

Blue rubber bleb nevus syndrome

Blue rubber bleb nevus syndrome (BRBNS) is a very rare condition. Herein we report the case of a 14-year-old girl who presented with chronic anemia that had required multiple blood transfusions and iron supplementation for many years. She had also received steroids. Physical examination revealed multiple small lesions on the dorsum of her feet (● Fig. 1). Computed tomography (CT) of the abdomen showed multiple intraluminal lesions in the small bowel and colon (● Fig. 2).

A gastroscopy was performed (● Fig. 3), followed by a colonoscopy and uneventful polypectomy of the colonic lesions (● Fig. 4; ● Video 1).

Histology revealed a cavernous hemangioma consistent with BRBNS. The patient underwent an exploratory laparotomy 1 week later during which she was found to have multiple lesions affecting the entire small bowel (● Fig. 5), some of which had also caused intussusceptions of the small bowel.

An enterotomy was performed; she underwent further polypectomy of the internal lesions and resection of small-bowel wall to remove lesions that were involving the muscle layer. She was discharged from the hospital on the seventh postoperative day and 1 year later is doing well, having required no further blood transfusions.

BRBNS was first reported by Gascoyen in 1860 [1] but the individual who made this disease famous was William Bennett Bean; it is also known as “Bean syndrome” [2]. The condition consists of multiple venous malformations that affect the skin and multiple visceral organs. The most common site for gastrointestinal tract involvement is the small bowel [3]. Skin lesions typically present at birth, whereas the gastrointestinal manifestations develop mostly in late adulthood. The commonest presenting symptom is spontaneous gastrointestinal bleeding, but the degree of bleeding may range from occult



Fig. 1 Multiple small black cystic lesions on the feet of a 14-year-old girl who presented with chronic anemia due to blue rubber bleb nevus syndrome.

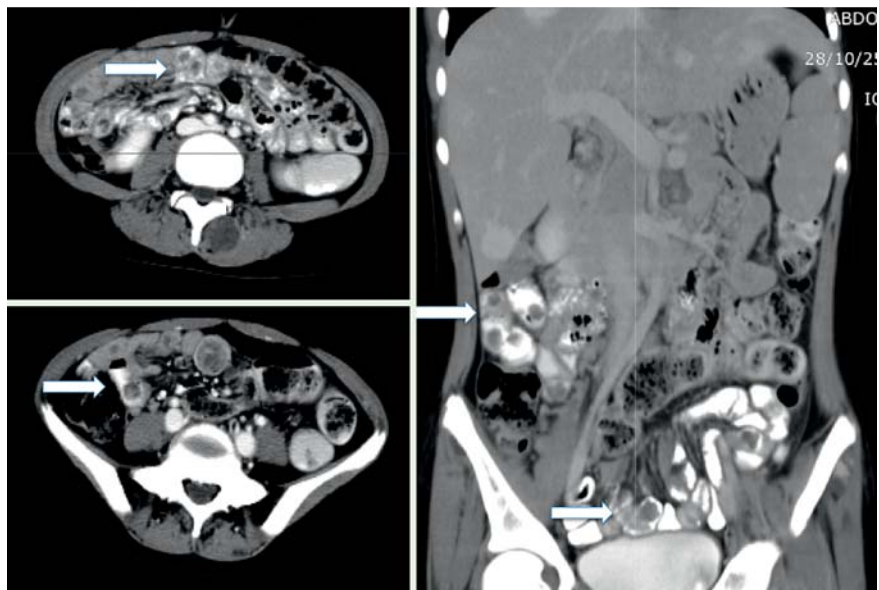


Fig. 2 Computed tomography (CT) scan of the abdomen showing multiple intraluminal polypoid lesions in the small bowel (white arrows).

to profuse bleeding [4]. Various combinations of endoscopic and surgical treatment, such as endoscopic polypectomy or injection [5], or enterotomy with excision of the small-bowel lesions [4] have been shown to be effective forms of management in refractory cases.

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Competing interests: None

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Video 1

Colonoscopic polypectomy of the lesions in the colon was performed using a standard snare with electrocautery.



Fig. 3 Appearance at esophagogastroduodenoscopy showing multiple submucosal lesions (white arrows) in: **a** the fundus of the stomach; **b** the body of the stomach; **c** the duodenum.



Fig. 5 Appearance of the small bowel at laparotomy showing multiple blebs within the wall of the small bowel.

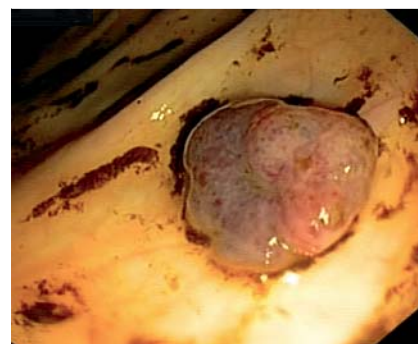


Fig. 4 Colonoscopic view showing one of the multiple small cystic polyps in the colon.

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Bibliography

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