HIV-1 infection presenting as Guillain-Barré Syndrome

Arvind BAMANIKAR [1], Isra Heryantee RASHID [2], Dayangku Siti Nur' Ashikin PENGIRAN TENGAH [2]
[1] Jerudong Park Medical Centre, BG3122, Brunei Darussalam and
[2] Department of Medicine, RIPAS Hospital, Brunei Darussalam

ABSTRACT

Human immunodeficiency virus (HIV) infection causes numerous non-opportunistic as well as opportunistic neurologic illnesses. Neurologic manifestations include Guillain Barré Syndrome (GBS) which may present atypically. In areas endemic for HIV, the manifestation of GBS with a motor syndrome and axonal neuropathy is not uncommon. We report a 36-year-old man who presented with a subacute limb weakness associated with HIV infection.

Keywords: Acquired immune deficiency syndrome, demyelination, neuropathy, human immune deficiency virus

INTRODUCTION

Guillain Barré Syndrome (GBS) is an immune-mediated inflammatory disorder affecting nerve roots (radiculopathy) and peripheral nerves (neuropathy) with an incidence of 1.1 to 1.8/100,000. [1] Typically patients present with progressive limb weakness accompanied by sensory disturbance such as distal paraesthesiae and a demyelinating neuropathy seen on nerve conduction studies (acute inflammatory demyelinating poyneuropathy, AIDP). Many variants exist such as pure motor, pure sensory, axonal (as opposed to the classic demyelinating variety) and the Miller-Fisher variant which consists of ophthalmoplegia, sensory disturbance and ataxia. Progression is usually maximal by the third week and then the condition plateaus before slowly improving. There is a significant mortality (0.5-1% in most studies) and morbidity for GBS and sequelae such as cardio-respiratory compromise, thrombo-embolism, painful neuropathy, constipation, etc should be carefully addressed in patient management.

Diagnosis is ultimately clinical after exclusion of other aetiologies such as compressive, infiltrative and infective. Nerve conduction studies are helpful especially in differentiating between axonal and demyelinating

Correspondence author: Arvind BAMANIKAR
Jerudong Park Medical Centre, BG3122,
Brunei Darussalam
Phone: +673-2611433
Fax: +673-2612421
Email-drarvindjpmc@gmail.com

neuropathies (of particular use in epidemiological study). Cerebrospinal fluid (CSF) protein is helpful if elevated but may be normal particularly in early stages of the illness. The pathogenesis of GBS is thought to be autoimmune in which a particular component of the peripheral nerve or nerve root e.g. the myelin sheath cross-react with antibodies generated by infection e.g. *campylobacter jejuni*. We report a 36-year-old man who presented with a sub-acute limb weakness associated with HIV infection.

CASE REPORT

A 36-year-old Chinese man working as a welder was initially admitted to a private hospital after two days of symmetrical upper and lower limb weakness. This was gradual in onset but progressing. Two weeks earlier, he had had a two day history of a self-limiting illness consisting of fever, abdominal pain and loose stool. Other than this, his only other relevant past medical history was poliomyelitis in childhood affecting the left lower limb. He was not taking any medications. He was unmarried, a teetotal, 20-pack year smoker and was from a neighbouring country.

On initial examination he was alert and orientated. Cranial nerve examination was normal. He had a wasted left lower limb typical of previous poliomyelitis. Tone in the limbs was reduced. He had symmetrical weakness of both proximal lower limbs (Medical research Council [MRC], grade 3/5) and distal upper limbs (grade 4/ 5). Knee reflexes were absent. The upper limb reflexes were sluggish but elicitable. There was no sensory loss. Other systems examination was normal. Notably there was no hypotension or respiratory compromise.

Initial investigations included blood count, inflammatory markers, routine biochemistry, creatinine kinase and thyroid function, all of which were normal. Anti-nuclear antibody (ANA), syphilis, hepatitis B, and HIV testing were also carried out as is routinely done in this hospital. ANA, RPR (rapid plasma reagin) test for syphilis and Hepatitis B surface antigen were all negative. HIV antibody (1 and 2) screening serology was positive although this required further confirmation with Western Blotting. Ultrasound scan of the abdomen was normal. He had magnetic resonance imaging (MRI) of the whole spine which was normal. The following day, lumbar puncture was done and this showed clear, colourless cerebrospinal fluid (CSF) with 12 white blood cells/µml (all lymphocytes), 17 red blood cells/ µml, glucose 4.87 mmol/L (no serum glucose available) and protein 0.34g/L.

Five days after admission, he was transferred to the Government tertiary hospital for further care under the Neurology Unit. His symptoms had continued to progress. He had difficulty sitting and standing unaided. By the following day he had markedly deteriorated with distal weakness of 1-2/5 in all four limbs. In the upper limbs, biceps and triceps reflexes were preserved but supinator reflexes were lost. His lower limb reflexes were all absent. Sensation was intact. His cardiovascular and respiratory systems remained normal.

