

UNILATERAL LIMB HYPERTROPHY

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Summary

Klippel-Trenaunay syndrome as a cause of unilateral hypertrophy of limb is described while other possibilities in such a situation are reviewed.

Unilateral hypertrophy of the limb is an intriguing clinical entity. Various etiologies have been mentioned as responsible^{1,2}, Klippel-Trenaunay syndrome being one of the rare ones^{3,4,5}. It is, therefore, important to know fully its classical clinical manifestations. Hence we report a case of unilateral hypertrophy of the left lower limb due to this syndrome.

Case Report

A 19 year old male, reported with progressive enlargement of the left leg since childhood and consequent inconvenience in walking. No other member of the family had similar disease.

Examination of the legs revealed conspicuous hypertrophy of the left leg as compared to the right. The length of the left thigh from anterior superior iliac spine to tibial prominence was 55 cm., while that of the right was 54 cm. The length of the left leg was 40 cm. in contrast to 36 cm. of the right leg. Circumference of the left leg just above the ankle was 24 cm. and on the right it was only 18 cm. Accordingly his height while standing on the left leg was 170 cm. as compared to 145 cm while standing on the right leg.

Skin over the left leg had erythematous, raised eruption of the size of 7.5 cm. × 7.5 cm., Partial blanching could be elicited in these lesions. Varicose veins were also prominent.

On radiological examination a marked contrast in soft tissue shadows of the left and the right leg was seen, the former showing hypertrophy. Radiological measurement of the left leg bone was 41 cm. and right leg 39 cm. (Fig. 1). No appreciable abnormality

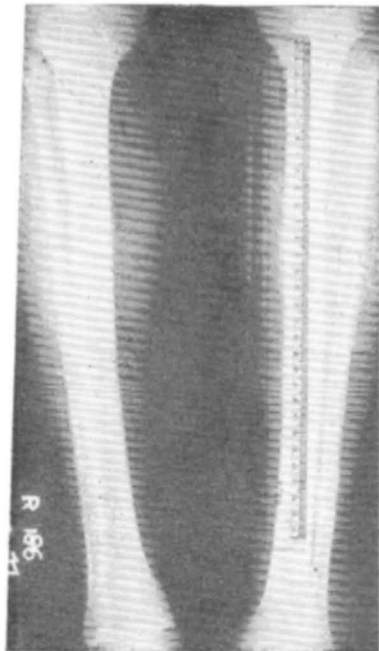


Fig 1

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was seen on venogram of the affected limb (Fig. 2).

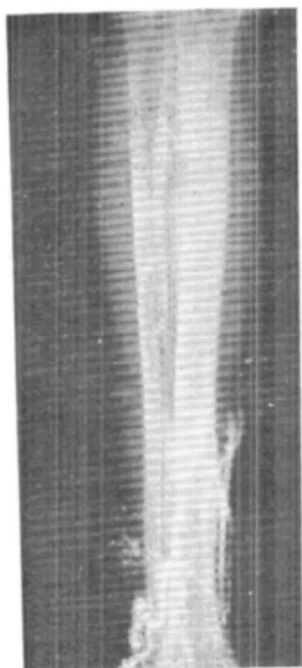


Fig 2

Histopathological features of the skin lesions were characteristic of nevus flammeus.

Discussion

The salient clinical features in the patient under review were an early onset on the disease, progressive hypertrophy and lengthening of the left leg and an associated nevus flammeus. These features characterise Klippel-Trenaunay syndrome. The hypertrophy of the bones and soft tissue were complimented by radiograph. Hence the need to utilize this procedure in such cases. It needs to be emphasised that this entity should be thought of while considering causes of unilateral hypertrophy of the limb^{1,2}, such as primary and secondary lymphoedema, neurofibromatosis and vascular abnormalities. These causes should be considered only when Klippel-Trenaunay syndrome is

incomplete i. e., when classical features are lacking.

Primary lymphoedema may manifest either as congenital hereditary lymphoedema - Nonne-Milroy's, or Meige type and lymphoedema praecox and lymphoedema tarda. These diagnosis depend upon history of an early or late onset respectively and characteristic solid nonpitting oedema. Filarial lymphoedema, tubercular lymphoedema, edema due to deep vein thrombosis of the leg and recurrent erysipelas cause secondary lymphoedema. The diagnosis of neurofibromatosis is made in the presence of multiple fibromas, cafe-au-lait spots and other associated systemic abnormalities. Vascular abnormalities such as arteriovenous fistula and agenesis of common iliac vein are diagnosed by venogram^{6,7}.

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