

Klippel-Trenaunay Syndrome: A Rare Cause of Recurrent Lower GI Bleeding

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ABSTRACT

Klippel-Trenaunay syndrome is a non-hereditary congenital abnormality characterized by a clinical triad of soft tissue hypertrophy of an extremity, varicosities and cutaneous hemangiomas or Lymphangiomas. Hematochezia is an uncommon but potentially serious complication of Klippel-Trenaunay syndrome.

We report here a case of a young female who presented to us with intermittent bleeding per rectum and iron deficiency anemia. The patient gave history of excision of hemangioma in the right leg 12 years ago. Her upper GI endoscopy revealed grade-I esophageal varices and colonoscopy showed vascular malformation from rectum to sigmoid. CT Angio revealed splenic haemangiomas with multiple vascular malformations in GI tract.

Key Words: Klippel-Trenaunay syndrome, Lower GI Bleed, Haemangioma, Anemia

INTRODUCTION

Klippel-Trénaunay syndrome (KTS) is rare congenital malformation characterized by bony or soft tissue hypertrophy, usually affecting one extremity, hemangiomas and / or lymphangiomas and varicoses or venous malformations [1,2]. Vascular malformation involving gastrointestinal tract (GIT) have been reported and can be a source of significant morbidity and mortality [2]. Visceral hemangiomas in KTS may involve organs such as the GIT, liver, spleen, urinary bladder, kidney, lung and heart [3].

We report here a case of a young female presenting with intermittent bleeding per rectum and iron deficiency anemia with significant past history of repeated blood transfusions and excision of vascular malformation from right gastrocnemius muscle. Physical examination and CT Angiography along with colonoscopic findings confirmed the diagnosis of KTS.

CASE REPORT

A 33 year old female patient presented to our Gastroenterology department with bleeding per rectum and iron deficiency anaemia. She gave history of repeated episodes of bleeding per rectum along with multiple blood transfusions since child hood. There was history of excision of haemangioma in the right lower limb from between the two heads of gastrocnemius muscle 12 years ago. One year after the surgery she noticed tortuosity of the right lower limb veins along with pain.

Physical examination revealed severe pallor with tortuous veins in right lower limbs.

Laboratory data showed severe anemia with hemoglobin of 6.5 gms /dl, hematocrit 20%, MCV-68 fl, MCH-22 pg and peripheral smear showed picture of iron deficiency anemia. Platelet count and coagulation parameters were normal. CT Angio of abdominal aorta showed multiple large tortuous vascular channels in the liver, mesentery, recto sigmoid and right gluteal soft tissue. Moderate splenomegaly with small splenic haemangiomas. Invasive angiography revealed Portal vein occlusion with periportal collaterals with diffuse venous vascular structures in pelvis and retroperitoneum. UGI endoscopy

showed grade-I esophageal varices and colonoscopy revealed varices extending from rectum to sigmoid colon along with grade-II intero external hemorrhoids for which banding was done. The patients bleeding stopped and the patient is doing well on oral Iron supplements.

DISCUSSION

KTS is a rare congenital disease with a reported incidence of 2-5/1,00,000 population. KTS patients presenting with GI bleed is also an uncommon presentation. Our case had recurrent hematochezia which required transfusions. KTS is a progressive disease & may have life threatening bleeds also.

