Endoscopic Approaches to Blue Rubber Bleb Nevus Syndrome (BRBNS)

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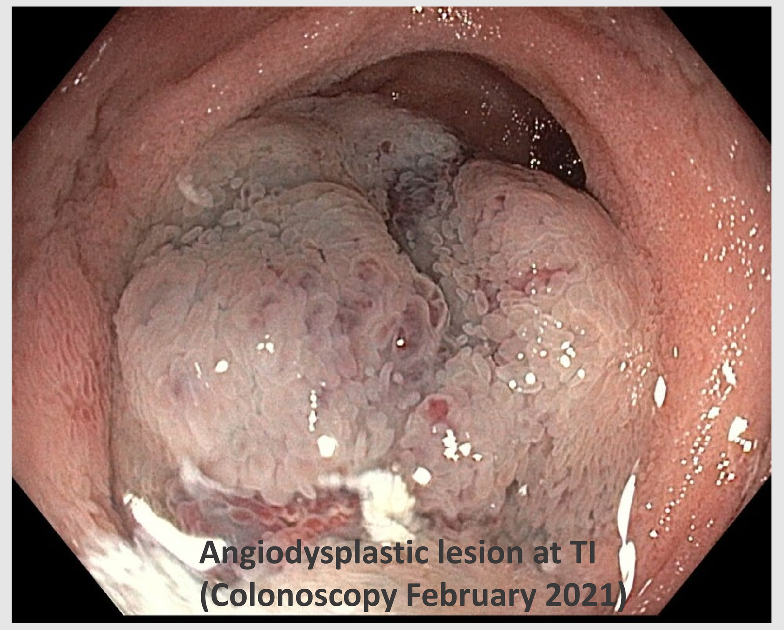
EUS findings 12/17/2020

Multiple endoscopic interventions are available for management of BRBNS

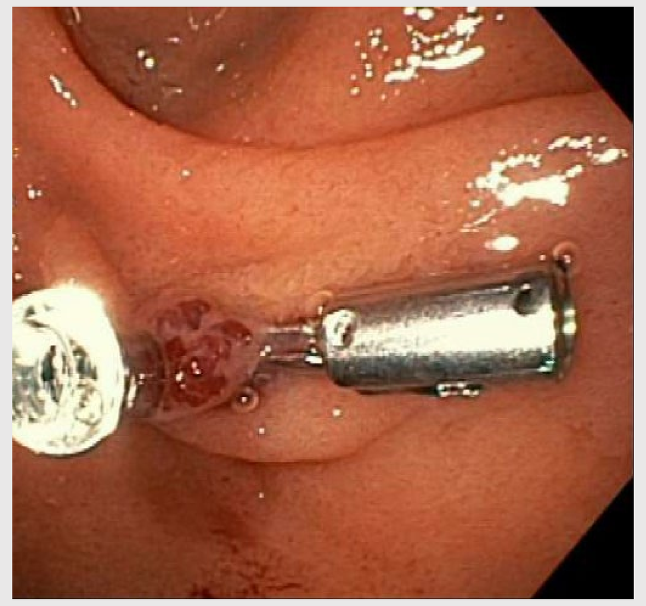
Introduction:

- Blue rubber bleb nevus syndrome (BRBNS) is a rare but potentially life-threatening disorder characterized by cutaneous and gastrointestinal tract vascular malformations and chronic iron deficiency anemia.
- Patients present on a spectrum from mild anemia to severe hemorrhage or intestinal obstruction/intussusception.
- No standard of care exists for therapy in these patients, and the optimal endoscopic technique for management of Arterio-venous malformation (AVM) related bleeding has not been elucidated.
- **Here we present a case illustrating the use of different endoscopic therapies for BRBNS.**



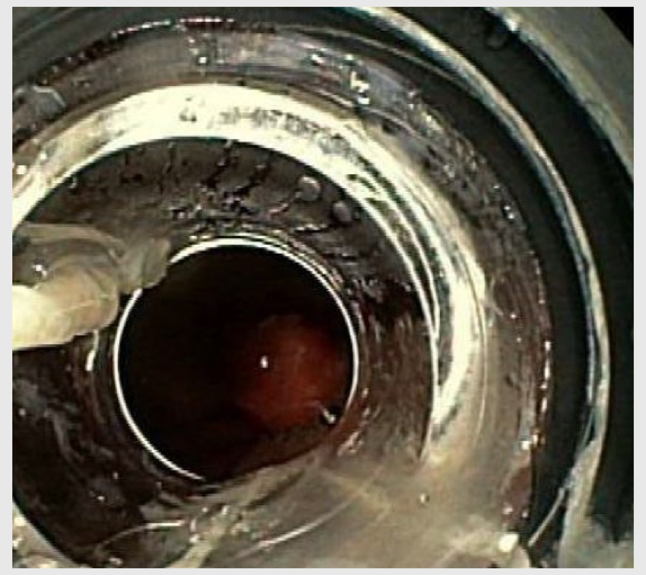
Angiodysplastic lesion at TI (Colonoscopy February 2021)

