

GUILLAIN-BARRÉ SYNDROME ASSOCIATED WITH ACUTE HEV HEPATITIS

Rawshan Ara Khanam, Mohammad Omar Faruq, Rawshan Ali Basunia, ASM Areef Ahsan

Department of Critical Care Medicine (ICU), Ibrahim Medical College & BIRDEM

Abstract

Guillain-Barré Syndrome (GBS) otherwise known as Acute Inflammatory Polyneuritis, characterized by acute progressive limb weakness and areflexia, is the prototype of a post infectious autoimmune disease. Two-thirds of the cases of GBS emerge from viral or bacterial infection. In August 2006, a 20 year old man presented at ICU, BIRDEM Hospital with a history of brief icteric illness followed by progressive bilateral symmetrical hypotonic areflexic muscular weakness, bilateral infra-nuclear facial palsy and bulbar weakness. Later on, he was diagnosed as a case of GBS and acute hepatitis E. Up till now, only three cases of GBS associated with hepatitis E have been reported in the medical literature world wide. This is probably the 4th case to be reported.

Ibrahim Med. Coll. J. 2008; 2(1): 32-34

Indexing words: Guillain-Barré Syndrome, acute hepatitis E

Introduction

The incidence of the typical GBS has been reported to be relatively uniform between 0.6 and 4 cases per 100 000 per year throughout the world¹. In two thirds of cases there are occurrences of gastroenteritis or flu like illness within six weeks of onset of GBS²⁻³. *Campylobacter jejuni* is reported to be the most frequent antecedent pathogen followed by cytomegalovirus⁴. Association of GBS with viral hepatitis is well documented. Most of the reported associations are with acute hepatitis B⁵⁻⁶ and hepatitis A⁷. Few cases also have been reported with Hepatitis C⁸. Association of GBS with HEV hepatitis is rare. Only three such cases have been published so far⁹⁻¹¹.

Case Report

In August 2006, a Bangladeshi 20 year old man employed in a plastic factory was admitted in the ICU, BIRDEM with the complaints of jaundice, weakness of limbs and dyspnoea. About fifteen days prior to admission (PTA), he developed low grade fever, anorexia, nausea and vomiting. Seven days PTA he noticed yellowish discoloration of skin, sclera, and

urine. Five days PTA, he felt weakness of both lower limbs progressing to both upper limbs over the next two days. He had no bladder or bowel dysfunction. Three days PTA, he developed dysphagia to both solid and liquid food accompanied by respiratory distress. Then he got admitted in an ICU out side of BIRDEM. There he was intubated and kept on mechanical ventilation (MV) and was finally transferred to ICU, BIRDEM. The patient had no prior history of receiving vaccination within in few months PTA.

On admission, he was conscious and mildly icteric. Respiratory rate was 24/min, pulse 100/min, blood pressure 150/80 mm of Hg and temperature 99° F. Systemic examination revealed mild tenderness in right hypochondrium and soft enlarged tender liver about 2 cm from the right costal margin. Neurological examination showed bilateral lower motor neuron (LMN) type facial palsy with evidence of dysphagia. Muscle power was 2/5 in all four limbs with diminished muscle tone. All the deep tendon and superficial reflexes were absent. Planter responses were equivocal. All sensory functions including touch, temperature and vibration were intact and there was no sign of meningeal irritation.

Address for Correspondence:

*Prof. Rawshan Ara Khanam, Department of Internal Medicine, Ibrahim Medical College & BIRDEM
122 Kazi Nazrul Islam Avenue, Shahbag, Dhaka-1000*

Laboratory investigations showed a total WBC count of 18700/cmm with Poly-85%, Lympho-12%, Mono-02% and Eosin-01%. Serum bilirubin was 3.6 mg/dl, ALT-2509U/L, AST-290 U/L, and Alkaline Phosphatase -580 U/L, with normal levels of urea, creatinine, electrolyte and coagulation profile. His anti HEV IgM was positive (ELISA, sample OD 2.735, cut off value 0.328) with negative viral markers for hepatitis B, hepatitis C and hepatitis A.

Ultrasonography revealed hepatomegaly suggestive of acute hepatitis. Nerve conduction studies revealed both demyelinating and axonopathic sensory motor polyneuropathy. CSF study showed albumin 190 mg/dl with "0" cell count – suggestive of albuminocytologic dissociation.

After diagnosis, the patient was advised for immunomodulation by either intravenous immunoglobulin (IvIg) or plasmapheresis. Although advised, patient could not be given IvIg or plasmapheresis for financial constraints. However, his muscle power showed significant improvement within one week of admission and reached 4/5 at lower limbs and 3/5 at upper limbs by two weeks of admission. Patient was successfully extubated from MV seven days after admission.

Discussion

GBS is a disease of peripheral nervous system, which is caused by aberrant immune response, directed against some components of peripheral nerves¹². The targeted antigens may be gangliosides present on plasma membrane of cell (e.g. GM-1, GD-1a), Schwann cell neurons (nerve growth cone region). There are four common sub types based on clinical and neuro-physiological studies e.g. 1. Acute Inflammatory Demyelinating Polyneuropathy (AIDP), 2. Acute Motor Axonal Neuropathy (AMAN), 3. Acute Motor Sensory Axonal Neuropathy (AMSAN), 4. Millar Fisher's Syndrome¹³.

Established diagnostic criteria exists. Once the clinical diagnosis is made, sub type classification can be done by using neuro-physiological study. Such study also confirms that the disease is a peripheral neuropathy¹⁴⁻¹⁵. Nerve conduction study in this case was suggestive of AMSAN subtype.

