

A forethought about obscure gastrointestinal bleeding: an unusual ileal mass



► **Fig. 1** Endoscopic view of the lesion.

The patient was a 32-year-old woman admitted for hematochezia, with a hemoglobin of 67 g/L and fecal occult blood test of 4+. Emergency gastroscopy and colonoscopy were unremarkable. A double balloon enteroscopy revealed a lobulated ileal mass located 70 cm proximal from the ileocecal valve (► **Fig. 1**, ► **Video 1**). Positron emission tomography/computed tomography (PET/CT) showed that the lesion was located in the pelvic segment of the small intestine, with a standardized uptake value index of 3.6 (► **Fig. 2**). Surgery was recommended owing to the patient's low hemoglobin levels and risk of recurrent gastrointestinal bleeding. A gross specimen revealed a lobulated, polypoid mass in the ileum, measuring up to 1.5 cm × 0.8 cm (► **Fig. 3**).

A final diagnosis of neuromuscular and vascular hamartoma of the small intestine was made. Postoperative pathology revealed a mixed component of blood vessels, nerve fibers, and smooth muscle consistent with that of a hamartoma (► **Fig. 4**). The haphazard arrangement of the vascular structures and muscular tissues were confirmed by CD31 and desmin staining, respectively. Aberrant nerve bundles and ganglion cells were highlighted by S100 staining (► **Fig. 5**).

Neuromuscular and vascular hamartoma is a rare gastrointestinal lesion first described in 1982 by Fernando and McGo-



► **Video 1** Retrograde double-balloon enteroscopy was performed using the water-exchange method.



► **Fig. 2** Positron emission tomography/computed tomography revealed a lesion with increased uptake in the pelvic segment of the small intestine.



► **Fig. 3** Macroscopic view of the lesion after surgical resection.

vern [1]. Clinical symptoms can be non-specific and can range from chronic abdominal pain and intermittent intestinal obstruction to gastrointestinal bleeding. This condition can occur as single or multiple strictures or a polypoid mass. It is mainly composed of disorganized fascicles of blood vessels, smooth muscle, and bundles of non-myelinated nerve fibers with scattered abnormal ganglion cells, and occurs focally within a segment of the small intestine [2]. Given that similar histological features seen in cryptogenic multifocal ulcerous stenosing enteritis or diaphragm disease of the small bowel, the hamartomatous nature of neuromuscular and vascular hamartoma has been argued [3]. However, our case demonstrated a rare single, lobulated polypoid lesion of the small intestine

with histological features consistent with neuromuscular and vascular hamartoma and cannot be attributed to other reactive diseases.

Endoscopy_UCTN_Code_TTT_1AP_2AD

Funding

Huashan Hospital
<http://dx.doi.org/10.13039/501100010120>
 2020QD008

Competing interests

The authors declare that they have no conflict of interest.

The authors

Yujen Tseng^{1,2}, Lishuang Lin³, Tiansheng Huang⁴, Zhongguang Luo¹

- 1 Department of Digestive Diseases, Huashan Hospital, Fudan University, Shanghai, China
- 2 Huashan Rare Disease Center, Huashan Hospital, Fudan University, Shanghai, China
- 3 Department of Pathology, Huashan Hospital, Fudan University, Shanghai, China
- 4 Department of Digestive Diseases, Shanghai Guanghua Hospital of Integrated Traditional Chinese and Western Medicine, Shanghai University of Traditional Chinese Medicine, Shanghai, China

Corresponding author

Zhongguang Luo, MD

Department of Digestive Diseases, Huashan Hospital, Fudan University, 12 Wulumuqi Middle Rd., Jing'an District, Shanghai 200040, China
 luozg8@126.com

References

- [1] Fernando S, Mcgovern V. Neuromuscular and vascular hamartoma of small bowel. *Gut* 1982; 23: 1008–1012
- [2] Hemmings C. Neuromuscular and vascular hamartoma arising in a Meckel's diverticulum. *Pathology* 2006; 38: 173–174
- [3] Setaffy L, Osuna M, Plieschnegger W et al. Cryptogenic multifocal ulcerous stenosing enteritis (CMUSE), and neuromuscular and vascular hamartoma (NMVH): two sides of the same coin? *Endoscopy* 2015; 47: 345–348

Bibliography

Endoscopy 2022; 54: E726–E727

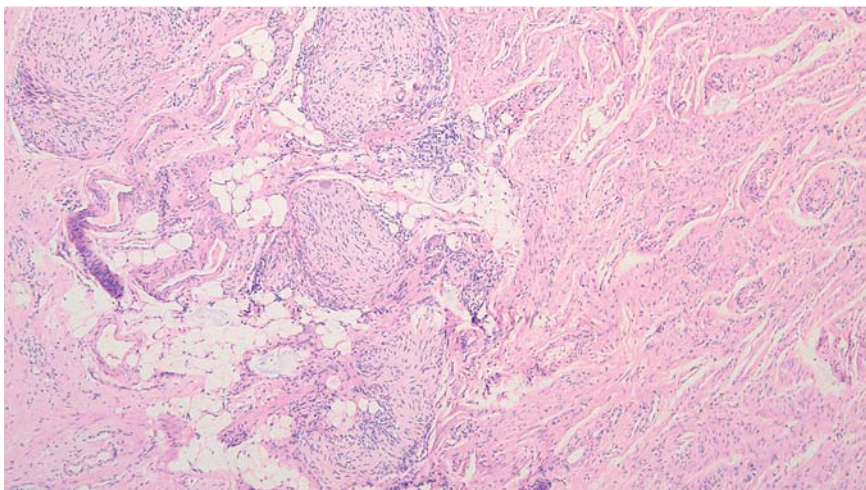
DOI 10.1055/a-1759-2256

ISSN 0013-726X

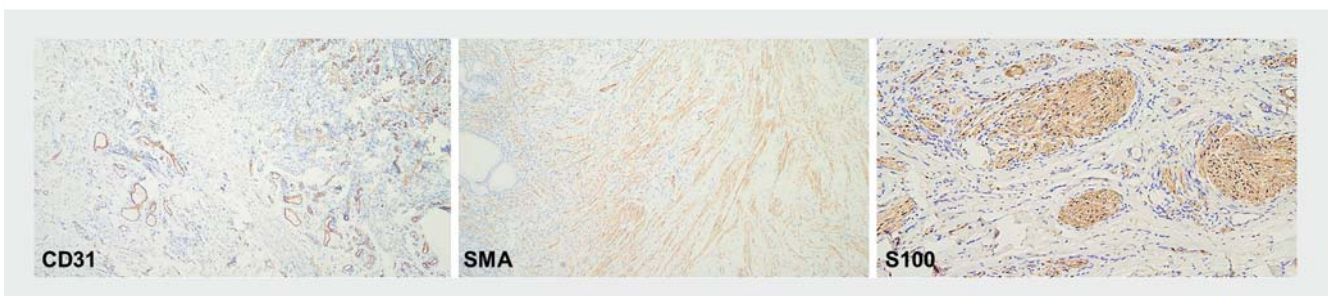
published online 10.3.2022

© 2022. Thieme. All rights reserved.

Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany



► **Fig. 4** HE staining showed an aberrant arrangement of vascular structures, muscular tissues and nerve bundles.



► **Fig. 5** Immunohistochemistry confirmed the presence of vascular, muscular and nerve tissues