



Blue Rubber Bleb Nevus Syndrome (BRBNS) Of Esophagus.

Gastroenterology

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ABSTRACT

Blue Rubber Bleb Nevus Syndrome (BRBNS) is a rare disorder that consists mainly of abnormal multifocal venous malformations affecting the gastrointestinal tract, skin and soft tissues. Patients with this syndrome develop Multiple distinctive skin lesions, anaemia from chronic GI bleeding, and require blood transfusion and iron supplementation lifelong. Mortality depends on the GI involvement because it is difficult to treat GI bleeding [1,2]. The incidence of this syndrome is very low, and only approximately 200 cases have been described in the literature. Isolated GI involvement in BRBNS is very minimal and common site is small intestine. We describe a previously unreported case of isolated Blue Rubber Bleb Nevus Syndrome of Esophagus.

KEYWORDS

Blue Rubber Bleb Nevus Syndrome, GI involvement, Esophagus.

CASE REPORT:

A 63-year-old woman was admitted with history of sporadic episodes of melena, fatigue and anemia. she had no other history and physical examination was normal. Laboratory data showed anemia with haemoglobin of 8.1g/dL, liver function tests, coagulation profile, colonoscopy, abdominal ultrasound was normal. Upper GI endoscopy showed multiple scattered bluish submucosal blebs throw out the esophagus with no stigmata of recent haemorrhage. We inferred this was related to BRBNS of esophagus. Since there was no active bleeding or oozing endoscopic sclerotherapy was not performed. Patient was observed in ward and anemia did not progress. Patient was started on iron supplementation and advised regular follow up.

DISCUSSION:

Blue rubber bleb nevus syndrome is very rare condition which consists of multiple venous malformation involving several organs, most commonly the skin and the GI tract. This condition was first described by Gascoyen[3], Bean[4] named this syndrome. The venous malformations may cause gastrointestinal bleeding. However the most important concern is the high probability of fatal GI bleeding or chronic severe anemia[5]. The presence of multiple discrete cutaneous, gastrointestinal, musculoskeletal vascular lesions should suggest the diagnosis of Blue rubber bleb nevus syndrome.

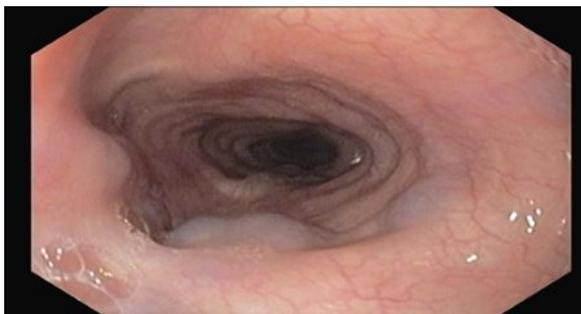
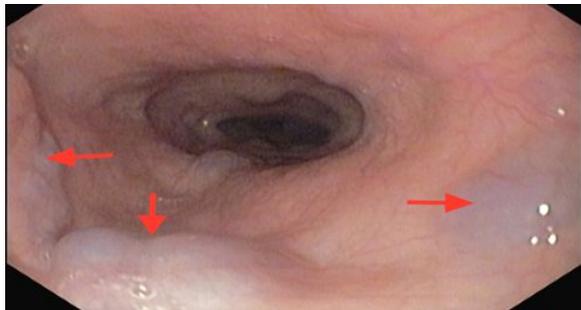


Figure 1 and 2: Upper GI scopy showing multiple scattered submucosal bluish rubbery blebs throughout the esophagus.

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