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An Uncommon Presentation of Klippel-Trenaunay Syndrome Presenting with Acute Hip Pain

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Authors' contributions

This work was carried out in collaboration between all authors. Author HS designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors AB and SD managed the analyses of the study. Author ZE managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Klippel-Trenaunay syndrome (KTS) is a syndrome that affects the development of blood vessels, soft tissues and bones. We report a rare case of KTS presenting with spontaneous acute hip pain.
Case Presentation: A 38-year-old male patient presented to the emergency department with a complaint of right hip pain and elevated temperature of one moth duration. These symptoms disappeared spontaneously after two days of hospitalization. Computerized tomography (CT) scan revealed a common iliac vein hypoplasia and collateral circulation associated with asymmetric psoas muscles. A diagnosis of KTS was made based on the vascular abnormalities and associated soft tissue changes.
Conclusion: Our case illustrates a new case of KTS presenting with spontaneous acute hip pain.

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Keywords: Klippel-Trenaunay syndrome; pain; hip; malformation.

1. INTRODUCTION

Klippel-Trenaunay syndrome (KTS) is an uncommon syndrome that affects the development of blood vessels, soft tissue, and bone. This rare congenital disease has three principle characteristic features: 1. a red birthmark termed port-wine stain; 2. an abnormal overgrowth of soft tissue and bone; and 3. venous malformations [1,2]. We report an unusual case of KTS first presenting with acute hip pain. The diagnosis of KTS was made based on the presence of vascular and soft tissue abnormalities and after ruling-out other potential diagnoses.

2. CASE REPORT

A 38-year-old male with no significant medical history, presented to the emergency department with a complaint of right-sided hip pain. He also reported right inguinal pain with associated with fever of one month's duration. On physical examination he was found to have a temperature of 38℃, pain in the right hip on mobilization but without limitation of movement, and left limb enlargement when compared to the right limb. He was hospitalized with a presumptive diagnosis of septic arthritis. Antibiotics were not initiated pending the results of CBC study. He was prescribed supportive therapy and bed rest. Hip and lower limbs radiographs did not show hypertrophy or bony changes in either hip or limb.. Ultrasound of the right hip revealed an hypertrophied left psoas muscle. Laboratory blood tests did not reveal any abnormalities. A CT scan revealed left external iliac vein hypoplasia with collateral circulation (Fig. 1). The CT also revealed left psoas muscle enlargement compared to the right (Fig. 2). After eliminating other possible diagnoses such as sepsis, inflammatory disease, and microcrystalline disease, the diagnosis of KTS was made based on the presence of vascular abnormalities and the soft tissue hypertrophic changes. The patient's symptoms resolved spontaneously with two days bed rest and no further treatment was required. After 8 months of follow-up observation the patient remained stable requiring no treatment.

3. DISCUSSION

KTS was first described by Klippel and Trenaunay in 1900 as vascular lymphatic malformation with hypertrophy of the affected limb's bony and soft tissue [1]. A restrictive diagnostic criterion was proposed

