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A Novel Endoscopic Treatment for Blue Rubber Bleb Nevus Syndrome

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Abstract

Introduction: Blue Rubber Bleb Nevus Syndrome (BRBNS), also known as Bean Syndrome, is a congenital disorder characterized by numerous, cutaneous and internal venous malformations, most commonly involving the skin and gastrointestinal tract (GI). (1) Patients with GI manifestations present with iron deficiency anemia due to recurrent GI hemorrhage from the oozing venous malformations. Due to the rarity of the condition, there are no defined guidelines on the management of BRBNS, further highlighting the need for case reports to dictate therapy.

Case Presentation: We present a case of an 83-year-old male who presented with a GI bleed due to an unknown etiology. Workup was initiated with an esophagogastroduodenoscopy (EGD) and a colonoscopy. The initial colonoscopy had poor prep and difficult visualization due to excessive bleeding. This led to extensive testing as the patient continued to have ongoing blood loss, which included a tagged red blood cell scan, angiography with attempted embolization, and magnetic resonance enterography. Finally, the patient was able to repeat a colonoscopy with appropriate prep, and findings consistent with BRBNS were identified. The patient was treated with six hemoclips of the actively bleeding spots, with resolution of his symptoms. The patient was followed for approximately two years, and has not had recurrence of GI bleed, with a stable hemoglobin.

Discussion: Blue Rubber Bleb Nevus Syndrome (BRBNS), is a congenital disorder characterized by numerous, cutaneous and internal venous malformations. The treatment of GI BRBNS is determined by the severity of the disease, taking frequency and volume of hemorrhage, as well as extent of intestinal involvement, into consideration. Treatment options include medical management with agents such as sirolimus, endoscopic therapy with band ligation, and sclerotherapy, and surgical resection of the affected bowel. A Pubmed/MedLINE literature review elucidated one other case of endoscopic hemoclip as treatment for BRBNS, with the resolution of patient symptoms for 5 years. Endoscopic hemocliping is a successful, novel, and clearly underutilized treatment option for BRBNS as currently there is no standard of treatment for Blue Rubber Bleb Nevus Syndrome.

Introduction

- Blue Rubber Bleb Nevus Syndrome (BRBNS), also known as Bean Syndrome, is a congenital disorder characterized by numerous, cutaneous and internal **venous malformations** (VMs)
- VMs most commonly involve the skin and gastrointestinal (GI) tract
- Patients with GI manifestations present with iron deficiency anemia due to recurrent GI hemorrhage from the oozing venous malformations
- Due to the rarity of the condition, there are no defined guidelines on the management of BRBNS, further highlighting the need for case reports to dictate therapy

Case Presentation

- 83-year-old male presented to the emergency department (ED) with **altered mental status** and urinary incontinence for 5 days, as well as **melenas for 3 weeks**
- Patient history was significant for Abdominal Aortic Aneurysm status post endovascular repair, on triple anticoagulation with Eliquis, Plavix, and Aspirin
- Patient was hemodynamically stable in the ED, with laboratory studies significant for a **hemoglobin of 3.9 g/dl** and a positive stool guaiac test. An extensive work up was conducted.
- EGD -> Negative for acute bleeding. Colonoscopy -> Inconclusive
- Tagged Red Blood Cell Scan -> Bleeding in the **right upper quadrant of the abdomen**, suggestive of an active small bowel bleed. This gave a general location of the bleed, but not a definitive source.
- Interventional Radiology attempted to embolize the bleed -> Superior mesenteric and celiac angiogram -> Negative for bleeding and a source of bleeding
- Magnetic Resonance Enterography -> Unable to provide definitive source of bleeding, however **abnormal mucosal or wall thickening at cecal and ascending colon junction** could not be excluded
- Patient continued to have bloody bowel movements with drops in hemoglobin, requiring multiple transfusions (Figure 5)
- Repeat colonoscopy -> Multiple small, vascular, blue raised lesions visualized from hepatic flexure, extending into the cecum, including involvement of the ileocecal valve.** (Figures 1-2)
- Six hemoclips were deployed on actively oozing lesions, achieving hemostasis** (Figures 3-4)
- The patient did not have any further episodes of hematochezia after the intervention, with a stable hemoglobin.
- The patient continues to be monitored with repeat hemoglobin, and has remained stable around 11 g/dl**

