

Case Report

Young adult with varicosities and recurrent lower gastrointestinal bleeding

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ABSTRACT

A 30 years gentleman presented with anaemia since last ten years due to recurrent rectal bleeding. He has had multiple admissions with transfusions in the preceding 10 years without a clear clinical diagnosis. His physical examination revealed gross pallor, prominent veins on the lateral side of the left lower limb with minimal hypertrophy and a hemangiomas patch over the left buttock. Colonoscopy revealed a few dilated tortuous submucosal vessels in the rectum and sigmoid colon. A MR angiogram showed absent left popliteal vein and angiomas lesions in the rectum and sigmoid colon. He was diagnosed to have Klippel-Trenaunay syndrome with hemangiomas of the lower gastrointestinal tract causing bleeding. He underwent a low anterior resection with stapled anastomosis, which has effectively controlled his bleeding. Adults with gastrointestinal bleeding due to congenital venous malformations commonly have visible external stigmata, which are often pointers of rare syndromes.

Keywords: Klippel-Trenaunay syndrome, Lower gastrointestinal bleeding

INTRODUCTION

Klippel-Trenaunay syndrome (KTS) is an uncommon congenital vascular malformation. A triad of bony or soft tissue hypertrophy, usually affecting one extremity, haemangiomas and/or lymphangiomas, and varicosities or venous malformations helps to clinch a clinical diagnosis of KTS.¹ In patients with KTS, vascular malformations involving the gastrointestinal tract have been reported, which can be a source of recurrent bleeding causing significant morbidity and mortality. Clinical manifestations vary from occult to massive life threatening bleeds. Segmental bowel resections may be required at times in patients with clinically significant gastrointestinal bleeding.² Due to diffuse nature of intestinal hemangiomas, endotherapy is often not useful.³ We report a case of KTS with lower GI bleeding due to diffuse vascular malformations involving the rectum and part of sigmoid colon. This gentleman was labelled as a

case of allergic proctitis and was managed conservatively because of the lack of awareness of this entity.

CASE REPORT

A thirty years old gentleman reported to our OPD with complaints of intermittent bright red rectal bleeding of ten years duration. He has remained transfusion dependent because of intermittent haematochezia resulting in severe anaemia. However, these episodes of haematochezia had never resulted in any hemodynamic instability.

Physical examination revealed generalized pallor, prominent varicosities on lateral aspect of the left lower limb and a haemangiomas patch on the left buttock. The left lower limb was longer than the right. There were numerous hyper-pigmented lesions on the lower aspect of both lower limbs. Abdominal and digital rectal

examination was normal. Proctoscopy revealed erythematous mucosa but no haemorrhoids. A provisional diagnosis of KTS was considered.

Laboratory evaluation demonstrated significant anaemia with haemoglobin of 3 gm/dl. The platelet count and coagulation parameters were normal. Upper GI endoscopy revealed no abnormality. On colonoscopy, dilated tortuous submucosal vessels in rectum and lower sigmoid colon consistent with angiomatous malformation were seen. A Doppler evaluation of lower limbs revealed dilated left short saphenous vein with dilated venous channels in lateral aspect of thigh draining into deep femoral vein. The left popliteal vein was absent. This confirmed the diagnosis of KTS.

The main concern was whether the angiomatous lesions were limited to rectum or extending into mesorectum as well. An MR Angiogram was done which revealed that the common iliac, internal iliac and external iliac arteries and veins were normal. There were obvious angiomatous lesions confined to the wall of the upper rectum and sigmoid colon. There were no pelvic hemangiomas lesions and AV fistulas. Femoral vein was present but popliteal vein was absent.

Because of the significant disability of his rectal bleeding, a low anterior resection and coloanal stapled anastomosis was carried out after thorough explanation of the morbidity of the procedure to the patient.

