis within 10 days, very weak at presentation, unevoked motor nerves on NCS, the

involvement of cranial nerves, and requiring ventilator support. In this case, despite the patient presented several risk factors for poor prognosis, the patient gradually recovered satisfactorily over a few weeks. This recovery was probably due to early diagnosis and rapid treatment.

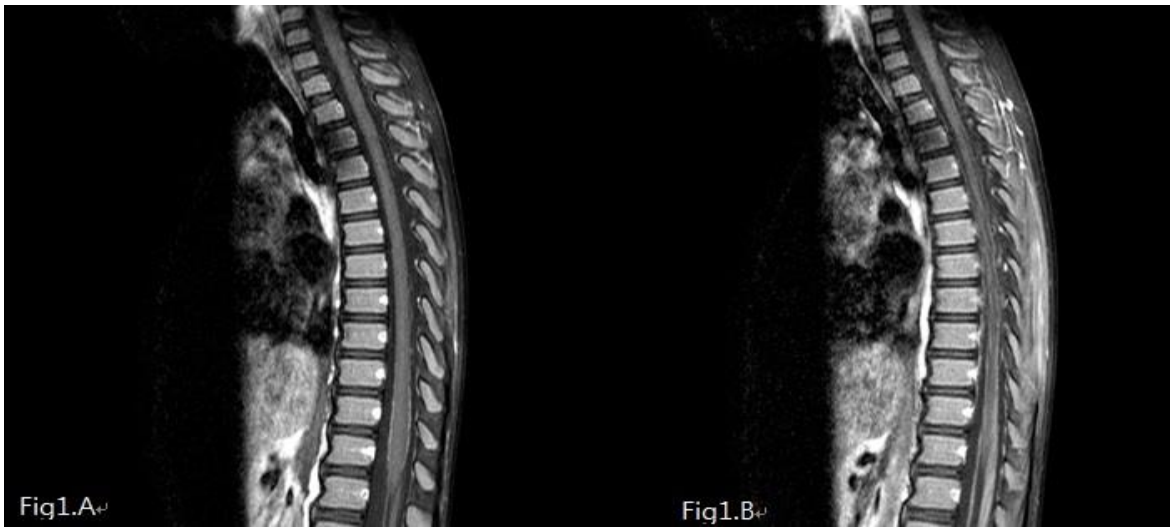
table1. The results of needle EMG and F-wave studies

EMG Summary Table

	Spontaneous					MUAP			Recruitment
Muscle	IA	Fib	PSW	Fasc	CRD	Amp	Dur	PPP	Pattern
R. Biceps brachii	N	None	None	None	None	N	N	N	N
R. Vastus medialis	N	None	None	None	None	N	N	N	N
R. Gastrocnemius (medial)	N	None	None	None	None	N	N	N	N

F-wave

Nerve	F min (ms)
R Median - APB	No Response
R Median - APB	No Response
L Tibial - AH	No Response



Magnetic resonance imaging of thoracic and lumbar spine. A. Normal conus medullaris at D12-L1 was observed B. Diffuse enhancement of cauda equine was present and definite nodularity or enlargement was not observed.

Sensory NCS

Nerve / Sites	Onset Lat ms	Peak Lat ms	Pk Amp µV	Distance mm	Velocity m/s
R Median - Digit III					
Palm	1.20	1.82	17.5	70	58
R Ulnar - Digit V					
Wrist	1.51	2.29	11.1		
R Sural - Ankle (Calf)					
Calf	1.82	2.71	7.5	90	49
L Sural - Ankle (Calf)					
Calf	1.72	2.60	5.6	90	50
R Superficial peroneal – Ankle					
Lat leg	1.77	2.71	9.5	90	48
L Superficial peroneal – Ankle					
Lat leg	2.08	3.02	9.4	90	43

Motor NCS

Nerve / Sites	Latency ms	Amplitude mV	Duration ms	Distance mm	Velocity m/s
R Median - APB					
Wrist	2.19	6.6	4.90		
Elbow	6.67	1.4	9.48	155	35
R Ulnar - ADM					
Wrist	1.93	5.9	5.68		
B.Elbow	4.43	4.0	5.73	125	50
R Peroneal - EDB					
Ankle	5.05	1.7	10.83		
Fib head	8.18	0.9	14.90	190	61
L Peroneal - EDB					
Ankle	4.22	2.8	8.13	70	
Fib head	8.13	2.7	9.06	200	51
R Tibial - AH					
Ankle	3.02	11.1	8.44	80	
Pop fossa	24.11	0.1	2.81	250	12
L Tibial - AH					
Ankle	3.44	10.2	8.28	80	
Pop fossa	12.76	0.3	6.56	250	27

ENoG

Nerve / Sites	Onset Lat (ms)	Pk Amp (mV)	Duration(ms)	Area (mVms)
R Facial - Fonralis,Orb Oculi, Nasalis,Orb Oris				
Frontalis	5.68	0.4	8.65	1.6
Oculli	2.66	0.7	7.08	2.6
Nasalis	3.33	0.5	7.40	2.0
Oris	3.44	0.8	19.95	5.8
L Facial - Fonralis,Orb Oculi, Nasalis,Orb Oris				
Frontalis	5.63	0.5	15.63	2.9
Oculli	2.66	0.9	7.08	2.6
Nasalis	3.23	0.9	8.07	2.9
Oris	3.70	0.6	12.55	3.5

The results of sensory, motor and facial nerve conduction studies