

# Abnormal Tc99m sulesomab in Klippel-Trenaunay syndrome

Shabina Begum, Ewa Nowosinska, John R. Buscombe  
Nuclear Medicine Royal Free Hospital, London, United Kingdom

[Received 31 XII 2010; Accepted 18 II 2011]

## Abstract

Tc-99m sulesomab is indicated in bone and joint infection, but reading of scans can be affected by pre-existing conditions. This case report describes a case of Klippel-Trenaunay syndrome (KTS) which results in vascular malformations of one or more limbs. Tc-99m sulesomab imaging demonstrated persistent blood pool activity up to 20 hours post injection. However, despite this, septic arthritis could be identified with confidence in the same limb.

**Key words:** Klippel-Trenaunay syndrome, Tc-99m sulesomab

Nuclear Med Rev 2010; 13, 2: 84–86

## Introduction

In 1900, the French physicians Klippel and Trenaunay described a rare syndrome: Klippel-Trenaunay syndrome (KTS) [1]. The syndrome is an autosomal-dominant disorder characterized by a triad of cutaneous vascular nevi, soft tissue, or bony hypertrophy, and varicose veins or venous malformations involving one or more extremities and often associated with chylous ascites [2]. Currently, Klippel-Trenaunay syndrome can be diagnosed on the basis of any two of these three features [3].

We report a case of extensive left leg KTS with minimal symptoms and its affect on imaging with Tc-99m sulesomab for septic arthritis.

*Correspondence to:* John Buscombe  
Nuclear Medicine Addenbrookes Hospital  
Hill Road, Cambridge CB2 0QQ, United Kingdom  
Tel: (+44) 1223 217185, fax: (+44) 1224 274596  
e-mail john.buscombe@addenbrookes.nhs.uk

## Case report

A 51-year-old woman was referred to our institution for a three-phase bone scan of the left knee joint to exclude post arthroscopic joint infection. She was a known case of Klippel-Trenaunay syndrome for five years but had with symptoms except in her of left knee in which she suffered a lateral meniscal tear. She had a low-flow venous malformation along the whole length of the left thigh, calf, and short saphenous vein on duplex ultrasound imaging. Several small varicose veins were present in both the postero and lateral aspect of the left thigh extending to the calf and foot. She had no limb deformity or gigantism, and no true birthmarks on either leg or buttock. MRI of the left knee joint showed innumerable abnormal vessels throughout the region of the knee in an intra- and extra-articular distribution suggesting arterio-venous malformation.

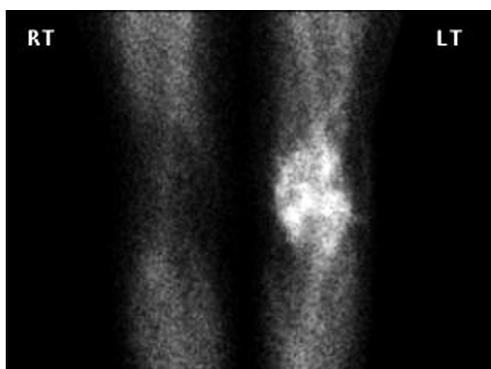
She had a history of intermittent pain, swelling, and locking of the left knee over 5 years and her venous malformation was stable. She was advised to use graduated compression stockings but she continued to suffer locking and pain in that knee. To reduce her symptoms she underwent an arthroscopy. Debridement of the degenerative meniscal tear was performed and generalized arthritic changes were also noted. After this procedure she developed pain, swelling of the joint, and pyrexia. On physical examination the left knee was swollen and bruised but was not noted to be warmer to the touch than the right knee.

She was referred to our department for a three-phase bone scan to identify if septic arthritis of the left knee was a possibility. Increased uptake was noted in the first two phases of the Tc-99m MDP scintigraphy involving the whole left knee, and in the three-hour static phase there was increased uptake seen in the medial aspect of the tibia corresponding to the osteolytic lesion in the margin of medial tibial plateau seen on X-ray (Figures 1–3). These findings suggested that the joint could indeed be infected and a confirmatory Tc-99m sulesomab scan was arranged. Local images of the knee were performed 1 and 5 hours after injection of 793 MBq of Tc-99m sulesomab. Both sets of images showed diffusely increased activity in the left leg compared to the right (Figure 4). However, in addition there were small areas of focal uptake throughout the leg seen above and below the knee. These changes were attributed to the patient's KTS.

In addition to these abnormalities there was increasing focal accumulation of Tc-99m sulesomab around the left knee joint



**Figure 1.** Osteolytic lesion seen in left-sided medial tibial plateau, suggestive of infection.



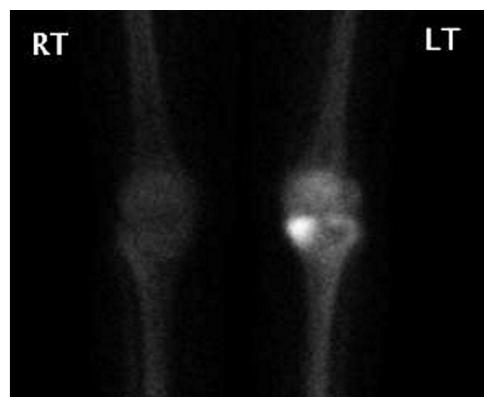
**Figure 2.** Blood pool images showing intense uptake in the tissues surrounding the left knee.

consistent with a septic arthritis or severe inflammation of the left joint with normal activity seen in the right knee.