Endoscopic Photos

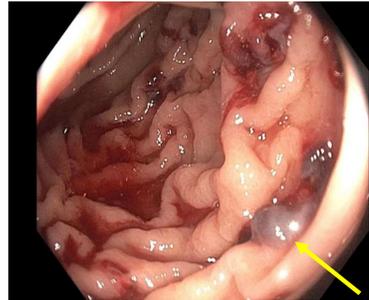


Figure 1. Ascending Colon with Blue Rubber Blebs

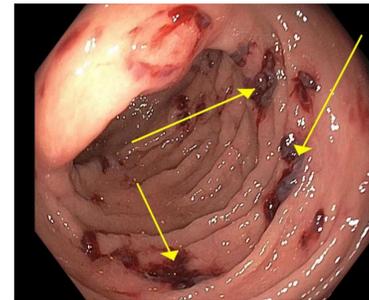


Figure 2. Cecum with Blue Rubber Blebs

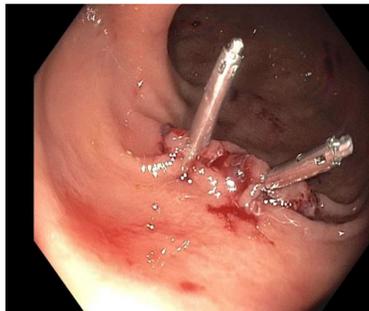


Figure 3. Cecum with Hemoclipped Blue Rubber Blebs

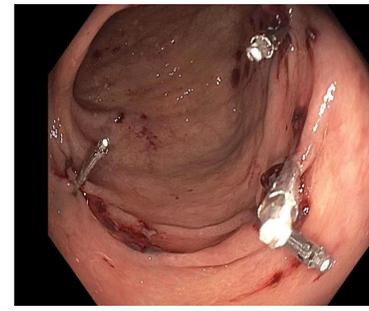


Figure 4. Cecum with Hemoclipped Blue Rubber Blebs

Laboratory Values

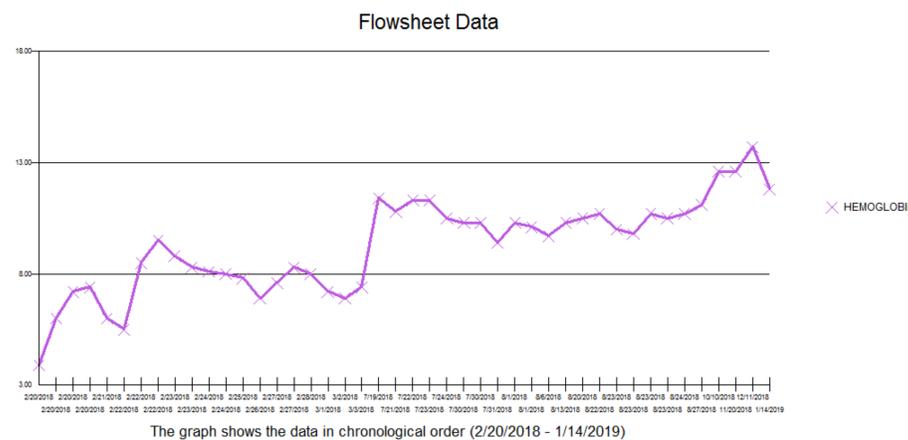


Figure 5. Patient Hemoglobin Trend. Hemoglobin of 3.9 at admission. Hemoglobin of 8 upon discharge, with a subsequent stable baseline of approximately 11 throughout the past two years.

Discussion

- BRBNS is a **rare** disorder, with approximately 200 cases reported in the literature (2-4)
- Cutaneous manifestations present at birth or early childhood
- Visceral involvement tends to present in early adulthood (5)
- Lesions most commonly arise in the **skin and GI tract**; however, they have been found in the heart, spleen, liver, CNS, eye, thyroid, parotid gland, muscles, lungs, and bladder (6-8)
- The most common site of GI tract involvement is the small bowel**; however, lesions may develop anywhere from the mouth to the anus
- While there have been many case reports discussing treatment options for BRBNS with GI involvement, there are **no guidelines regarding the management of this condition**
- The treatment of GI BRBNS is determined by the severity of the disease, taking frequency and volume of hemorrhage, as well as extent of intestinal involvement, into consideration
- Mild or intermittent GI Bleed
 - Iron supplementation and blood transfusions
- Severe cases
 - Many medical, endoscopic and surgical approaches to treatment have been taken for severe cases.
 - Medical management
 - Sirolimus (9-11)
 - Angiogenesis inhibitor
 - Reduces the need for blood transfusions in these patients
 - Adverse drug reactions have not been observed
 - Endoscopic treatment
 - Polypectomy
 - Band ligation
 - Sclerotherapy
 - Surgical
 - Definitive treatment can be achieved with surgical resection of the affected bowel
 - The patient described in this case achieved hemostasis for approximately two years following placement of six hemoclips**
 - A PubMed/MedLINE search elicited **one case** of treatment of BRBNS with hemoclips
 - Kanai et. al describes a case of a 19-year-old patient with BRBNS who was treated with hemoclips and remained stable for 5 years (12)
 - Endoscopic hemocliping is a successful, novel, and clearly underutilized treatment option for BRBNS as currently there is no standard of treatment for Blue Rubber Bleb Nevus Syndrome**

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