

A CASE OF THE KLIPPEL-TRENAUNAY-PARKES WEBER SYNDROME

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Abstract. An extreme case of Klippel-Trenaunay-Parkes Weber syndrome is presented. The patient had extensive cutaneous naevus involving the left side of the body and consisting of naevus flammeus, hemangioma cavernosum, and naevus verrucosus. The left extremities were longer and there were multiple arteriovenous connections between a and v subclavia and a and v radialis. Secondary symptoms were sciatica, varicosity and osteoporosis.

Key words: Klippel-Trenaunay disease; Angiomatosis; Arteriovenous malformations

The principal symptoms of the Klippel-Trenaunay syndrome are angiomatous tissue changes, enlargement of limbs, hypertrophy of soft tissues, and varicosity (3, 4, 6, 11).

Angiomatosis may occur in the form of a naevus flammeus, cavernous hemangioma or lymphangioma. The anomalies are unilateral, though some bilateral lesions have also been reported (8).

In 1907, Parkes Weber reported an arteriovenous anastomosis in a hypertrophic limb (9). Later observations have shown that the varicosity commonly associated with this condition is caused by agenesis of the deep veins or by congenital arteriovenous anastomosis (1).

The exact mechanism of the bone and soft tissue hypertrophy is still obscure. Many investigators see the increase in the blood supply or the increased blood oxygen saturation caused by arteriovenous fistulae as an explanation for this enlargement (12, 13). However, in the majority of the cases no anastomoses or varicosities are to be found (10).

Lengthening of the extremities is usually associated with soft tissue hypertrophy, but sometimes also with atrophy (14). Secondary symptoms may be vertebral scoliosis, ischias, stasis dermatitis, edema and thrombosis (11).

Cases of familial transmission are reported by some investigators (5, 7). The mode of inheritance is still unknown, but it seems likely to be of the in-

complete dominant type. Involvement of a genetic factor is a possibility, when one considers the numerous naevoid tissue formations associated with this syndrome.

Angiomatosis may be located in any organ. The roentgenographic features of the syndrome are thoroughly described by Calenoff et al. (2).

The following case is presented as a typical example of this syndrome with its numerous symptoms.

CASE REPORT

The patient was a 47-year-old unmarried lumberman from Northern Finland. At birth he had extensive cutaneous naevus involving the left side of the body from the shoulder to the ankle. The skin lesions persisted and at puberty became enlarged on the chest. The left extremities grew more rapidly than the right, and remained longer. Muscular strength was weaker on the left side and there were dilated veins along the left arm. Vertebral scoliosis and sciatica on the right side gradually became evident. The patient was exempted from military service, but was in continuous employment as a lumberman.

Clinical findings:

The patient had extensive unilateral naevus, dark brown in colour, involving the left side of the body (Figs. 1, 2), and forming longitudinal streaks on the left extremities. On the left side of the chest it formed an elaborate pattern, with cavernous hemangioma, capillary hemangioma and papillomatosis, and verrucosis continuing to the back as transversal streaks. One cavernous hemangioma was also found on the left ankle. On the palmar skin of the left hand the colour of the striated hyperkeratotic naevus faded towards the thumb and forefinger. There was varicosity on the left arm. A shunt sound was heard along the arteria subclavia and arteria radialis. Blood pressure in the right arm was 190/100 mmHg. Arteriography revealed multiple arteriovenous connections between a and v subclavia and a and v radialis (Fig. 3).

Cardiac catheterization was performed. The relation of the pulmonary circulation to the systemic circulation was 1.3:1 and the volume of the shunt flow 2.1 l/min. The pressure measured in the right ventricle was 42/7 mmHg and in the right atrium, 6 mmHg. The wedge pressures, "pulmonary capillary pressures", were 18 and 12 mmHg. The oxygen

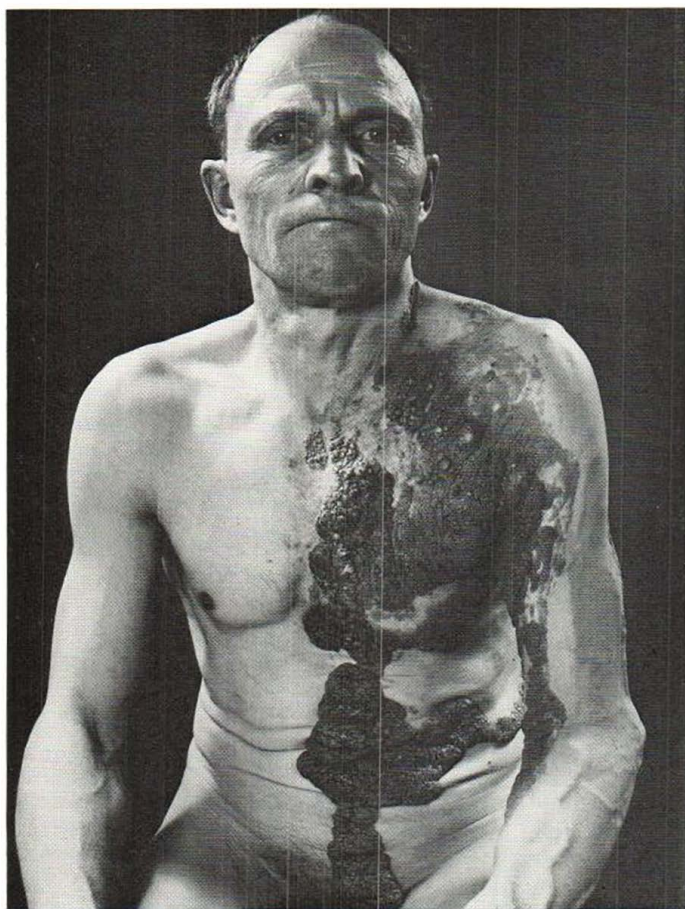


Fig. 1. The Klippel-Trenaunay-Parkes Weber syndrome. Extensive naevus unius lateris with papillomatous and verrucous structures. On the chest there are cavernous hemangiomas and naevus flammeus areas. There is venous dilatation of the right arm.

