

Klippel-Trenaunay syndrome in a new born with multiple congenital anomalies

Sir,
Klippel-Trenaunay syndrome (KTS), as originally defined in 1900, is comprised of the triad of a port-wine stain extending the full length of a limb, venous varicosities of the same limb, either congenital or of onset in infancy and overgrowth of all the tissues of the affected limb, particularly bone.^[1] Today, this term is generally used for any case where there is an association of port-wine stain and increased limb size, whether or not bony overgrowth is present and whether or not venous varicosities are apparent.^[2] One or several macular telangiectatic vascular naevi are almost invariably present at birth.^[3] In addition to these main features, a wide range of associated conditions and complications have been described. Other vascular malformations such as angiokeratomas and lesions resembling granuloma telangiectaticum are not uncommonly associated with KTS.^[2]

A 26-year-old mother delivered a female infant by vaginal delivery at full term. There was no history of medication for any systemic disorder taken during pregnancy or of maternal illness. The father of the baby was a 28-year-old male and he was in good health. The family history was unremarkable. Cutaneous examination of the baby, on the second day of birth, revealed a macular, telangiectatic vascular hemangioma of the 'port-wine' variety, pink to purple in color, present in a splotchy distribution over the entire right lower extremity extending over the right gluteal region and onto the medial aspect of the right thigh and partially involving the genitals. The lesion was diffusely spread over the anterolateral aspect of the right lower extremity with only a limited involvement of the medial aspect. The right limb was grossly edematous and hypertrophied and there was a perceptible difference in the limb lengths and mid-calf circumferences [Figure 1]. Multiple, markedly dilated and tortuous venous varicosities were present in and around the right knee joint.

Small brownish-black angiokeratomatous nodules were irregularly distributed over the underlying hemangioma. The right foot was distinctly bigger in size with respect to the left foot. Syndactyly was present in between the second and third, and the fourth and fifth right toes along with bilateral congenital talipes equinus varus deformity. A Doppler study performed showed a normal pattern of venous and arterial blood flow in the affected leg. Ultrasonography of the abdomen and chest X-ray were normal. Hematological profile including platelet count was normal. A diagnosis of Klippel-Trenaunay syndrome with congenital talipes equinus varus with syndactyly and angiokeratomas was made.

The etiology of KTS is unknown. Manifestations of this syndrome begin at birth, or shortly after birth with most of the patients displaying cutaneous hemangiomas. As the child begins to walk, varicosities appear and become more obvious. Limb and soft tissue hypertrophy usually manifests several years later and involves a single lower extremity. In our patient, all the three components, i.e. hemangioma/port-wine stain, varicosity and limb hypertrophy were present at birth



Figure 1: Port-wine stain over the right lower extremity along with angiokeratomas and syndactyly between the second and third, and the fourth and fifth right toes (a marked hypertrophy of affected limb can be noticed)

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which itself is a unique presentation and not reported till date. In addition, but relatively not frequent, congenital orthopedic anomalies such as polydactyly, syndactyly, oligodactyly, compensatory scoliosis^[3] and hip dislocation can occur in some cases. Equinus deformity of the feet as present in our case too, is the rare orthopedic anomaly. Many patients with KTS can be managed conservatively with elastic support garments. The usual agents used for the treatment of hemangiomas, like prednisolone, interferon alpha are of no benefit in KTS. Recently, pulse dye laser therapy and surgical correction to debulk the excessive tissue have been found effective.^[4] Most of the cases of KTS are diagnosed after the different cutaneous changes start manifesting as the child grows. Jalil *et al.*^[5] have recently reported a case of KTS diagnosed at four years of age.

This case highlights the presence of various developmental defects apart from three classical components of KTS, which have been diagnosed just after birth. Awareness of the syndrome and knowledge of its associated anomalies are important for appropriate follow-up and subsequent management.

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