A 64-year-old Japanese woman visited our hospital in July 2013 with a complaint of intermittent abdominal pain for 1 month. She had a history of maculopapular rashes on the neck over a period of 14 years. Colonoscopy using indigo carmine dye spraying revealed a few yellowish-white polypoid lesions (Fig. 1a,b) and yellowish-white nodular or granular mucosal lesions in the ascending colon (Fig. 1c) and the cecum (Fig. 1d). Histological examination of the biopsy specimens taken from the bowel lesions revealed marked mast cell infiltration with prominent eosinophils in the lamina propria (Fig. 2a,b). The mast cells were highlighted by immunostaining for KIT (Fig. 2c). Histology of biopsy specimens taken from the maculopapular rash on the neck also confirmed increased mast cells in the lamina propria, which were again highlighted by KIT (Fig. 2d).

On the basis of these findings, we diagnosed the patient as having indolent systemic mastocytosis involving the colon and skin. No evidence of mast cell infiltration was found in the bone marrow. We treated the patient with oral histamine receptor 1 and 2 antagonists. The medications resulted in prompt resolution of her abdominal pain and the maculopapular rash on her neck.

Systemic mastocytosis is a rare disease characterized by a clonal neoplastic proliferation of mast cells that accumulate in the bone marrow, skin and/or other extracutaneous organs, such as the gastrointestinal tract [1]. Several reports of systemic mastocytosis involving the colon have been described in Western countries [2–5]. In these reports, nodular mucosal lesions [2,4,5], multiple areas of pigmentation [3], polypoid lesions [4], aphthous ulcers, erosions [4], granularity, erythema, and even normal-appearing mucosa [4,5] have been described as the characteristic endoscopic findings of systemic mastocytosis involving the colon. To the best of our knowledge, this is the first report of systemic mastocytosis involving the colon in a Japanese patient.

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Fig. 1 Colonoscopic views in a 64-year-old Japanese woman who had a long history of maculopapular rashes on the neck and a 1-month history of intermittent abdominal pain showing: a a few yellowish-white polypoid lesions in the ascending colon, with; b a shallow depression in the center; c nodular mucosal lesions in the ascending colon; d nodular mucosal lesions in the cecum.

Shuji Kochi1, Shotaro Nakamura2, Yumi Oshiro3, Koichi Kurahara1, Keisuke Kawasaki1, Hiroki Yaita1, Tadahiko Fuchigami1

1 Division of Gastroenterology, Matsuyama Red Cross Hospital, Ehime, Japan
2 Department of R/D for Surgical Support System, Center for Advanced Medical Innovation, Kyushu University, Fukuoka, Japan
3 Department of Pathology, Matsuyama Red Cross Hospital, Ehime, Japan

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Fig. 2  Histological findings of the biopsy specimens. a, b, c Biopsy of one of the colonic lesions showing mast cell infiltration with eosinophils in the lamina propria: a stained with hematoxylin and eosin (H&E), original magnification ×20; b stained with H&E, original magnification ×40; c immunostained for KIT, which highlights the mast cells, original magnification ×40. d Biopsy of the skin lesion showing increased mast cells in the lamina propria that are highlighted by the KIT immunostain (original magnification ×20).

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Corresponding author
Shuji Kochi, MD
Division of Gastroenterology
Matsuyama Red Cross Hospital
1 Bunkyo-cho, Matsuyama-shi
Ehime 790-8524
Japan
Fax: +81-89-9269916
decorin@intmed2.med.kyushu-u.ac.jp