□ CASE REPORT □

Fisher Syndrome Associated with Immune Thrombocytopenic Purpura

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Abstract

We describe a 51-year-old man with Fisher syndrome (FS) and immune thrombocytopenic purpura (ITP) that developed after upper respiratory infection (URI). Laboratory investigations demonstrated immunoglobulin (Ig) G class of anti-GQ1b autoantibody and reduced platelet count with platelet-associated IgG, which spontaneously improved in parallel with neurologic symptoms. Thus the possible association of ITP should be considered when encountering a patient with FS. This patient suggests that there may be a certain infectious agent causing URI, leading to the co-occurrence of FS and ITP.

Key words: Fisher syndrome, immune thrombocytopenic purpura, upper respiratory infection

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Introduction

Fisher syndrome (FS), an immunological disorder characterized by ophthalmoplegia, ataxia, and areflexia, is considered to be a variant of Guillain-Barré syndrome (GBS) (1). Immune thrombocytopenic purpura (ITP) is also considered an autoimmune disorder characterized by platelet depletion and mucocutaneous bleeding (2). Although associations between GBS and ITP have been reported (3-8), there have not been any reports of FS associated with ITP. We describe a patient with FS and ITP that developed after upper respiratory infection (URI).

Case Report

A 51-year-old man with a 9-day history of low-grade fever, dry cough, and sore throat developed numbness of the distal extremities and a floating sensation 5 days after spontaneous improvement of infectious symptoms. He had been receiving treatment with candesartan cilexetil for IgA nephropathy and hypertension since the age of 45.

On admission, his blood pressure was slightly elevated (147/86 mmHg), body temperature was normal, and there was no purpura. Neurologic examination demonstrated hori-

zontal gaze nystagmus, restriction in upward gaze, paresthesia with a glove and stocking type distribution, diminished tendon reflexes, and limb and truncal ataxia. Urinalysis demonstrated microscopic hematuria and proteinuria. On hematological examination, the WBC count was 6,770/ μ L with a normal differential count, hemoglobin 13.6 g/dL, and platelet count 68,000/ μ L. Intensive investigations for coagulopathy including fibrinogen, D-dimer, and lupus anticoagulant were all normal. Blood chemistry tests including thiamine were also normal. Immunological investigation was positive for platelet-associated immunoglobulin (Ig) G (PA-IgG, 511 ng/ 10^7 cells; normal range <45 ng/ 10^7 cells), and IgG class antibodies against ganglioside GT1a, GT1b, and GQ1b. Investigations for infectious agents including mycoplasma, Epstein-Barr virus, Cytomegalovirus (CMV), and *Helicobacter pylori* (HP) were all negative. Cerebrospinal fluid investigations did not show any abnormal findings including cell count or protein level. Magnetic resonance image of the brain and cervical spinal cord did not show any remarkable findings. Neurologic symptoms gradually improved two days after admission without any treatment, and platelet count and PA-IgG level also improved in parallel with neurologic symptoms (Fig. 1).

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Figure 1. Clinical course of the patient. Upper respiratory symptoms developed on day 0 and continued for nine days. Platelet count on day 7 was normal. Five days after recovery from upper respiratory symptoms, sensory disturbance and floating sensation developed and gradually worsened. On admission (day 19), reduced platelet count with a high level of PA-IgG was observed and gradually recovered with improvement of neurologic symptoms.

Discussion

The present patient developed FS and thrombocytopenia after URI. Although our patient had no purpura, we considered the patient as having mild ITP because of the presence of PA-IgG without any other cause of thrombocytopenia. Neurologic symptoms, platelet counts, and titer of PA-IgG all improved spontaneously in parallel. The possible association of ITP should be considered when encountering a patient with FS.

Although FS and ITP may appear with an autoimmune-mediated disorder, the reason for the association between FS and ITP in the present patient is not clear. GQ1b is a ganglioside that is expressed on nerve tissue and anti-GQ1b autoantibody is frequently detected in FS (1). Meanwhile, a major target of ITP is the glycoprotein IIb/IIIa complex, which is expressed on the platelet surface (2). As previously discussed in cases of GBS associated with ITP, ganglioside constituents of platelets differ from those of nerve tissue, and glycoprotein IIb/IIIa complex is not expressed in nerve tissue (6). It is unlikely that the association of ITP with GBS or FS can be simply attributed to the common antigen between the nerve tissue and platelets.

FS occurs subsequent to a wide variety of infections. *Campylobacter jejuni* and *Haemophilus influenzae* are related to development of the disease in some patients with FS, however, the causative agent remains unclear in the majority of the patients (9). On the other hand, it is also considered that some infectious agents including HP, CMV or varicella-zoster virus is related to disease development in some patients with ITP (10). Interestingly, previously re-

ported patients with GBS and ITP (3-8) as well as the present patient had preceding URI. Although the mechanism of association with GBS/FS and ITP is unclear, there is the possibility that a certain infectious agent causing URI may have epitopes of both ganglioside constituent of nerve tissue and platelet-surface glycoprotein, leading to the co-occurrence of GBS/FS and ITP.

The authors state that they have no Conflict of Interest (COI).

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