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Severe Guillain-Barré Syndrome which Responded to Repeated Courses of Intravenous Immunoglobulin: A Case Report

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ABSTRACT. Guillain-Barré Syndrome is an idiopathic acute inflammatory demylinating polyneuropathy with good prognosis. We described a case of a 56-years old male patient was presented with flaccid quadriplegia and respiratory failure due to Guillain-Barré Syndrome which was successfully treated with repeated courses of intravenous immunoglobulin. Further studies are needed on this modality of treatment especially in severe form of Guillain-Barré Syndrome.

Keywords. Severe Guillain-Barré Syndrome, Respiratory failure, Intravenous immunoglobulin

Introduction

Guillain-Barré Syndrome (GBS) is an idiopathic acute inflammatory demyelinating polyneuropathy characterized by progressive muscle weakness and a relexia with spontaneous remission being the rule[1]. It is a leading cause of acute paralysis. It occurs world-wide in patients of all ages and both sexes[2, 3]. It has an annual incidence of 1.2 cases per 100,000 population[4] and a mortality of 3-8%[5-7]. About one-third of patients require intensive care unit admission for assisted ventilation and management of autonomic cardiovascular instability[5]. Several studies have shown the efficacy of intravenous immunoglobulin (IVIg) in the treatment of GBS [8-10].

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We described a case of a 56-years old male who presented himself with severe GBS (flaccid quadriplegia ad respiratory failure) that had been successfully treated with repeated courses of IVIg.

Case Report

A fifty-six (56) years old Saudi male patient was admitted to our hospital with a history of acute respiratory distress. His problem started 2 weeks before admission with a history of upper respiratory tract infection followed 10 days later by weakness in both lower limbs that progressively ascended to the upper limbs followed by the development of severe difficulty in breathing within one week of the start of his illness. His systemic review didn't reveal significant abnormalities. He has been a diabetic for 2 years and his blood glucose is well controlled with oral hypoglycemic agents. He had a history of abdominal surgery, 10-years ago for removal of colonic mass. He is a clerk, married with 4 children, and is a non-smoker. His family and allergic history were unremarkable. On examination, the patient was in severe respiratory distress, tachycardia (pulse 140/min.), blood pressure 140/90 mmHg, and his neurological examination revealed flaccid quadriplegia grade II. Cranial nerves' examination was intact and fundi were normal. Heart and chest examinations were clear. Abdominal examination revealed non-tender hepatomegaly with liver span of 16 cm. The patient was intubated and his initial investigations showed normal complete blood count, serum urea and creatinine, liver function tests, chest X-ray, and controlled blood glucose. His electrocardiography showed persistent sinus tachycardia. Lumber puncture was performed and it showed raised cerebrospinal fluid protein with normal cells. Electrophysiological nerve study's findings were consistent with the diagnosis of GBS with demyelinating feature. Cytomegalovirus and Epstein-Bar virus serology tests were negative. Abdominal ultrasound showed adenocarcinomata secondaries. The patient was treated with IVIg course (0.4 gm/kg/day) for 5 days, and beta-blocker (metoprolol) was added to control his sinus tacchycardia in addition to other supportive measures. The patient didn't show significant improvement neither in motor nor respiratory function. To control the underlying immunological abnormalities another course of IVIg (0.4 gm/kg/day) for 5 days was considered after 2 weeks. The patient showed remarkable improvement in his respiratory function within a few days and we were able to wean him from the ventilator. Also, he showed progressive improvement in motor function and was able to walk around without support. An oncologist's opinion was taken regarding the patient's liver metastasis. After 3 months of hospital's admission, the patient was discharged home in good general condition, breathing spontaneously and walking unassisted. He is on regular follow-up for 2 years now after his illness. He is doing fine without any motor or respiratory sequelae and without any relapses.

Discussion

GBS is an autoimmune disease of peripheral nerves. Suffers may require intensive care and may be permanently disabled[11]. GBS has a more benign course in children [12] and severe forms of adult GBS had been reported in patients with mean age of 48 years [6]. Increasing age and severe rapidly progressive disease had been found to be significantly associated with poor outcome[4, 7, 13]. Antecedent infection whether respiratory or gastrointestinal has been reported in approximately two-third of patients[1, 7]. Our patient was in the mid-fifties with a history of proceeding respiratory tract infection and rapidly progressive disease with respiratory failure within one week of the onset of his illness. Autonomic dysfunction is a well organized feature of GBS and is a significant source of mortality[14]. It has been described in 11% of patients with severe

GBS[6]. Our patient had persistent sinus tacchycardia due to autonomic dysfunction. The course of GBS is believed to be immune mediated, a variety of clinical and experimental data have implicated both humoral factor and cell mediated immune phenomena which damage myelin and/or myelin-producing Schwann cells[15]. The main modalities of therapy for GBS is to control the immune abnormality by either plasma pheresis (PE) or IVIg. Intravenous immunoglobulin has been advocated for therapy of GBS as it is effective, safe, and easy to use[8, 10, 16]. Combination of PE and IVIg in severe disease didn't confer a significant advantage over single therapy, but it increases cost and risk [9]. Our patient showed dramatic response to repeated course IVIg without sequelae or relapses. Farcas *et al*.[17] had described a remarkable recovery after the second course of IVIg in a case of severe GBS. This raises the possibility that repeated courses of IVIg might be helpful in severe GBS. Further studies are needed on this modality of therapy especially in severe forms of GBS.

