Case Report

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Lower Gastrointestinal Bleeding in Klippel-Trenaunay Syndrome: A Case Report

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Abstract:

Klippel-Trenaunay syndrome (KTS) is a rare, vascular malformation. The major clinical presentation is an overgrowth involving the extremities. The diagnosis of KTS was based on an imaging study, which revealed vascular malformations. The author's report is of a 36-year Thai man with; hypertrophy of the right lower extremity, whilst having suffered from two-months of bloody defecation. The magnetic resonance imaging showed venous malformations with soft tissue hypertrophy of the affected limb, genitalia and rectum. The patient was diagnosed with: KTS accompanied by gastrointestinal complications.

Keywords: gastrointestinal bleeding, Klippel-Trenaunay syndrome

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Case report

A 36-year Thai man presented with a scrotal mass eight years in age. He had also further developed painless hematochezia, over a period of two months. Upon physical examination, a well, defined violaceous plaque, with multiple reddish papules resembling a port wine stain, was found over his right thigh. He reported as having had this lesions since time of birth. The scrotal mass was 4 to 5 cm in diameter, irreducible, soft and non-pulsatile on palpation. He also had multiple non-pulsatile, subcutaneous swelling extending from his right groin to the dorsum of his right foot with disproportion between both lower extremities (Figure 1).

The results of the blood work were as follows: Complete blood count, hemoglobin 7.6 g/dl (13.0-18.0), hematocrit 29.7% (40-54), mean corpuscular volume 61.1 fL (83-97), red cell distribution width 28.9% (11-16), white blood cell count 5,150 cells/mm^3 (4,500-10,000) (neutrophils 44.4%, lymphocytes 40.8%, eosinophils 5.6%, monocytes 8.2%, basophils 1%), platelet count 258,000/mm^3 (150,000-450,000), serum creatinine 0.84 mg/dl (0.67-1.17), and liver function tests, aspartate transaminase 11 U/L (0-40), alanine transaminase 8 U/L (0-41), albumin 44 g/l (35-52). A colonoscopy was performed. The study revealed a generalized, erythematous wall of submucosa along the sigmoid colon running through the anal canal.

A magnetic resonance imaging (MRI) of the lower extremities (Figure 2a) showed multiple, vascular dilatations along the subcutaneous region at the lateral aspect of the right thigh to the right leg, involving the anteroposterior muscle compartments. The deep, superficial venous systems were dilated on the right side. In contrast, the visualized arteries, along, with its major branches were not dilated.

An MRI of the lower abdomen showed dilated vascular channels in the subcutaneous layer of a lateral aspect of the right pelvic, scrotal sac and the right, lateral surface of the penis. The rectum was edema of the wall and also appeared to have some varices (Figures 2b-c).

The patient was diagnosed with Klippel-Trenaunay syndrome (KTS) with gastrointestinal (GI) complication and iron deficiency anemia. He received a compression stocking for the varicose veins, corrected anemia and was scheduled for follow-up of the scrotal mass and clinical symptoms.

Figure 1 A patient's clinical picture demonstrated a disproportion between both legs and thighs and a presentation of large violaceous plaque on the right lateral thigh

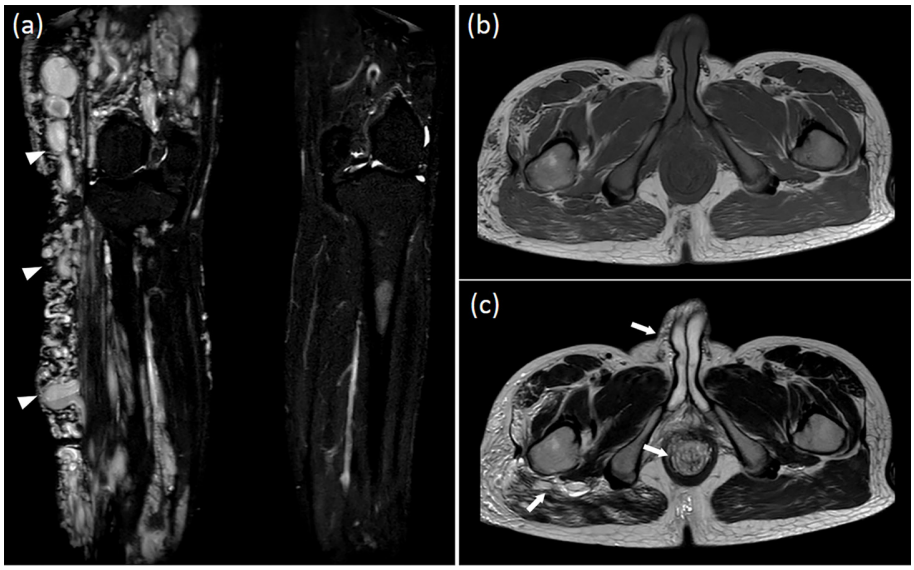

Figure 2 The magnetic resonance imaging. (a) A coronal view showed superficial and deep venous dilatation along the lateral side of the right thigh and leg (arrowheads) (STIR sequence). (b, c) An axial view showed dilated vascular vessels in the lateral aspect of the right pelvic and scrotal sac with varices in the rectum (arrow). (b, T1W-TSE image; c, T2W-TSE image)

Discussion

Klippel-Trenaunay syndrome is a congenital limb capillary-venous (CVM) or capillary-lymphatic-venous (CLVM) malformation.[1] The diagnosis requires two features out of a triad of capillary malformation, atypical varicose veins or venous malformation and the hypertrophy of soft tissues or bone.[2] KTS is classified into two types, based on the cutaneous lesion as well as degree of CLVM. Simple KTS has blotchy or segmental cutaneous lesions, while complex KTS has well, demarcated geographic lesions and is associated with more severe vascular and lymphatic malformation.[3]

CVM and CLVM can arise from both the superficial and deep venous system causing varicose veins, cellulitis, venous stasis and ulcers.[4] Limb hypertrophy from abnormal vascular supply causes limb length discrepancy, which results in an abnormal gait.[5] The primary treatment of KTS is the use of a compression stocking, because of the low flow nature of the CVM. Polidocanol microfoam sclero therapy is also an option in small venous malformations.[6]

GI bleedings from vascular malformations and varices are uncommon in KTS, however, it is the leading cause of morbidity for this disease. The primary source of bleeding, in the GI tract, is most commonly caused from the distal colon and rectum.[7-9] Clinical bleeding is often of an intermittent behavior, which could cause iron deficiency anemia. Computed tomography and MRI scans can evaluate the visceral involvement of KTS.[10] Endoscopic studies may also demonstrate submucosal and mucosal CVM, however, can not evaluate deeper levels.[8] In patients, who have mild anemia, the only treatment is observation coupled

with iron supplementation. In contrast, in the cases of life–threatening cases, embolization or surgical procedures are the main courses of treatment.[11,12]

Conclusion

The authors reported a Thai patient with Klippel–Trenaunay syndrome, who had GI bleeding, jointly with iron deficiency anemia. General physicians should consider the complications of KTS when experiencing similar patients, especially those, who have had unilateral limb hypertrophy and skin vascular malformations.

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