Clinical History:

The patient came with complaint of swelling and pigmentation of left lower limb. On local examination, varicose veins were found on the left lower limb on the lateral side. Brodie Trendelenburg showed saphenofemoral junction competence and Pratts test showed competent deep venous system.

Imaging Findings:

CT-angiography and venography of both lower limbs was done with continuous 1 mm axial sections. CT-scan from D10 vertebra up to ankle joint immediately after contrast injection. Special reformation CT-angiography of abdominal aorta, aortic bifurcation, iliac, femoral and popliteal artery was done. Multiple venous collateral channels were noted in the left lower limb, i.e. left thigh, left leg and foot. They were arising from the left common femoral vein with an average diameter of around 10 mm, running in lateral aspect of the left lower limb in subcutaneous plane and showing communication with multiple collaterals in the left lower limb. There was thickened subcutaneous tissue with muscle hypertrophy involving the left lower limb. Left common iliac veins, left external iliac veins, left common femoral veins, left popliteal veins, left anterior and posterior tibial veins appeared normal with good contrast opacification.

Discussion:

A. Background: Klippel-Trenaunay syndrome is a congenital vascular anomaly with cutaneous haemangiomas, limb hypertrophy (both soft tissue and bony components) and varicose veins as its pathognomic findings. Neither its genetic predisposition nor its exact incidence is known. There are various postulates which explain its pathogenesis. As per Servelle theory, varicosities and tissue hypertrophy is due to venous obstruction. Angiopoetin-2 antagonism which regulates vascular remodelling has also been implicated. At molecular level, VG5Q expression which regulates angiogenesis may also be involved in the pathogenesis.[2]

B. Clinical perspective: The classic triad includes port wine stain (nevus flammeus), bone and soft tissue overgrowth and varicosities. One must also keep in mind complications such as deep vein thrombosis which presents as painful swollen limbs and also pulmonary thromboembolism which presents as breathlessness.

C. Imaging perspective: The goal of imaging is to look for any arteriovenous shunts and to assess severity and extent of malformations. Colour duplex scan of lower limbs is done to evaluate the venous system and rule out any complications.
(thrombosis, shunts, insufficiency). Bone overgrowth, as indicated by bone length, is measured using plain X-rays and CT scannograms. Abdominal ultrasound and CT are done to look for visceral involvement and evaluate venous collaterals which is helpful in surgery planning. Angiography reveals lower leg superficial varicosities with its pathognomic "marginal vein of Servelle" noted on the lateral aspect of thigh and calf. However, the final diagnosis is still purely clinical, done based on its pathognomic findings.

D. Outcome:
Management of Klippel-Trenaunay syndrome is largely conservative. Surgery is indicated for cases which have complications such as DVT and cellulitis [3]. Compression garments are helpful for managing lymphoedema and venous insufficiency.

E. Conclusion:
The manifestations of KTS are highly variable [1]. Its close differential is Parkes-Weber syndrome which has significant arteriovenous shunting. Imaging is not helpful for diagnosing but instead for planning management and detecting complications. Management is mostly non-surgical.

**Differential Diagnosis List:** Klippel-Trenaunay syndrome without any complications, Parkes-Weber syndrome, Proteus syndrome

**Final Diagnosis:** Klippel-Trenaunay syndrome without any complications

**References:**
Description: Dilated vein noted in lateral aspect of left thigh extending distally communicating with iliac vein. Origin: Viral P
Description: 3D reconstruction Origin: Viral P
Description: Left lower limb appears swollen with areas of portwine stain on it. Origin: Chaitnya P, New civil Hospital, surat
Figure 3

a

Description: Left limb appears swollen with dilated venous channels with muscle hypertrophy. Origin: Chaitnya P, New civil Hospital, Surat

b

Description: Left limb appears swollen with dilated venous channels with muscle hypertrophy. Origin: Chaitnya P, New civil Hospital, Surat
Description: Left limb appears swollen with dilated venous channels with muscle hypertrophy. Origin: Chaitnya P, New civil Hospital, Surat