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## “Klippel: Trenaunay Syndrome ( KTS) & It’s Management”

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### ABSTRACT:

Klippel–Trenaunay Syndrome(KTS) is a congenital disorder with a rare incidence to affect at least 3-5 in 100,000 people worldwide. Alternative names given for Klippel-Trenaunay Syndrome are Klippel-Trenaunay-Weber syndrome. It is a rare sporadic disease characterized by clinical triad of capillary malformation; soft tissue and bony hypertrophy; and atypical varicosity. Symptoms of Klippel-Trenaunay Syndrome (KTS) include pain, swelling, lymphedema, bleeding, superficial thrombophlebitis, and deep vein thrombosis. This type of syndrome was first described more than hundred years ago, exact incidence has not been estimated yet. We report a case of a 21-year-old Male who presented to us with the symptoms of varicose plaques over left lower limbs & was diagnosed as a case of KTS. CT-Angiography IV contrast Vascular Doppler Ultrasound, skeletal X-ray, and magnetic resonance imaging of the Limb indicated a Klippel–Trenaunay disease.

**KEY WORDS:** Klippel-Trenaunay Syndrome, Varicose veins, Capillary Malformations, compression stockings.

## 1.INTRODUCTION

Klippel–Trenaunay Syndrome (KTS) is a congenital disorder with a rare incidence of 3-5/100,000. It is a rare sporadic disease characterized by clinical triad of capillary malformation; soft tissue and bony hypertrophy; and atypical varicosity[1]. KTS is now termed capillary-lymphatic-Venous malformation (CLVM). This disease is subject to significant morbidities such as bleeding, deep vein thrombosis, and embolic complications. Other authors prefer to separate Klippel–Trenaunay–Weber Syndrome (KTWS) with the usage of the term Parkes–Weber Syndrome to describe the condition in those patients who have true arteriovenous malformations in addition to KTS [2]. This article describes a case study of a young presenting with Klippel–Trenaunay including a review of the syndrome and treatment recommendations.

## 2.CASE REPORT

A 21 year's old young born out with small stain on superficial to thigh since birth. When age of this patient will be increased, this stain enlarged progressively. He reported that the dilatation over the limb increased during walking and standing. Superficial varicosities noted in Lt leg and on the posterior aspect of thigh. Deep varicosities noted in vastus lateralis and lateral head of gastrocnemius muscle.

There are tubular vascular channels extending to the left lower limb (anterior, posterior and lateral aspect) variably affecting the thigh, calf extending to the foot along with intrapelvic extension predominantly in the prevesical, perirectal spaces and the left lateral pelvic wall.

Altered density areas are also seen in the left scrotum along with multiple foci of calcification/phleboliths. Similar multiple phleboliths are also seen in the pelvis.

In the left lower limb, the lesion is predominantly subcutaneous in location with extension into the intermuscular fascial plane as well as intraarticular extension into the left knee joint. Associated intramuscular extension is seen in the left gluteus maximus, left vastus lateralis, the lateral gastrocnemius and minimally in the soleus. These muscle show fatty streaks / fatty atrophy within. The lesion exhibits multiple enhancing as well as non-enhancing areas. Probable drainage is seen into the left common iliac, external as well as internal iliac veins which appear dilated. Lymphatic / venous edema is seen in the perirectal, presacral, left lateral wall in the pelvis. Tortuous non-enhancing channels are also seen in the posterior gluteal fat adjacent to the gluteal cleft bilaterally.



**Figure 3:** (a) T1-weighted coronal image, (b) T2-weighted coronal image, (c) short-tau inversion recovery coronal image diffuse irregular tortuous channels in the subcutaneous plane involving the entire thigh appearing hypointense on T1, hyperintense on T2-weighted and short-tau inversion recovery images with flow voids suggestive of venous collaterals. Furthermore, hypertrophy of muscle of both compartments noted

### 3.DISCUSSION

KTS was described by two French physicians, Klippel and Trenaunay in 1900.[3] It is a triad of capillary malformation, venous/lymphatic varicosity with soft tissue, and bony hypertrophy. Some authors use the term KTWS to describe the conditions affecting those individuals who have significant arteriovenous malformations as one of the component of their KTS[4].

### 4.CLINICAL MANIFESTATION SPECTRUM

The severity of KTS can be classified based on the type of dysplasia of the blood vessels involved[14]: (1) venous dysplasia; (2) arterial dysplasia; (3) arterial and associated venous dysplasia (a) with no

AV-shunt and (b) with AV-shunt; and (4) mixed angiodysplasias (a rare form of KTS).

