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Klippel - Trenaunay syndrome: Association with absence of ipsilateral testis

Sir,

We wish to report a case of Klippel-Trenaunay Syndrome (KTS) with absence of ipsilateral testis, a hitherto unreported association. The KTS, first described by Klippel and Trenaunay in the year 1900, classically consists of a triad of port-wine stain of limb, varicose veins and hypertrophy of soft tissues and bone of the same side. However, the current definition includes port-wine stain and increased limb size, irrespective of the presence of bony overgrowth and varicosity of veins. Though the specific genetic abnormality causing the disease has not been identified till date, a para-dominant mode of inheritance is speculated.¹ Port-wine stain or macular telangiectatic vascular nevus is the earliest, most common and characteristic clinical lesion. It usually involves the lower limb causing hypertrophy of bone and soft tissue, characterized by an increase in the length and girth.

A 16 year old male presented with multiple erythematous macules and plaques involving posterior aspect of left lower limb and left lower abdomen. The macules were present since birth and the plaques, appearing as masses of soft tissue hypertrophy, were

noticed by him since last 5 years. There was a history of bleeding from the plaques with the slightest trauma. He also had scoliosis and an abnormal gait.

On examination, he was found to have lengthening of the left lower limb and varicosities of veins over the same side (Figure 1). The areas of soft tissue hypertrophy were nontender but there was a history



Figure 1: Port wine stain, dilated veins, soft tissue hypertrophy and scoliosis



Figure 2: Absence of ipsilateral testis



of pain during walking and also profuse hemorrhage after trauma. There was no evidence of entrapment syndromes like trigger finger or carpal-tunnel syndrome. On auscultation over the affected limb, no bruit was audible. However, the left testis was found to be absent (Figure 2). No mass, suggestive of an undescended testis, was palpable in the lower abdomen, inguinal canal or scrotal sac. On ultrasonography of the abdomen and pelvis, no soft tissue shadow was found along the path of descent of the testis.

A plethora of associations have been reported with the KTS. They include hypertrichosis, unilateral enlargement of the tongue, trigger finger and carpal-tunnel syndrome, verrucous epidermal nevi, polydactyly and syndactyly.² It may also be associated with widespread venous abnormalities of the internal organs like the lungs, urinary bladder, kidney, rectum and colon.³ Occasionally, blue naevus and macrocephaly coexist with this condition.

However, absence of the ipsilateral testicle has not been reported with this condition. Any developmental anomaly causing incomplete descent of the testis could not be confirmed because as a rule, by the age of sixteen years, such an incompletely descended testis would have been irreversibly degenerated.⁴

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Pretibial myxedema, ophthalmopathy and acropachy in a male patient with Graves' disease

Sir,

The thyroid gland plays an important role in the maintenance of normal cutaneous function. Five per cent of men and 15% of women with thyroid dysfunction show cutaneous changes.¹ Graves' disease is the most common cause of hyperthyroidism, affecting 15-50 of 100000 people per year, mostly females.² Classical findings include thyroid swelling, exophthalmos, pretibial myxedema and acropachy.² We report a man presenting with all the features.

A 29-year-old man presented with a history of asymptomatic swelling over the legs for the past 2 years. It was gradual in onset and progressive in nature. There was no history of trauma, excessive sweating, heat intolerance or loss of appetite. The patient had undergone thyroidectomy 2½ years back for Graves' disease. Skin examination revealed multiple well-defined plaques and nodules of varying size situated bilaterally over the tibia. The overlying skin was waxy and indurated with some areas of hyperpigmentation (Figure 1). There was clubbing of the nails with thickened periungual skin (Figure 2). The patient also had exophthalmos.

Routine blood and urine examinations were normal. Thyroid function tests showed decreased T3 (0.30 ng/ml) and T4 (2.10 mg/ml) and increased TSH (39.41 mIU/ml) levels, suggestive of hypothyroidism. A skin biopsy specimen from the pretibial skin showed normal epidermis, marked dermal pallor due to mucin deposition and dispersed stellate shaped fibroblasts, findings suggestive of myxedema.

The patient was put on oral thyroxin and received 5 doses of intralesional triamcinolone, 0.5-1 ml per lesion, with marked improvement in the skin lesions. Later, a high potency steroid ointment was used.

