Chronic inflammatory demyelinating polyneuropathy presenting as Guillain–Barre syndrome: A case presentation

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ABSTRACT

Guillain–Barre syndrome (GBS) and chronic inflammatory demyelinating polyneuropathy (CIDP) are immune-related neuropathies and differentiated mainly by disease course. GBS is characterized by a monophasic course, with a clinical nadir within 4 weeks of symptom onset, whereas CIDP typically demonstrates a slowly progressive course with gradual worsening over more than an 8-week period. Both may share common features such as demyelinating polyneuropathy, raised protein in cerebrospinal fluid, and conduction delay or block in nerve conduction study. Here, we present a case who was diagnosed and managed as GBS based on history, clinical presentation, and electrodiagnostic criteria on initial visit. Later on, we revised our diagnosis to acute-CIDP when the patient deteriorated after 8 weeks. This subset of the patient (up to 16% of CIDP) may initially present as GBS. Distinguishing between GBS and CIDP is crucial as prognosis and treatment differ.

KEY WORDS: Neuropathies; Monophasic; Demyelinating; Nerve Conduction Study; Deterioration

INTRODUCTION

Guillain–Barre syndrome (GBS) is an autoimmune acute polyradiculopathy that usually presents as ascending lower motor neuron paralysis with or without sensory involvement. It evolves over hours to a few days. Chronic inflammatory demyelinating polyneuropathy (CIDP) is usually gradual over few months or longer but shares many features with the common demyelinating form of GBS such as elevated cerebrospinal fluid (CSF) protein and electrodiagnostic findings. However, few patients of CIDP may present similar to GBS and are termed as acute-onset CIDP (A-CIDP). Distinction between GBS and A-CIDP is important as treatment and outcome differ in both and maintenance treatment is generally required in later. Here, we present a case who was initially diagnosed as GBS but later turned out to be a case of A-CIDP.

CASE REPORT

A 26-year-old lady was admitted in our hospital with complain of weakness both upper and lower limbs. She was apparently well till 10 days ago when she developed difficulty in climbing stairs, getting up from sitting/squatting. Simultaneously, she had difficulty in mixing food, lifting bag, and combing her hairs. All symptoms progressively worsened over a period of 1 week to the extent that she was bedridden. At the onset of illness, there was also tingling sensation distally. She was not suffering from any systemic illness. There was no history of any recent illness such as fever or diarrhea. Bladder and bowel were intact. General examination did not reveal any significant findings. Examinations of other systems were within normal limits. CNS examination: Higher functions were intact. The patient was conscious and oriented. Minimal state examination was 30/30. There was no mood or psychotic symptoms and all cranial nerves were intact. Motor
examination revealed flaccid quadriparesis (Medical Research Council Grade 1 in lower limbs and Grade 2 in upper limbs) [Table 1]. No involuntary movements were observed. Joint position sense was absent distally in both upper and lower limbs. Superficial abdominal reflexes were present in all four quadrants of abdomen, but deep tendon reflexes were absent bilaterally in both upper and lower limbs. Plantar reflexes were bilaterally flexor. There were no meningeal signs or signs of cerebellar dysfunction. Spine and skull were normal. Baseline investigations were normal. Nerve conduction study showed acquired primary demyelinating and secondary axonal sensorimotor polyradiculopathy involving all four limbs [Table 2].

Thus, we made diagnosis of GBS (acute progressive symmetrical flaccid quadriplegia with absent deep reflexes and demyelinating features in nerve conduction velocity [NCV]). IV immunoglobulin was started for 5 days (total of 2 gm/kg body weight) along with supportive care keeping watch on progression of disease. On the 8th day, after completion of treatment, there was some improvement in the power of muscles. The patient could move her upper limb (power increased from Grade 2 to Grade 3) and power in lower limb increased from Grade 1 to Grade 2. Physiotherapy was done aggressively with monitoring of vitals. The patient was observed for another 2 weeks then discharged with advice to continue physiotherapy and to be on regular follow-up. 25 days after discharge (9th week of disease), the patient was brought by attendants as she complained of sudden deterioration in the strength of her limbs. On examination, power in both upper and lower limb had decreased bilaterally (almost Grade 1 proximally and distally) and JPS was absent till knee in lower limbs and till wrist in upper limbs. The patient was referred to neurologist for opinion where the patient was thoroughly investigated. Extensive blood test, CSF examination, and repeat NCV were performed.