References

[1] **Hahn AF.** Guillain-Barré syndrome. *Lancet* 1998; **352:** 635-641.

[2] **Hughes ARC**. *Guillain-Barré Syndrome*. London: Springer-Verlag, 1990.

[3] **Ropper JH, Wijdicks EFM, Truax BT.** *Guillain-Barré Syndrome. Volume 34 of Contemporary Neurology Series.* Philadelphia, PA USA: FA Davis, 1991.

[4] **Rees JH, Thompson RD, Smeeton NC, Hughes RA.** Epidemiological study of Guillain-Barré syndrome in southeast England. *J Neurol Neurosurg Psychiatry* 1998; **64(1):** 74-77.

[5] **Hahn AF.** Management of Guillain-Barré Syndrome (GBS). *Baillieres Clin Neurol* 1996; **5(3):** 627-644.

[6] **Ng KK, Howard RS, Fish DR, Hirsch NP, Wiles CM, Murray NM, Miller DH.** Management and outcome of severe Guillain-Barré Syndrome. *QJM* 1995; **88(4):** 243-250.

[7] **(No authors listed).** A prospective study on the inidence and prognosis of Guillain-Barre syndrome in Emilia-Romagna region, Italy (1992-1993). Emilia-Romagna Study Group on Clinical and Epidemiological Problems in Neurology. *Neurology* 1997; **48(1):** 214-221.

[8] **Sater RA, Rostami A.** Treatment of Guillain-Barré syndrome with intravenous immunoglobulin. *Neurology* 1998; **51 (6 Supp. 5):** S9-15.

[9] **(No authors listed).** Randomised trial of plasma exchange, intravenous immunoglobulin, and combined treatments in Guillain-Barre syndrome. Plasma Exchange/Sandoglobulin Guillain-Barré Syndrome Trial Group. *Lancet* 1997; **349:** 225-230.

[10] **Fasanaro AM, Pizza V, Rossi V.** Immunoglobulins i.v.: a new approach to the treatment of Guillain-Barré syndrome. *Minerva Med* 1996; **87(1-2):** 17-20.

[11] **Hadden RD, Hughes RA.** Guillain-Barré syndrome: recent advances. *Hosp Med* 1998; **59(1):** 55-60.

[12] **Gurses N, Uysal S, Cetinkaya F, Islek I, Kalayci A**. Intravenous immunoglobulin treatment in children with Guillain-Barré syndrome. *Scan J Infect Dis* 1995; **27:** 241-3.

[13] **Ropper AH.** The Guillain-Barré syndrome. *N Engl J Med* 1992; **326(17):** 1130-1136.

[14] **Hund EF, Borel CO, Cornblath DR, Hanley DF, McKhann GM.** Intensive management and treatment of severe Guillain-Barré syndrome. *Crit Care Med* 1993; **21(3):** 433-446.

[15] **Saida T, Saida K, Dorfman SH, Silberberg DH, Sumner AJ, Manning MC, Lisak RP.** Experimental allergic neuritis induced by sensitization with galactocelebroside. *Science* 1979; **204:** 1103-1106.

[16] **van der Meche FG.** The Guillain-Barré syndrome: diagnosis, treatment and prognosis. *Acta Neurol Belg* 1994; **94(2):** 124-127.

[17] **Farcas P, Avnun L, Frisher S, Herishanu YO, Wirgiun I.** Efficacy of repeated intravenous immunoglobulin in severe unresponsive Guillain-Barré syndrome. *Lancet* 1997; **350(9093):** 1747.

حـالة خطرة لمرض الجيلـين باري التي استجابت لجرعات متكررة من البروتين الممنع

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المستخلص. مرض الجيلين باري هو التهاب حاد للأعصاب. أسبابه غير معروفة وعادة ما تكون نسبة الشفاء فيه عالية. لقد تم وصف حالة رجل يبلغ من العمر ٥٦ سنة أصيب بمرض الجيلين باري بصورته الخطرة مع شلل رباعي للأطراف وقصور في التنفس وقد تم شفاءه بعد علاجه بجرعات متكررة من البروتين الممنع . يجب عمل دراسات مستقبلية لهذه الطريقة من العلاج لإثبات فعاليتها في حالات الجيلين باري الخطرة .