VIDEO CASE REPORT

Blue rubber bleb nevus syndrome in a 10-year-old child treated with loop ligation facilitated by double-balloon enteroscopy

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Blue rubber bleb nevus syndrome is an extremely rare systemic vascular disorder (with only 200 cases published to date), characterized by multiple cutaneous and GI venous malformations.[1] Patients present with fatigue, iron-deficiency anemia, and occult or overt obscure GI bleeding. Patients are usually managed conservatively with iron supplementation and/or blood transfusions. However, endoscopic (argon plasma coagulation,

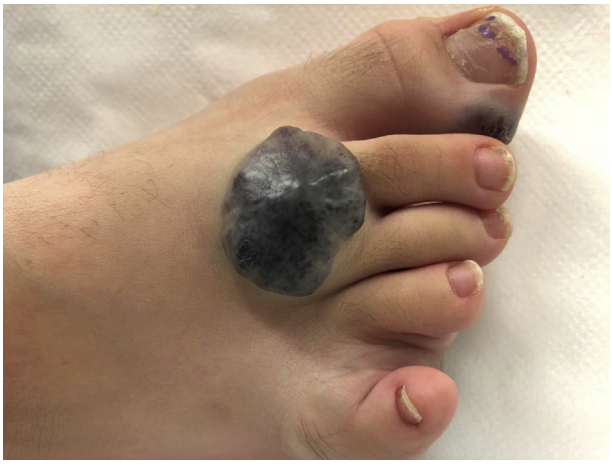

Figure 1. Cutaneous lesions.

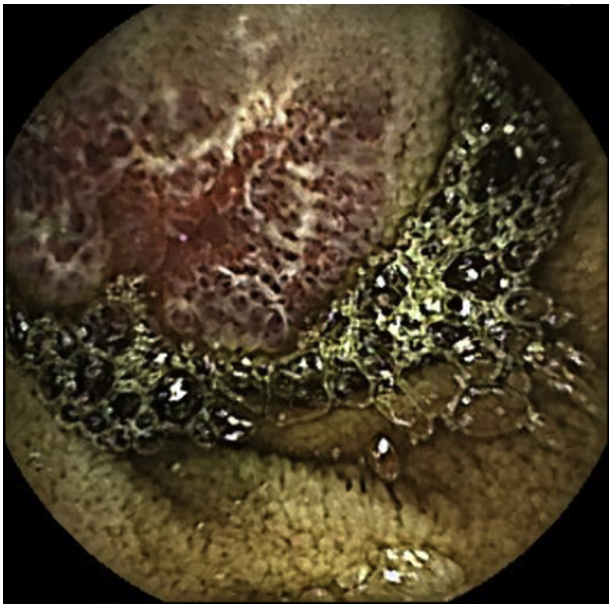

Figure 2. Small-bowel lesion seen in capsule.

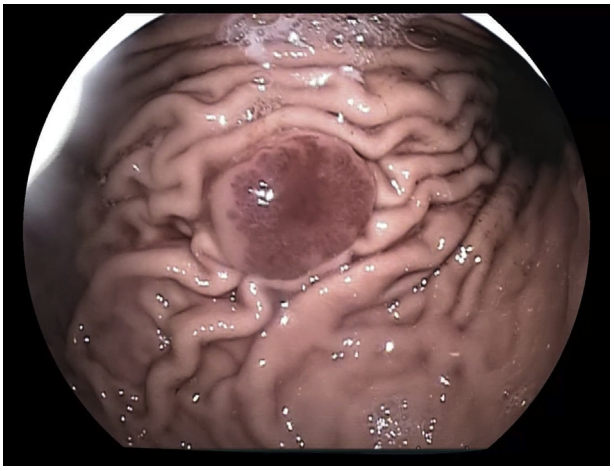

Figure 3. Gastric lesion.

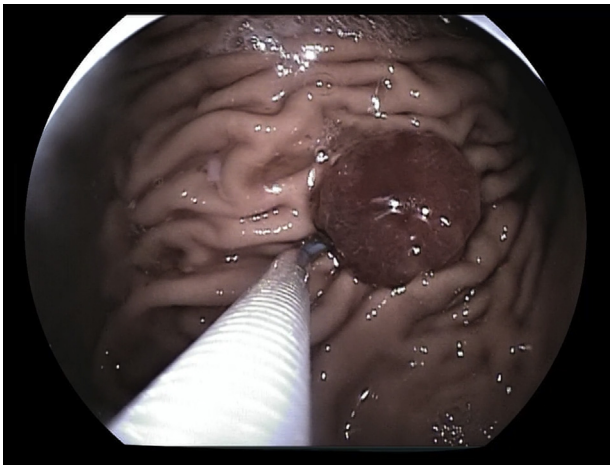

Figure 4. Loop ligating devise applied around gastric lesion.