As the patient presented with relapsing polyradiculopathy with temporal progression beyond 8 weeks along with albuminocytological dissociation and evidence of demyelination in NCV [Table 3] diagnosis of acute CIDP was made. She was initiated on pulse therapy (weekly methylprednisolone) and rituximab as steroid-sparing agent which she tolerated well. Just after the first dose of therapy, the patient started improving and was able to walk with aid in 10 days. After 3 months of therapy, the patient regained almost normal power. She is going to complete almost 1 year by attendants as she complained of sudden deterioration in the strength of her limbs. On examination, power in both upper and lower limb had decreased bilaterally (almost Grade 1 proximally and distally) and JPS was absent till knee in lower limbs and till wrist in upper limbs. The patient was referred to neurologist for opinion where the patient was thoroughly investigated. Extensive blood test, CSF examination, and repeat NCV were performed.

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**DISCUSSION**

During initial presentation of our patient, we made diagnosis of GBS based on pattern of evolving weakness, areflexia, and demyelinating features in NCV![1,2] GBS manifests as rapidly evolving areflexic motor paralysis with or without sensory disturbance. The usual pattern is ascending paralysis that typically evolves over hours to few days (nadir within 4 weeks) and is frequently accompanied by tingling dysesthesias in the extremities and the legs are frequently more affected than arms.[3] It is a monophasic autoimmune process often triggered by an upper respiratory or gastrointestinal infection and most common cause of flaccid paralysis worldwide.[5] In
our case, there was slight improvement with IVIG, but the patient deteriorated again after 9 weeks of onset of disease. During the second admission, she was considered as a case of CIDP due to chronic relapsing nature.\textsuperscript{[5,6]} This neuropathy (CIDP) shares many features with the common demyelinating form of GBS such as elevated CSF protein levels and the Edx findings.\textsuperscript{[7]} Onset in cases of CIDP is usually gradual over a few months or longer (\textgtr 8 weeks), but in few cases, the initial attack is indistinguishable from that of GBS when the entity is known as acute-CIDP. The diagnosis of acute-onset CIDP should be considered when a patient thought to have Guillain–Barré syndrome deteriorates again after 8 weeks from onset of disease (which happened with our patient) or when deterioration occurs 3 times or more.\textsuperscript{[8]}

Around 16\% of CIDP patients, described as A-CIDP, show rapidly progressive weakness within 4 weeks from onset and 18\% of CIDP patients present with relapsing weakness.\textsuperscript{[9,10]} This A-CIDP should be differentiated from 8\% to 16\% of GBS patients who show one or more deteriorations after initial improvement, described as treatment-related fluctuations (TRF).\textsuperscript{[8]} TRF is defined as improvement in the GBS disability scale of at least one grade after completion of immunotherapy (immunoglobulin/plasmapheresis) followed by a worsening of the disability scale of at least one grade within the first 2 months after disease onset.\textsuperscript{[11]} Our patient deteriorated after 2 months of disease onset so she did not fit in the criteria of TRF. Why it is important to differentiate between TRF (GBS) and acute-CIDP? It has prognostic and treatment implications. Deterioration in GBS may improve with new course of IVIg and patients of acute-CIDP need chronic maintenance therapy.\textsuperscript{[8]} Moreover, patients of CIDP respond to glucocorticoids, whereas GBS does not.\textsuperscript{[3,12]} Our patient responded drastically to methylprednisolone and is on maintenance therapy. At times, patients of CIDP benefit from treatment with immunosuppressive agents. Our patient was reasonably investigated for vasculitis, collagen vascular disease (especially systemic lupus erythematosus), chronic hepatitis, HIV infection, amyloidosis, inflammatory bowel disease, lymphoma, etc., to rule out secondary CIDP.

CONCLUSION

Few patients (16\%) of CIDP may present like GBS and responds to immunotherapy. If a patient of GBS deteriorates after 8 weeks of onset of disease or relapses $>3$ times, then acute-CIDP should be considered. It is important to differentiate between the two as treatment and outcomes are different.

REFERENCES