Guillain-Barré Syndrome Complicated by Myocarditis

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Abstract

Guillain-Barré syndrome (GBS) is an autoimmune disease affecting the peripheral nerves, and is frequently associated with triggering events several weeks prior to the onset of symptoms. We report the case of a 68-year-old female who was diagnosed with GBS and subsequently developed myocarditis. She was treated with inotropic support and intravenous immunoglobulin (IVIG), and her condition improved. This presentation of GBS complicated by myocarditis is very rare. We examined the literature regarding this association.

Key Words: Guillain-Barré Syndrome, myocarditis, cardiomyopathy.

Introduction

GBS is an autoimmune disease affecting the peripheral nervous system. It is associated with triggering events several weeks prior to the onset of symptoms. The most common triggers are upper respiratory and gastrointestinal infections. Known complications include respiratory failure, autonomic dysfunction, and syndrome of inappropriate antidiuretic hormone secretion (SIADH); there have been rare reports of GBS complicated by myocarditis. We report the case of a 68-year-old female who was diagnosed with GBS and subsequently developed myocarditis. This presentation of GBS complicated by myocarditis is very rare. We examined the literature regarding this association.

Case Summary

Our patient is a 68-year-old woman who initially presented to her internist with a one-week history of flu-like symptoms. This was diagnosed as bronchitis and treated with levofloxacin. Several days later, she presented to the emergency department, complaining of bilateral lower extremity weakness, lower back pain, and bilateral buttock pain. Vital signs were within normal limits. Neurological exam revealed 4/5 strength in both lower extremities and the left upper extremity, and 5/5 strength in the right upper extremity with absent deep tendon reflexes in all four extremities. The patient had a right up-going plantar reflex and decreased sensation in her right lower extremity. Laboratory tests were unremarkable except for serum sodium of 126 mmol/L. Initial electrocardiogram (Fig. 1A) was unremarkable, and initial chest X-ray (Fig. 1B) showed no acute disease. Lumbar puncture revealed no cells and mildly elevated protein (55 mg/dL), consistent with early GBS. Initial electromyogram showed an absent H reflex (the most sensitive test for early GBS), and repeat electromyogram showed diffuse demyelinating polynuropathy, consistent with GBS.

The patient was admitted and developed progressive weakness. She later developed respiratory failure and autonomic instability, requiring intubation and monitoring in the intensive care unit. Subsequently, she manifested a new incomplete right bundle branch block and a left anterior fascicular block on electrocardiogram (Fig. 1C). Creatine phosphokinase remained normal and troponin-I increased to a maximum of 1.8 ng/mL. Transthoracic echocardiography revealed global left ventricular dysfunction. There was a moderate pericardial effusion localized around the right heart and apex. Subsequent cardiac catheterization demonstrated normal coronary arteries (Fig. 2), and a severely decreased left ventricular ejection fraction (35%) was seen on ventriculography (Fig. 3). This presentation was consistent with acute myocarditis, and so the patient was given inotropic support. After a long and complicated hospital course, and treatment with IVIG, the patient’s condition improved, and she was discharged to a rehabilitation facility, with significant residual lower extremity weakness and vocal cord dysfunction.

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Myocarditis is most commonly the result of an infectious process, although it may also result from hypersensitivity to drugs, chemicals, radiation, or physical agents. While almost every infectious agent is capable of producing myocarditis, the United States clinically significant acute myocarditis is most commonly caused by viruses, especially Coxsackie virus B and echovirus. The clinical expression of myocarditis ranges from the asymptomatic state associated with limited and focal inflammation to fulminant fatal congestive heart failure due to diffuse myocarditis associated with transient ST and T wave abnormalities, atrial and ventricular arrhythmias, heart failure, and death. Myocarditis may also be associated with AV conduction defects, and rarely, Q waves (1–6). Patients with myocarditis may complain of pleuritic chest pain; however, the pain may be more typical of ischemia and simulate an acute myocardial infarction with electrocardiographic changes and elevated cardiac enzymes, suggesting myocardial necrosis.