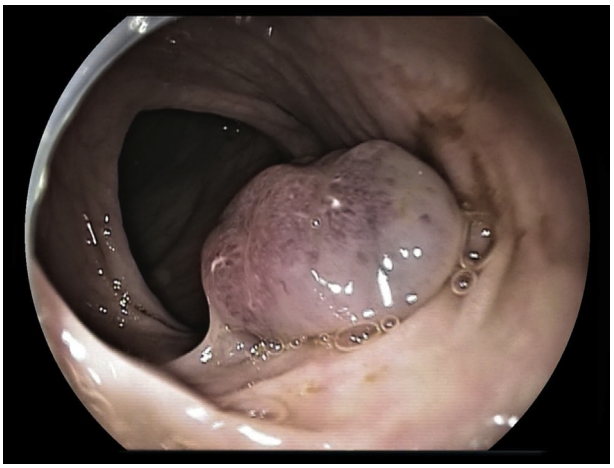

Figure 5. Colonic lesion.

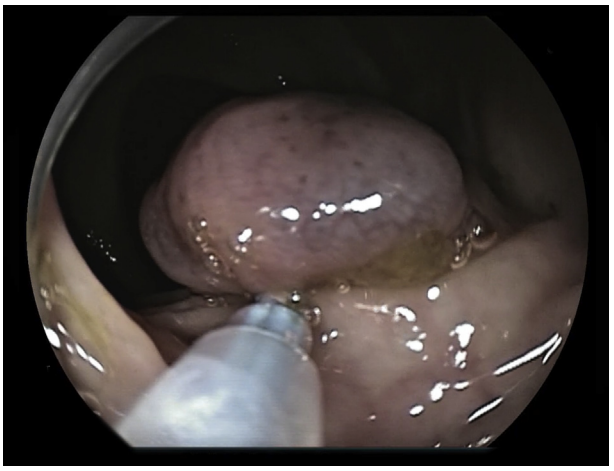

Figure 6. Loop ligation of colonic lesion.

sclerotherapy, polypectomy, and ligation), radiologic, and surgical approaches are preferred for severe cases.[2,3] Systemic therapy with sirolimus appears promising.[4]

A 10-year-old girl with iron-deficiency anemia and multiple cutaneous lesions was diagnosed with blue rubber bleb nevus syndrome at a local hospital (Fig. 1). The patient was investigated with a small-bowel capsule endoscopy that revealed vascular lesions throughout the small bowel (Fig. 2). Consequently, the patient was referred to our institution for further management and consideration of small-bowel endotherapy owing to blood transfusion dependence and rectal bleeding.

An anterograde double-balloon enteroscopy was performed with the patient under general anesthesia. Two 20-mm vascular lesions were identified in the gastric body (Fig. 3). A loop ligating device (PolyLoop; Olympus, Tokyo, Japan) was applied around the base of each lesion, tightened, and completely detached (Fig. 4; Video 1, available online at www.VideoGIE.org). After loop ligation was deployed, deflation of the lumen facilitated optimal device use and successful treatment. No further vascular malformations were found in the duodenum, jejunum, and proximal ileum. Although transfusion requirements decreased over the following 6 months, a follow-up retrograde double-balloon enteroscopy was performed because of persistent anemia. Six lesions were identified: 2 in the transverse colon, 1 in the cecum, and 3 in the distal ileum. Loop ligation was used for the 2 transverse colonic (Figs. 5 and 6) and ileal lesions (Fig. 7). The 2 remaining lesions were too flat, and loop ligation was not technically feasible. Protruding, raised lesions are usually most suited for treatment by this technique, whereas loop ligation of flat lesions has a high failure rate because application of the loop device is

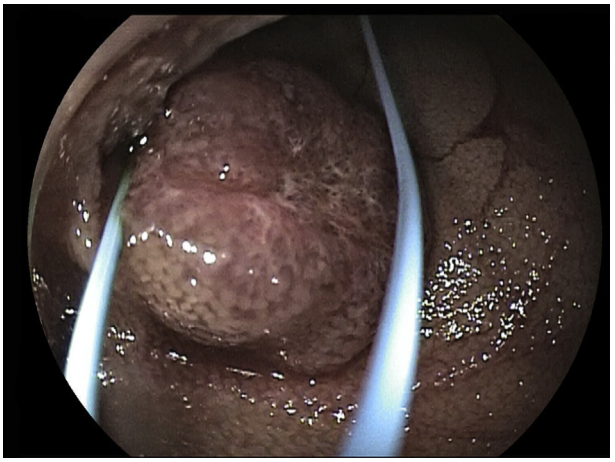

Figure 7. Small-bowel lesion.

challenging. No immediate, early, or late adverse events occurred. At 1-year follow-up, guided by the patient's clinical response (ie, anemia and transfusion requirements), no further GI investigation or intervention has been deemed necessary.

Double-balloon enteroscopy–facilitated loop ligation appears to be a safe and minimally invasive option in patients with blue rubber bleb nevus syndrome, reducing blood transfusion dependence.

DISCLOSURE

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