With his progression, the clinical picture was consistent with a rapidly progressive polyneuropathy on the background of a prodromal illness. The main differential diagnosis for this was GBS. Given the positive HIV serology, the possibility of this being HIV-related was highly likely. Other causes of a progres-

sive polyneuropathy needed consideration such as infective e.g. tuberculosis, other viruses (Epstein-Barr, cytomegalovirus) and infiltrative e.g. lymphomatosis. Normal MRI spine ruled out a compressive lesion.

He was put on close observations including cardiac and blood pressure monitoring in view of the potential for cardio-respiratory instability such as arrhythmia, hypo- and hypertension and respiratory failure. Low molecular weight heparin was commenced for thrombo-prophylaxis. He was also commenced on a five day course of intravenous (IV) immunoglobulin at a dose of 0.4g/kg body weight per day. Over the next three days, he stabilised and began showing an objective improvement in limb strength. By the fifth day of treatment he was virtually back to normal although reflexes remained abnormal. Nerve conduction studies were unavailable at the time of his admission.

Repeat HIV testing was confirmed to be positive. The patient was informed of the positive result and a full history was taken for risk factors. He admitted to having unprotected intercourse with a previous partner and sex workers (all heterosexual encounters). The diagnosis of HIV and its management was explained and he was discharged with a diagnosis of GBS associated with HIV infection.

DISCUSSION

Our case highlighted the association of HIV infection with an interesting neurological manifestation. Our patient had features consistent with GBS. The absence of other infections or atypical cells on CSF microscopy and culture, the normal MRI spine and the excellent response to IV immunoglobulin further strengthened the diagnosis. However, we were unable to determine whether it was demyelinating or axonal. Interestingly in many patients with HIV-associated GBS, the neuropathy is pure motor [2] and nerve conduction studies show the neuropathy to be axonal [3, 4] which differs from classic AIDP. Nerve conduction studies in our patient would have been a useful additional finding had they been available.

The incidental diagnosis of HIV was important as it provides a likely aetiological explanation and will be influential to his future management. GBS was first reported in a patient with AIDS in 1985. [4] In larger case series from Africa where in some parts HIV is endemic, there is a high prevalence of HIV infection among those with GBS (Zimbabwe 55% and Tanzania 30.5%). [5, 6]

There are numerous neurological complications of HIV and AIDS relating to the disease itself such as dementia and as sequelae of immunosuppression such as opportunistic infection and malignancy. Peripheral neuropathy is the commonest neurological complication. [7] Whilst this category includes GBS, the most frequent neuropathy encountered is a chronic distal polyneuropathy are often caused by anti-retroviral treatment. Other neuropathies include cytomegalovirus (CMV) related polyradiculopathy, mononeuritis multiplex and autonomic neuropathy.

The pathophysiology of HIV-associated GBS is not entirely clear. It often presents prior to the advent of AIDS and while CD4 count remains high and hence in some cases at least is unrelated to immunosuppres–

sion. [5, 8]

In conclusion, HIV should be considered in patients with a diagnosis of GBS. Intravenous immunoglobulin appears to be a helpful therapy with this combination. Whilst widespread HIV screening may not always be appropriate, there should be a high index of suspicion particularly if the GBS is purely motor and axonal even if no other features of HIV are present.

REFERENCES

1: Brannagan TH 3rd, Zhou Y. HIV-associated Guillain-Barré syndrome. J Neurol Sci. 2003; 208:39-42.

2: Verma A. Epidemiology and clinical features of HIV-1 associated neuropathies. J Peripher Nerv Syst. 2001; 6;8-13.

3: Perry GJ. Peripheral neuropathies associated with human immunodeficiency virus infection. Ann Neurol 1988; 23:S49-53.

4: Thornton CA, Latif AS, Emmanuel JC. Guillain-Barré syndrome associated with human immunodeficiency virus infection in Zimbabwe. Neurology 1991; 41:812-5.

5: Howlett WP, Vedeler CA, Nyland H, Aarli JA. Guillain-Barre syndrome in northern Tanzania: a comparison of epidemiological and clinical findings with western Norway. Acta Neurol Scand 1996; 93:44-9.

6: Pardo CA, McArthur JC, Griffin JW. HIV neuropathy: Insights in the pathology of HIV peripheral nerve disease. J Peripher Nerv Sys. 2001; 6:21-7.

7: Kumar S, Alexander M, Markandeyulu V, Gnanamuthu C. Guillain-Barré syndrome presenting in the anti-HIV seroconversion period. Neurol India 2003; 51:559.

8: Berger JR, Difini JA, Swerdloff MA, Ayyar DR. HIV seropositivity in Guillain-Barré syndrome. Ann Neurol 1987; 22:393-4.

9: Hughes RA, Swan AV, Raphaël JC, Annane D, van Koningsveld R, van Doorn PA. Immunotherapy for Guillain Barre Syndrome: a systematic review. Brain 2007; 130:2245-57.