Guillain-Barré Syndrome with HIV Infection: A Case Report

Siew-Hee Ting, Jiann-Horng Yeh, and Hou-Chang Chiu

Abstract- A 56-year-old woman had distressing dysesthesia of acute onset in the extremities, followed by generalized paralysis and respiratory failure. Electrophysiological studies showed demyelinating polyradiculoneuropathy. Human immunodeficiency virus (HIV) antigen was positive in her serum. Cerebrospinal fluid analysis showed pleocytosis and elevated protein concentration, consistent with a diagnosis of Guillain-Barré syndrome (GBS) in HIV infection. According to her history of regular blood donation, GBS was presumably induced during the seroconversion period of an underlying HIV infection. Plasmapheresis successfully ameliorated neurological deficits of GBS. HIV-related GBS might present exclusively with sensory features, followed by a fulminant course. For GBS patients with pleocytosis in their CSF, prompt screening for HIV infection should be done.

Key Words: Guillain-Barré syndrome, Human immunodeficiency virus, Cerebrospinal fluid

Acta Neurol Taiwan 2003;12:139-142

INTRODUCTION

The spectrum of neurological complications in acquired immunodeficiency syndrome (AIDS) caused by human immunodeficiency virus (HIV) includes AIDS dementia complexes, vacuolar myelopathy, and peripheral and autonomic neuropathy. Neurological disorders are among the main clinical features of AIDS and HIV disease[1]. About 10% to 35% of HIV infected individuals develop a clinical neuropathy[2]. Histopathological abnormalities within nerves are found in over 95% of patients dying of AIDS[2]. HIV related peripheral neuropathies includes acute inflammatory demyelinating polyradiculoneuropathy, chronic inflammatory demyelinating polyradiculoneuropathy, multiple mononeuropathy, distal symmetrical polyneuropathy, lumbosacral polyradiculopathy, autonomic neuropathy and mononeuropathies[3]. Guillain-Barré Syndrome (GBS) has been reported as one of the initial manifestations of HIV infection[4].

The clinical and electrophysiological findings in HIV-associated GBS were no different than that of non-HIV-associated forms, except for cerebrospinal fluid pleocytosis[5]. We report a patient with HIV-associated GBS presenting with severe distressing neuropathic pain, followed by a fulminant paralytic course.

CASE REPORT

A previously well 56-year-old woman had donated

From the Department of Neurology, Shin Kong Wu Ho-Su Memorial Hospital, Taipei, Taiwan.
Received March 7, 2003. Revised April 16, 2003.
Accepted June 30, 2003.
Reprint requests and corresponding to: Jiann-Horng Yeh, MD. Department of Neurology, Shin Kong Wu Ho-Su Memorial Hospital, 95 Wen-Chang Road, Taipei, Taiwan.
E-mail: M001074@ms.skh.org.tw

blood every 3 months for the several past years. The date of the last donation was 3 December 2001. On 28 February 2002, she developed acute onset of ascending numbness of her limbs. The numbness was initially noted in her left lower limb below the knee, and followed by left upper limb below the wrist. Few hours later, it extended to her right lower limb below the knee and right upper limb below the wrist. Severe camping pain in bilateral calf muscles and urinary frequency were also present. There was no motor weakness.

The patient was admitted to Shin Kong Wu Ho-Su Memorial Hospital on 1 March 2002. Neurological examination showed that the pinprick sensation was decreased on both upper limbs below the mid-forearm and both lower limbs below the midthigh. The joint position sense and vibration sense of the toes were impaired. Diffuse tenderness of the paraspinal muscles and bilateral calf muscles were noted. The muscle power of the limbs was preserved, with generalized areflexia. No pathological reflex was recorded.

On the second day of admission, muscle power of the neck and distal limbs were mildly reduced to Medical Research Council grade 4 (Fig.) and that of the proximal limbs worsened to grade 3. There was no progression of sensory impairment. The pain was distressing and persistent.