## Discussion

Klippel-Trenaunay syndrome (KTS) is a rare congenital malformation characterised by the triad of capillary malformations (port-wine stains), varicose veins, and hypertrophy of the soft tissues and bones [4, 5]. The absence of arteriovenous shunting differentiates this entity from other congenital vascular malformations of venous predominance such as Parker-Weber syndrome [6]. The diagnosis is confirmed by the presence of at least two of the three clinical features, although most patients will have all three manifestations [1, 3, 7, 8].

The aetiology of KTS remains unknown although it is thought to be a mesodermal defect that leads to an alteration in local regulation or the production of growth factors affecting angiogenesis. This vascular overgrowth is responsible for the increase in blood



**Figure 3.** Static Tc-99m MDP image confirming intense uptake in the left medial tibial plateau, suggesting infection.

flow, producing hyperthermia and anomalous growth patterns [9] and the increased flow seen on both Tc-99m MDP and Tc-99m sulesomab imaging.

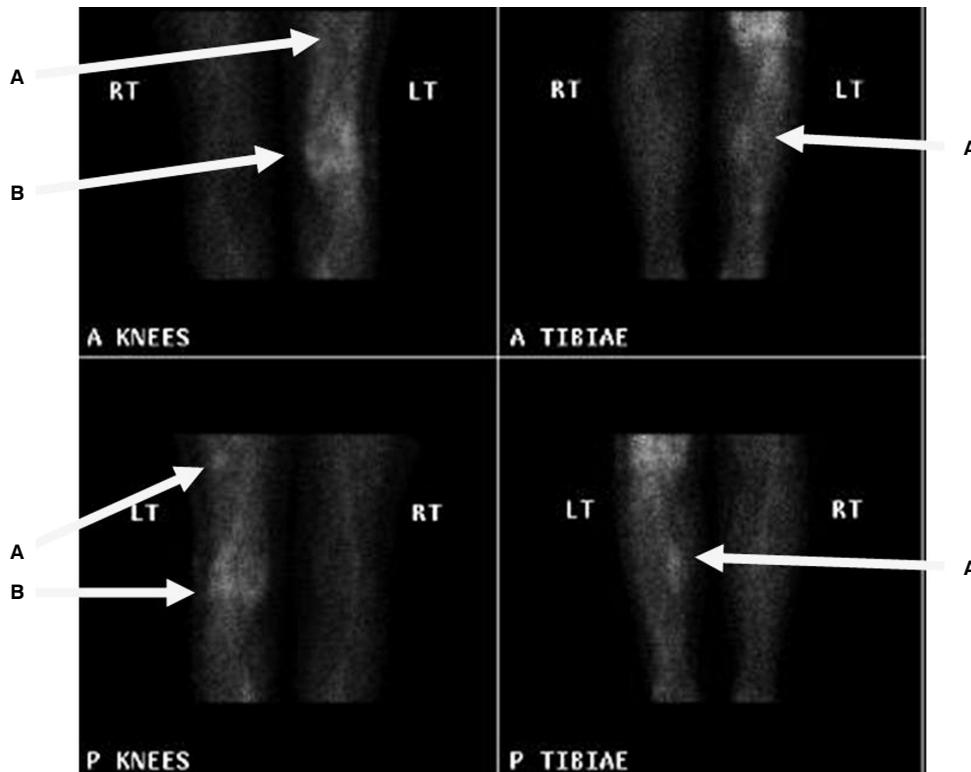
The exact prevalence of this syndrome is unknown. It is estimated that there are 900 affected individuals worldwide, but many cases may be unrecognised or unreported [10]. Most cases of KTS are sporadic and diagnosed at birth or early childhood, with males and females being affected equally. There is no known racial predilection found in KTS [3].

Hypertrophy is the most variable of the three classic features of KTS [3], although up to 30% of patients do not present with hypertrophy [11]. Enlargement of the extremity consists of bone elongation, circumferential soft tissue hypertrophy, or both [3, 12, 13]. The lower limb, upper limb, or both are affected in 95%, 5%, and 15% of cases with KTS, respectively; however, the majority of cases display unilateral changes [14]. Most series describe hypertrophy of tissue/bones in the involved limb [3, 6, 15], but our patient showed no evidence of limb deformity or gigantism, or true birthmarks on either leg. There were, however, osteolytic lesions in the margin of medial tibial plateau on X-ray, and tropic changes were observed secondary to venous stasis as reported by Servelle et al. [15].