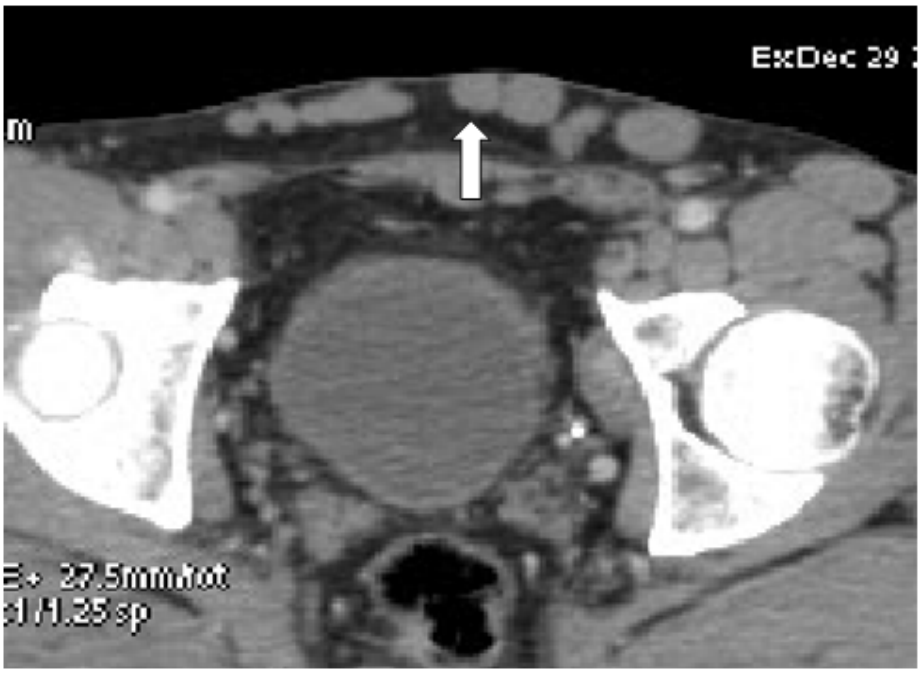

Fig. 1. CT scan showing left external iliac vein hypoplasia

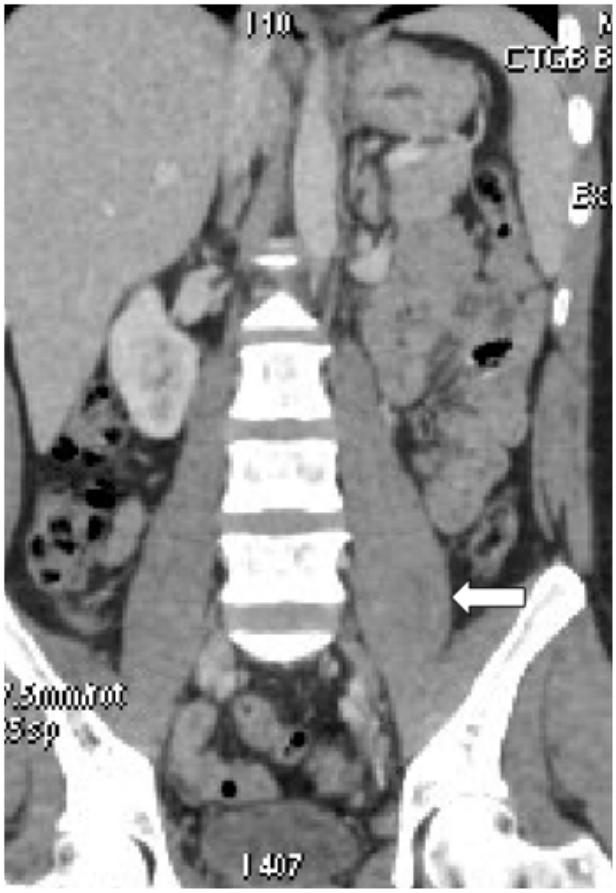

Fig. 2. CT scan showing asymetric psoas muscles

by Oduber et al. comprising 2 major features: vascular malformation (capillary, venous, arteriovenous, and lymphatic malformations) and disturbed growth of bone and/or soft tissues (hypertrophy and rarely hypotrophy) [2]. Several theories for pathophysiology of KTS have been proposed. Baskerville et al. suggested a mesodermal abnormality during fetal development leading to the maintenance of microscopic arteriovenous communications in the limb bud [3]. An increase in blood flow may lead to nevus formation, superficial varices, and hypertrophy of the juxtaepiphyseal cartilage [4]. Most cases of KTS are sporadically reported. A few cases of a familial background have also been reported [5,6]. However, there was no family history of KTS in our patient's case. It is a rare condition with no gender predominance [7]. This disease is characterized by a wide range of symptoms with a broad spectrum of severity [8]. Signs and symptoms can range from negligible hypertrophy of the limbs, as reported in our case, or only slight varicosities to life-threatening complications, Symptoms can also include: pain and discomfort, leg length discrepancy, erysipelas, venous thrombosis, pulmonary embolism and/or gastrointestinal bleeding [9,10]. KTS is more commonly found to be unilateral than bilateral [7,11]. In the series published by Sung et al. of the 19 patients with KTS, 15 patients had malformation of one limb only. The lower limbs were affected in 13 patients (68%). Pain was noted in 11 cases [8]. Our new case of KTS presented with spontaneous right hip pain but the malformation was found to be on the left side. We found no explanation for the right-sided pain localization other than the possibility of postural guarding and muscle spasm due to limb discrepancy and our patient's response to bed rest. A diagnosis of KTS can be made if two of the following findings are present: hypertrophy of the soft tissue, bone overgrowth and vascular malformation. Our case is unique in the initial presentation of spontaneous and acute intense pain.

4. CONCLUSION

KTS is an extremely rare disorder but should be included in the rule-out diagnosis when there is a presence of pain and venous malformations.

CONSENT AND ETHICAL APPROVAL

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images. The case study was approved by the hospital local ethics committee.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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