

Ileocecal junction perforation by colonic T-cell lymphoma in a patient with primary Sjögren's syndrome

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Abstract

Primary Sjögren's syndrome (pSS) is associated with an increased risk of lymphoma, especially non-Hodgkin's lymphoma. The rarest pathological subtype is T-cell lymphoma. We herein report a case of a 52-year-old man with a 17-year history of pSS who was admitted to our hospital with chronic epigastric pain and a positive fecal occult blood test. Colonoscopy revealed multiple colonic ulcers, and histological and immunological studies demonstrated the T-cell origin of this lymphoma. However, the patient rejected all treatments. He developed recurrent intestinal obstruction and infection for 3 years until an intestinal perforation occurred. The right half of the colon was resected and colostomy was performed. However, the patient died of an intestinal fistula and intraperitoneal infection 40 days postoperatively. This case highlights the rarity of the correlation between T-cell lymphoma and pSS.

Keywords

Sjögren's syndrome, T-cell lymphoma, colon ulcer, perforation, mortality, case report

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Introduction

Primary Sjögren's syndrome (pSS) is generally considered to be a benign autoimmune disease that always involves lymphocytic infiltration of the exocrine glands. A complication of pSS is lymphoproliferative malignancy, especially non-Hodgkin's lymphoma (NHL).¹ T-cell lymphoma is

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relatively uncommon. No more than 30 cases of concurrent T-cell lymphoma and pSS have been reported to date, and among these, various histological subtypes have been reported.^{2,3} Furthermore, no reports have described T-cell lymphoma affecting the gastrointestinal tract in patients with pSS. The present report describes a case of peripheral T-cell lymphoma that manifested as multiple colonic ulcers in a patient with pSS.

Case report

In August 2010, a 52-year-old man with a 17-year history of pSS was initially admitted to our hospital with chronic epigastric pain, intermittent diarrhea, and a positive fecal occult blood test. The patient had a history of oral NK/T-cell lymphoma and had undergone radiotherapy 30 years ago. The patient remained well for 17 years until developing dry eye, dry mouth, dental caries, and hypokalemic paralysis. The patient was diagnosed with pSS according to the American-European Consensus Criteria and was treated with oral prednisolone (30 mg/day with tapering to 15 mg/day as the maintenance dose). Physical examination revealed moderate tenderness over the patient's right lower abdominal quadrant. Laboratory tests revealed a positive fecal occult blood test, renal function inadequacy with a high creatinine level (189 $\mu\text{mol/L}$), a high erythrocyte sedimentation rate (95 mm/h), hyperglobulinemia at 42.8 g/L (reference range, 20–40 g/L), a high immunoglobulin G (IgG) level of 16.9 g/L (reference range, 7.0–16.0 g/L), a high IgA level of 4.96 g/L (reference range, 0.7–4.0 g/L), and a low C3 level of 0.62 g/L (reference range, 0.9–1.8 g/L). However, the C4 level was normal. Protein electrophoresis revealed an elevated γ globulin level of 35.5%. Serological tests revealed positive antinuclear antibodies (3200 \times), anti-SSA antibodies, anti-SSB antibodies, and anti-Ro52 antibodies but negative anti-DNA

antibodies, anti-ribonucleoprotein antibodies, anti-Sm antibodies, and antineutrophil cytoplasmic antibodies. Colonoscopy revealed multiple discrete colonic ulcers. The histologic examination demonstrated dense infiltration of small lymphocytes with thickened walls. The patient underwent symptomatic treatment and intensive surveillance. In March 2011, the patient's laboratory results revealed aggravated hyperglobulinemia at 50 g/L, an IgG level of 28.8 g/L, and an IgA level of 9.5 g/L. Repeat colonoscopy revealed the progression of multiple colonic ulcers. The final immunologic phenotypic profile demonstrated CD3+, CD7 \pm , CD45+, CD56–, cyclin D1–, CD79a (part +), CD20+, CD68+, BcL-2+, and CD30– cells, suggestive of T-cell lymphoma. Furthermore, lymphadenectasis was found in the left inguinal region, and the histologic examination also suggested T-cell lymphoma. However, no lymphadenectasis was observed in the abdomen or lungs. The patient refused to undergo bone marrow aspiration.