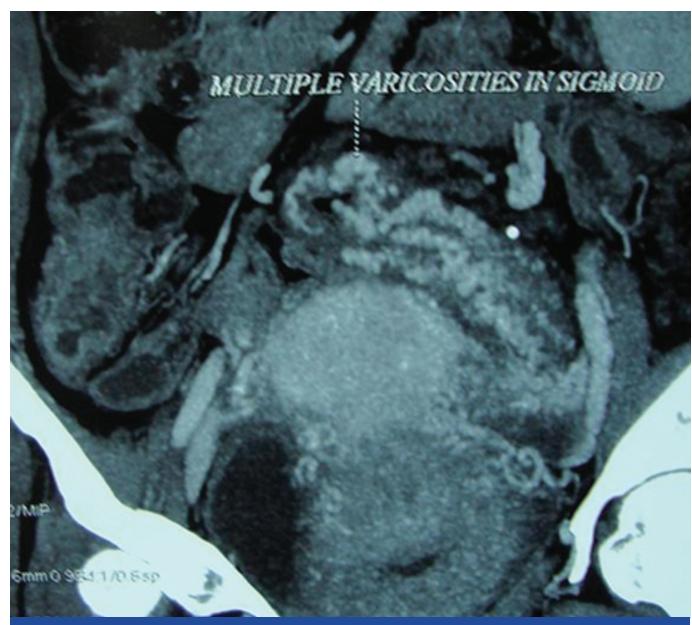
Involvement of the GI tract may be more common in KTS than previously believed (occurring in perhaps as many as 20% of patients) and may go unrecognized in patients without overt symptoms [4]. The most common bleeding sites in the GI system are the distal colon and rectum. Jejunal haemangiomas and esophageal varices as bleeding sources caused by prehepatic portal hypertension were reported in the literature [2] GI haemorrhage usually begins in the first decade of life and tends to be intermittent [5] However, the progressive nature of KTS warrants that physicians consider invasive surgical operation or angiographic intervention such as embolization of the bleeding vessel during the ongoing follow-up when there is a patient with transfusion-dependent anemia, life-threatening bleeding episodes, and/or poor quality of life due to severe anemia [2].

Endoscopic therapy is usually preferred for localized lesions or postoperative residual disease. A literature search showed that KTS patients with colonic haemangiomas may benefit from endoscopic interventions. The neodymium: yttrium-aluminum- garnet laser for residual lesions, partial colectomy for visceral haemangiomas [6] and argon laser photocoagulation for hemangiomas involving the distal 7 cm of the anorectum [7] were found effective in patients with KTS.

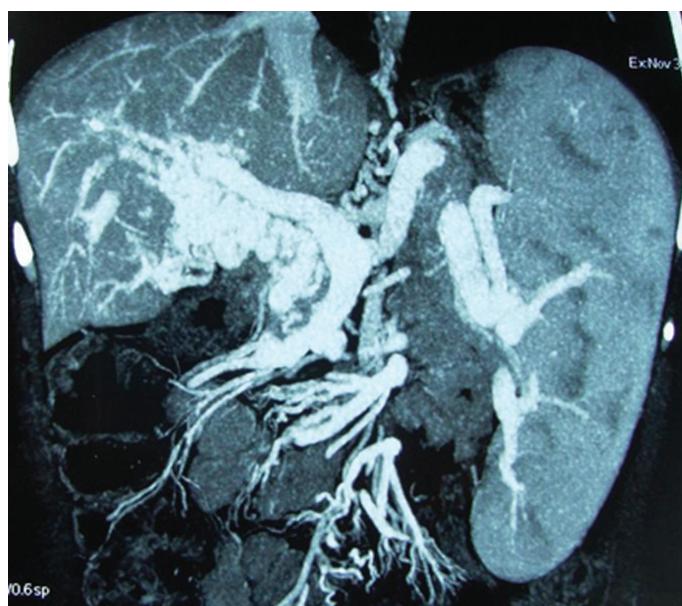
Due to the progressive nature and wide extension of KTS lesions, endoscopic therapies have limited value in the management .Angiographic interventions should be used preoperatively for visualizing the vascular anatomy and determining the disease extent [8].



[Table/Fig-1]: Varicosities & soft tissue hypertrophy on posterior aspect of leg.



[Table/Fig-4]: CECT abdomen showing multiple varicosities in sigmoid colon.



