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Klippel Trenaunay Weber Syndrome Associated with Abdominal Hamartoma with Undescended Testis

K. Biswas
A.K. Sarkar
S. Sarkar
B. Mukhopadhyay
S. Ganguly

Klippel Trenaunay Weber Syndrome is a non-heritable disorder consisting of a macular vascular nevus (port-wine nevus) in combination with bony and soft tissue hypertrophy and venous varicosities(l). Though a few cases have been reported(2-4), but to the best of our knowledge, the association of an abdominal hamartoma with undescended testis and appendage in this disorder has not been documented.

Case Report

A $2^{1/2}$-year-old boy born of non-consanguinous parents was admitted to our hospital in October, 1991 with history of progressive enlargement of left lower limb, buttocks, perineum; pink stains at different sites of the left side of the body since birth

From the Departments of Pediatrics, Pediatric Surgery and Pathology, Institute of Postgraduate Medical Education and Research and S.S.K.M. Hospital, Calcutta.

Reprint requests: Dr. Amiya K. Sarkar, Flat A-1/7, Kalindi Housing Estate, Calcutta 700 089.

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and progressive abdominal swelling since one year of age. Birth history, milestones of development and family history were normal. Physical examination revealed a pale, alert child with hypertrophy of the left lower limb, buttocks and perineum. There was extensive portwine staining over the left side of the trunk extending from inframammary region and over the lateral aspect of die left limb *(Fig. 1)*. Varicose veins were present over the medial aspect of the left foot. Left testis was undescended and not palpable. Abdominal examination showed a huge, nontender, irregular and ill-defined retroperitoneal mass extending over right and left lumbar, umbilical and left iliac fossa. It had no mobility and was dull on percussion. Anthropometric measurements confirmed that the left lower limb was bigger than the right (length L:R 39:37 cm, Circumference 3 cm above knee joint L:R 29:17 cm). Skull circumference was 47 cm.

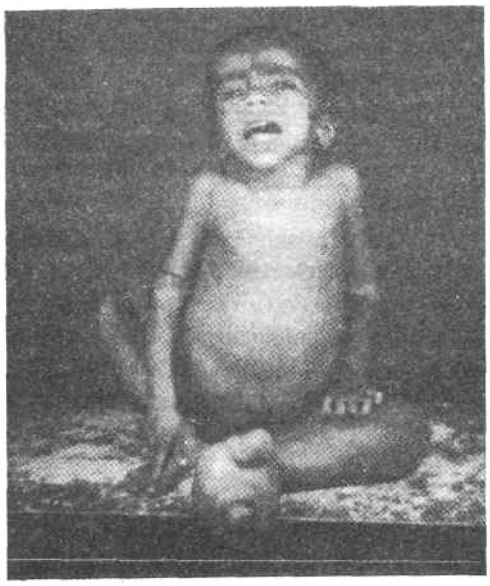

Fig. 1. Extensive portwine staining-left side of the trunk and lateral aspect of left lower limb with hypertrophy of the limb, buttocks and perineum.

Investigations revealed a hemoglobin of 7.5 g/dl, total leucocyte count of 13,000/cumm with polymorphonuclear leucocytosis. Bleeding time, clothing time, absolute platelet count and other biochemical parameters were normal. Skeletal survey showed left femur, tibia and fibula larger than their contralateral counterpart by 7 mm. Soft tissue overgrowth of the left lower limb was more than the right. Abdominal ultrasonography showed a huge septated heterogeneous mass in the pre- and paravertebral regions. Intravenous urography revealed an extrarenal mass with pressure effects on both ureters. Barium meal follow through examination showed a homogeneous mass with extrinsic pressure effects to shift the descending colon upwards and the pelvic colon to the right. CT scan of the abdomen confirmed a huge heterogeneous mass occupying the left lower abdomen and the pelvis. Angiogram showed multiple arterial communications with the tumor mass without any arteriovenous fistula. On laparotomy a firm, encapsulated mass was found in the left iliac fossa, left side of the pelvis extending to the left lumbar region. The mass was retroperitoneal pushing the sigmoid colon to the right and compressing the rectum and urinary bladder from behind. Testis and the *vas deferens* could not be identified separately from the mass during operation. The mass was respected completely keeping the capsule intact. The tumor mass contained necrotic material. Histopathological studies of the mass revealed hamartoma containing many blood vessels, nerve tissue, fatty tissue with degenerative changes and inflammatory cell infiltration *(Fig. 2)*. There was also an undescended testis with epididymis and vas deferens adherent to this hamartoma

(Fig. 3). No tumor was seen within the undescendcd testis.

