

Klippel - Trenaunay Syndrome - A Case Report

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1. Introduction

- Klippel- Trenaunay Syndrome (KTS) is a rare congenital anomaly characterized by a triad of varicose veins, port-wine stain, and hypertrophy of bone and soft tissues.
- It is considered as angio-osteo-hypertrophic syndrome.
- The incidence of KTS is 3-5 /1,00,000 population¹.

KTS classically comprises a triad of:

- Port wine nevi
- Bony or soft tissue hypertrophy of an extremity (localized gigantism)
- Varicose veins or venous malformations of unusual distribution
- The diagnosis of KTS is usually made when any two of the three features are present. Capillary malformations may be absent in the atypical form.

2. Case Report

- 3 year 9month old boy born of non-consanguineous marriage presented with skin lesions over the right leg.
- On examination, he had large encrusted hemangioma over right anterolateral aspect of thigh (5 x 4cm), one on lateral aspect of leg (2x3cm) and multiple small hemangiomas over the right leg (Fig no 1).
- Varicosities and hypertrophy of right lower limb was also noted. Contrast MRI right lower limb showed low flow vascular cutaneous malformation (Fig no 2)
- MRI angiography showed superficial varicoid drainage without involvement of deep venous system. Surgical excision of hemangioma was done followed by oral propranolol therapy.



Figure 1: Hemangiomas at presentation

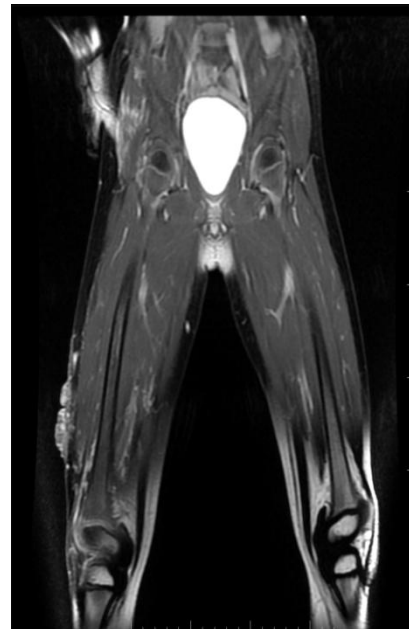


Figure 2: MRI showing hemangioma



Figure 3: Post surgical excision

3. Discussion

- Klippel-Trenaunay Syndrome, first described in 1900 by two French doctors, Klippel and Trenaunay, is a triad of vascular malformation, venous/lymphatic varicosity and hypertrophy of soft tissue and bones.
- The exact aetiology of KTS is unknown. It is generally a sporadic disorder, although paradominant inheritance pattern has been suggested.

Volume 9 Issue 2, February 2020

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- lower limb is the most common affected side and generally follows a benign course.
- Complications due to hemangioma include ulceration, bleeding, and secondary infection.
- Complications of varicosities include paresthesia, ulcers, dermatitis, pulmonary embolism, thrombophlebitis, haemorrhage and cellulitis.
- Hypertrophy of a limb may lead to vertebral scoliosis and gait abnormalities. It can also cause degenerative joint disease.
- Treatment includes either surgical excision or laser treatment of hemangioma. Currently, the flashlamp-pumped pulsed dye laser is the treatment of choice in vascular lesions.
- Sclerotherapy/vein ligation or stripping, and compression therapy for chronic venous insufficiency.
- Regarding limb hypertrophy, heel inserts are generally sufficient for limb length discrepancies of 1.5 cm or less. If projected leg length discrepancy exceeds 2.0 cm at skeletal maturity, it can be treated by epiphysiodesis in the growing child.

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