Exceptional Association of a Cerebral Sinus Thrombosis and a Guillain-Barre Syndrome: A New Case Report and Review of the Literature

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Abstract

Background: The association of Guillain-Barre syndrome and cerebral sinus thrombosis is uncommon. Case Presentation: We report a 37-year-old patient hospitalized in medical ICU for respiratory distress following a Guillain-Barre syndrome. He had symptomatic treatment in addition to plasma exchange. In the presence of clonic movements, a brain venography magnetic resonance showed a thrombophlebitis of the left lateral sinus, and hence a low-molecular-weight heparin treatment was begun. Immunological, thrombophilia and serological tests were negative. After a favorable evolution, he was transferred to the neurology department. Conclusion: The combination of a Guillain-Barre syndrome and a cerebral sinus thrombosis would suggest a common process. A rigorous investigation, including the use of imaging, is necessary in front of any unusual clinical sign during a GBS.

Keywords
Cerebral Sinus Thrombosis, Guillain-Barre Syndrome, Plasma Exchange, Venography Magnetic Resonance

1. Introduction

Although a rare disease (5 cases/1,000,000 inhabitants), cerebral sinus thrombosis (CST) is a cause of stroke (0.5% of strokes), characterized by the extreme diversity of their clinical presentation and etiologies [1] with a mortality of less than 5% during the acute phase [2]. Its etiology and risk factors can be classified as infectious (local, regional or general) and non-infectious affections (general,
mechanical or hemodynamic, prothrombotic, inflammatory, hematological, medical or unidentified causes) [3]. Guillain-Barre syndrome (GBS) is an immune-mediated disease affecting the peripheral nerves with an inflammatory cell infiltration. GBS has an incidence of 1 - 2 cases/100,000 per year [4] with a mortality of 3% - 7% [5], a large spectrum of short-term and long term complications and significant morbidity since 40% of patients were admitted to the rehabilitation centers [6]. In two-thirds of cases, GBS is a typical case of acute respiratory or gastrointestinal (bacterial or viral) infectious disease [7]. Local or systemic infections are admitted risk factors in the occurrence of CST and GBS; however, no etiological link between the two pathologies has been elucidated [8].

We report an unusual association of CST and GBS in a 37-year-old man and review of published similar cases.

2. Case Presentation

The case is a 37-year-old male patient with no past medical history. The patient had been well until four weeks before this admission when untreated gastroenteritis developed. Two weeks later, he presented symmetric lower limbs weakness and tingling which was brusque, bilateral, synchronous and spreading to the arms. On examination, the temperature was afebrile (37.4˚C), the blood pressure 132/70 mmHg, the pulse 90 beats per minute, the respiratory rate 18 breaths per minute, and the oxygen saturation 98% while the patient was breathing ambient air. He was conscious (Glasgow Coma Scale was 15) with tetraplegia and hypoglossal nerve palsy. Strength was symmetric and graded as 3 out of 5 in the proximal and distal muscles of the arms, 0 out of 5 in the proximal and distal muscles of the legs. The remainder of the clinical examination (cardiovascular, respiratory and abdominal) was normal. Electromyography (EMG) showed very low motor conduction velocities at the upper limbs and undetectable at the lower limbs with low amplitudes, low sensitivity conduction velocities with very low amplitudes, scattered and elongated F waves, compatible with acute inflammatory demyelinating polyneuropathy (Guillain-Barre syndrome). Cerebrospinal fluid (CSF) showed increased albumin with normal CSF white blood cell count. Laboratory analysis demonstrated normal blood cell counts (leukocytes 9430/mm³, platelets 305.10³/mm³, red cells 4.7.10⁶/mm³, and hemoglobin 13 g/dl), as was the result of the renal function test (urea 0.3 g/l, creatinine 6 mg/l). The aspartate aminotransferase and alanine aminotransferase were elevated at 70 IU per liter and 353 IU per liter respectively. A cerebral magnetic resonance imaging (MRI, without venography) performed before admission was normal. Plasma exchange was started on the first day of hospitalization in the neurology department. Twenty-four hours later, the patient was referred to our medical ICU because of respiratory distress requiring mechanical ventilation. The patient had symptomatic treatment and treated with plasma exchange. After cessation of sedation, the patient presented clonic movements of the right upper limb and the right side of the face. A cerebral angio-MRI showed a thrombophlebitis of
the left lateral sinus (Figure 1), hence the initiation of curative anti-coagulation based on low molecular weight heparin associated with sodium valproate. An immunoassay and thrombophilia assessment were normal. Human immunodeficiency virus and hepatitis serologies were negative. After a stay of 23 days in the medical ICU with a good clinical evolution, the patient was transferred to the neurology department. Then, he was discharged from the hospital three weeks later and is alive after a follow-up of 90 days.

3. Discussion

The occurrence of CST has been reported as a complication of intravenous immunoglobulin (IVIG) administration in an immune thrombocytopenia purpura [9], a humoral immunodeficiency (Bruton’s disease) [10], a polymyositis [11] and an isolated IgG1 deficiency [12]. In the GBS, only one case of CST was published [8]. The latter case was hospitalized for a neurological symptomatology of a headache, dizziness, swallowing disorders, vomiting and unilateral facial and hypoglossal paralysis.

Cerebral thrombosis of the right transverse sinus has been demonstrated on an angio-MRI and an anticoagulant therapy has been initiated. The clinical condition of the patient was worsened by the installation of respiratory distress with paralysis and areflexia of the lower limbs. The diagnosis of GBS has been made in the presence of clinical features, CSF analysis and the EMG findings. Viral serologies were negative.

Figure 1. An obstruction of the left lateral sinus on an angio-MRI in a 37-year-old patient hospitalized for Guillain-Barre syndrome.
While our patient was treated by plasma exchange (PE). However, during 20,485 procedures on the Swedish registry and 144,432 procedures on the Canadian apheresis registry, no CST was reported [13] [14]. Subacutely, CST may occur without or with few nonspecific symptoms in about 30% of patients [15] [16]. Clinically, our patient had a glosso-pharyngeal nerve palsy, but the initial MRI was normal, although at the beginning (before the 5th day), false negatives could be seen at MRI, an isosignal in T1 and hyposignal in T2, which necessitates the use of sequences T2* and angio-MRI vein for the confirmation of the diagnosis [17]. There is no evidence that CST can cause GBS or vice versa; as a result, this exceptional association may suggest a possible common infectious process that triggers GBS and progresses through the meningeal spaces to the cerebral veins and sinuses to induce CST [8].

4. Conclusion

The combination of a GBS and a CST would suggest a common process. A rigorous investigation, including the use of imaging, is necessary in front of any unusual clinical sign during GBS.

Competing Interests

The authors declare that they have no competing interests.

Informed Consent

The patient gave informed consent.

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The staff at the medical ICU and neurology department.

References


List of Abbreviations

GBS: Guillain-Barre Syndrome
CST: Cerebral Sinus Thrombosis
EMG: Electromyography
ICU: Intensive Care Unit
CSF: Cerebrospinal Fluid
MRI: Magnetic Resonance Imaging
IVIG: Intravenous Immunoglobulin
IgG: Immunoglobulin G
PE: Plasma Exchange