On 3 March 2002, the patient developed shallow breathing associated with paradoxical chest wall movement. The arterial blood gas analysis showed pCO_2 of 31 mmHg (reference: 35-45 mmHg), pO_2 of 112.5 mmHg (reference: 80-100 mmHg), HCO_3- of 22.9 mmol/L (reference: 21-28 mmol/L), pH of 7.47 (reference: 7.38-7.44), and oxygen saturation 98.8% (reference: 97 ± 2%). Both lung fields were clear on chest X-ray. Under the impression of respiratory failure, she received endotracheal tube intubation. Her muscle power progressively deteriorated in the following days. On 5 March, her neck and limb muscles were completely paralyzed except for grade 3 power in the distal lower limbs. Screening for serum HIV antigen by enzyme-linked immunosorbent assay (ELISA) was positive. The CD4 was 14.7% (reference: 32-61%) and CD8 was 56.2% (reference: 18-42%). The rapid plasma reagin (RPR) test for syphilis was nonreative. An autoimmune survey including antinuclear antibody, anti-ds DNA, and rheumatoid factor were all within normal limit. Tumor markers for plasma α-feto-

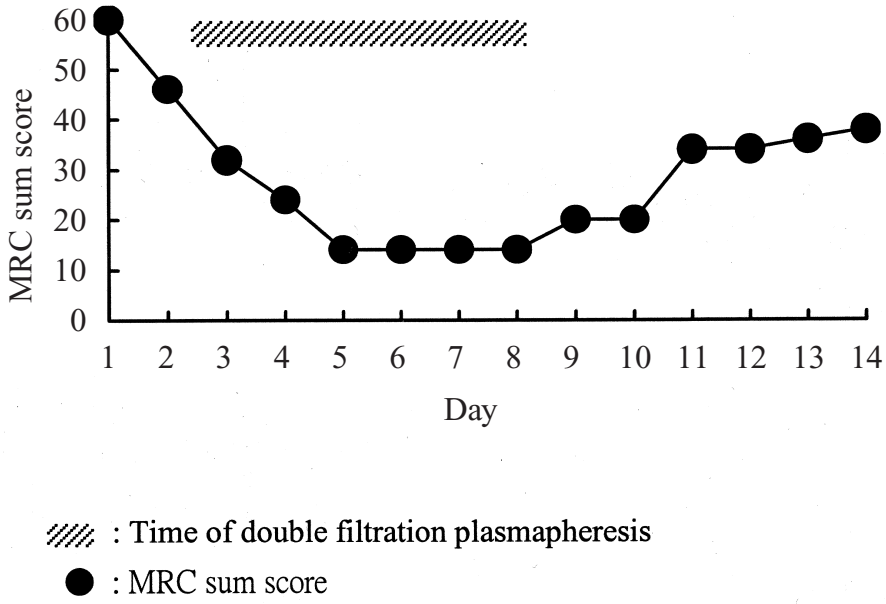

Figure. Change in Medical Research Council (MRC) sum score and time of double filtration plasmapheresis in a patient with HIV-related Guillain-Barré syndrome. The MRC sum score is a summation of the MRC grades (range, 0-5) given in full numbers of the following muscle pairs: upper arm abductors, elbow flexors, wrist extensors, hip flexors, knee extensors, and foot dorsiflexors. The MRC sum score ranges from 0 (total paralysis) to 60 (normal strength).

protein, CA-153, CA-125, CA-199, and carcinoembryonic antigen were all within normal limit. Cerebrospinal fluid (CSF) analysis showed elevated protein of 246.8 mg/dL (reference: 20-50 mg/dL) and pleocytosis of 20 cells (reference: < 5 cells), with a lymphocyte predominance (L/N = 99/1). A nerve conduction study done on 3 November showed conduction block, slowed conduction velocities, and prolonged F-wave latencies for all sampled nerves. The results were consistent with demyelinating polyradiculoneuropathy (Table 1).