Capillary malformations are the most common cutaneous manifestation of KTS [3]. It typically involves the enlarged limbs, but skin changes may be seen on other parts of the body [12]. Varicose veins are present in the majority of the patients with KTS [13]. The lower extremity varicosities tend to be more prominent laterally [16]. An enlarged tortuous, superficial venous trunk can be identified in the affected leg laterally above the knee and the thigh [11, 13]. This represents the abnormal persistence of an embryonic lateral vein, which normally regresses at 8 weeks of gestation [17].

Our patient showed low-flow venous malformation along the whole length of the left thigh, calf, and short saphenous vein on duplex scan. Several small varicose veins were present in the both postero and lateral aspect of the left thigh extending to the calf and foot. Innumerable abnormal vessels coursing throughout the region of the knee in an intra and extra articular distribution on MRI suggested arteriovenous malformation.

However, despite all these vascular abnormalities, the use of two-phase imaging with Tc-99m sulesomab allowed identification



**Figure 4.** Five-hour Tc-99m sulesomab images showing a vascular malformation in the thigh and varicosed vein in the leg (arrowed "A"), and the intense uptake in the left knee due to septic arthritis (arrowed "B").

of the presence of infection, showing that pattern recognition and changing patterns of activity over time still play a significant role in the skills required by the nuclear medicine physician.

## Conclusions

Klippel-Trenaunay syndrome is a rare condition, which can have a direct appearance on both the blood pool phase of the bone scan and Tc-99m sulesomab imaging. It is still possible, despite these abnormalities, to identify co-existing infection with the use of dual-phase Tc-99m sulesomab imaging.

## Acknowledgements

Dr S. Begum was supported by the British Council and the International Atomic Energy Agency.

## References

- Gloviczki P, Stanson AW, Stickler GB et al. Klippel-Trenaunay syndrome: the risks and benefits of vascular interventions. *Surg* 1991; 110: 469–479.
- Cohen MM Jr. Klippel-Trenaunay Syndrome. *Am J Med Genet* 2000; 93: 171–175.
- Jacob AG, Dricoll DJ, Shaughnessy WJ, Stanson AW, Clay RP, Gloviczki P. Klippel-Trenaunay syndrome; spectrum and management. *Mayo Clinic Proc* 1998; 73: 28–36.
- Klippel M, Trenaunay P. Du noevus variqueux osteo-hypertrophique. *Arch Gen Med (Paris)* 1900; 185: 641–672.
- Szilagyi DE, Smith RF, Elliott JP, Hageman JH. Congenital arteriovenous anomalies of the limbs. *Arch Surg* 1976; 111: 423–429.
- Lindenauer SM. The Klippel-Trenaunay syndrome: varicocity, hypertrophy and hemangioma with no arteriovenous fistula. *Ann Surg* 1965; 162: 303–313.
- Gloviczki P, Hollier LH, Telander RL, Kaufman B, Bianco AJ, Stickler GB. Surgical implications of Klippel-Trenaunay syndrome. *Ann Surg* 1983; 197: 353–362.
- Noel AA, Gloviczki P, Cherry KJ, Rooke TW, Stanson AW, Driscoll DJ. I surgical treatment of venous malformations in Klippel-Trenaunay syndrome. *J Vasc Surg* 2000; 32: 840–847.
- Bakersville PA, Ackroyd JS, Browse NL. The etiology of Klippel-Trenaunay syndrome. *Ann Surg* 1985; 202: 624–627.
- Taybi H. Klippel Trenaunay syndrome. In: Taybi H, Lachman RS (eds). *Radiology of syndromes, metabolic disorders and skeletal dysplasias*, 4<sup>th</sup> Ed. Mosby 1996 St. Louis, Missouri USA; 276–279.
- Roebuck DJ, Howlett DC, Farzer CK. Pictorial review; the imaging features of lower limb Klippel-Trenaunay syndrome. *Clin Radiol* 1994; 49: 346–350.
- Berry SA, Peterson C, Mize W, Zachary C, Blasco P, Hunter D. Klippel-Trenaunay syndrome. *Am J Med Genet* 1998; 79: 319–326.
- Baskerville PA, Ackroyd JS, Lea Thomas M, Browse NL. The Klippel-Trenaunay syndrome: clinical radiological, and haemodynamic features and management. *Br J Surg* 1985; 72: 232–236.
- Dohil MA, Baugh WP, Eichenfield LF. Vascular and pigmented birthmarks. *Pediatr Clin North Am* 2000; 47: 783–812.
- Servelle M. Klippel-Trenaunay's syndrome — 768 operated cases. *Ann Surg* 1985; 201: 365–373.
- Sooriakumaran S, Landham TL. The Klippel-Trenaunay syndrome. *J Bone Joint Surg Br* 1991; 73: 169–170.
- Meine JG, Schwartz RA, Jannigerck. Klippel-Trenaunay Weber syndrome. *Cutis* 1997; 60: 127–132.