In rare cases, where nerve conduction studies cannot be done because of lack of recordable action potential,

nerve biopsy can differentiate the subtypes. However this is done only for research purposes¹⁶⁻¹⁷. CSF studies shows raised protein and paucity of cell in 80% cases (albuminocytologic dissociation), in 20% cases CSF study is usually normal in first few days of illness¹⁸⁻¹⁹. In this case, albuminocytologic dissociation was present when the CSF was studied four days after admission.

The treatment of GBS is multidisciplinary. Both plasma exchange and high dose of immunoglobulin are effective in reducing both the severity of the disease and the neurological deficits. Steroids are not effective in GBS²⁰. A randomized controlled trial showed that IvIg has similar efficacy to plasma exchange and it is recommended as the first line of treatment for severe GBS in most hospitals because of its greater convenience²¹. There are also case reports of spontaneous recovery of GBS without IvIg or plasmapheresis²². One of the three reported cases with HEV⁹ as well as this case recovered spontaneously without Immunomodulation.

References

1. Hubhcs RAC, Rees JH. Clinical and epidemiological features of Guillain-Barré Syndrome. *J Infect Dis* 1997; **176**(suppl): S92-98.
2. Winer JB, Hughes RAC, Anderson MJ, Jones DM, Kangro H, Watkin RFP. A prospective study of acute idiopathic neuropathy. *J Neurol Neurosur Psychiatry* 1988; **51**: 613-18.
3. Guillain- Barré Syndrome study group. Guillain-Barré Syndrome: an Italian multicentre case control study. *Neurol Sci* 2000; **21**: 229-34.
4. Schmidt-Ott R, Schmidt H, Feldmann S, Brass F, Krone B, Gross U. *Clin Vaccine Immunol* 2006; **13**(7): 779-83.
5. Berger JR, Ayyar R, Sharemata WA. Guillain-Barré Syndrome complicating acute hepatitis B. A case with detailed electrophysiological and immunological studies. *Arch Neurol* 1981; **38**: 366-8.
6. Ray G, Ghosh B, Bhattacharya R. Acute hepatitis B presenting as Guillain-Barré Syndrome. *Indian J Gastroenterol* 2003; **22**(6): 228.
7. Chitamber SD, Fadnis RS, Joshi MS, Habbu A, Bhatia SG. *J Med Virol* 2006; **78**(8): 1011-4.
8. Lacaillc F, Zvlberberg H, Hagege H, Roualdes B, Meyrignac C, Chousterman M, Girot R. Hepatitis C associated with Guillain-Barré Syndrome. *Liver* 1998; **18**(1): 49-51.

9. Sood A, Midha V, Sood N, Guillain-Barré Syndrome with Acute Hepatitis E. *AM J Gastroenterol* 2000; **95**(12): 3667-8.
10. Rajesh K, Bhoi S, Manish K, Sharma B, Singh BM, Gupta BB. Guillain-Barré Syndrome with Acute Hepatitis E - a rare association. *J Indian Acad Clin Med* 2002; **3**: 389-91.
11. Kamani P, Bajjal R, Amarapurkar D, Gupte P, Patel N, Kumar PH, Agal S. Guillain-Barré Syndrome associated with Acute Hepatitis E. *Indian J Gastroenterol* 2005; **24**: 216.
12. Palazzuoli A, Lenzi C, Iovine F, Carrea A, Nuti R. *Neurol Sci* 2006; **26**(6): 447-50.
13. Richard AC Hughes, David R Cornblath, *Lancet* 2005; **336**: 1653-66.
14. Hadden RDM, Cornblath DR, Hughes RAC. et al. Electrophysiological classification of Guillain-Barré Syndrome: Clinical association and outcome. *Ann Neurol* 1998; **44**: 780-88.
15. Ho TW, Mishu B Li CY, et al. Guillain-Barré Syndrome in northern China. Relationship to campylobacter jejuni infection and anti glycolipid antibodies. *Brain* 1995; **118**: 597-605.
16. Hall SM, Hughes RAC, Payan J, Atkinson PF, McColl I, Gale A. Motor nerve biopsy in severe Guillain-Barré Syndrome. *Ann Neurol* 1992; **31**: 441-44.
17. Barciano J, Figols J, Garcia A, et al. Fulminant Guillain-Barré Syndrome with universal excitability of peripheral nerves. A clinicopathological study. *Muscle Nerve* 1997; **20**: 846-57.
18. Paradiso G, Tripoli I, Galicchio S, Fejerman N. Epidemiological, clinical and electro diagnostic findings in childhood Guillain-Barré Syndrome. A reappraisal. *Ann Neurol* 1999; **46**: 701-07.
19. Hughes RAC. Guillain-Barré Syndrome. Heidelberg: springerverlag; 1990.
20. Cosi V, Versino M. Guillain-Barré Syndrome. *Neurol Sci* 2006; **27**(suppl 1): 347.
21. Van der Meche FGA, Schmitz PIM, Dutch. Guillain-Barre Study group. A randomized trial comparing intravenous immunoglobulin and plasma exchange in Guillain-Barre Syndrome, *N Engl J Med* 1992; **326**: 1123-29.
22. Lundkvist I, Van Doorn PA, Vermeulen M, Brand A. Guillain-Barré Syndrome, spontaneous recovery. *Clin Immunol Immunopathol* 1993; **67**(3 pt 1): 192-8.