This patient refused all medical and surgical treatments and subsequently experienced recurrent intestinal obstruction and infection. In May 2013, colonoscopy revealed irregular enormous ulcers with prominent peripheral nodular protrusion from the ileocecal junction to the descending colon. The largest ulcer was located in the cecum and had a diameter of >3 cm. Laboratory results revealed decreased hyperglobulinemia at 33 g/L, an IgG level of 18.9 g/L, and an IgA level of 9.07 g/L. One month after the colonoscopy, the patient developed intense abdominal pain, and an abdominal radiograph suggested intestinal perforation. The right half of the colon was resected and colostomy was performed, which revealed a perforation that had occurred in the ileocecal junction. Unfortunately, the patient died of an intestinal fistula, intraperitoneal infection, and tumor bleeding 40 days postoperatively.

Discussion

Lymphomas develop in approximately 5% to 8% of patients diagnosed with pSS.⁴ Different kinds of lymphomas have been investigated by many researchers to date. Mucosa-associated lymphoid tissue lymphomas occur at the highest incidence,⁵ while T-cell lymphomas associated with pSS are uncommon. Theander et al.⁵ reported that merely 1 of 12 patients (0.08%) had T-cell lymphoma, which was consistent with the findings of a study conducted in China.⁶ T-cell lymphomas that affect the gastrointestinal tract have not been reported to date. In addition, the risk of lymphoma has been shown to increase with time after the diagnosis of pSS.^{4,5}

Several predictors of lymphoma development have been identified. Although these indexes can help to predict the occurrence of lymphoma, only a few can be readily accessed. In addition, many different biomarkers should be closely monitored in these patients. A study of 244 patients with pSS revealed that hypergammaglobulinemia, anemia, and parotid enlargement were risk factors for future development of NHL.⁴ The results of the present study indicate that hypergammaglobulinemia and peripheral lymphadenectasis may suggest the onset of lymphoma. However, to what extent hypergammaglobulinemia can predict the occurrence of lymphoma remains unclear.

The mechanisms underlying the development of lymphoma in patients diagnosed with pSS are unknown. Some viruses and bacteria have been reported to be responsible for NHL development,⁷ such as human T-cell leukemia virus type 1. However, the serological status of this virus was not tested in our patient.

The pathological diagnosis of lymphoma and especially determination of its association with pSS remains difficult. Hence, the understanding of primary gastrointestinal

T-cell lymphomas remains limited. The presence of monoclonal immunoglobulin heavy chain gene rearrangements can be used for early detection of malignant lymphoma and prediction of its progression.

There is still no consensus on the difference between the characteristics of lymphomas that complicate autoimmune disorders and the characteristics of lymphomas in general. Accordingly, whether these should be given different treatments remains unclear. The clinical stage has been shown to be significantly important for patients with colonic lymphoma.⁸ Therefore, surgery should be considered an appropriate option for patients with localized tumors or complications from diseases. In the present case, chemotherapy was suggested when the patient was first diagnosed with T-cell lymphoma. However, the patient refused this treatment. When the patient developed intestinal perforation after 2 years, his lymphoma had progressed to an advanced clinical stage. Although the patient underwent a palliative operation, he died of postoperative complications.

Peripheral T-cell lymphoma has an overall inferior prognosis when compared with aggressive B-cell lymphomas. In patients with peripheral T-cell lymphoma not otherwise specified, the 5-year overall survival rate ranges from 25.6% to 35.0%.^{9–12} However, no data are available for T-cell lymphoma associated with pSS.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics and consent statements

This study was approved by the Ethics Committee of Meitan General Hospital. The patient provided consent to publish and report his data.

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