Case Report

Klippel-Trenaunay Syndrome: A case report
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The ORION Medical Journal 2007 May;27:470-471

Introduction
Klippel-Trenaunay Syndrome (KTS) is an eponym denoting a slow-flow, capillary-lymphatic venous malformation in association with soft tissue and/or skeletal overgrowth. The syndrome was described first in 1990 and characterized by the classical triad of capillary malformation, venous varicosities and limb hypertrophy. KTS affects the lower extremity in 95% of patients, the upper extremity in 5% of patients and least commonly the trunk. Superficial varicosities result from incomplete valves and deep venous anomalies. Various form of lymphatic anomalies including lymphedema and macro cysts are present in 50% of affected individuals. Limb overgrowth is obvious at birth and hypertrophy may worsen during childhood. In contrast, Parkes Weber Syndrome (PWS) is defined as a fast-flow lesion consisting of an arterio-venous capillary and venous malformation with hypertrophy. Pain, limb swelling and cellulitis may occur. Thrombophlebitis, dislocation of joints, gangrene of the affected extremity, congestive heart failure, hematuria secondary to urinary tract hemangioma, rectal bleeding from lesions of the gastrointestinal tract, pulmonary lesion and malformation of the lymphatic vessels are infrequent complications. Supportive care includes compression bandage for varicosities; surgical treatment may help carefully selected patients. Leg-length differences should be treated with orthotic devices to prevent the development of spinal deformities, corrective bone surgery may be needed to treat significant leg-length discrepancy. We encountered one patient of such rare nonheritable disorder in a local clinic and later on to Medical College Hospital for further management. We felt interested to publish as a case report as it was a rare disorder.

Case
A new born baby was born with swelling of the left lower limb. Baby was delivered by cesarean section and had breach presentation. He was 35 wks of gestational age and cried immediately after birth. No history of consanguinity was present and similar disease in the family. Mother was not suffering from any other disease during pregnancy. Swelling was gradually increasing in size. After two days of birth the baby was ill looking with temperature of 102°F. There was no other organomegaly. Local examination was done and found whole left lower limb swollen. Skin was more shiny than that of the right. No scar or sinus was present. Consistency of the swelling was soft and nontender. No local temperature was raised.

There was hyperpigmented lesion on the later aspect of the left limb. Length of the left limb was more than the right. Left limb was 25 cm and right 22 cm. Left lower limb circumference was 22 cm but right 9 cm. Varicosity of vein was observed in the left. Movement was restricted on the same side. The baby had no other congenital anomaly. X-ray of the left leg was done and found soft tissue swelling of the entire limb without bony hypertrophy.

Discussion
The risk of KTS in the general population is 1 in 14480. In that respect we encountered this case suddenly in our private practice. No history of consanguity or family incidence is observed like other studies. KTS affects the lower extremity in 95% patients, the upper extremity in 5% of patients and least.

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commonly the trunk. Lower extremity is involved in our case which is similar for the other reports. In our patient bony involvement is absent. Limb hypertrophy in KTS or PWS is secondary to the vascular anomaly located on the affected limb. The lymphatic malformation associated with KTS is known to cause localized soft tissue and skeletal hypertrophy. Some authors consider KTS to be a risk factor for Wilms tumor. Other do not consider KTS and Wilms tumor to be significantly associated. Our case has no such association with Wilms tumor because this case is detected early when it has not developed the tumor. Patients with isolated hemi hypertrophy have an increased risk of developing other cancer (5.9%). Intelligence is usually not impaired in KTS. However, when there are hemangioma on the face (facial hemangiomatosis), there may also be hemangiomas in the brain which can cause seizures (convulsion) or mental retardation, such problems are rare in KTS. Our patient gets no time to evaluate the intelligence status because the baby is very young to comment as well as died at the age of 6 days. The cause of death is probably septicemia and hypovolemia.

Conclusion
It could be mentioned that we found interest to report this case as it was a rare disorder. Although, it was rare, it might be encountered in our practicing life. Immediate meticulous conservative and surgical measures could be of benefit and increased life expectancy of the patient.

References