INTRODUCTION:
Blue rubber bleb nevus syndrome (BRBNS) is a rare vascular anomaly syndrome consisting of multifocal venous malformations. The malformations are most prominent in the skin, soft tissues, and gastrointestinal (GI) tract, but may occur in any tissue. The cutaneous lesions of BRBNS are generally small, measuring less than 1–2 cm, and blue to purple in colour. Bean was the first to draw attention to the unique quality of these compressible cutaneous lesions that he labelled as “blue rubber-bleb nevi.” The GI lesions of BRBNS are more clinically relevant than the skin and soft tissue lesions. Patients usually exhibit GI bleeding at an early age that continues throughout their life. Massive sudden haemorrhage rarely occurs. Rather, patients are chronically anaemic, requiring lifelong iron replacement and repeated blood transfusions. Although there are reported cases that appear to have an autosomal dominant transmission, most cases are sporadic.

CASE REPORT:
An 18-Year-old female admitted in civil hospital Ahmedabad in surgical OPD with complain of fatigue with off & on episode of bleeding per rectal since 4-5 year. Patient having history of operation for meningomyelocele haemangioma and haemangioma over right shoulder at age of 1 year. Patient having history of multiple blood transfusion for iron deficiency anaemia. Patient having history of taking octreotide and thalidomide 100mg OD since 2 yr. No family history of similar complain.

On physical examination there is pallor of conjunctiva, lips multiple bluish skin lesions over sole of left lower limb (1-2cm.) which was soft, non-tender, compressible. Bilateral chest was clear, heart sound was normal, no focal neurological deficit. Abdomen was soft, non-tender.

Laboratory findings-Hb-6.0 mg/dl, MCV-25pg, MCHC-35g/dl, WBC-6000, Platelets-1,90,000, Serum ferritin-9, Renal and Liver function test was within normal limits. Faecal occult test was positive. Ultrasoundography was normal.

On upper GI scopy there was single bluish lesion at D1-D2 junction.

On colonoscopy there were 3 bluish lesions (size 1-2cm) found at sigmoid colon, descending colon, transverse colon with no active bleeding, biopsy taken which shows histology of blue rubber bleb nevus syndrome.

On capsule endoscopy shows multiple bluish lesions in proximal jejunum and ileum.

On exploratory laparotomy there were total 14 lesions found from DJ junction to IC junction. 3 lesions removed by wedge resection. 6 lesions removed by resection & anastomosis which were nearby to each other, approx. 20-25 cm small bowel resected which include distal jejunum and proximal ileum. 5 lesions were small so scleroscent injected.

Lesion found on colonoscopy was managed by coagulation and hemoclip. Patient discharged un-eventful and on regular follow-up patient is asymptomatic.

MANAGEMENT:
After correction of anaemia patient taken to elective surgery. On exploratory laparotomy there were total 14 lesions found from DJ junction to IC junction. 3 lesions removed by wedge resection. 6 lesions removed by resection & anastomosis which were nearby to each other, approx. 20-25 cm small bowel resected which include distal jejunum and proximal ileum. 5 lesions were small so scleroscent injected.

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(E) Resected small bowel with lesions.

**DISCUSSION:**

The lesions of BRBNS represent a specialized type of venous malformation. They are typically small, circumscribed, and multifocal. Although they may be present in any body tissue, they predominate in the skin and GI tract. The gastrointestinal lesions may manifest clinically with bleeding and secondary IDA and may also present with severe complication such as rupture, intussusception, gangrene. The bleeding is typically chronic continuous bleeding rather than sudden haemorrhage.

The cutaneous lesions are usually asymptomatic and do not require treatment. Symptomatic lesion, lesion in joints and for cosmetic reason treatment may be required, including surgery, sclerotherapy, coagulation.

The treatment of GI disease depends upon severity and extent of involvement.

If bleeding is minor conservative treatment such as blood transfusion, iron supplementation given because of high recurrence of the disease. Pharmacologic agents have been used in an attempt to control the chronic blood loss. Antiangiogenic agents such as corticosteroids, low dose sirolimus and interferon alpha also have been tried. As for other causes of GI bleeding octreotide has also been attempted.

For more significant haemorrhage or other complication surgical resection, endoscopic sclerosis, photo coagulation, haemoclip, band ligation has been proposed.

Once an operative approach is chosen, complete visualization of the entire gastrointestinal mucosa from the mouth to the anus must be performed.

The prognosis of depends on which organs are involvement and the extend of involvement. Most patients can live a long life with the disease but the quality of life is limited due to GI bleeding, oral drug therapy, blood transfusion.

**REFERENCES:**