Blue Rubber Bleb Naevus Syndrome

V Anumakonda

Citation

V Anumakonda. *Blue Rubber Bleb Naevus Syndrome*. The Internet Journal of Gastroenterology. 2006 Volume 5 Number 1.

Abstract

We report for the first time a case of Blue rubber bleb naevus syndrome associated with ischemic colitis. This young man had incidental triad of BRBS, ischemic colitis and Gilbert's syndrome.

CASE REPORT

A 36-year-old man presented with abdominal cramps, diarrhoea and several episodes of profuse rectal bleeding of 1 week duration. There was no associated fever, sweats or weight loss. He is known Gilbert's syndrome and had appendicectomy. He had no known diabetes, hypertension, ischemic heart disease or hyperlipidaemia. There was no significant family or drug history.

Vital signs were normal. On physical examination, the abdomen was mild left iliac fossa tenderness, no organomegaly and bowel sounds were normal. Digital rectal examination revealed some fresh blood but no haemorrhoids and a normal prostrate was palpable. He had multiple blue naevi on his buttocks, back and legs. Investigations showed c-RP 10 mg/l, bilirubin 82 mol/l, alkaline phosphate, aspartate transaminase, and gamma-glutamyl transferase were normal at 91,23, 26 IU/l respectively. Haemoglobin was 13.5g/dl, WBC 8.34 x 103/mm3, and platelets 162 x 103/mm3 and normal urea and electrolytes. Multiple stool cultures were negative. A flexible sigmoidoscopy demonstrated blue naevi in the recto-sigmoid area (figures A, B), confluent inflammation of the descending and sigmoid colon (figure C), and relative rectal sparing of inflammation, with ulceration. The patient was treated with oral mesalazine empirically, pending histological confirmation.

Figure 1

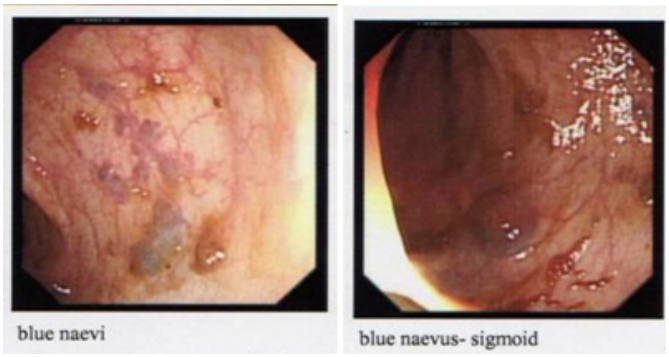
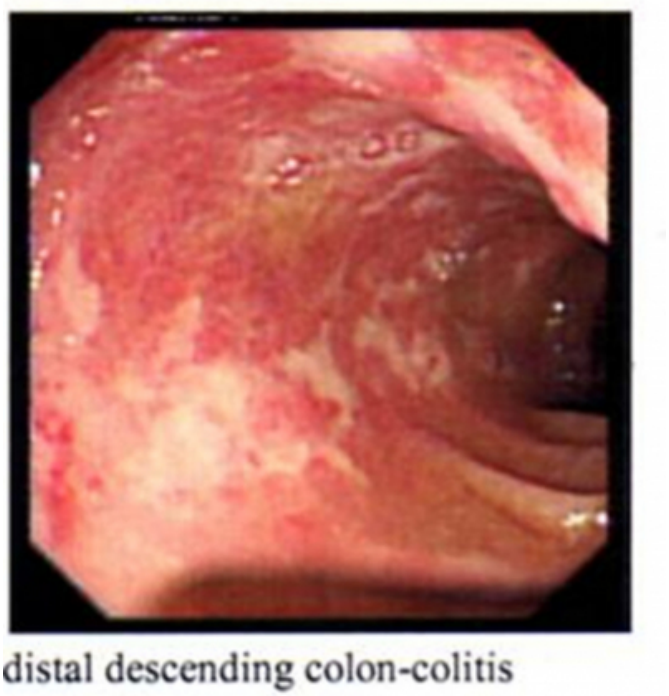


