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## The Klippel Trenaunay Weber Syndrome Presenting with Cutaneous Bleeding

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**Abstract.** A patient with the Klippel Trenaunay Weber syndrome presented with prolonged bleeding from the finger tips during strenuous exercise. Tissue fibrinolysis studies showed increased fibrinolytic activity suggesting that the blood vessels in the affected limb may be functionally as well as anatomically abnormal.

**Key words:** Klippel Trenaunay Weber syndrome; Bleeding; Fibrinolysis

The Klippel Trenaunay Weber syndrome as described by Klippel & Trenaunay in 1900 (6, 7) is characterized by vascular naevoid abnormalities confined to a limb. It is associated with hypertrophy which may be gross, and varicose veins.

### CASE REPORT

The patient is a 43-year-old building labourer whose presenting complaint is that when using his arms energetically (when digging, for example) he may bleed profusely from the tip of his right forefinger. The bleeding usually stops after sustained pressure has been applied to the finger, but on three occasions he has attended the casualty department because of persistent bleeding. On each visit the bleeding was such that cautery was required to quench it.

On examination the patient was healthy except for the physical signs in his right arm which was enlarged, and showed marked varicosities throughout the whole limb.

In addition he had multiple telangiectases extending

from the neck throughout the limb but which were very heavily grouped on his fingertips (Fig. 1). Blood pressure was 130/85 mmHg in both arms. No arterial or cardiac bruits were present but a venous hum was audible over the right arm.

### Investigations

Normal hematopoietic, renal and liver functions were revealed. Chest X-ray was normal and soft tissue radiograph failed to demonstrate any abnormal vascular channels in the bones of the right arm. Twenty-four hour urinary oestriol excretion was within our normal range.

Blood was drawn from right and left antecubital veins; the euglobin lysis time was estimated and found not to differ significantly (right arm 345 min; left arm 360 min). However, skin biopsies taken from corresponding sites in right and left arms were assayed for tissue fibrinolytic activity (2) and this was found to be increased in tissue from the right arm (47% vs. 24%).

Because of the increased skin fibrinolytic activity observed in the right arm, we prescribed oral epsilon aminocaproic acid and the patient has had no further bleeding in the past 5 months.

### DISCUSSION

Cases of unilateral hypertrophy and hemanomatous malformations were described in the mid-nineteenth century (1), but Klippel & Trenaunay in 1900 (6, 7) grouped together the triad of signs now called eponymously after them and F. Parkes Weber.

The condition has been associated with paraplegia (5) and even malignant change (3). It appears to be more common in boys, with the arm affected twice as frequently as the leg; the head and trunk are affected in 10% of cases (4, 10).

We were surprised to find no difference in venous fibrinolytic activity between right and left arms. As venous endothelium produces fibrinolytic activators, we might expect increased fibrinolytic activity in the affected arm with more numerous venous channels. However, if the veins per unit area were to produce fewer activators than normal, the amount of fibrinolytic activity seen in the abnormal limb would be similar to that in the normal limb, thus explaining our finding and giving support to the concept that the veins in the abnormal limb are functionally as well as anatomically abnormal.

Bleeding from telangiectatic vessels is not usually a problem with these patients. The mechanism of action of the bleeding could be a failure of contraction in an abnormal part of the vessel wall.

Fibrinolytic activators are produced by vascular—particularly venous—endothelium and the

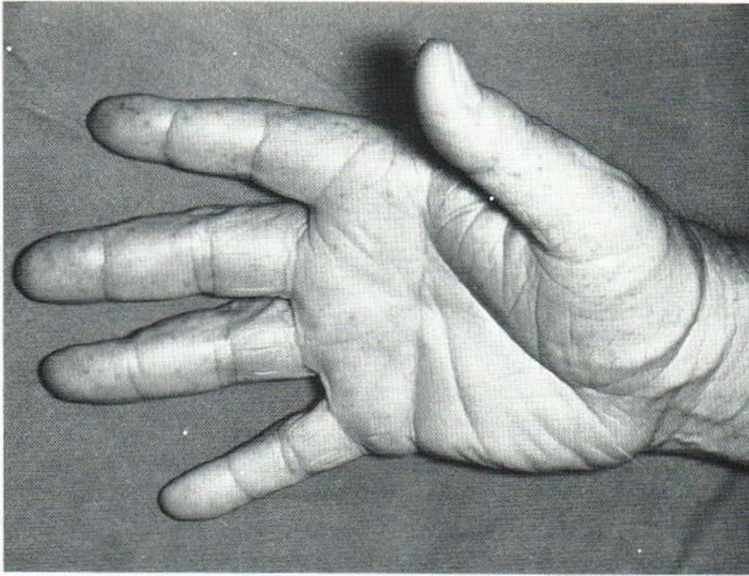


Fig. 1. The subject's right hand showing fingertip telangiectases.

biopsy from the abnormal arm showed increased fibrinolytic activity.