Based on the diagnosis of Guillain-Barré Syndrome, she underwent double filtration plasmapheresis starting on 2 March, to 8 March, for a total of five sessions. On 13 March, she began to gradually regain muscle power of her neck and was able to move her head. Nevertheless, there was no significant improvement of the sensory impairment. A follow-up nerve conduction study showed further decrease in nerve conduction velocities and prolonged F-latencies on all examined nerves (Table 2). She was extubated on 14 March. On the next day, she was transferred to another hospital for further care of the underlying HIV infection.

DISCUSSION

A 56-year-old woman presented with acute onset of sensory deficits and neuropathic pain, followed by a fulminant course of paralytic weakness. Respiratory failure developed on the fourth day after the onset of symptoms. Electrophysiological studies showed demyelinating polyradiculoneuropathy. The CSF analysis showed pleocytosis and elevated protein concentration. The overall findings of our patient were compatible with the diagnosis of GBS.

Our patient donated her blood on 3 December 2001, and the blood screen for HIV was negative. Three

Table 1. *Nerve conduction studies (Day 4 after symptom onset)*

Nerve	DL (ms) R/L (UL)	Amp (mv) R/L (LL)	NCV (m/s) R/L (LL)	F-wave (ms) R/L (UL)
Motor nerve				
Median	4.7/4.5 (4.2)	3.8-1.6*/ 4.4-3.3* (3.0)	31.1/36.4 (49.2)	34.3/32.7 (25.3)
Ulnar	3.5/3.0 (3.4)	4.6-3.8*/ 5.4-4.0* (3.0)	40.5/45.0 (51.2)	29.3/28.7 (25.9)
Peroneal	5.0/4.3 (5.1)	3.1-2.7*/ 6.1-3.3* (2.0)	36.8/34.4 (41.0)	45.2/NR (48.4)
Tibial	4.0/4.5 (5.1)	10.3-6.9*/ 12.4-8.8* (5.0)	34.2/34.4 (42.0)	49.3/44.7 (48.4)
Sensory nerve				
Median	3.2/2.8 (2.7)	6.5/11.0 (10.0)	43.8/50.9 (48.7)	
Ulnar	2.8/2.3 (2.8)	6.4/7.5 (10.0)	46.4/56.5 (47.9)	
Sural	2.9/2.9 (3.3)	19.0/13.0 (5.0)	44.8/44.8 (41.6)	

DL: distal latency; R: right; L: left; UL: upper limit; LL: lower limit; NCV: nerve conduction velocity; Amp: amplitude; *distal-proximal; NR: no response.

Table 2. *Nerve conduction studies (Day 15 after symptom onset)*

Nerve	DL (ms) R/L (UL)	Amp (mv) R/L (LL)	NCV (m/s) R/L (LL)	F-wave (ms) R/L (UL)
Motor nerve				
Median	5.8/5.3 (4.2)	2.5-1.4*/ 2.6-2.2* (3.0)	15.3/30.0 (49.2)	62.5/60.8 (25.3)
Ulnar	4.5/3.3 (3.4)	4.2-3.1*/ 4.7-4.6* (3.0)	30.9/35.5 (51.2)	62.8/61.2 (25.9)
Peroneal	8.7/7.3 (5.1)	3.3-3.0*/ 2.5-1.4* (2.0)	22.9/22.3 (41.0)	74.8/73.5 (48.4)
Tibial	9.2/7.0 (5.1)	10.4-7.4*/10.0-7.6* (5.0)	31.4/29.5 (42.0)	75.7/72.0 (48.4)
Sensory nerve				
Median	3.8/3.1 (2.7)	5.0/ 6.0 (10.0)	36.8/45.2 (48.7)	
Ulnar	2.7/2.7 (2.8)	3.9/15.0 (10.0)	48.1/48.1 (47.9)	
Sural	3.6/3.8 (3.3)	15.0/12.0 (5.0)	36.1/34.2 (41.6)	

DL: distal latency; R: right; L: left; UL: upper limit; LL: lower limit; NCV: nerve conduction velocity; Amp: amplitude; *distal-proximal.

months later, seropositivity for HIV was found. This implied the GBS was induced during the seroconversion period of HIV infection, which is consistent with a previous report in literature[3].