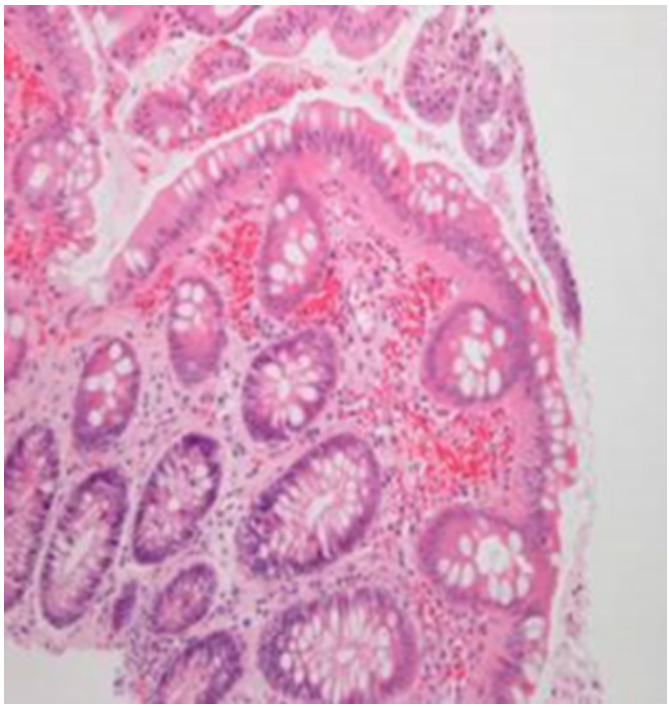
Figure 2



The histopathology specimens of the sigmoid / descending colon revealed an ischemic colitis with red cell

extravasations, haemosiderin deposition and fibrosis (Figures D).

Figure 3



A colonoscopy 6 weeks later, revealed the previously visualised blue naevi in the recto-sigmoid colon but the inflammation in the sigmoid and descending colon had completely resolved. The patient had no further rectal bleeding and remained asymptomatic during his follow up over a period of 6 months.

DISCUSSION

Blue rubber bleb nevus syndrome (BRBS) associated with ischemic colitis, first to be reported in literature

Cutaneous angiomas in BRBS are located mainly on the upper limbs, trunk, and perineum, which empty on pressure but quickly refill when the pressure is removed.([1], [2]) The GI angiomas can be found anywhere, but mostly affects the small bowel. They can cause chronic iron deficiency, although may present as an acute bleed or rarely, with a chronic consumptive coagulopathy ([3]). Occasionally, they may cause complications such as intussusception or volvulus ([4]).

BRBNS is managed conservatively with iron replacement therapy and transfusions. Surgery can be performed to remove the vascular malformations. Endoscopic laser photocoagulation has been successfully used for lesions in the alimentary tract ([5]).Individual case reports have suggested that systemic treatment with corticosteroids, interferon and vincristine may also be effective. More recently, subcutaneous octreotide in the presence of active lesion proliferation or disseminated intravascular coagulation has been used successfully ([6], [7]).

References

1. Laura K Gilbey, Carlos E Girod. Blue rubber bleb nevus syndrome*: Endobronchial involvement presenting as chronic cough. Chest. Chicago: Aug 2003.Vol.124, Iss. 2; pg. 760
2. Sumi Y, Taguchi N, Kaneda T. Blue rubber bleb nevus syndrome with oral hemangiomas. Oral Surg Oral Med Oral Pathol 1991;71:84-6.
3. H. Apak; T. Celkan; A. Özkan; I. Yildiz; E.H. Aydemir; S. Ozdil; S. Kuruoglu. Blue Rubber Bleb Nevus Syndrome Associated with Consumption Coagulopathy: Treatment with Interferon. Dermatology; 2004; 208, 4; pg. 345.
4. Tyrrel RT, Baumgartner BR, Monte mayor KA. Blue rubber bleb nevus syndrome: CT diagnosis of intussusception. AJR Am J Roentgenol 1990; 154:105-6
5. Morris L, Lynch PM, Gleason WA Jr, et al: Blue rubber bleb nevus syndrome: Laser photocoagulation of colonic haemangioma on a child with microcytic anaemia. Paediatric Dermatol 1992, 9:91-94.
6. Nardone G, Rocco A, Balzano T, et al. The effficacy of octreotide therapy in chronic bleeding due to vascular abnormalities of the gastrointestinal tract. Aliment Pharmacol Ther. 1999; 13:1429-36.
7. Buonamico P, Sabba C, Garcia-Tsao G, et al. Octreotide blunts postprandial splanchnic hyperaemia in cirrhotic patients: a double-blind randomized echo-Doppler study. Hepatology. 1995: 21 (1): 134-9.

Author Information

V. Anumakonda
Registrar, Kings college Hospital