Post-Varicella Acute Inflammatory Demyelinating Polyradiculoneuropathy in a 51-Year-Old Filipino Male: A Case Report

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ABSTRACT

Primary varicella zoster infection is commonly observed in school-aged children. There are increasing reports of adults also being affected. *Varicella zoster* infection has a myriad of clinical complications. The rarer of these complications is Guillain-Barre syndrome (GBS) or acute inflammatory demyelinating polyradiculoneuropathy with less than 50 cases in reported literature. We report the case of a 51-year-old Filipino male who presented with bilateral lower extremity weakness two weeks after a primary varicella infection. Cerebrospinal fluid (CSF) analysis showed elevation of CSF protein at 69 mg/dL (NV 15 to 45 mg/dL). CSF varicella virus Immunoglobulin G was 1.8 mIU/ml (NV 1.3 mIU/ml) and Immunoglobulin M was at 1 mIU/ml (NV 0.9 mIU/ml). Nerve conduction velocity studies mainly showed a demyelinating form of neuropathy involving motor (predominantly) and sensory nerves. The objective finding in this case, as well as the clinical history, is indicative of a demyelinating sensorimotor polyneuropathy after a varicella infection. According to our awareness and considering its rarity, this was the first Filipino case to be reported with varicella infection and GBS.

Keywords: varicella, acute inflammatory demyelinating polyradiculoneuropathy, Guillain-Barre Syndrome.

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INTRODUCTION

Varicella zoster virus is the etiologic agent of varicella (primary infection) and herpes zoster (reactivation of latent infection). It is commonly observed among school-aged children. In tropical climates, more cases are reported in adults. Although varicella is most often a relatively benign and self-limited childhood illness, the disease can be associated with a variety of serious and potentially lethal complications in both immunocompetent and immunocompromised persons (1, 2).

The incidence of neurologic complications associated with varicella is estimated to be 1-3 per 10,000 cases. The rarer of these complications is the GBS or acute inflammatory demyelinating polyradiculoneuropathy (AIDP) with less than 50 cases in reported literature (1-4).

We report the case of a 51-year-old Filipino male who presented with bilateral lower extremity weakness and numbness indicative of demyelinating polyradiculoneuropathy two weeks after a primary varicella infection.

CASE

Our patient is a 51-year-old male admitted due to progressive numbness and weakness in the lower extremities.

In early March 2015, he had intermittent undocumented fever, followed shortly after by the appear-

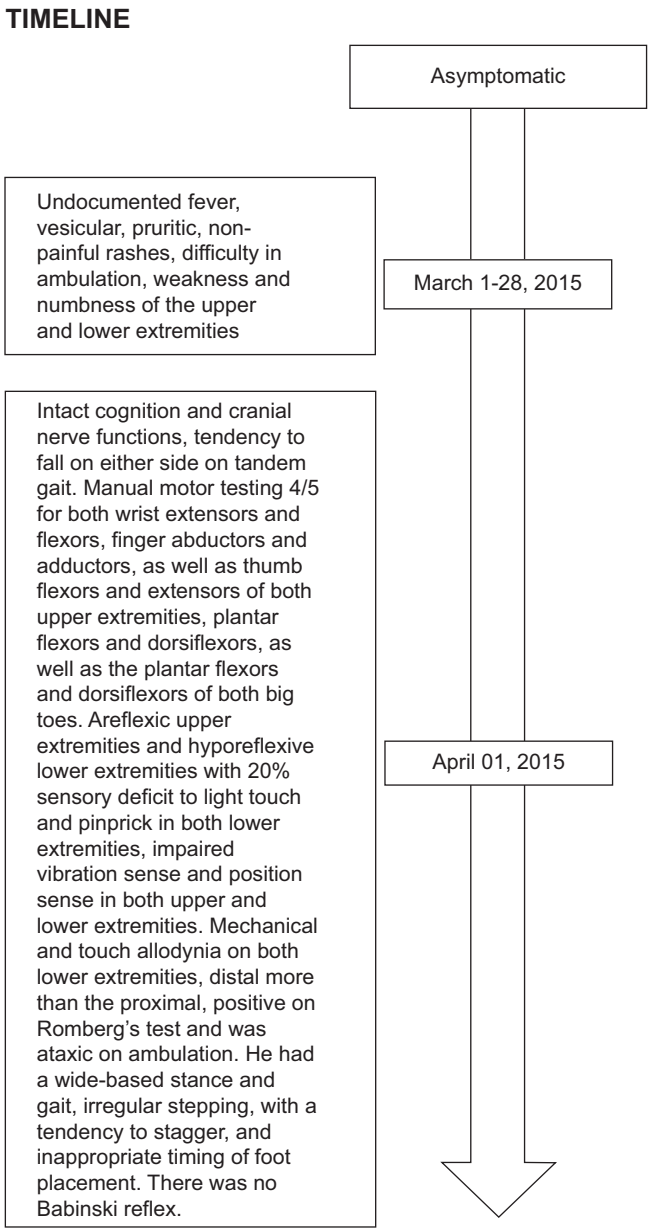

Figure 1. Course of the Illness.

