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# Klippel-Trenaunay Syndrome: A Case Report.

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**ABSTRACT**

Klippel-Trenaunay syndrome (KTS) is a rare congenital syndrome of vascular malformations and soft tissue and bone hypertrophy. Vascular malformations can affect multiple organ systems. Here we present a case of a four and half year old male child with gradually progressive marked right upper limb and ipsilateral chest wall swelling and large varicosities.

**Key words:** Port-wine stain, varicose veins and Gigantism.

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## INTRODUCTION:

In 1900, French physicians Klippel and Trenaunay first described a syndrome[1] in 2 patients presenting with a port-wine stain and varicosities of an extremity associated with hypertrophy of the affected limb's bony and soft tissue. They termed the syndrome naevus vasculosus osteohypertrophicus. In 1907, Parkes Weber[2], unaware of Klippel and Trenaunay's report, described a patient with the 3 aforementioned symptoms as well as an arteriovenous malformation of the affected extremity. He termed the process hemangiectatic hypertrophy. However, in our case, there was no such association of arteriovenous fistula.

## CASE REPORT:

A four and half year old male child presented with gradually progressive marked right upper limb and ipsilateral chest wall swelling (Fig.1) and large varicosities. On clinical examination the swelling was leathery hard and there was minimal tenderness over the swelling. Left upper limb appeared to be normal. The patient was later subjected to radiological and hematological investigations. Hematological investigations were within normal limits. CT Angiography was performed and showed overgrowths of soft tissue of right upper limb, extending up to involvement of the lateral thoracic wall. Multiple dilated varicose veins were noted in the right upper limb and lateral thoracic wall involving the superficial venous system (Fig2). Arterial system appeared to be normal and no evidence of arteriovenous malformation was seen. Multiple small and large, well-defined cystic areas were seen in the right upper limb as well as along the chest wall, probably representing dilated lymphatic spaces, lymphoceles (Fig 3). No underlying bony abnormality was seen.

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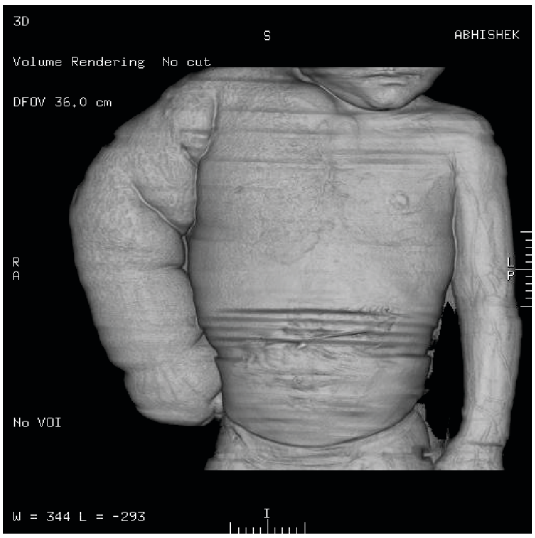


**Fig.1-** Volume rendered 3D image showing marked right upper limb and ipsilateral chest wall swelling.

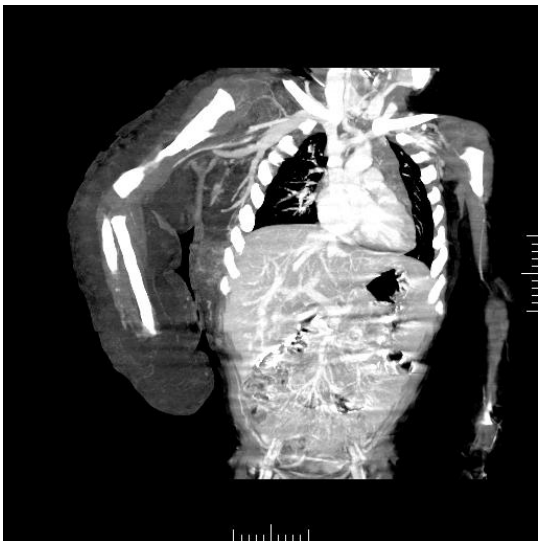

**Fig.2** CT Angiography coronal reformatted image showing overgrowth of soft tissue of right upper limb extending up to involve the lateral thoracic wall and multiple dilated varicose veins in the right upper limb and lateral thoracic wall involving the superficial venous system.

