Guillain-Barré syndrome with autoimmune hemolytic anemia following acute viral hepatitis


*Istituto di Clinica Medica I A. Ferrata
**Istituto Scientifico C. Mondino, Clinica Neurologica
***Istituto di Anatomia ed Istologia Patologica, Università di Pavia

A patient with Guillain-Barré syndrome associated with autoimmune hemolytic anemia following post-transfusional acute viral HBsAg-positive hepatitis improved dramatically and subsequently recovered from both diseases after plasmapheresis and steroid therapy. This case supports an autoimmune mechanism of Guillain-Barré syndrome.

Key-Words: Guillain-Barré syndrome — autoimmune hemolytic anemia — acute viral hepatitis — plasmapheresis

Introduction

The pathogenesis of Guillain-Barré syndrome (GBS) is still unknown, but experimental evidence has accumulated in support of the possibility that the nerve damage is immunologically mediated [1]. In fact there are many similarities between neural lesions in experimental allergic neuritis (EAN), an immuno-mediated disorder that occurs in animals inoculated with peripheral nerve antigens, and those seen in GBS. These lesions are virtually indistinguishable one from another [2, 3]. Moreover, some workers have succeeded in passively transferring primary demyelination to normal animals by injection of EAN serum [4]. Others have suggested the hypothesis of an imbalance between T cell subsets, with decreased T suppressor activity induced by circulating immune complexes [5, 6].

Our case report of simultaneous onset of GBS and immunologically-mediated hemolytic anemia supports the immunopathogenetic mechanism of GBS.

Case report

A 63 year old man suffering from mitral valve stenosis of rheumatic origin underwent valvulotomy because of atrial fibrillation and repeated left ventricular failure on 1 June 1983. Some days after surgery hematemia and melena from a peptic ulcer of the duodenum (shown by gastroduodenoscopy) occurred, necessitating a total of 8 blood transfusions. He was discharged on 14 June 1983. 15 days later fatigue and anorexia began. On 10 August he was admitted to the local hospital due to increasing jaundice, dark urine and weight loss. The physical examination showed principally hepatomegaly (2 cm beyond lower costal margin) and evident jaundice. The values of SGOT (467 mU/ml), SGPT (651 mU/ml) and total serum bilirubin (11.3 mg/100 ml) were high. Since the serum contained HBsAg, a diagnosis of acute viral hepatitis was made. At the end of August 1983 normochromic anemia (Hb 9.9 gr/dl) was detected with high unconjugated biliru-
bin levels (see fig. 1), while SGOT and SGPT had became almost normal. At the same time the patient complained of neurological troubles, namely paresthesias and increasing weakness of the limbs so that within 3-4 weeks he was unable to climb stairs or dress. The only treatment given was packed red cell transfusions.

On 10 October he was transferred to our Institution. Physical examination showed only mild jaundice, hepatomegaly (2 cm beyond lower costal margin), while neurological examination showed weakness mainly of proximal muscles (2.5-3.5 according to MRC grading), mild hypotrophy of involved muscles, diffuse tendon areflexia; no sensory deficits were observed, although the paresthesias persisted. Hb was 12.0 gr/dl, with increased reticulocytosis (fig. 1), unconjugated bilirubin was 3.2 mg/100 ml. Bone marrow smears showed erythroid hyperplasia. The direct Coombs test was positive (IgG ++ , Cd + - ), haptoglobin was 43 mg/100 ml. HBsAg was not demonstrable in the serum, although HbcAb and HBeAb were detected. Circulating immune complexes at low rates (1:4) were present. Other humoral parameters were normal. Cerebro spinal fluid (CSF) examination showed high protein levels (107 mg/100 ml) with CSF cells lower than 1/mm³; electric focusing of CSF revealed a double T, with a similar serum and CSF protein pattern, as happens in course of blood-CSF barrier damage. Electromyography (EMG) and electroneurography (ENG) showed spontaneous activity, increased length and amplitude of motor unit potentials owing to increased percentage of polyphasic potentials with reduced interference pattern at maximal effort in deltoid, anterior tibial and femoral quadriceps muscles; motor conduction velocity of the SPE was only slightly reduced but with reduced M response and increased distal latencies. Sural nerve biopsy (fig. 2) showed marked loss of small myelinated fibers, with aspects of regeneration of myelin sheaths in some fibers scattered fibers showed wallerian degeneration. No significant modifications of the vasa nervorum or amyloid deposits were observed. Biopsy of deltoid muscle showed typical neurogenic changes of muscular fibers.