Series of EGD's December 2020:



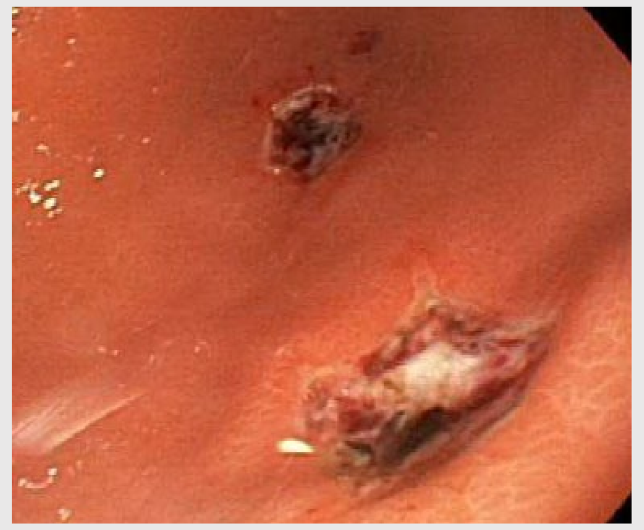
December 16th:

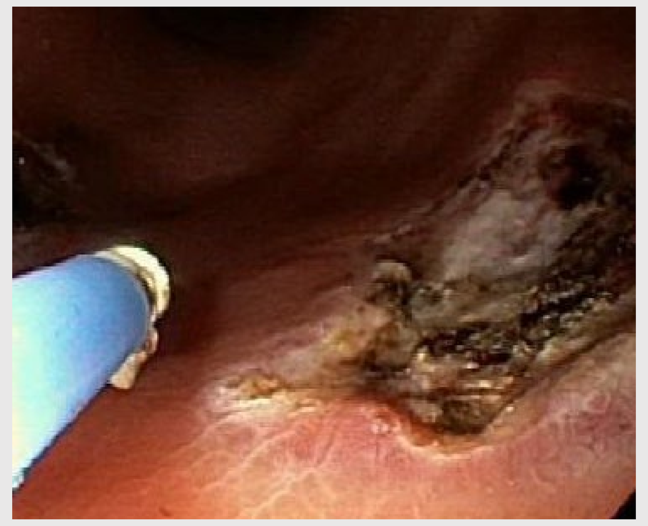
- Three non-bleeding angiodysplastic lesions in the duodenum
- Two hemostatic clips placed in angiodysplastic lesion in D2.



December 17th:

- EUS: intramural (subepithelial) lesion was found in the body of the stomach consistent with BRBNS.
- Largest two of the lesions were successfully banded





December 18th:

- Repeat EGD done for a 3-point drop in Hgb, hematemesis and melena post procedure
- Non-bleeding post banding gastric ulcers with pigmented material. Treated with argon plasma coagulation (APC)

Case:

- 23-year-old lady with known Blue Rubber Bleb Nevus Syndrome
- Cutaneous vascular malformations diagnosed at age six when patient had PICA and severe anemia s/p multiple cutaneous resections, right hand first digit amputation as well bowel resections x2 (secondary to intussusception March 2012 and Ileal perforation May 2012).
- On maintainance sirolimus therapy but unable to take it regularly due to social and financial limitations.
- Presented for evaluation of GI involvement.

Outpatient EGD 12/16/2020
- Gastric and duodenal hemangiomas seen
- **Hemostatic clips placed**
- Subsequently had postprocedural bleeding

EGD 12/17/2020
- No clear evidence of bleeding from clipping
- **Banding x2 done (hemangiomas)**
- Melena and hematochezia post-procedure
- HGB drop from 8.8 to 5.7

EGD 12/18/2020
- Post banding ulcers noted with pigmented material in gastric body
- **APC done**

Outpatient EGD 02/2021
- No interim signs of bleeding
- Persistent duodenal bulb lesions noted
- **s/p hemostatic injection (sodium tetradecyl sulphate**

Discussion:

- From anti-angiogenic systemic therapies to endoscopic treatments to surgical resection, case reports from 1990 to date have reported benefit with sirolimus therapy + **pre-emptive endoscopic treatment** with bipolar cautery, APC, band ligation and sclerosing/hemostatic injection(1)(2)(3)(4).
- Surgery is usually reserved as a last resort; following surgery lesions can appear elsewhere along the GI tract.
- In our patient, due to affordability of antiangiogenic treatment, finding an endoscopic approach that worked for her became even more prudent.
- Although band ligation was attempted, the resultant **bleeding from post banding ulcers seemed to be even worse than her baseline slow rate of bleeding**.
- Hemostatic injection appeared to be an effective measure for her, with up-trending hemoglobin following this and no further signs of overt GI bleeding in over a year.
- **GI care is an important adjunct to the overall management of a patient with BRBNS along with consultation with hematologist.**

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References:
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