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A Case of Acquired Chronic Inflammatory Demyelinating Polyneuropathy

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ABSTRACT

BACKGROUND AND IMPORTANCE

This case report is of a patient who is suffering from an unclear autoimmune demyelinating polyneuropathy with progressive weakness in upper limb with no sensory involvement. Her symptoms did not resolve after initial steroid treatment but after a course of IVIG, her symptoms improved. On her next presentation, she also had sensory involvement and IVIG treatment led to her recovery.

CLINICAL PRESENTATION

A 19 years old female presented with progressive upper and lower limb weakness for last 4 months, inability to walk, gait disturbance, and dizziness. On examination, there was no sensory loss, normal tone and reflexes, decreased power on both side without wasting or fasciculation.

CONCLUSION

Multifocal motor neuropathy and multifocal acquired demyelinating sensory and motor neuropathy have an overlapping presentation with other autoimmune polyneuropathies. The diagnosis is difficult without sensory involvement. Further studies are required to distinguish between these autoimmune polyneuropathies, as the treatment of these diseases is expensive and hazardous if complications occur.

KEY WORDS

Case, Acquired, Inflammatory, Demyelinating, Polyneuropathy .

CASE REPORT

BACKGROUND AND IMPORTANCE

This case report is of a patient who is suffering from an unclear autoimmune demyelinating polyneuropathy with progressive weakness in upper limb and later spreading to lower limbs. Although her

symptoms were successfully managed on both presentation but her diagnosis, remain unclear. Due to absence of contrasting features on presentation, the initial diagnosis of the patient was unclear. After a course of IVIG, the patient shows great improvement. The patient presented again with similar complain after few months now also with sensory involvement. She was again treated with IVIG that led to the recovery of the patient.

CLINICAL PRESENTATION

A 19 years old female with no prior comorbidities presented in ER with progressive weakness in upper limbs for last 4 months, lower limbs for last 2 months and dizziness for last 1 week. At first, it started as weakness in the left hand and later it involved the right hand. She was unable to hold grip from both hands. For last 2 months, she was unable to walk, which started as weakness in lower limb progresses from gait disturbance to unable to walk at present. On further inquiry, all of her developmental milestones were delayed. On examination, bulk, tone, and reflexes in both limbs were intact, but power was decreased in upper limb (3/5 R), (2/5 L) as well as lower limbs 3/5 on both sides. The modality of sensation was also impaired in both upper and lower limbs. The finding of Nerve Conduction Study shows motor polyneuropathy with condition block. From these finding, the probable diagnosis was Lewis Sumner Syndrome or MADSAM. She had similar episode 5 months back but sensation were intact in both limbs in that episode.

All routine labs were normal. MRI brain was normal. MRI cervical spine show osteophytic complexes at C2/C3, C3/C4, and C4/C5 levels with mild disc bulge along with osteophytic disc complex causing thecal indentation. Right-sided neural foramina was narrowed without significant nerve root impingement. The report of NCS shows bilateral symmetrical conduction block in median and ulnar nerves in non-compressive site. These findings were suggestive of multifocal motor neuropathy with conduction block in contrast to MADSAM in this visit. The serum anti GMI antibodies cannot be done because it is not available in our country. CSF Protein was slightly elevated. She was treated with IVIG, which improves her symptoms. She may have further episodes after that, but her

follow-up was lost. Nerve conduction study is shown below in the table.

Nerve Conduction Studies 22-11-2016

Mncv	Site/segment	Latency Ms	Amplitude Mv	Durate Ms	Area Mvms	Distance Mm	Velocity M/s
Medianus r	Abp-wrist	3.4	6.3	6.7	21.1		
	Wrist-elbow	20.1	0.94	21.9	5.6	260	15.6
Ulnaris r	Adm-wrist	6.0	2.6	11.4	16.7		
Tibialis r	Abd brevis-malleolo med	5.2	2.0	8.2	11.0		
	Malleolo med-cavo	29.8	0.411	6.0	1.5	390	15.9
Peroneus r	Pedidio-caviglia	5.4	2.2	7.2	7.3		
	Caviglia-sottocaput	14.0	0.718	8.3	6.4	275	32.2
	Sottocaput-sopracaput	26.9	0.645	27.4	26.4	100	33.5
Peroneus r	Pedidio-caviglia	2.6	2.2	4.4	2.8		
	Caviglia-sottocaput	4.7	1.5	3.8	1.7	100	48.0
Tibialis l	Abd brevis-malleolo med	5.5	3.7	8.4	12.3		
	Malleolo med-cavo	15.0	0.401	8.5	7.2	360	37.8
Peroneus l	Pedidio-caviglia	4.9	3.5	7.2	13.0		
	Caviglia-sottocaput	11.7	1.4	7.2	2.8	270	39.3
	Sottocaput-sopracaput	14.2	1.8	7.2	4.1	100	40.0
Peroneus l	Pedidio-caviglia	2.6	1.9	5.6	4.0		
	Caviglia-sottocaput	4.4	1.3	4.3	4.2	100	55.4
Medianus l	Abp-wrist	3.3	5.5	7.2	11.4		
	Wrist-elbow	12.9	0.344	7.8	4.2	260	26.9
Ulnaris l	Adm-wrist	4.0	2.2	9.1	13.6		
Sncv	Site/segment	latency ms	amplitude mv	durate ms	area mvms	distance mm	velocity m/s
Medianus r							
Suralis r							
Medianus l							

