

## Picture Story

# Klippel Trenaunay syndrome

K Jagadish Kumar<sup>1</sup>, Abhishek Chavan<sup>2</sup>, Mitun Jain<sup>2</sup>, V G Manjunath<sup>3</sup>, Perumal Satya<sup>2</sup>

*Sri Lanka Journal of Child Health*, 2012; 41(4): 209-210

(Key words: Klippel Trenaunay syndrome; KTS; girl)

### Introduction

Klippel Trenaunay syndrome (KTS) is a non-heritable rare disorder characterized by the triad of macular vascular naevus (port-wine naevus), bony and soft tissue hypertrophy and venous varicosities<sup>1</sup>. Incidence of KTS is about 2-5 per 100,000<sup>2</sup>. The vascular malformation is usually limited to a single extremity, though multiple extremities can be involved<sup>2</sup>. Legs are more often affected than arms<sup>2</sup>. It affects males more often than females<sup>2</sup>.

### Case report

A 12 year old developmentally normal girl presented with asymptomatic reddish skin lesions over right upper limb since birth. Dermatological examination revealed port wine stain over the entire right upper limb and the upper chest wall (Figures 1 and 2). Prominent superficial and engorged veins were also seen over the right axilla (Figure 2).

Physical examination revealed hypertrophy of the whole right upper limb. The circumference of right arm was 2 cm more than the left arm and the right forearm 1.5 cm more than the left forearm. She had a limb length discrepancy with the right upper limb being 1 cm longer than the left. There was no hyperthermia or bruit over the right upper limb and Branham sign was negative. The other limbs were normal. She had no macroglossia, abdominal wall defects, ear lobe creases or café-au-lait macules. Ophthalmological and systemic examinations were normal. Her intelligence quotient was normal. Her platelet count and chest x-ray were normal. Doppler ultrasound scan of the right upper limb revealed normal anatomy and flow pattern in the arteries,

<sup>1</sup>Professor of Paediatrics, <sup>2</sup>Resident in Paediatrics, <sup>3</sup>Associate Professor of Paediatrics, JSS Medical College, JSS University, Mysore, India

(Received on 12 December 2011; Accepted after revision on 27 January 2012)

increased flow velocity in the veins of the hand and dilated subcutaneous veins. There were no arteriovenous malformations. CT scan of the thorax was normal.



**Figure 1:** Port wine naevus over right upper limb and upper chest wall



**Figure 2:** Hypertrophy of the R upper limb with port wine naevus and varicose veins in the R axilla

## Discussion

Most patients with KTS demonstrate all 3 signs of the clinical syndrome viz. port-wine stain, varicose veins, and bony and soft tissue hypertrophies. In a series of 252 patients at the Mayo Clinic, 63% had all 3 features and 37% had 2 of the 3 features. Port-wine stain was seen in 98% of patients, varicosities or venous malformations in 72% and limb hypertrophy in 67%<sup>1</sup>. In KTS the capillary haemangioma or port-wine stain usually presents first and is typically of the naevus-flammeus type but cavernous haemangiomas or lymphangiomas may also occur<sup>1</sup>. Unlike strawberry haemangiomas, the port-wine stain haemangioma possesses neither a proliferative nor a regressing phase. Varicose veins in KTS are congenital<sup>1</sup>. Hypertrophy is mostly related to soft tissue and fat overgrowth, although bony hypertrophy may be present. Increased length of limbs implies bony hypertrophy, while increased girth implies soft-tissue overgrowth<sup>2</sup>. Local hyperaemia and augmented

arterial flow related to the vascular abnormalities are postulated to be the cause for overgrowth<sup>2</sup>. In Parkes Weber syndrome, the same triad is present as in KTS but the limb hypertrophy is caused by multiple arteriovenous fistulae and hence a continuous bruit may be present. Application of a tourniquet often results in bradycardia (Branham sign)<sup>2</sup>.

## References

1. Janniger C K. Klippel-Trenaunay-Weber Syndrome. Medscape Updated: May 27, 2011. Available from: <http://emedicine.medscape.com/article/1084257>
2. Suchithra G, Madhu.R, Srinivasan. MS. Klippel Trenaunay Syndrome. *e-Journal of the Indian Society of Teledermatology* 2008; **2**; 7-14.