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CASE REPORT | ENDOSCOPY

# Familial Occurrence of Enteric Muco-Submucosal Elongated Polyp

Yuto Shimamura, MD, Kenji Nakamura, MD, Takeshi Okamoto, MD, Noriatsu Imamura, MT, Naoki Ishii, MD, and Yoshiyuki Fujita, MD

*Division of Gastroenterology, St. Luke's International Hospital, Tokyo, Japan*

## ABSTRACT

We report 2 cases of enteric muco-submucosal elongated polyps (EMSEPs) that presented with gastrointestinal bleeding. The 2 patients are siblings. They both had a history of percutaneous coronary intervention for coronary artery disease and were on dual antiplatelet therapy. They underwent endoscopic resection of the polyps, which displayed identical endoscopic and histological features compatible with EMSEP. This is the first report of familial occurrence of EMSEP, suggesting possible genetic involvement. It is also important to note that the use of antiplatelet agents appears to be a predisposing factor for gastrointestinal bleeding from EMSEP.

## INTRODUCTION

Enteric muco-submucosal elongated polyps (EMSEPs) are rare, benign gastrointestinal polyps with an elongated, slender, worm-like projection that consists of normal mucosal and submucosal layers containing dilated vascular and lymphatic structures surrounded by edematous connective tissue. This entity has not been well described in the past, and its etiopathogenesis is unknown.[1-3]

## CASE REPORT

### Case 1

A 67-year-old man presented with melena. He denied abdominal pain and other gastrointestinal symptoms on presentation. His past medical history was significant for coronary artery disease treated with percutaneous coronary intervention, after which he remained on dual antiplatelet therapy consisting of aspirin and clopidogrel. His laboratory results were significant for severe anemia with a hemoglobin level of 6.1 g/dL. The results of the esophagogastroduodenoscopy and colonoscopy were unremarkable. Capsule endoscopy revealed a bleeding polypoid lesion in the proximal jejunum. Subsequently, antegrade double balloon enteroscopy revealed a solitary, 32 x 30 x 10-mm pedunculated, worm-like polyp in the proximal jejunum consistent with the lesion seen on capsule endoscopy (Figure 1). The polyp was removed by endoscopic mucosal resection after applying a clip to the base of the stem. The histopathology revealed normal small intestinal mucosal lining overlying the submucosa, which consisted of edematous loose connective tissue with a prominent vascular and lymphatic component (Figure 2).

### Case 2

A 64-year-old man, the younger brother of the above patient, presented with melena. The patient was also on dual antiplatelet therapy consisting of aspirin and clopidogrel following percutaneous coronary intervention for coronary artery disease. His other medical history includes hypertension, dyslipidemia, and chronic kidney disease. The patient was discharged after being evaluated by esophagogastroduodenoscopy, colonoscopy, and capsule endoscopy, none of which suggested a source of bleeding. The patient presented again 6 weeks later with melena and severe anemia requiring blood transfusion (hemoglobin level: 6.6 g/dL). At this time, a repeat capsule endoscopy showed a bleeding site in the proximal jejunum. The culprit lesion, a 20 x 10 x 20-mm

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**Correspondence:** Yuto Shimamura, Division of Gastroenterology, St. Luke's International Hospital, 9-1 Akashi-cho, Chuo-ku, Tokyo, 104-8560, Japan (yutshi@luke.ac.jp).

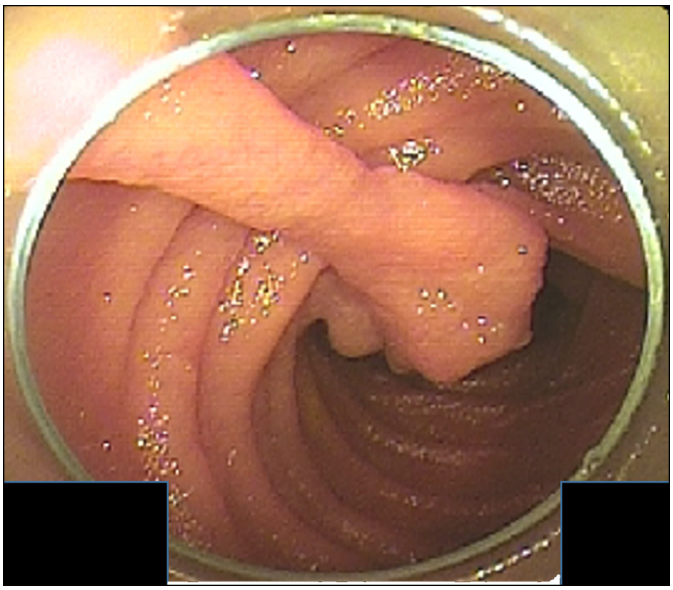

**Figure 1.** Double-balloon enteroscopy revealed a long, slender, worm-like polyp in the proximal jejunum.

pedunculated, slender, worm-like polyp, was reachable by push enteroscopy. It was removed by polypectomy after applying a clip to the base of the stem (Figure 3). The histopathological diagnosis was similar to that of case 1, which was compatible with EMSEP.