### 5. CAPILLARY MALFORMATION

This is the most common cutaneous presentation of KTS. Usually, capillary malformations are seen in the hypertrophied limb, though dermatological changes can be noted in any location of the body. The most common site of capillary malformations is the lower limb and is usually seen in around 95% cases. The dermatological vascular malformation is usually present as a flat, red or purple colored capillary hemangioma. The overlying skin is usually hard, dry, thickened, and hyperpigmented.[5]

## 6.HYPERTROPHY

Hypertrophy is the most variable of the three classic features of KTS. Enlargement of the extremity can be either bone elongation, circumferential softtissue hypertrophy, or both. On clinical examination, hypertrophy often manifests as a leg-length discrepancy, although any limb may be affected. Significant limb length discrepancy, defined as that amount that would necessitate orthopedic intervention, is relatively uncommon, occurring in only 14% of patients in one study.[4]

## 7.VARICOSE VEINS

The majority of KTS patients have varicose veins[6]. Both the superficial and deep venous systems are capable of developing venous malformations. The majority of superficial venous abnormalities start with the ectasia of tiny veins and can either be massive venous malformations or persisting embryologic veins [7]. Aneurysmal dilatation, aplasia, hypoplasia, duplications, or venous incompetence are all examples of deep venous disorders. The pathologic process of the vascular system is associated to complications. Stasis dermatitis, thrombophlebitis, and cellulitis are among the problems, and thrombosis, coagulopathy, pulmonary embolism, congestive heart failure, and bleeding from aberrant arteries in the kidney, genitalia, stomach, and lungs are among the more serious ones.

## 8.OTHER ORGAN ASSOCIATION

The central nervous system, genitourinary tract, and gastrointestinal tract are the many organs involved. In all the complications of different visceral vascular malformations, bleeding from the rectum and bladder is a life-threatening emergency condition with a documented prevalence of 1%. [15] The various clinical signs and symptoms include:

8.1.Gastrointestinal tract:Disseminated intravascular consumptive coagulopathy and bleeding that progresses from slight or covert bleeding to severe, life-threatening haemorrhages are both involved (DIC). The distal colon and rectum are the most common sites of involvement, and esophageal variceal bleeding has also been reported[16]. Splenic hemangiomas are a sign of splenic involvement, according to [17].

8.2.Genitourinary tracts:Usually, it gets involved in really grave situations. Renal hypertrophy and recurring, painless, gross hematuria are its defining features. [18].

**8.3.Skeletal manifestation:**These are frequently caused by variations in leg length, and a few of them include ipsilateral hip dislocations and scoliosis[19]. Polydactyly,[20] oligodactyly, macrocephaly, blue nevi, pulmonary vein varicosities, cerebral aneurysm,[21] and pulmonary embolism are just a few of the developmental anomalies that are frequently present[22].



## 9. DIAGNOSIS

Diagnosis of KTS is mainly clinical. Investigations in KTS should focus on evaluation of the type, extent and severity of the malformation, and on confirming the absence of any clinically significant arteriovenous shunting. Examination of limb and pelvic lesions is complemented by detailed colour duplex scanning of the venous system of the leg to establish patency, incompetence, thrombosis, arteriovenous shunting and any anomalies such as hypoplasia. Plain X-rays of the long bones (scanograms) are most helpful to measure bone length. MRI is helpful in differentiating bone, fat, muscle hypertrophy and lymphedema. When ablation of a dilated superficial embryonic vein is intended, CT- Angiography IV-Contrast, venography, and computed tomography scans are necessary to evaluate the deep venous system and collateral [8] There are tubular vascular channels extending to the left lower limb (anterior, posterior and lateral aspect) variably affecting the thigh, calf extending to the foot along with intrapelvic extension predominantly in the prevesical, perirectal spaces and the left lateral pelvic wall.

Altered density areas are also seen in the left scrotum along with multiple foci of Calcification/phleboliths. Similar multiple phleboliths are also seen in the pelvis. In the left lower limb, the lesion is predominantly subcutaneous in location with extension into the intermuscular fascial plane as well as intraarticular extension into the left knee joint. Associated intramuscular extension is seen in the left gluteus maximus, left vastus lateralis, the lateral gastrocnemius and minimally in the soleus. These muscle show fatty streaks / fatty atrophy within. The lesion exhibits multiple enhancing as well as non-enhancing areas. Probable drainage is seen into the left common iliac, external as well as internal iliac veins which appear dilated. Lymphatic / venous edema is seen in the perirectal, presacral, left lateral wall in the pelvis. Tortuous non-enhancing channels are also seen in the posterior gluteal fat

adjacent to the gluteal cleft bilaterally.



