

## Case Report

# Klippel-Trenaunay Syndrome : Radiological Aspect

Rajesh K Badhan<sup>1</sup>, Varinder Singh Bhatti<sup>2</sup>, Gurpreet Singh<sup>1</sup>

<sup>1</sup> Assistant Professor, <sup>2</sup> Junior Resident,

Department of Radiodiagnosis, Govt. Medical College and Rajindra Hospital, Patiala, Punjab, India

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**Corresponding Author:**

**Dr. Rajesh K Badhan**

Email ID – rkbadhan@yahoo.co.in      Contact No- 9417149907

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## Abstract

Klippel – Trenaunay Syndrome (KTS) is a rare complex congenital angio-osteo-hypertrophic syndrome. Features of KTS are Capillary Malformation, Soft Tissue/Bone Hypertrophy and Varicose Veins or Venous Malformation (1, 2). The lower extremity is the site of involvement in approximately 95% of patients (3).

## KEYWORDS

Klippel- Trenaunay Syndrome (KTS), Colour Doppler , Vascular Malformation, Hemihypertrophy, Varicosities

## INTRODUCTION

KTS is a rare disorder, with estimated incidence of 3-5/100000. There is no sex predilection (4). Superficial venous malformation of the unilateral lower limbs or varicosities in unusual distribution are the most common findings. Persistence of embryonic vein of Sevelle is also a feature suggestive of KTS. Although conventional venography is a gold standard procedure, but due to its invasive nature, it has been largely replaced by other non-invasive radiological modalities like Colour Doppler and Computed Tomography Scan(CT) or Magnetic Resonance Imaging (MRI) Venography (5,6).

## CASE REPORT

A 14 year old male brought to Rajindra Hospital Patiala for right leg swelling since birth. On clinical examination, patient was provisionally diagnosed as a case of varicose veins. Then patient was referred to Radiodiagnosis Department for Colour Doppler study of lower limbs on 11-09-2021. On physical examination there was increase in girth and length of right lower limb along with overlying few cutaneous lesions. Multiple venous prominences along the

buttock, lateral part of thigh, anterior aspect of knee and leg of right lower limb noted (Fig. 1).



Figure 1 : Clinical image showing varices and hypertrophy of right lower limb with subcutaneous skin changes.

Supplementary X-Ray of hip and both lower limbs taken to know bone length status which shows lengthening of right lower limb bones (Fig. 2).



Figure 2: X-Ray Bilateral hip joints and thigh - AP view shows increased length of right femur.

Colour Doppler of right lower limb revealed multiple dilated, tortuous vascular channels in subcutaneous and intermuscular planes suggestive of varicosities more so on anterolateral compartment (Fig. 3A,3B,3C).

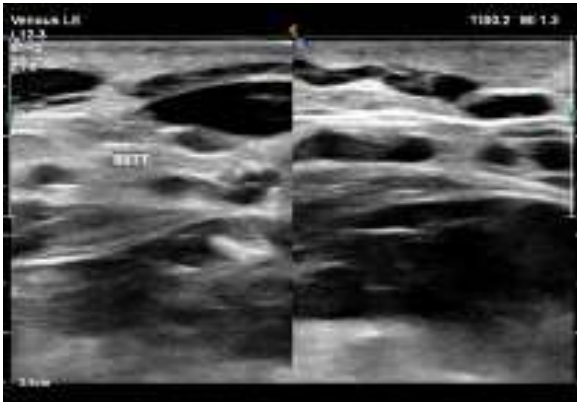


Figure 3 A : Colour Doppler of right buttock shows multiple anechoic dilated channels suggestive of superficial varicosities

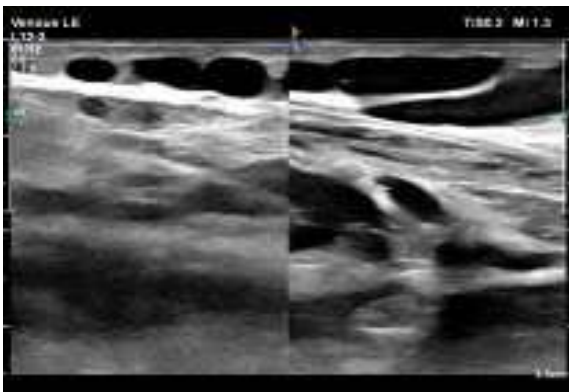


Figure 3 C: Colour Doppler images shows varicosities in subcutaneous and intramuscular plane

No intersaphanous anastomotic channels were identified. The deep venous system did not show any evidence of thrombosis or venous insufficiency. Right saphenofemoral and saphenopopliteal junctions were competent. Colour Doppler of left lower limb venous system was unremarkable. Colour Doppler of bilateral lower limb arterial system was unremarkable.

MRI angiography & venography shows marked subcutaneous thickening and hypertrophy of right buttock, thigh and leg. There were multiple flow void tortuous channels corresponding to varicosities (Figure 4)



Figure 4 : MRI shows diffuse subcutaneous thickening of right buttock and thigh along with multiple tortuous void channels corresponding to varicosities.

## CONCLUSION

There are various causes of varicose veins in the lower extremities such as venous insufficiency of the saphenofemoral junction, saphenopopliteal junction, or usual perforating veins. Traditionally, Colour Doppler has been used for evaluation of varicose veins. Sometimes, varicose veins arise from an unexpected anatomic source; in these cases, CT or MRI venography can provide an overview of the varicose veins. Colour Doppler with complementary CT venography is useful for determining the precise cause of varicose veins. Radiologists should be familiar with the complete range of primary causes of varicose veins in the lower extremities and with their radiologic manifestations and should recognize the complementary role of CT venography in their evaluation(7). A high degree of suspicion should be required for diagnosis of KTS in the presence of few salient features on imaging like history since birth, unilateral lower limb involvement and atypical distribution of varices(5). Being a complex genetic origin, KTS cannot be cured in the complete sense of word. Based on radiological characterisation of different components of the syndrome bone shortening procedures, foam sclerotherapy, vein ligation, debulking surgery are some procedures that could be offered to some cases amenable to treatment. Hence, a radiological protocol to pursue management planning is required to characterize the different components of this syndrome. Skeletal survey should be an essential tool in detection of associated skeletal abnormalities. Color Doppler ,CT or MRI plays a great role in diagnosis, classification, characterisation of soft tissue involvement and accurate evaluation of venous and lymphatic components of the syndrome(3).

### Abbreviations

KTS : Klippel-Trenaunay Syndrome

CD : Colour Doppler

CT: Computed Tomography

MRI: Magnetic Resonance Imaging

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