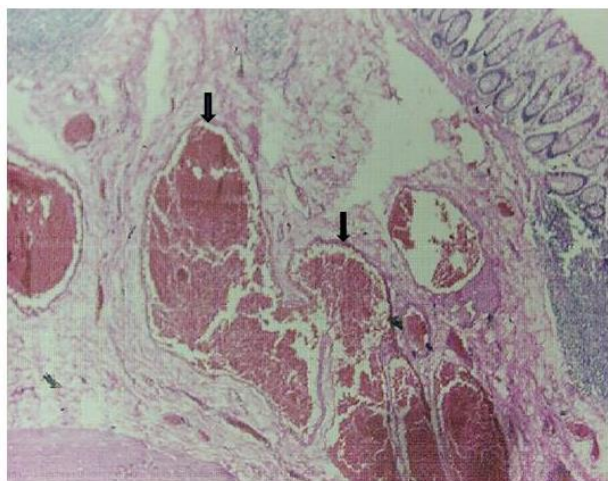


Figure 1: Dilated vascular channels in sub mucosa (black arrows) with normal mucosa and muscularis.

Pathological examinations of the recto sigmoidectomy specimen confirmed vascular malformations confined to the submucosa (Figure 1). Postoperative convalescence was unremarkable and he was discharged on the 7th postoperative day. He was advised compressive stockings for his varicosities.

At follow up after 18 months, he has normal haemoglobin and has been completely free from rectal bleeding.

DISCUSSION

Hemangiomas in various organs of the gastrointestinal tract have been reported in patients with KTS.⁴ Although gastrointestinal bleeding begins in early life, it is often intermittent in nature.⁵ Diffuse hemangiomas involving the distal colon and rectum are the commonest sites of gastrointestinal bleeding, reported in 1-12.5% of KTS cases.⁵ The clinician and the endoscopist should be vigilant in picking up these lesions on scopy. Due to diffuse nature of the lesions, an incorrect diagnosis of inflammatory bowel disease can be made. Hemangiomas of GI tract are associated with various syndromes e.g. KTS, Osler Rendu Weber syndrome, etc with characteristic other lesions elsewhere on the body. Hence a complete clinical examination would help to make a timely and correct diagnosis.

The pathogenesis of visceral hemangiomas remains unclear. Servallo claimed that shunting of blood flow into a dilated internal iliac vein lead to hindrance of drainage of vesical, genital and rectal veins, with subsequent venous malformations.² Shunting occurred secondary to interruption of the superficial femoral or popliteal veins of the affected limb.

Issues that needed resolution in this case were a) confirming that the vascular malformation is indeed the cause of bleeding, b) ruling out similar lesions elsewhere in the gastrointestinal tract and c) deciding on requirement for surgery. Radiological investigations play an important role in assessing visceral involvement and possibly to identify the likely source of bleeding. Clinically this patient presented with haematochezia, so it was considered likely that the source was in distal colon or rectum. Colonoscopy was useful to localize the anomalous vessels and rule out other coincidental colonic lesions. The role of MR Angiography has not been well defined in KTS but has the potential of assessing the vascular malformations non-invasively with better accuracy.⁶ We could preoperatively rule out pelvic hemangiomas lesions and AV fistulas with the help of MR Angiography. Thus MR Angiography is a useful preoperative tool in defining the abnormal vascular anatomy and extent of disease. Submitting a young gentleman to a major resection particularly a rectal resection was a difficult decision. Nevertheless due to extensive nature of the disease local sclerotherapy was not possible. Sphincter saving low anterior resection, if feasible, avoids a permanent stoma and restores satisfactory anal function. It is imperative to avoid impotence by careful preservation of sacral nerves. This is possible since the rectal dissection can be done within the mesorectum as mesorectum is usually devoid of vascular malformations. Fortunately in our patient there was no major morbidity postoperatively.

Despite the reasonably obvious external stigmata of KTS in this gentleman, he remained undiagnosed for a long period due to lack of awareness of this entity. Young

adults presenting with severe gastrointestinal bleeding, venous malformations should be high on the list of differential diagnosis. With this in mind, if careful examination is performed for external stigmata, spot diagnosis of rare entities causing gastrointestinal bleeding can often be made.

CONCLUSION

Young adults presenting with varicosities, hemangiomas and recurrent lower gastrointestinal bleeding, a spot diagnosis of Klippel-Trenaunay syndrome can be made. MR angiography helps to localise the disease prior to intervention. Surgery is a one time, curative and safe option for localised disease.

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