A Case of Relapsing - Remitting CIDP with Sixth Nerve Palsy

Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an immune-mediated neurological illness affecting the peripheral nervous system.[1] It has a diversified clinical presentation categorised as monophasic, relapsing-remitting, or chronic progressive.[2] Cranial nerve involvements are not uncommon in CIDP. Cranial nerves are usually irreversibly affected.[3,4] We report a case of relapsing-remitting CIDP with recurrent sixth nerve palsy.

Case Summary

A 30-year-old male patient presented with a recurrent neurological deficit for the past 13 years. Initially, he developed bilateral lower limb proximal weakness and about two weeks later developed distal weakness of the lower limbs and proximal weakness of upper limbs followed by distal weakness of upper limbs. Simultaneously the patient also developed paresthesia involving bilateral lower limb followed by upper limb. After one month of onset of illness, symptoms started improving, and in about two months, he recovered completely.

In the next five years, he had five similar events, which improved without any treatment. In the next five years, he was symptom-free. In the past three years, he has had four similar events, in one event in addition to his usual symptoms he also had horizontal diplopia. The diplopia was worse on looking at distance and towards the left side. For the above complaints, he was treated with pulsed steroids, and he improved completely within one week. He was on an oral steroid for six months following which he stopped the treatment on his own. After two months of stopping steroids, he developed horizontal diplopia. The diplopia was worse on looking at distance and towards the right side. He also had paresthesia involving both upper limbs and distal weakness of all four limbs. There was no other cranial nerve involvement. No h/o joint pain/rash/hypopigmented or hypoesthetic skin patches/long-term drug use/toxin exposure. No h/o headache/vomiting/fever/bowel/bladder symptoms/erectile dysfunction/lightheadedness/addiction/any comorbid illness. There was no significant family history. His physical examination showed a right sixth nerve palsy, distal weakness of all four limbs and absent deep tendon reflexes. He had a remarkable response to intravenous pulsed steroid therapy and was discharged on oral steroid therapy and then started on azathioprine in the follow-up. The oral steroid was gradually tapered and stopped.

Motor nerve conduction study revealed reduced velocities and reduced amplitude with prolonged latencies in the right median and right tibial nerve. There were reduced velocity and prolonged latencies with normal CMAP with conduction block in the left median, right peroneal and right tibial nerve. All other nerves tested were not recordable. F waves and sensory nerve conduction study revealed were not recordable in all nerves tested. The nerve conduction study was suggestive of demyelinating sensory-motor polyneuropathy involving all four limbs [Table 1]. CSF study showed albumino-cytological dissociation (cell count 4 lymphocytes, Protein: 98 mg% Sugar: 111.2 mg%). Other routine hemogram and biochemistry, HbA1C, serum protein electrophoresis for M band and serum ACE were normal. Serum viral markers, vasculitis profile, and ELISA for Borrelia burgdorferi and brucella were negative. In MRI brain, 3rd, 5th, and 9th cranial nerves appeared hyperintense on T2W and showed post-contrast enhancement [Figure 1]. X-ray chest, USG abdomen, ECG was normal.

Discussion

A young male patient, without any comorbid illness presented with 13 years history of recurrent lower motor neurone type quadriparesis with sensory involvement without bowel-bladder involvement. He also had two events of lateral rectus palsy (left side followed by right side). Initial events had a spontaneous recovery. Later events were treated with pulsed steroid therapy. The nerve conduction study (NCS) revealed a sensory and motor demyelinating polyneuropathy with conduction block in multiple nerves consistent with CIDP. Cerebrospinal fluid demonstrated

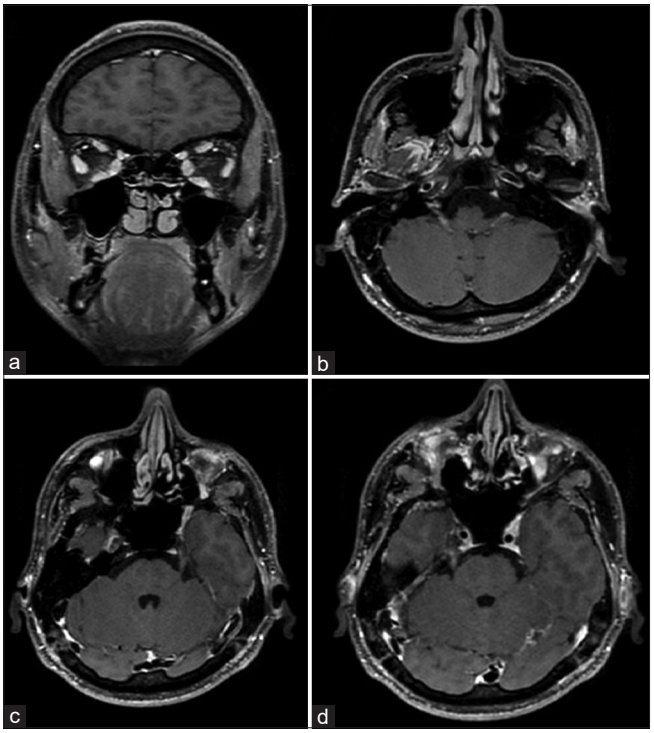

Figure 1: T1W post contrast images show (a) thickening and enhancement of the left maxillary nerve (b) enhancement of cisternal segment of the left trigeminal nerve (c) enhancement of cisternal segment of the right trigeminal nerve (d) enhancement of right 9th cranial nerve

