Skin vascular malformations and recurrent melena which refers to a nevus syndrome

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DESCRIPTION

A 14-year-old North Indian boy presented with a one-year history of multiple hospitalisation anamnesis for recurrent melaena and anaemia. He had received iron supplementation and whole-blood transfusions of more than 10 units for the last 1 year. On physical examination, we observed multiple dark blue, soft, lobulated lesions (5–20 mm) on his palm, sole and extremities which were present for the last 2 years (figure 1). With the application of direct pressure the contained blood emptied leaving behind a wrinkled sac. Doppler ultrasonography of the above-mentioned skin lesions suggested multiple dilated venous lesions filled with blood without any arterio-venous communication. Mucous surfaces of the patient were free of bleb-like lesions and were severely pale. His haemoglobin level was 7 g/dl, with hypochromic microcytosis indicating iron deficiency. Occult blood analysis of the stool was positive on many occasions. Oesophagogastroduodenoscopy revealed bluish venous malformations in stomach and proximal duodenum. Colonoscopy revealed multiple papular venous malformations of approximately 1–2 cm in size located at the distal, ascending colon and transverse colon (figure 2). In order to search for additional lesions, we carried out double balloon enteroscopy and identified multiple venous malformations in the jejunum and ileum. He was treated with multiple sessions of endoscopic sclerotherapy of the

Figure 2 Colonoscopy revealed bluish venous malformation in ascending colon.

gastrointestinal (GI) venous lesions in addition to the blood transfusion and iron supplementation. Our patient's case draws attention to a rare vascular anomaly syndrome predominately involving the skin and whole GI tract. This patient was a case of blue rubber bleb nevus syndrome who was successfully treated with multiple endoscopic therapeutic sessions, transfusion and haematinics supplementation.^{1 2}

Figure 1 Multiple dark blue, soft, lobulated lesions (5–20 mm) on his palm, sole and back of the neck.

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Learning points

- ▶ Patients with recurrent melaena and severe anaemia with simultaneous involvement of skin vascular malformations need visualisation of the whole gastrointestinal (GI) tract by combination of modalities such as oesophagogastroduodenoscopy, colonoscopy and enteroscopy in order to investigate the extent of GI tract involvement by vascular malformations and eradication of vascular lesions either by endoscopic therapy or by other modalities and prevent the patient from future recurrent GI blood loss and severe anaemia.
- ▶ The skin lesions of Blue rubber bleb nevus syndrome (BRBNS) are usually small, multiple, measuring less than 1–2 cm, and blue to purple in colour. Although skin is most commonly involved in BRBNS, the GI lesions may be clinically more relevant.
- ▶ Regular follow up with hemoglobin and stool for occult blood should be done in these patients.

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Competing interests None.

Patient consent Obtained.

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