CASE REPORT

Ulcereative Colitis Associated with Isolated Unilateral Hypoglossal Nerve Palsy

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Abstract

We herein describe a rare case of ulcerative colitis associated with unilateral hypoglossal nerve palsy. A 64-year-old woman developed severe active ulcerative colitis and was treated with prednisolone. The dose of oral prednisolone was reduced to 7.5 mg/day, following which the patient noticed slight dysphagia and a speech disturbance. She was diagnosed with unilateral hypoglossal nerve palsy, which was thought to be caused by mononeuritis. She was treated with intravenous methylprednisolone at a dose of 500 mg/day, which improved the neuropathy. When the neuropathy occurred, the patient was in a mildly active stage of ulcerative colitis. We concluded that the mononeuritis observed in the present case was likely an extraintestinal manifestation of ulcerative colitis.

Key words: hypoglossal nerve palsy, ulcerative colitis, neuropathy

(intern Med 51: 3135-3137, 2012)
(DOI: 10.2169/internalmedicine.51.8616)

Introduction

Ulcerative colitis (UC) is a chronic disorder characterized by inflammation of the colonic mucosa. However, UC should be considered as a systemic disease, as it is associated with the clinical manifestations involving organs outside of the alimentary tract. On the other hand, there are few reports of neuropathy associated with UC, while other extraintestinal manifestations (EIMs) have been described (1-4). We herein present a rare case of UC associated with unilateral hypoglossal nerve palsy.

Case Report

A 64-year-old woman was diagnosed with ulcerative colitis (UC) in August 2009 and had sustained a clinical remission following the administration of oral mesalamine at a dose of 4 g daily. However, iritis developed in March 2010 and she was treated with betamethasone eye drops. Approximately one half month later in April, the patient experienced severe active rectal bleeding, diarrhea and abdominal pain. She visited the outpatient clinic of the Department of Internal Medicine of Matsue Seikyo Hospital at the end of April, at which time, the Rachmilewitz clinical activity index score was 19. Colonoscopy findings revealed multiple deep ulcers (Fig. 1), and the patient was considered to be suffering from a severe active episode of UC. She was admitted and treated with intravenous prednisolone at a dose of 50 mg once daily and granulocytapheresis (GCAP) twice a week, after which the abdominal symptoms gradually improved and remission was achieved two months later. Furthermore, the colonoscopy findings showed mucosal healing in the large intestine. Treatment with azathioprine was then started at the beginning of June, and the dose of oral prednisolone was gradually reduced to 7.5 mg once daily by September.

At the beginning of September, 2010, the patient noted slight dysphagia and a speech disturbance and visited the outpatient clinic of the Department of Neurology three days later. A neurological examination revealed deviation of the

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Received for publication July 16, 2012; Accepted for publication August 19, 2012
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torque to the right side from atrophy of the right lingual muscle (Fig. 2); no other abnormal neurological findings were detected. She was diagnosed with right hypoglossal nerve palsy and therefore was readmitted to the hospital. Computed tomography and magnetic resonance imaging examinations found no abnormalities in the brain, while a swallowing function examination demonstrated an abnormality of molding foods due to a lack of tongue function. In addition, the cerebrospinal fluid (CSF) protein level was elevated to 49 mg/dL, whereas no oligoclonal bands or myelin basic proteins were detected and the Immunoglobulin (Ig) G index was within normal limits at 0.61. The level of C-reactive protein (CRP) was elevated to 1.57 mg/dL and the erythrocyte sedimentation rate (ESR) was 51 mm/hour. Tests for vitamins B1 and B12, blood sugar, hemoglobin A1c, immunoglobulin, complement, antinuclear antibodies, cytoplasmic and perinuclear antineutrophil antibodies (C-ANCA, P-ANCA) and D-dimer in the serum were either normal or negative. A virological examination for cytomegalovirus, herpes virus and human T-cell leukemia virus found no activity. It was therefore concluded that the right hypoglossal nerve palsy was caused by mononeuritis due to an immune-mediated response. Treatment with intravenous methylprednisolone was initiated at a dose of 500 mg/day for five days, after which the dose of oral prednisolone was increased again to 30 mg/day. The patient’s tongue deviation began to gradually improve and completely resolved one month later. At the same time, the CSF protein level decreased to 36 mg/dL (within the normal range), and the CRP and ESR levels decreased to 0.20 mg/dL and 20 mm/hour, respectively. Thereafter, no abnormal neurological findings were seen for one year.

The patient’s abdominal symptoms remained unchanged and colonoscopy was again performed at the end of October, which showed diffuse erosive mucosa in the sigmoid colon and rectum, while UC was found to be exacerbated on the endoscopic findings.

Discussion

UC should be considered a systemic disease, as it is associated with clinical manifestations involving organs outside of the alimentary tract. Recent studies have focused on probable manifestations of UC in the nervous system, although few have noted neuropathy, and other EIMs have been reported (1-4). To the best of our knowledge, there are no reports of neuropathy of the hypoglossal nerve occurring in association with UC, although some studies have noted the occurrence of cranial nerve disorders, such as optic neuritis, and sensorineural hearing loss (1, 5-11). Furthermore, one study found that demyelinating diseases occur more frequently in patients with UC than in non-UC patients (12). Moreover, it is noteworthy that demyelination occurring during anti-tumor necrosis factor alpha therapy with infliximab has been reported (3, 13).

Initially, we considered the possibility that the neuropathy observed in the present case was associated with demyelinating disease. However, the laboratory data excluded that possibility, and no abnormal neurological findings were demonstrated thereafter. In addition, other causes of neuropathy were excluded, including viral infection, collagen disease, vasculitis and cerebrovascular disease. Therefore, we concluded that the isolated hypoglossal nerve palsy observed in this case was a form of immune-mediated neuropathy. In fact, immunosuppressive therapy with the administration of intravenous methylprednisolone proved to be very effective in treating right hypoglossal nerve palsy in our patient.

Some EIMs related to UC may not be correlated with the disease activity of colitis, although most tend to follow the clinical course. It is interesting that the UC was shown to be exacerbated on both laboratory data and colonoscopy findings when neuropathy occurred in our patient. In other words, the patient was in a mild active stage of UC when the symptoms of neuropathy appeared, although the dose of oral prednisolone was reduced to 7.5 mg/day. Moreover, Gondim et al. proposed that primary immune-mediated neuropathy is likely a type of EIM of UC (14). We concluded
that isolated mononeuritis in the unilateral hypoglossal nerve was a type of EIM related to UC in the present case.

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

References