concentrations showed a relatively large shunt from the left side to the right side above the atrium. The left extremities were longer and there was muscle and bone atrophy on the same side. The left thigh was 2 cm less in circumference than the right one, and the left leg correspondingly 1 cm less. The patient had compensatory vertebral scoliosis and disc degeneration (L III-IV) with sciatic syndrome on the right side.

Bone roentgenography revealed changes on the left side. The skeletal structures were normal except for the disc degeneration. The left clavicle and scapula and the bones of the left upper extremity showed osteoporosis and diffuse radiolucent areas. The left hip near the acetabulum had cystic radiolucencies and sclerotic bone islets. Similar islets were also found in the tibia and femur.

Laboratory findings

Hb, Hcr, MCHC, Leucocytes, ALAT, ASAT, bilirubin, Alk. phosph., blood sugar, creatinine, calcium, cholesterol were normal.

ECG showed partial RBBB.

Skin specimens were taken from the papillomatous naevus of the chest and from the verrucous lesion on the left shoulder.

Histologic diagnoses were papilloma fibroepithelialis and naevus verrucosus.

No treatment was attempted for the vascular and naevoid tissue anomalies.

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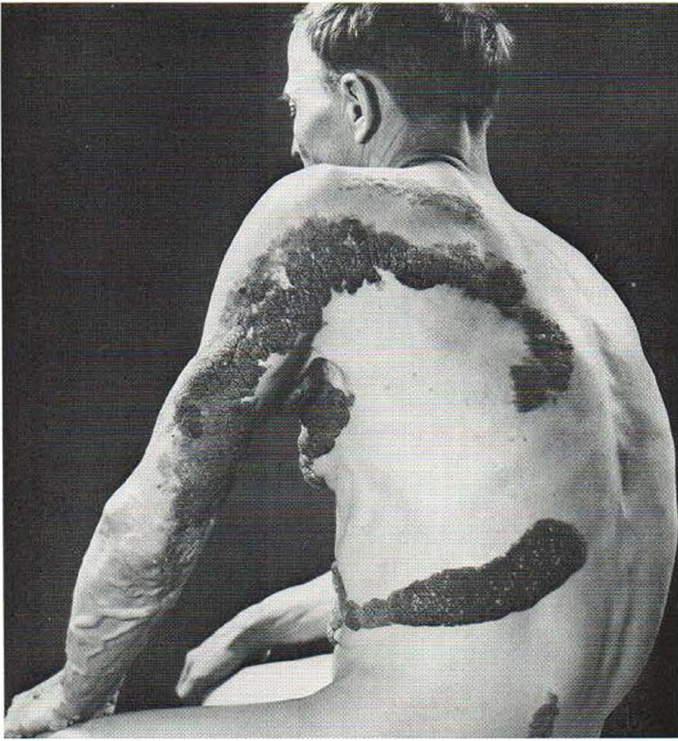


Fig. 2. Transversal naevoid streaks on the left back.

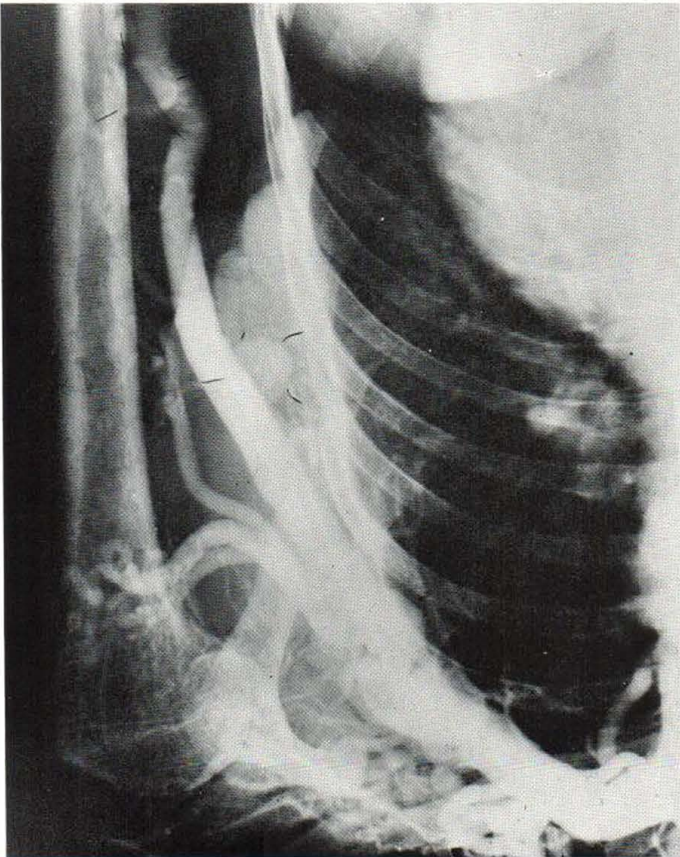


Fig. 3. Arteriography: The subclavial artery is dilated and there are arteriovenous communications between the subclavial artery and vein and between the radial artery and vein.

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