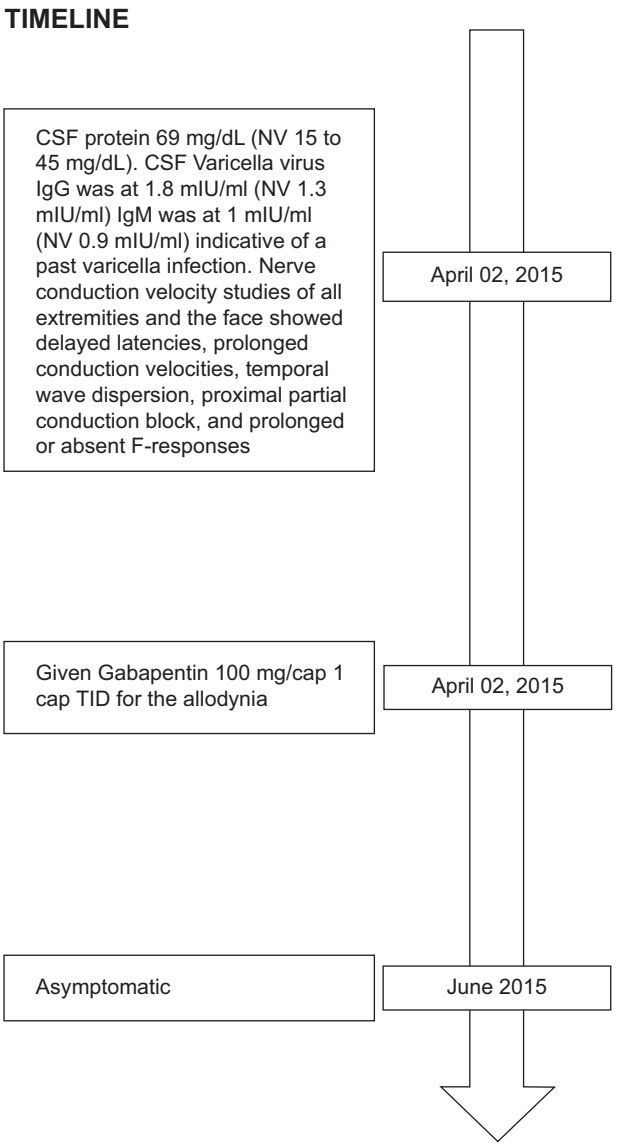

Figure 1. Continued

ance of vesicular, pruritic, non-painful rashes on his upper back, which eventually spread to his face, chest, and extremities. Interval history showed lysis of the fever after three days, but there was worsening of the rashes.

Two weeks after the appearance of rashes, the patient began to experience numbness in both his soles and palms. He also noted difficulty in walking and making precise steps as he easily lost balance. He preferred to stay seated but was still able to ambulate and maintain a standing position with assistance. No weakness of the extremities was reported at this time.

He sought consultation at a clinic and was assessed with a nonspecific nerve problem. No workup was done. He was given Vitamin B complex, which did not relieve his symptoms. Four days later, he noted progression of the numbness from his soles to the lower third of his distal legs. This was now accompanied by weakness of his distal legs, feet, and handgrip. He could no longer ambulate and maintain a standing position. He also experienced dull pain on the palmar area of both hands extending to his forearms, upper arms, and shoulders, most prominent whenever he tried to reach out for an object. No disturbances in bladder or bowel functions, slurring of speech, facial asymmetry, double vision, or dysphagia were noted. He has a history of hypertension and is maintained on losartan 40 mg/tab 1 tab BID. He also has a family history of hypertension and diabetes. No other family members presented with symptoms similar to the patient. Fasting blood sugar of this patient showed a normal result at 98.10 mg/dL (NV <126 mg/dL).