In a study of GBS in Tanzania, the mean duration from onset to maximum weakness was shorter in the HIV seropositive group than the seronegative group, and the seropositive group had more extensive neurological involvement[6]. Sensory involvement was less common in the seropositive group[6]. In our patient, respiratory failure occurred on the fourth day after the onset, followed by a fulminant course with generalized paralysis reaching a nadir on the fifth day. Nevertheless, dysesthetic pain was the presenting feature for GBS in our patient, unlike patients in the Tanzanian series.

GBS is a motor-predominant neuropathy that presents with variable clinical features. In a prospective study by Ropper and Shahani, around 55% of patients had characteristic pain early in the illness[7]. Moulin at al reported that in some patients, pain preceded weakness by a mean of 6.1 days and in other patients, weakness preceded pain by a mean of 5.3 days[8]. Three discrete pain syndromes were associated with GBS, as described by Moulin at al[8]. The most common one was deep aching or throbbing pain, followed by dysesthetic extremity pain and myalgic-rheumatic extremity pain[8]. In our patient, the dysesthetic pain preceded motor paralysis by two days and was disabling.

The clinical course of HIV-related GBS was similar to that of non-HIV related GBS in terms of clinical manifestations and response to immune therapy[9]. Plasma exchange and intravenous immunoglobulin are equally effective in the treatment of GBS[10,11]. Our patient responded well to plasmapheresis. Successful extubation was achieved 12 days after starting plasmapheresis treatment. These findings confirm the efficacy of plasmapheresis for the treatment of HIV-related GBS.

HIV-related GBS might present with exclusive sensory features, followed by a fulminant course. For GBS patients with pleocytosis in the CSF, a prompt search for HIV infection should be done. If initial serology is negative, a follow up test is warranted.

REFERENCES

1. Howlett WP, Nkya WM, Mmuni KA, et al. Neurological disorders in AIDS and HIV disease in the northern zone of Tanzania. AIDS 1989;3:289-98.
2. Manji H. Neuropathy in HIV infection. Current Opinion in Neurology 2000;13:589-92.
3. Mendell JR, Kissel JT, Cornblath DR. Diagnosis and management of peripheral nerve disorders. Peripheral Neuropathy Associated with HIV Infection. Oxford: Oxford University Press, 2001:400-28.
4. Colon CP. HIV infection as Guillain-Barré syndrome in Lusaka, Zambia. Transaction of the Royal Society of Tropical Medicine & Hygiene 1989;83:109.
5. Benatar MG, Eastman RW. Human immunodeficiency virus-associated pure motor lumbosacral polyradiculopathy. Arch Neurol 2000;57:1034-9.
6. Howlett WP, Vedeler CA, Nyland H, et al. Guillain-Barré syndrome in northern Tanzania: a comparison of epidemiological and clinical findings with western Norway. Acta Neurol Scand 1996;93:44-9.
7. Ropper AH, Shahani BT. Pain in Guillain-Barré syndrome. Arch Neurol 1984;41:511-4.
8. Moulin DE, Hagen N, Feasby TE, et al. Pain in Guillain Barré syndrome. Neurology 1997;48:328-31.
9. Wuest D, Goldfinger D. Plasmapheresis in the treatment of acute relapsing inflammatory demyelinating polyradiculoneuropathy associated with human immunodeficiency virus infection: a case report. J Clin Apheresis 1988;4:149-51.
10. Vedeler C, Wik E, Nyland H. The long-term prognosis of Guillain-Barré syndrome. Evaluation of prognostic factors including plasma exchange. Acta Neurol Scand 1997;95: 298-302.
11. Plasma Exchange/Sandoglobulin Guillain-Barré Syndrome Trial Group. Randomised trial of plasma exchange, intravenous immunoglobulin, and combined treatments in Guillain-Barré syndrome. Lancet 1997;349:225-30.