KLIPPEL - TRENAUNAY - WEBER SYNDROME (A Case Report)

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Summary

A case of Klippel - Trenaunay - Weber Syndrome in a 13 years old male with involvement of three extremities is reported. A brief review of literature is given.

Klippel-Trenaunay-Weber Syndrome is a rare disease characterized by three major abnormalities namely, (1) Hemangioma of the nevus flammeus type (2) Varicose veins and (3) Hypertrophy of soft tissue and bones[1]. This entity was initially described in 1900 by Klippel and Trenaunay[2] who defined it as a vascular nevus of lower limb with segmental distribution, varicosities in the affected limb and hypertrophy of all tissues on the affected side. Later, Parkes - Weber[3,4] unaware of the earlier publications described similar cases but added another important feature that of arterio - venous fistula. He used the term hemangiectatic hypertrophy for this condition, discussed the whole spectrum of vascular anomalies and local hypertrophy.

Lindenauer[5] considered the above entities as separate ones e.g. syndrome associated without arterio - venous fistula as Klippel - Trenaunay Syndrome and with fistula as Parke - Weber Syndrome. Mullins et al[6] suggested that separating this into different entities has relevance only if it helps in deciding prognosis of the disease or therapy. He recommended that the condition be designated as Klippel - Trenaunay - Weber Syndrome.

Etiology of this condition which is a congenital anomaly remains uncertain. The hypertrophy of soft tissue and bones in patients with arterio-venous fistula can be explained on the basis of the increased blood supply but hypertrophy without A - V fistula can not be explained. Commonly there is involvement of one limb but infrequently involvement is seen bilaterally[6]. Sometimes there is involvement of both limbs on one side and rarely all four extremities may be affected to a variable degree[1]. Hemangioma is of nevus flammeus variety and has patchy distribution. Usually there is hypertrophy of the bones and soft tissues but uncommonly atrophic changes may be seen. Some cases may reveal osteoporotic changes in the involved extremities[7].

Lamer et al[8] have described the association of certain other anomalies like syndactilism and polydactilism. Familial cases of Klippel-Trenaunay-Weber syndrome are known and other transmissible diseases such as ichthyosis have been reported in association with this syndrome[9].

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A case of Klippel-Trenaunay-Weber syndrome with an uncommon presentation involvement of three extremities is reported. To the best of our knowledge, no such case has been reported from India in the past.

Case Report

13 years old male patient, son of a serving soldier reported to skin center, Base Hospital, Lucknow with history of red coloured patches on both lower limbs and left upper limb ; and progressively increasing swelling of affected limbs.

He was born normally. Mother's antenatal period was uneventful. Two other brothers were unaffected. At the age of 20 days patient was diagnosed to have pyloric obstruction which was managed conservatively. Past history was otherwise not significant. His milestones were normal.

At birth parents noticed a red patch on the right buttock which gradually increased and spread to the other limb also. At the age of six months similar cutaneous lesions were noticed on the left upper limb. About the same time, mild swelling appeared on the affected limbs which progressively increased. Examination revealed non-pitting swelling of the affected limbs, more marked in the lower limbs. Extensive areas on both lower limbs, left upper limb, buttocks and scapular region, were covered with nevus flammeus. There was no evidence of varicosity, ulceration, sympathetic overactivity, polydactily or syndactily, in the affected limbs. Systemic examination did not reveal any abnormalities.

Routine blood examination (including bleeding time, clotting time, platelet count), Fundoscopy, X-Ray skull and ECG were normal. X-Ray of affected limbs showed marked hypertrophy of soft tissue whereas no significant changes were observed in bones. Angiographic studies could not be performed due to unwillingness on the part of patient's parents.

Discussion

The Klippel-Trenaunay-Weber syndrome is a rare condition and usually involves a single extremity. Rarely multi-limb involvement can occur. Lamer[5] has reported a case with involvement of all four extremities. Our case had involvement of both lower limbs and one upper limb (Fig. 1). This is

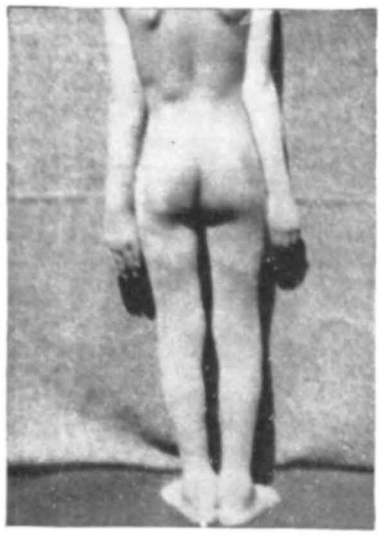

Fig. 1 Showing involvement of one upper limb and both lower limbs.

an uncommon presentation. The affected limbs showed marked hypertrophy of soft tissue as clearly evident in upper extremities (Fig. 2). In the lower limbs the soft tissue hypertrophy was particularly evident, in the lower part (Fig. 3). There was no obvious hypertrophy or atrophy of the skeleton. Hemangiomatous changes (Nevus flammeus) were seen at sites other than the limbs like scapular region.

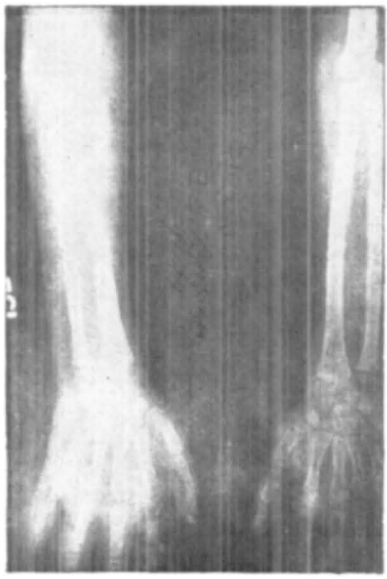

Fig. 2 Marked hypertrophy of soft tissue

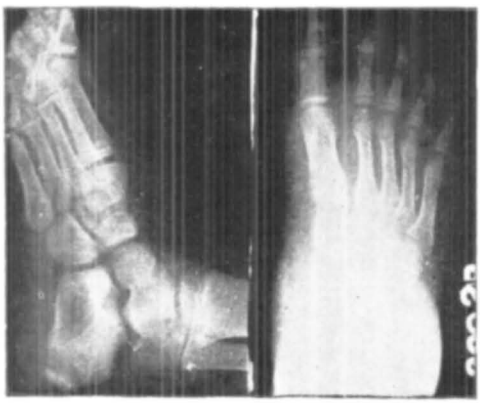

Fig. 3 Soft tissue hypertrophy of foot

Not much stress has been given on the sex distribution of this syndrome in literature. The cases described by Lamer et al[8] (one case) and Mullins et al[6] (two cases) were females. Our case is a male.

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