**Fig. CT- Angiography Film**

## 10.MANAGEMENT

Patients with KTS should be monitored at least annually and more frequently if clinical symptoms are present. If there is a progression of disease, imaging studies should be done, and proper intervention carried out if indicated.

A multidisciplinary approach to treatment and prevention of possible complications of KTS including pediatrician, internist, phlebologist, pediatric, Orthopedic, plastic and vascular surgeons, an interventional radiologist, cardiologist or vascular internist and a physical therapy physician provides optimal care for the patient.

## 11.SURGICAL MANAGEMENT

Surgery is typically saved for cases with symptoms. Pre-operative evaluation of the deep venous system utilising methods like computed tomography (CT) arteriography, duplex scanning contrast angiography, which can help determine the degree of vascular involvement, and the presence of arteriovenous fistulae are essential for the outcome of surgery. Numerous writers have also documented symptoms getting worse after surgeries like repeated ligations and stripping [9].

## 12.LESSOR TREATMENT & RADIOTHERAPY

Laser therapy is frequently used to cure port wine stains [10–11]. Ulceration instances may potentially benefit from laser therapy. Lesions that are raised or under the skin typically do not react well to laser therapy [11]. Patients with KTS can also employ endovenous laser therapy on their larger saphenous veins to treat varicosities.

In some cases of KTS, radiotherapy has been observed to be beneficial; the radiation may aid to cause hemangiomas to retreat. The consequences, however, typically take time to manifest [12].

## 13.PAIN MANAGEMENT

Up to 88% of patients with KTS report having some level of pain [13]. As a result, KTS patients commonly need pain management. The treatment of pain in these individuals uses causal treatment because the management concepts are based on the cause of the pain [13]. Compression stockings or surgery are used to treat pain caused by chronic venous insufficiency. Recurrent cellulitis-related pain is controlled by maintaining strict hygiene, using antibiotics and analgesics sparingly, and using analgesics as needed [13]. Surgical intervention is necessary to treat pain brought on by vascular abnormalities. Long-term treatment with the proper analgesics, such as opiates, may be used when the lesions interosseous cannot be eradicated [13].The vast majority of the time, traditional painkillers are ineffective at treating neuropathic pain. Anticonvulsants and antidepressants may be used in such circumstances. A multi-drug regimen

may be required, and it might even be decided that a brief course of steroids is essential [13].

## 14.CONSERVATIVE MANAGEMENT

The symptomatic management of KTS should take a conservative approach and make use of non-invasive techniques. Most KTS patients are encouraged to wear compression stockings, whether they are elastic or not. The most successful method for treating individuals with KTS has been determined to be the combination of elastic stockings and psychological support [23,24]. Along with additional conservative treatments including regular leg elevation, physiotherapy, lifestyle changes, and upkeep of rigorous hygiene, these



stockings are worn. Cellulitis and thrombophlebitis are frequently treated with analgesics, antibiotics, and corticosteroids. Anticoagulants may be acute thrombosis [25, 26].



employed either as a preventative measure before to surgery or in situations of

ineffective for treating larger deformities. [27] Due to the extremely high recurrence rate, vascular surgeries to treat venous insufficiency are typically ineffective. It is useless to ligate and remove huge venous lakes. Either ultrasound-guided foam sclerotherapy (USFS) or steam vein sclerosis (SVS) followed by foam sclerotherapy are less invasive and hence superior treatments. The USFS has been promoted as a very successful, minimally invasive, and painful ambulatory procedure that has produced outstanding results in KTS patients. [28]

## 16.CONCLUSION

Vascular abnormalities, soft tissue hypertrophy, and bone hypertrophy are the hallmarks of the extremely rare congenital syndrome known as KTS. The precise incidence and pathophysiology are unknown. Only by clinical means is a diagnosis made. The conservative management offers lifetime assistance. An adequate interdisciplinary approach to care is required since sickness affects several organs.

## 15.SCLEROTHERAPY

Although it is an excellent alternative for treating tiny defects, conventional sclerotherapy is



## 17. REFERENCE

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