This could simply be a reflection of the increased number of cutaneous blood vessels in the abnormal limb.

The response to epsilon aminocaproic acid which reduces fibrinolytic activity (8) supports the suggestion that an increase in tissue fibrinolytic activity might, in part, be an explanation for the persistent bleeding in such telangiectatic blood vessels.

#### ACKNOWLEDGEMENT

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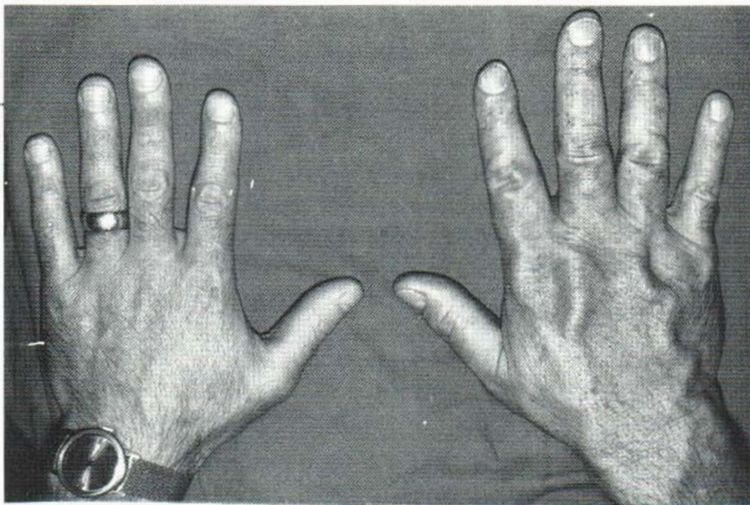


Fig. 2. Right and left hands compared.

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## Palmo-plantar Keratoderma Associated with Carcinoma of the Bronchus

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**Abstract.** A 63-year-old man presented with a 15-month history of palmo-plantar keratoderma and was found to have a bronchogenic carcinoma. The implications of this association are discussed.

**Key words:** Keratoderma; Carcinoma of the bronchus

Many disorders of keratinization are known to be associated with internal malignancy. Exfoliative dermatitis and acquired ichthyosis are particularly linked with lymphoma but have been described with a number of malignancies (12). Bazex's syndrome or acrokeratosis paraneoplastica (1) which is associated with carcinoma of the laryngo-pharyngeal region, may present with keratoderma but usually starts with erythema and psoriasiform scaling which involves the fingers, toes, ears and nose. The keratoderma has a characteristic violaceous colour

and the rest of the body may be affected in the latter stages of the disease. The Howel-Evans syndrome (7, 15) constitutes palmo-plantar keratoderma of autosomal dominant inheritance, associated with carcinoma of the oesophagus. Acanthosis nigricans may also present with keratoderma or sometimes with the so-called tripe palm anomaly (2, 3). It has been estimated that over 50% of adults presenting with acanthosis nigricans have an associated neoplasm (5). The acquired form of pachydermoperiostosis may also present with keratoderma and has been described in association with a variety of systemic illnesses and internal malignancies including carcinoma of the bronchus, adenocarcinoma of the lung and mesothelioma (12).

### CASE REPORT

#### *History*

A 63-year-old male fur liner presented in April 1981 with a 15-month history of gradual thickening of the palms and soles. This developed as isolated areas of involvement which became confluent on palmar and plantar surfaces. He complained of recent painful fissures on the palms and loss of sensation over the tips of his fingers.

Prior to this he had been well and denied any other symptoms. His family history was unhelpful. He smoked 60 cigarettes a day.

#### *Examination*

His palms showed gross diffuse hyperkeratosis with a finely corrugated surface (Fig. 1). The palmar creases appeared to be spared but were affected in places by painful fissures. His soles showed relatively minor changes of a similar nature. There was no sparing over the plantar creases. General examination was normal and there were no other signs of acropachy or peripheral neuropathy.

#### *Investigations*

Full blood count—normal. ESR 26 mm/h. Chest X-ray revealed a large mass in the left upper lobe (Fig. 2) indicative of carcinoma of the bronchus. Growth hormone levels were normal.

#### *Histopathology*

A biopsy from the right palm showed marked compact hyperkeratosis and acanthosis. A remarkable feature was regular "tenting" of the epidermis to create an appearance of pinnacles where there was loss of the otherwise increased granular layer and associated areas of parakeratosis. Prominent dilated capillaries were evident in the papillary dermis.

### DISCUSSION

The association of palmo-plantar keratoderma with carcinoma of the bronchus without other clinical