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Sensory neuronopathy with ataxic gait and fine motor difficulties: A case report

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Background

Sensory neuronopathy is known to be occurred by various causes including genetic disease, paraneoplastic syndrome, HIV infection, Sjogren's syndrome, intoxication, vitamin E deficiency, and even idiopathic origin. We report a patient of idiopathic sensory neuronopathy with ataxic gait and fine motor difficulty relapsing for five years.

Case

A 73 year old male patient with waxing & waning ataxic gait for 5 years was admitted in Neurology. In nerve conduction study, motor conduction study showed normal results in his four extremities, while the sensory nerves were not provoked in all extremities. After taking steroid and mycophenolate, his symptom was improved and he was discharged. Nine months later, he was admitted again with relapsing ataxic gait and fine motor difficulties. His clinical diagnosis was chronic inflammatory demyelinating polyneuropathy (CIDP) and they performed several laboratory studies and computed tomography (CT), which were all not remarkable. After immunoglobulin therapy, he was transferred to rehabilitation medicine department for proper evaluation and rehabilitation. We evaluated his muscle power and it showed grade 4 in all extremities. Berg Balance Scale score was 23/56. His gait was ataxic, which showed sway during ambulation and wide-based to keep his balance. He had difficulties in fine motor control. In the monofilament test, it showed decreased sensory protective function in the bilateral median, ulnar, and radial sensory area. He showed decreased vibration sense and position sense. We repeated motor and sensory nerve conduction studies and it showed same results with those of previous study.(Table 1, 2) We performed needle electromyography study to know the existence of axonal involvement in motor nerves, and there was no spontaneous activities and all muscles tested showed normal voluntary motor unit action potential. We concluded that his ataxic gait and fine motor difficulties were from sensory neuronopathy/ganglionopathy rather than CIDP. His ataxic gait was improved, his fine motor function was significantly improved, and he could use his chopsticks after 3 weeks of rehabilitation.

Conclusion

Sensory neuronopathy/ganglionopathy, a subset of peripheral neuropathy, is thought to be related with the degeneration of dorsal root ganglion (DRG). It is usually diagnosed as 'sensory ataxia' in neurology clinic, and the patient shows ataxic gait, proprioceptive sensory loss, decreased muscle reflex, and difficulties in fine motor controll. In nerve conduction studies, motor conduction study shows normal or reveals only mild

abnormalities and SNAPs are absent. We confirmed the patient's diagnosis with the motor and sensory nerve conduction study. The limitation of this study is that we could not perform MRI study of spinal cord to know the involvement of ganglion. Proper rehabilitation including occupational therapy helped the patient restoring the gait and fine motor function.

Table 1. Nerve conduction study

Motor Conduction Study

	Nerve	Distal latency(㎳)	Amplitude(㎷)	Conduction velocity(㎧)
Rt	Median	3.07	12.5	50.5
	Ulnar	2.50	10.2	52.6
	Tibial	4.17	15.4	42.1
	Peroneal	3.96	3.7	40.1
Lt	Median	3.44	9.1	50.5
	Ulnar	2.29	9.6	52.5
	Tibial	4.69	14.3	40.5
	Peroneal	4.48	3.9	40.3

Sensory Conduction Study

	Nerve	Distal latency(㎳)	Amplitude(㎶)	Conduction velocity(㎧)
Rt	Median	No response		
	Ulnar			
	Sup.Radial			
	LAC			
	Superficial Peroneal			
	Sural			
Lt	Median			
	Ulnar			
	Sup.Radial			
	LAC			
	Superficial Peroneal			
	Sural			

Table 2. Needle Electromyography

Needle Electromyography

Muscles		Insertional Activity	Spontaneous Activity				Voluntary Motor Unit Action Potential			Recruitment Pattern
			PSW	Fib.	CRD	Fas.	Amp.	Dur.	PPP	
Bil.	Biceps		0	0	0	0	N	N	N	C
	Flex. Carpi Radialis		0	0	0	0	N	N	N	C
	1st Dorsal Interossei		0	0	0	0	N	N	N	C
	Abd. Pollicis Brevis		0	0	0	0	N	N	N	C
	Tibialis Anterior		0	0	0	0	N	N	N	C
	Gastrocnemius		0	0	0	0	N	N	N	C