

The Klippel-Trenaunay Syndrome with Acro-angiodermatitis (Pseudo-Kaposi's Sarcoma)

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A patient with the Klippel-Trenaunay syndrome and pseudo-Kaposi elements is reported. This combination of symptoms has only been published very occasionally. Selective quantitative measurements of the peripheral tissue blood flow revealed a significantly increased cutaneous blood flow in the hypertrophied leg. This finding supports the assumption that a high perfusion rate and a high oxygen saturation are involved in the etiopathogenesis of the pseudo-Kaposi elements. *Key words: Acro-angiodermatitis; Cutaneous blood flow; Subcutaneous blood flow.* (Received May 8, 1984.)

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The Klippel-Trenaunay syndrome is a rare disorder characterized by cutaneous haemangioma, pronounced soft tissue and bone hypertrophy associated with varicose veins. It is usually confined to one extremity, most often the leg (1, 2, 3). Cutaneous haemangioma is often observed at or shortly after birth and the tissue hypertrophy and varicose veins usually develop during childhood or at puberty (1). It is generally assumed that the tissue hypertrophy is caused by a high peripheral tissue blood flow. Quantitative measurements of the tissue blood flow has, however, not previously been published.

We want to report a patient with this syndrome, who, at his affected leg, developed cutaneous haemangiomas which clinically and histologically resembled Kaposi's sarcoma. Previously, this combination has only been published 5 times in the literature (4). Furthermore, selective quantitative measurements of the peripheral tissue blood flow revealed a significantly increased cutaneous flow in a normal skin area of the affected hypertrophied leg.

CASE REPORT

The patient is a 36-year-old man who was first seen in our department at the age of 20 months, at which time he presented a naevus flammeus on the lateral part of his right leg. The right leg was larger than the left leg. At the age of 33 he was admitted to an ortopaedic department because of pronounced varicose veins, restlessness and leg cramps in his right leg. A varicectomy was performed, but there was recurrence of varicosities within a few months. A phlebography revealed partial obstruction of the deep veins. Anti-coagulation therapy and supportive bandages did not reduce his symptoms.

The patient was seen again at our dermatological department at the age of 36 because of two newly developed tumours on the lateral side of his right leg. The lesions had a red-bluish hue, an irregular, sharply demarcated border, and were slightly infiltrated. Clinically these lesions were indistinguishable from classical Kaposi's sarcoma. The right leg was 2 cm longer than the left leg, and the circumference was about 5 cm bigger at comparable levels. There were pronounced varicose veins, but except for this the skin was quite normal. The pulses of the extremities were equal. He complained of restlessness, leg cramps, and lower back pain presumably due to a compensatory scoliosis of the spine. Otherwise he was completely healthy. He was heterosexual with no previous history of venereal disease.

Histological examination of a biopsy specimen from a tumour showed focal parakeratosis of the epidermis and pronounced proliferation of capillaries, extravasation of erythrocytes and a moderate lymphohistiocytic infiltration. A factor 8 staining confirmed endothelial proliferation. These histological changes are suggestive of early Kaposi's sarcoma although not diagnostic.

Cutaneous and subcutaneous blood flow (CBF, SBF) was measured quantitatively by the ^{133}Xe washout method after epicutaneous atraumatic tracer labelling (5). The tissue-to-blood partition coefficient, λ_{Xe} was determined by a double isotope washout technique with ^{133}Xe and ^{131}I -Antipyrine (6).

There was no difference in the λ_{Xe} in symmetric normal looking skin, whereas the λ_{Xe} for subcutaneous tissue was significantly smaller on the right leg, 1.6 ml/g, compared to the left leg, 4.4 ml/g (normal range, 5.0 ± 1 SE ml/g). This finding indicated a massive subcutaneous edema. SBF was, however, not significantly influenced, being 3.04 ml/100 g/min. on the right side compared to 2.46 ml/100 g/min. on the left side. Normal SBF on the crus is 2.30 ± 0.85 SE ml/100 g/min. Venous pressure was not increased to a degree which affected the veno-arteriolar reflex mechanism in the subcutaneous tissue and a normal autoregulation of the SBF was present (7). CBF was, however, significantly increased in the affected right leg, 10.22 ml/100 g/min., compared to the left side 6.10 ml/100 g/min. (normal CBF being 4.62 ± 0.84 SE ml/100 g/min.).

The patient received topical steroid and X-ray with some beneficial effect. But during follow-up he developed new 1/2 cm large dark-blue lesions which both histologically and clinically resembled Kaposi's sarcoma.

DISCUSSION

The Klippel-Trenaunay syndrome may be associated with additional skin changes and symptoms such as edema, stasis dermatitis, ulcerations, phlebitis, cellulitis, and hyperhi-

drosis (2, 8). So far only very few patients with pseudo-Kaposi elements have been reported to have Klippel-Trenaunay syndrome.

Chronic stasis dermatitis of the foot with lesions clinically and histologically simulating Kaposi's sarcoma was described as acro-angiadermatitis by Mali and coworkers in 1965 (9). Similar lesions have been described in patients with arteriovenous fistules (10, 11) and Parkes-Weber syndrome (4).

The etiology of the pseudo-Kaposi elements is unknown. A relative high oxygen saturation of the affected tissue, which causes proliferation of small vessels and fibroblasts (4, 9) and a high perfusion rate—as found in the affected leg of this patient—seem to be involved in the etiopathogenesis of the pseudo-Kaposi elements.

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