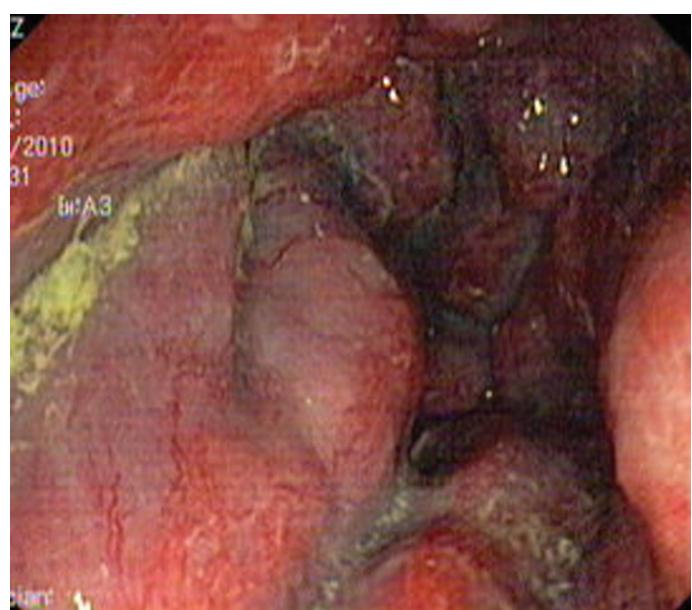
[Table/Fig-2]: (CT angio) showing Increased vascularity at porta ; dilated splenic vein ; increased vascularity at lower end of oesophagus.



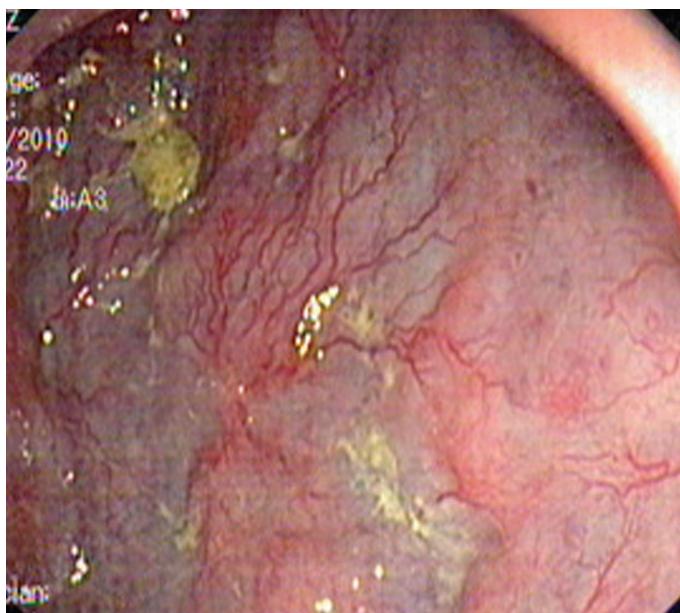
[Table/Fig-5]: CECT abdomen showing intramural vascularity of sigmoid colon



[Table/Fig-3]: CT Angio showing tortuous vessels (arrow head) arising from internal iliac artery.



[Table/Fig-6]: Colonoscopy showing Hyperemia & bluish nodular folds in rectum suggestive of ?Angiomatosis.



[Table/Fig-7]: Colonoscopy showing bluish tortuous vessels suggestive of rectal varices.

In one of the largest published series of KTS patients, haematochezia was reported in only six of 588 patients, although a few other cases may have gone unnoticed [5]. Management of colorectal intestinal hemangiomas in KTS will depend on the extent and severity of blood loss. When the entire rectum is severely involved, surgery is less attractive, as a permanent stoma will prove necessary.

Conservative management and iron supplements may be sufficient in those patients who present with occasional non-significant and non-debilitating bleedings. However, long-term treatment mostly requires surgical resection comprising proctocolectomy in the case of life-threatening colonic bleeding and coloanal anastomosis with preservation of anal function, especially in younger patients, or abdomino-perineal resection in the case of rectal bleeding due to the diffuse and sometimes progressive disease process [2].

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Vascular embolization can be considered if a distinct bleeding site is encountered. Endoscopic photocoagulation using argon laser is sometimes employed for the management of localized lesions or ablation of postoperative residual disease.

CONCLUSION

This report describes extensive cavernous haemangiomas of the colon in a patient with Klippel-Trénaunay syndrome as a rare cause of recurrent lower gastrointestinal bleeding. Apart from occasional hematochezia and mild anemia our patient was asymptomatic. In view of the extensive colonic involvement described, bleeding will most likely only subside following surgical resection of the colon. Hence, only oral iron supplementation was administered so far.

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