## DISCUSSION

The first case of enteric muco-submucosal elongated polyp (EMSEP) was reported by Matake et al in 1998, with subsequent reports mostly coming from Japan.[1-5] Although most muco-submucosal elongated polyps in the literature have been identified in the large intestine, there are also a few reports of muco-submucosal elongated polyps in the small intestine. EMSEP are typically found in the duodenum, the proximal jejunum, and the terminal ileum.[6] These polyps are characterized by its endoscopic features, which are elongated, slender, worm-like projection. Histologically, these are

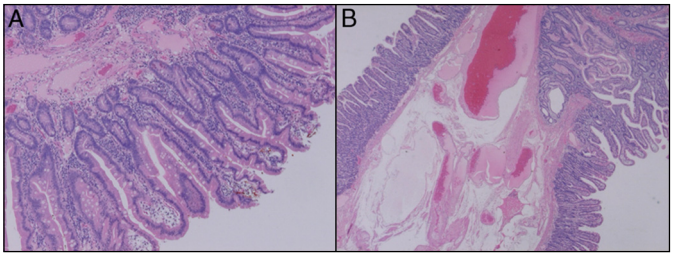


**Figure 2.** (A) Histopathological view showing the tip of the polyp covered by normal mucosa and dense submucosal layer with dilated vascular and lymphatic elements. (B) Histopathological findings of the base of the polyp with normal small intestinal mucosal lining overlying the submucosa, which consists of edematous loose connective tissue with prominent vascular and lymphatic components.

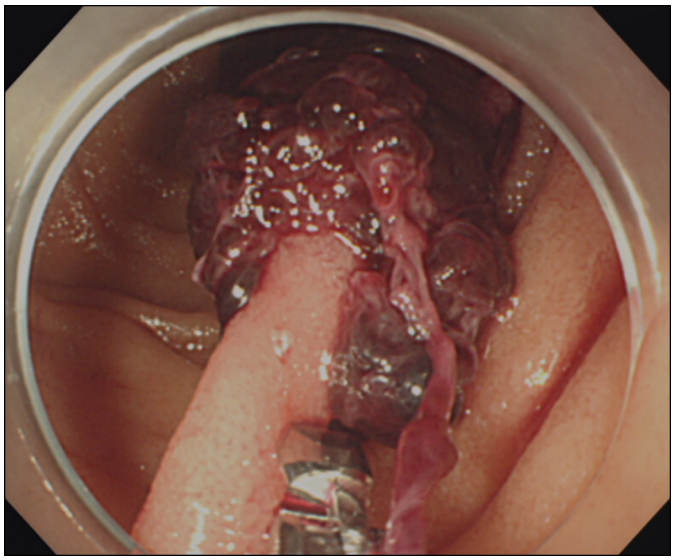

**Figure 3.** Push enteroscopy revealed a long, slender, worm-like polyp in the proximal jejunum.

composed of normal mucosal and submucosal layers containing the dilated lymphovascular components with edematous connective tissue.[7]

This entity has not been well described in the past, and its etiopathogenesis is unknown. There is a speculation that continuous peristalsis with mechanical irritation causes the submucosal layer to become elevated, leading to elongation. There are also reports suggesting an association between diverticulosis and EMSEP.[8,9] Differential diagnoses warranting consideration include inflammatory polyps, inverted colonic diverticula, filiform polyps associated with inflammatory bowel disease, residual stalks of a previously excised pedunculated adenoma, and autoamputation of a pedunculated polyp of any kind.[10] Typical endoscopic appearance, presence of submucosa with lymphovascular components with absence of mucosal inflammation, or fibromuscular proliferation in histology can differentiate from other entities.[7] Endoscopic resection is an option for symptomatic cases; however, there is no established treatment strategy for asymptomatic cases. Given reports suggesting an association between EMSEP and neoplasms, endoscopic resection may need to be considered.[11-13]

We report the cases of 2 siblings who presented with GI bleeding and were diagnosed with EMSEP in the proximal jejunum. They had similar past history of ischemic heart disease and were on dual antiplatelet therapy. They were treated successfully by endoscopic resection without complications and no rebleeding was seen in either case for more than 6 months. There are no reports of familial occurrence of EMSEP to date. These cases suggest a possible underlining familial disposition or genetic involvement. In addition, dual antiplatelet may increase the likelihood of bleeding from

these polyps. We speculate that more EMSEP will be diagnosed as advances are seen in diagnostic techniques for the small intestine. We also believe the increased use of antiplatelet agents may lead to an increase in the incidence of GI bleeding from EMSEP.

## DISCLOSURES

Author contributions: Y. Shimamura, T. Okamoto, and N. Imamura wrote the manuscript. N. Ishii and K. Nakamura critically reviewed the article. Y. Fujita gave approval of the version to be published. All authors have reviewed and approved this manuscript. K. Nakamura is the article guarantor.

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