F wave	M latency Ms	F-m latency Ms	F min latency Ms	F max latency Ms	F/m amplitude	F %
Medianus r	3.6	36.4	40.0	59.2	0.05	07
Ulnaris r	0.1	26.0	26.1	…..	0.22	0
Tibialis r	0.1	27.2	27.4	27.6	0.21	37
Peroneus r	5.1	34.9	40.0	…..	0.43	0
Tibialis l	0.1	39.9	40.0	…..	0.64	0
Peroneus l	4.7	35.3	40.0	…..	0.2	0
Medianus l	4.0	36.0	40.0	…..	0.17	0
Ulnais l	3.6	22.9	26.5	26.8	0.18	50

H reflex	M latency Ms	H latency Ms	H/m
Tibialis r	0.3	0.0	2.41
Tibialis l	4.3	40.0	0.22

Nerve Conduction Studies 6-5-2016

Mncv	Site/segment	Latency Ms	Amplitude Mv	Durate Ms	Area Mvms	Distance Mm	Velocity M/s
Medianus l	Abp-wrist	3.4	9.6	10.1	31.4		
	Wrist-elbow	14.2	0.97	27.2	4.3	250	23.2
Ulnais l	Adm-wrist	2.9	8.1	10.6	24.0		
	Wrist-down elbow	6.1	1.4	10.4	5.1	175	54.8
	Down elbow-up elbow	11.1	1.0	20.8	3.6	100	20.0
Tibialis l	Abd brevis-malleolo med	4.2	6.7	6.3	21.3		
	Malleolo med-cavo	12.3	6.1	8.5	14.5	390	48.0
Peroneus l	Pedidio-caviglia	4.3	3.4	6.3	18.2		
	Caviglia-sottocaput	10.1	3.2	14.5	13.1	260	45.1
	Sottocaput-sopracaput	12.1	3.0	15.1	12.5	100	49.7
Tibialis r	Abd brevis-malleolo med	3.9	5.1	8.6	26.9		
	Malleolo med-cavo	11.8	4.4	8.3	12.4	390	49.3
Peroneus r	Pedidio-caviglia	4.5	4.2	6.8	10.9		
	Caviglia-sottocaput	9.9	4.1	5.9	10.0	270	50.5
	Sottocaput-sopracaput	11.8	3.9	11.7	14.1	100	51.4
Medianus r	Abp-wrist	2.8	11.9	7.6	34.2		
	Wrist-elbow	10.0	4.2	14.1	15.7	265	37.0
Ulnaris r	Adm-wrist	2.6	4.6	14.7	20.4		
	Wrist-down elbow	11.4	0.653	24.1	2.3	170	10.4
	Down elbow-up elbow	16.3	0.259	21.8	1.6	100	20.3
Sncv	Site/segment	latency ms	amplitude mv	durate ms	area mvms	distance mm	velocity m/s
Medianus l	Iii finger wrist	3.2	20.1	0.8	3.70	140	43.7
Ulnaris l	Wrist v-finger	3.2	11.9	0.7	3.17	140	43.3
Suralis r	Leg malleolo lat	3.8	19.5	1.6	27.5	140	37.3
Suralis l	Leg malleolo	3.6	18.0	1.5	27.5	140	30.9
Medianus r	Iii finger-wrist	3.2	23.0	2.0	27.2	140	43.7
Ulnaris r	Wrist-v finger	3.3	11.2	2.3	8.36	140	43.5