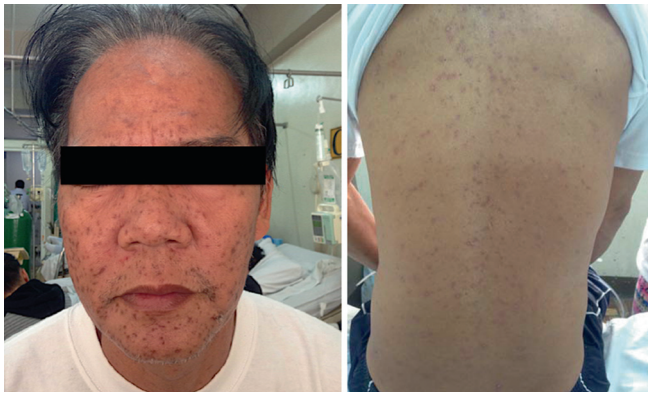

Figure 2. Multiple Generalized Hyperpigmented Lesions Over the Face and Back.

Clinical Findings

Physical and neurological examination showed multiple generalized hyperpigmented scars (Figure 2). He had intact cognition and cranial nerve functions. On tandem gait, he had a tendency to fall on either side. Manual motor testing revealed a score of 4/5 for both wrist extensors and flexors, finger abductors and adductors, as well as thumb flexors and extensors of both upper extremities. The rest of the upper extremity muscle groups were scored 5/5. For both lower extremities, a score of 4/5 was given to plantar flexors and dorsiflexors, as well as the plantar flexors and dorsiflexors of both big toes. He had no biceps, triceps, and brachioradialis reflex. He was hyporeflexive on both knees and ankles. On sensory examination, he had 20% sensory deficit to light touch and pinprick in both lower extremities with impaired vibration sense and position sense in both upper and lower extremities. We noted mechanical and touch allodynia on both lower extremities, distal more than the proximal. He tested positive on Romberg's test and was ataxic on ambulation. He had a wide-based stance and gait, irregular stepping, with a tendency to stagger, and inappropriate timing of foot placement. There was no Babinski reflex.

Diagnostic Assessment

Lumbar puncture and CSF analysis of this patient showed normal opening and closing pressure, as well as CSF glucose levels. There was elevation of CSF protein at 69 mg/dL (NV 15 to 45 mg/dL). CSF Varicella virus IgG was at 1.8 mIU/ml (NV 1.3 mIU/ml) IgM was at 1 mIU/ml (NV 0.9 mIU/ml)

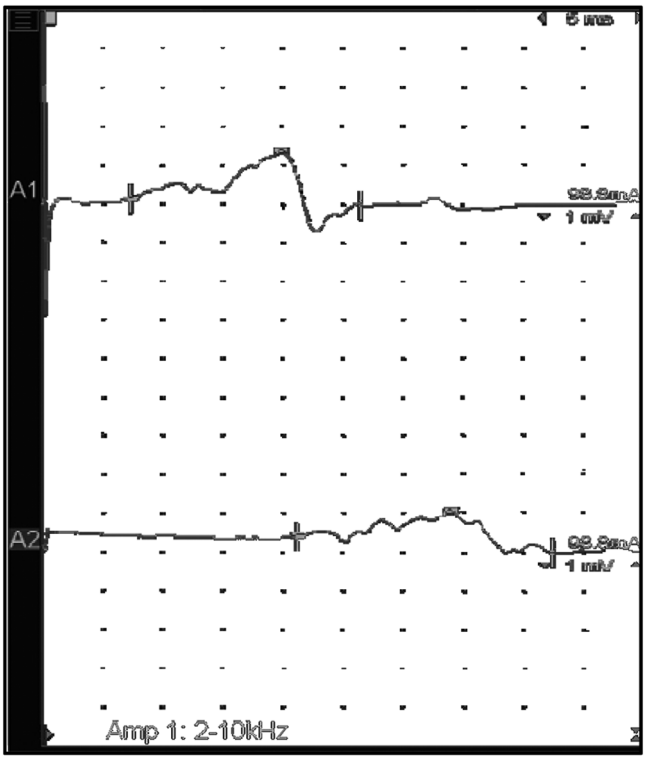

Figure 3. Delayed Latency, Temporal Wave Dispersion Seen on the Left Tibial Motor Nerve Study.

indicative of a past varicella infection. Nerve conduction velocity studies of all extremities and the face showed delayed latencies, prolonged conduction velocities, temporal wave dispersion, proximal partial conduction block, and prolonged or absent F-responses (Figure 3, Table 1).

There is electrophysiological evidence of diffuse sensorimotor polyneuropathy, demyelinating in nature which indicates the presence of acute inflammatory demyelinating polyneuropathy.

Therapeutic Intervention

The patient was closely monitored with spontaneous gradual improvement of symptoms while admitted. Gabapentin 100 mg/cap 1 cap TID was given to address his allodynia.

Follow-up and Outcomes

He was completely asymptomatic when he was last seen two months after discharge on June 2015.