DISCUSSION:

Klippel-Trénaunay syndrome consists of an enlarged extremity with associated cutaneous vascular lesions and underlying diffuse venous and lymphatic malformations. Although Klippel-Trénaunay syndrome generally involves only one of the lower extremities, bilateral involvement, upper extremity involvement, or extension into the trunk may occur.[7] Klippel-Trénaunay syndrome must be distinguished from Parkes-Weber syndrome, in which an enlarged extremity occurs that, is related to an underlying arteriovenous malformation.

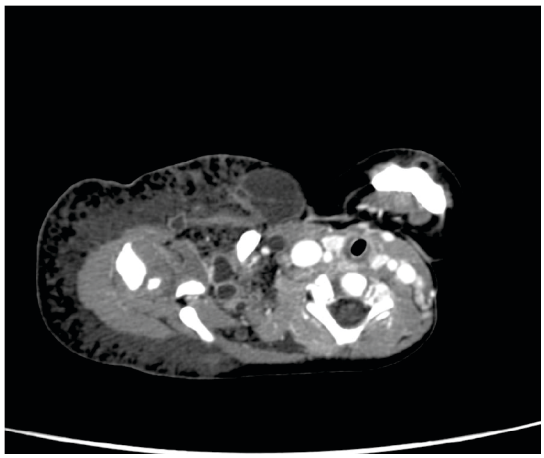

**Fig.3 -** CT Angiography, axial image showing multiple small and large well-defined cystic areas in the right upper limb as well as along the chest wall representing dilated lymphatic spaces, lymphoceles.

In Klippel-Trénaunay syndrome, enlargement of the extremity consists of bone elongation or circumferential soft-tissue hypertrophy.[9,10] The latter may be evident at birth or may become evident as the patient grows.[5] The cutaneous vascular lesion is generally a capillary malformation and usually involves the enlarged limb, although involvement of the whole side of the body or of the contralateral limb may be seen.[9,10] Prominent superficial varicose veins are present in a majority of patients with Klippel-Trénaunay syndrome.[7,9,10] Extremity pain, spontaneous cutaneous hemorrhage, chronic venous insufficiency,

or thrombophlebitis is commonly encountered.[7,8] Less commonly, deep venous thrombosis or pulmonary embolism may result.[7] Clinical sequelae of the lymphatic component of the syndrome include lymphangitis, cutaneous lymphatic vesicles, lymphorrhea, or mass effect from macrocystic portions of lymphatic malformations. Clinical findings of high flow rates (pulsatility, thrill, bruit) are absent in Klippel-Trénaunay syndrome.

At radiography, bone elongation contributing to leg length discrepancy, soft-tissue thickening, or calcified phleboliths may be seen. Venography usually demonstrates extensive dilation of superficial veins and enlarged perforating veins communicating with the deep venous system. In some patients, segmental absence or hypoplasia of the deep venous system is seen and must be distinguished from incomplete filling with contrast material at venography.[7] At lymphangiography, hypoplasia of the lymphatic system has been reported.[8] CT images demonstrate a lack of enlarged high-flow arterial structures, along with malformed venous and lymphatic lesions. CT venography can provide a global picture of the superficial varicosities, enlarged perforating veins, and absent or hypoplastic deep veins characteristic of Klippel-Trénaunay syndrome[7].

Treatment in a majority of patients with Klippel-Trénaunay syndrome is conservative and includes application of graded compressive stockings or pneumatic compression devices to the enlarged extremity. Percutaneous sclerosis of localized venous malformations or superficial venous varicosites may be indicated in some patients.[5] Surgical treatment may include epiphysiodesis to control leg length discrepancy, excision of soft-tissue hypertrophy, stripping of superficial varicose veins, or, less commonly, reconstructive surgery at sites of deep venous obstruction.[8,9,10] Care must be taken because the condition of patients with Klippel-Trénaunay syndrome may worsen if intervention is performed on dilated superficial collateral veins associated with deep vein hypoplasia.[6,7]

The constellation of extremity soft-tissue hypertrophy, combined diffuse venous and lymphatic malformations, and a cutaneous vascular lesion is characteristic of Klippel-Trénaunay syndrome. In addition to these findings, this case displays a marked interval progression of soft-tissue hypertrophy, which may occur with the growth of the patient. Lifelong clinical follow-up is mandatory in these patients because the natural history of venous and lymphatic malformations is one of progressive enlargement.

## CONCLUSION:

In our patient, the triad of marked enlargement of the right upper extremity and chest wall and findings of extensive combined venous and lymphatic malformations allowed the diagnosis of Klippel-Trénaunay syndrome.

Conflicts of interest: No.

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