f wave	m latency m/s	f-m latency m/s	f min latency ms	f max latency ms	f/m amplitude	f %
medianus l	3.2	36.8	40.0	…..	0.07	0
ulnaris l	2.6	37.4	40.0	…..	0.08	0
medianus l	4.6	35.4	40.0	45.6	0.03	14
tibialis l	4.3	35.7	40.0	…..	0.02	0
peroneus l	4.2	35.8	40.0	…..	0.03	0
tibialis r	3.9	36.1	40.0	…..	0.02	0

peroneus r	4.6	35.4	40.0	…..	0.07	0
medianus r	2.9	37.1	40.0	41.3	0.03	100
ulnaris r	2.6	37.4	40.0	…..	0.1	0

h reflex	m latency ms	h latency ms	h/m
tibialis r	4.2	24.0	0.07
tibialis l	4.0	40.0	0.04

DISCUSSION

Multifocal motor neuropathy (MMN), a type of chronic acquired demyelinating polyneuropathy (CADP), is a rare autoimmune demyelinating disorder characterized by muscle weakness and fasciculation more commonly in upper limbs. The diagnosis of MMN is based on clinical, laboratory and electrophysiological studies. It is a progressive asymmetric disease with loss of motor function of distal extremities without sensory involvement [1]. Patients may present with hand weakness, wrist drop, or foot drop, which progresses over the course of several years to involve other limbs .The disease is differentiated from chronic inflammatory demyelinating polyneuropathy by the absence of sensory deficit and the proximal location of Conduction block[2]. Due to the overlapping presentation of this potentially treatable neuropathy, the disease can be sometimes misdiagnosed as motor neuron disease. Electro physiologic studies reveals conduction block away from the usual nerve compression site, which is a hallmark of disease, more commonly in the ulnar (80%) and median (77%) nerves[3,4]. There may be a rise in anti – ganglioside GM1 antibodies in up to 40% of cases and slight increase in CSF protein usually less than 100mg/dl. In addition, the presence of anti-GM1 antibodies or the conduction blocks is not necessary to diagnose MMN[5]. The disease usually responds to intravenous Immunoglobulin and immunosuppressive drugs such as cyclophosphamide but not steroids or plasma exchange. Patients may respond to steroids, but it is no longer considered useful [6] . Cases have been reported of deterioration of disease after steroid treatment and plasmaparesis[7] [8]. Early IVIG treatment has been proven to postpone axonal degeneration and permanent motor deficits. It also improves muscle strength and reduces disability in up to 97% of patients4.

Lewis-Sumner syndrome also known as multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), is also a variant of chronic acquired demyelinating polyneuropathy (CADP), is an autoimmune demyelinating disorder characterized by chronic asymmetric sensorimotor neuropathy usually in the upper extremities[9]. The disease often has insidious onset with slow progression, involving the arms initially with later spread to legs. The Diagnosis of LSS can be established by electro-physiologic evidence of persistent multifocal mixed conduction block distally and nerve biopsy showing demyelinating changes[10]. There are also normal levels of CSF protein and titers of antibodies directed against myelin component. The treatment is corticosteroids and IVIG but response rate may vary from 25 to 79% with corticosteroid and 50 to 67% with IVIG while 10 to 20% of the patients may have refractory disease. Plasma exchange for refractory disease has been reported with dramatic improvement in symptoms of the patient[11]. The distinguishing features of LSS are the presence of sensory involvement, the absence of serum anti-GM1 antibodies[12]. Although cranial nerve involvement is not reported in MMN, 17-31% cases of LSS also show various cranial nerve involvement11 [13].

These two disease seems to have contrasting features but often the diagnosis is difficult to make, as the usual presentation of these diseases is often atypical or overlapping with other neuropathies. Several clinical features like age of onset, distribution of weakness, wasting and fasciculation, conduction block, distal involvement and recurrence is common is both diseases. Without clear sensory involvement, the diagnoses may remain unclear even after treatment. Only few of the above diagnostic features are usually present. Patients lacking the characteristic findings can be managed by their response to immune modulatory treatment.

CONCLUSION

Multifocal motor neuropathy, multifocal acquired demyelinating sensory and motor neuropathy, and other forms of autoimmune polyneuropathies have many common presenting symptoms. The diagnoses of these diseases are often unclear due to their overlapping symptoms and sometimes management is led by their response to

treatment. Without sensory involvement, it is difficult to distinguish between these two diseases. The investigations like anti – ganglioside GM1 antibody test is often inaccessible. Also the existing course of treatment like IVIG and plasma exchange are very expensive and can be life threatening for the patient if complication occurs. Further studies are required to distinguish between these and other similar diseases and to device proper treatment plan.

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