DISCUSSION

Varicella zoster infection may cause several neurological complications like myelitis, aseptic meningitis,

Table 1. A summary of the nerve conduction velocity abnormalities in this patient.

	Nerve Conduction Velocity Abnormalities	
	Motor	**Sensory**
Delayed latencies or absent response	Bilateral ulnar nerves Bilateral median nerves Peroneal nerves Tibial nerves	Bilateral median nerves Bilateral ulnar nerves Bilateral sural nerves Bilateral superficial peroneal
Prolonged conduction velocities or absent response	Bilateral median nerves Bilateral ulnar nerves Bilateral peroneal nerves Bilateral tibial nerves	Bilateral median nerves Bilateral ulnar nerves Bilateral peroneal nerves Bilateral sural nerves
Temporal dispersion	Bilateral median nerves Bilateral ulnar nerves Bilateral peroneal nerves Tibial nerves	
Partial conduction block	Bilateral median nerves Bilateral ulnar nerves Bilateral peroneal nerves Bilateral tibial nerves	
Prolonged or absent F wave response		Tibial nerves
Absent H-reflex response		Tibial nerves

meningoencephalitis, ventriculitis, herpes zoster ophthalmicus, and AIDP (5). Cell-mediated immunity and other degrees of immunosuppression are known to follow viral infections. There is depression of tuberculin reactivity in varicella infection suggesting the role of immunosuppression as a contributing factor in triggering an autoimmune disease of the peripheral nervous system (6,7). It is possible that viral infections may reduce suppressor T-cell activity thereby leading to a proliferation of few T and B lymphocytes, which are normally present in humans and recognize nervous system antigens (8). Reduction of the suppressor T cell subpopulation in patients with AIDP is consistent with this hypothesis. AIDP is a post-infective immune-mediated phenomenon, which causes rapidly progressive, diffuse proximal and distal weakness of the limbs accompanied by sensory loss and areflexia (9).

Symptoms of involvement of the nervous system following varicella develop between the fifth and twentieth day after the appearance of the rash, usually in the first half of the second week (10). Antibodies positive to gangliosides can be attributed to poor recovery or rapidly reversible weakness (11). This can be considered as the limitation of this study as the CSF examination was not determined.

The clinical picture and nerve conduction velocity study results of this patient are suggestive of a demyelinating type of sensorimotor polyneuropathy. Likewise, the appearance of vesiculopapular rashes following a history of fever is highly suggestive of a primary varicella infection.

REFERENCES

1. Anne G. Varicella and herpes zoster: Clinical disease and complications. *Herpes*. 2006;13:2A-7A.
2. Gnann Jr JW. Varicella-zoster virus: atypical presentations and unusual complications. *The Journal of Infectious Diseases*. 2002 Oct 15;186(1):S91-98.
3. Cresswell F, Eadie J, Longley N, Macallan D. Severe Guillain–Barré syndrome following primary infection with varicella zoster virus in an adult. *International Journal of Infectious Diseases*. 2010 Feb 28;14(2):e161-e163.
4. Guess HA, Broughton DD, Melton L3, Kurland LT. Population-based studies of varicella complications. *Pediatrics*. 1986 Oct 1;78(4):723-727.
5. Ranga S, Avati AV. Primary varicella zoster infection presenting as Guillain-Barre-like syndrome. A case report and review of the literature. *International Journal of Clinical Cases and Investigations*. 2012;4:82-84.
6. Starr S, Berkovich S. The depression of tuberculin reactivity during chickenpox. *Pediatrics*. 1964 May 1;33(5):769-72.
7. Lisak RP, Mitchell M, Zweiman B, Orrechio E, Asbury AK. Guillain–Barré syndrome and Hodgkin's disease: Three cases with immunological studies. *Annals of Neurology*. 1977 Jan 1;1(1):72-78.
8. Winer JB, Greenwood RJ, Hughes RA, Perkin GD, Healy MJ. Prognosis in Guillain-Barré syndrome. *The Lancet*. 1985 May 25;325(8439):1202-3.
9. Hughes RA, Aslan S, Gray IA. Lymphocyte subpopulations and suppressor cell activity in acute polyradiculoneuritis (Guillain-Barré syndrome). *Clinical and Experimental Immunology*. 1983;51(3):448.
10. McKendall RR, Klawans HL. Nervous system complications of varicella-zoster virus. *Handbook of Clinical Neurology*. 1978;34:161-83.
11. Rajabally YA, Uncini A. Outcome and its predictors in Guillain–Barré syndrome. *J Neurol Neurosurg Psychiatry*. 2012 Jan 1:jnnp-2011.