Discussion

The triad of defects in Klippel Trenaunay Weber syndrome (nevus vasculosus osteohypertrophicus) constitutes nevus flammeus type of hemangioma, vericose veins and hypertrophy of soft tissue and bone. Though usually unilateral,

Fig. 2. Photomicrograph showing a hamartoma composed of blood vessels, fatty tissue and smooth muscle tissue (H&E × 80).

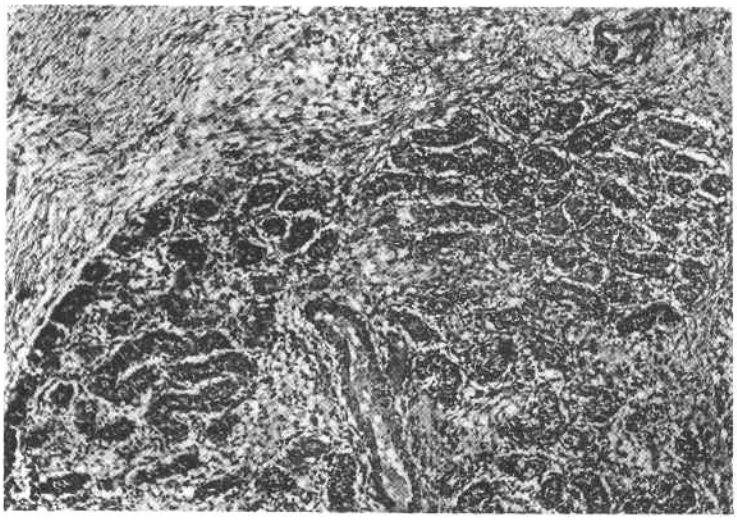

Fig. 3. Photomicrograph showing infantile testicular tissue (H&E × 80).

this anomaly may affect two or all four limbs including a portion of trunk and face. The affected part may be larger at birth or the rapid growth may only gradually become apparent. Legs are more often affected than arms. Rarely there may be atrophy of the limb rather than hypertrophy(2). The cutaneous hemangioma of the nevus flammeus type often has a patchy distribution. In addition to its distribution over the involved lower extremity, it may extend to the buttock and trunk. Infrequently there may be other hemangiomas away from the affected limb. This syndrome appears to be somewhat more commoner in boys(2). Thrombophlebitis, dislocation of joints, congestive cardiac failure, recurrent bouts of cellulitis, gangrene of the affected extremity(l), hematuria secondary to bladder hemangiomas(4), colonic and rectal hemangiomas(2), pulmonary lesions and malformations of the lymphatic vessels are infrequent complications. The syndrome may also be associated with syn/polydactyly(2), megalencephaly, mental retardation(l), Sturge Weber syndrome(5), myotonic dystrophy(6) and portal hypertension(7). One of the features of Klippel Trenaunay Weber syndrome is the presence of hemangiomata. Though the argument as to whether hemangiomas are 'hamartomas' or true 'neoplasms' has not been fully settled, most agree in favor of considering hemangiomas as hamartomas(8). In that case the association between a hamartoma and Klippel Trenaunay syndrome is not exceedingly uncommon. Moreover, undescended testis though not recorded cannot be considered unusual in the present case since a number of congenital anomalies are often associated with this type of disorder. Arteriograms and venograms of the limb may delineate the extent of anomaly. Surgical correction or palliation is often difficult. Swelling may be controlled with elastic garments or wearing of a fitted pneumatic graduated compression lymphatic pump at night(9).

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