Table 1: Nerve Conduction Study

Motor Nerve Conduction Study

Nerve/Sites	Latency (ms)	Amplitude (ms)	Segments	Distance (mm)	Latency Difference (ms)	Velocity (m/s)
Right Median - APB						
Wrist	5.94	2.5	Wrist - APB			
Elbow	15.42	1.9	Elbow - Wrist	240	9.48	25
Right Ulnar - ADM						
Wrist	NR	NR	Wrist - ADM			
Right Median - APB						
Wrist	5.42	6.0	Wrist - EDB			
Elbow	27.14	2.7	Elbow - Wrist	240	21.72	11
Left Ulnar - ADM						
Wrist	NR	NR	Wrist - ADM			
Left Peroneal - EDB						
Ankle	NR	NR	Ankle - EDB			
Fib head	NR	NR	Fib head - Ankle		NR	
Left Tibial - AH						
Ankle	NR	NR	Ankle - AH			
Popliteal fossa			Popliteal fossa - Ankle		NR	
Right Peroneal - EDB						
Ankle	6.88	5.1	Ankle - EDB			
Fib head	39.48	0.7	Fib head - Ankle	320	32.60	10
Right Tibial - AH						
Ankle	11.61	3.9	Ankle - AH			
Popliteal fossa	48.44	0.7	Popliteal fossa - Ankle	400	36.82	11

Sensory Nerve Conduction Study

Nerve/Sites	Onset Latency (ms)	Peak Latency (ms)	Amplitude (μV)	Segments
Right Median - Digit II (Wrist)	NR	NR	NR	Wrist - Dig II
Right Ulnar - Digit V (Wrist)	NR	NR	NR	Wrist - Dig V
Left Median - Digit II (Wrist)	NR	NR	NR	Wrist - Dig II
Left Ulnar - Digit V (Wrist)	NR	NR	NR	Wrist - Dig V
Left Sural - Ankle (Calf)	NR	NR	NR	Calf - Ankle
Right Sural - Ankle (Calf)	NR	NR	NR	Calf - Ankle

F Wave

Nerve	F wave latency (ms)
Right Median - APB	NR
Right Ulnar - ADM	NR
Right Median - EDB	NR
Right Tibial - AH	NR
Left Ulnar - ADM	NR
Left Peroneal - EDB	NR
Left Tibial - AH	NR
Right Peroneal - EDB	NR

APB: Abductor Pollicis Brevis, ADM: Abductor Digiti Minimi, EDB: Extensor Digitorum Brevis, AH: Abductor Hallucis, NR: No Response

albumino-cytologic dissociation. The patient was managed as a case of relapsing-remitting CIDP with recurrent sixth nerve palsy (Left followed by right).

The relapsing-remitting course of CIDP is seen, ranging from 5% to 51% in various studies.[5,6] It was predominant seen in the juvenile age group.[7] Our patient also had the juvenile onset of symptoms. The prognosis regarding response to IVIg was better in the relapsing-remitting course of CIDP. Our case report describes a rare presentation of CIDP, i.e., relapsing-remitting CIDP with recurrent sixth nerve palsy. Cranial nerve involvement is well reported in CIDP.[5,8] Ophthalmoplegia in CIDP is seen in about 3-8% of cases in various case series.[9] They might be restricted to cranial nerve involvement or followed by subsequent limb involvement. IgG anti-GQlb gangliosides, which is seen in the Guillain-Barre syndrome with ophthalmoplegia is usually not seen in CIDP. One case of CIDP with recurrent ophthalmoplegia had reported the presence of anti-GM1

antibodies.[10] We did not test for antiganglioside antibody profile in our patient.

In previous case reports of CIDP with cranial nerve involvement, the patients had a complete recovery with IVIg.[11,12] Our patient also had a full recovery, but with pulsed steroid alone. The published randomised trials and systematic review suggested that pulsed steroids are better than IVIg in attaining remission and lower rates of serious adverse effects.[13-15] The patient was compliant with the treatment of oral steroid. He was started on azathioprine, and the oral steroid was gradually tapered and stopped. The patient is in follow up with us for the past 1.5 years without any new relapse.

Conclusion

In conclusion, CIDP with cranial nerve involvement needs detailed evaluation, including neuroimaging, to rule out the varied differential diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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