To our knowledge, fewer than ten cases of GBS complicated by myocarditis have been reported in the medical literature (Table). Two cases were seen in children; one associated with infectious mononucleosis (1), and another with an upper respiratory tract infection (2). Four cases were seen in adults; one associated with gonococcal urethritis (4), another with Lyme disease (6), a third occurring with a relapse of GBS (5), and finally a case of hypersensitivity myocarditis probably secondary to the concurrent administration of phenytoin and IVIG (3).

The precise causal mechanism for myocarditis was unclear in most of the above cases. It was noted by Reimers et al. (6), in his reported cases of GBS, that myocarditis could be caused by direct invasion of the myocardium by Borrelia burgdor-
feri. It is also possible that the same inflammatory or autoimmune response to an infectious process was responsible for both peripheral nerve and myocardial damage. In one other case, an epileptic patient on phenytoin died after being treated with IVIG for GBS. On autopsy, he was found to have a hypersensitivity myocarditis, presumably from the interaction between phenytoin and IVIG (2).

Many patients with GBS develop autonomic instability associated with blood pressure fluctuations, postural hypotension, and cardiac dysrhythmias secondary to demyelination of the vagus and glossopharyngeal nerves, the sympathetic chain, and the white rami communicantes (2). Some GBS patients believed to have autonomic instability, based on the presence of tachycardia, may actually be developing acute myocarditis.

**Conclusion**

GBS is an autoimmune disease affecting peripheral nerves and is often associated with autonomic instability. Although rare, GBS has been known to be complicated by myocarditis, which is potentially fatal. If the clinician knows to include myocarditis in the differential diagnosis of the GBS patient with tachycardia and/or electrocardiographic changes, early recognition and proper treatment may prevent unnecessary morbidity and mortality.

**References**


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**TABLE**

Reported Cases of GBS Complicated by Myocarditis

<table>
<thead>
<tr>
<th>Report</th>
<th>Age/Sex</th>
<th>Associated Conditions</th>
<th>Clinical Outcome</th>
<th>Language</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gortner¹ (1984)</td>
<td>10 M</td>
<td>Infectious mononucleosis, membranous tonsillitis, nephritis</td>
<td>Mild neurologic deficits</td>
<td>German</td>
</tr>
<tr>
<td>Hodson et al.² (1984)</td>
<td>12 M</td>
<td>Upper respiratory tract infection, paralytic ileus, SIADH, autonomic instability</td>
<td>Death</td>
<td>English</td>
</tr>
<tr>
<td>Koehler et al.³ (1996)</td>
<td>43 M</td>
<td>Epilepsy, upper respiratory tract infection, hypersensitivity to phenytoin and IVIG</td>
<td>Death</td>
<td>English</td>
</tr>
<tr>
<td>Marshall et al.⁴ (1975)</td>
<td>23 M</td>
<td>Gonococcal urethritis, papilledema</td>
<td>Full recovery</td>
<td>English</td>
</tr>
<tr>
<td>Okuma et al.⁵ (1999)</td>
<td>35 M</td>
<td>Upper respiratory tract infection 2 years prior leading to GBS, with re-presentation two years later with an acute relapse of GBS associated with myocarditis</td>
<td>Full recovery</td>
<td>Japanese</td>
</tr>
<tr>
<td>Reimers et al.⁶ (1993)</td>
<td>38 M</td>
<td>Lyme disease</td>
<td>Death</td>
<td>English</td>
</tr>
<tr>
<td>Goldman and Makaryus (Current Report, 2006)</td>
<td>68 F</td>
<td>Upper respiratory tract infection/bronchitis</td>
<td>Significant residual lower extremity weakness, vocal cord dysfunction</td>
<td>English</td>
</tr>
</tbody>
</table>

SIADH = syndrome of inappropriate antidiuretic hormone; IVIG = intravenous immunoglobulin; GBS = Guillain-Barré syndrome.