"SEGMENTAL PURPURA" —A SIGN OF DEEP VENOUS THROMBOSIS

By H. L. Chan and O. T. Khoo

SYNOPSIS

Purpura developing in a cutaneous area corresponding to the distribution of tributaries distal to a thrombosed vein is not a generally recognised clinical sign. Two cases of its occurrence are reported here, one in the upper limb, and the other, the lower limb. The segmental nature is distinctive. The mechanism of its production is discussed. When present, it develops early in the course of the thrombosis, and awareness of this sign will lead to increased confidence in the diagnosis of acute deep venous thrombosis.

INTRODUCTION

Deep venous thrombosis (D.V.T.) in the extremities is often silent and may not be evident until an embolus has lodged in the pulmonary artery (Uhland, 1964).

Local clinical manifestations of D.V.T. include pain and tenderness, and a measurable increase in circumference of the affected limb. Prominent veins, oedema, and mottled cyanosis may be found distal to the site of thrombosis. The thrombosed vein may be tender, and Homan's sign positive. An increase in local temperature with delayed cooling on exposure has been reported (Provan, 1965). Lowenberg described a sphygmomanometer cuff pain test. In severe cases, diminished arterial pulsations have been detected on the affected side.

Any or all of these signs may be absent. "Segmental purpura", that is, petechiae and ecchymoses occurring in an area of skin from which the thrombosed vein receives its tributaries, is not generally recognised as a sign of acute phlebothrombosis. There is no mention of this sign in the standard texts in medicine and dermatology (Harrison, 1970; Cecil-Loeb, 1971; Rook, 1972). However, in severe iliofemoral venous thrombosis ("phlegmasia cerulea dolens"), petechiae and ecchymoses, and even gangrene may develop (DeBakey, 1949).

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The purpose of this paper is to document two instances of segmental purpura following acute D.V.T. and to discuss the mechanisms for its production and its significance.

Case 1

B.H.B., a 14-year old female Chinese was first seen on 16.6.73 with a history of periodic fever of 2 months duration, associated with pallor. There was no joint pain or skin rash. Examination at that time revealed anaemia and hepatosplenomegaly. The haemoglobin was 7·5 g.%, total white cell count was 9,700/cu.mm., and platelet count 215,000/cu.mm. ESR was very high, 159 mm. fall in the first hour. Urine examination showed albumin+, RBC 25-30 per HPF. Blood urea was 22 mg.%. Although blood for LE cells was negative on three occasions, rheumatoid factor was positive. She was treated as Systemic Lupus Erythematosus with prednisolone 10 mg. q.d.s. with improvement. She did well on maintenance prednisolone of 10 mg. t.d.s.

On 25.11.73 she was re-admitted with a 3-day history of sudden onset of pain and swelling over her right forearm followed by swelling and numbness of her right hand. She also noticed red spots over her right hand and forearm the same day.

Her general condition was good. She appeared "cushingnoid". Local examination showed:-

(1) petechiae and ecchymoses over the radial one half of right forearm, palm and dorsum of hand (see Fig. 1),

(2) swelling of forearm, thumb, and index fingers,

(3) mottled cyanosis of thumb and index fingers,

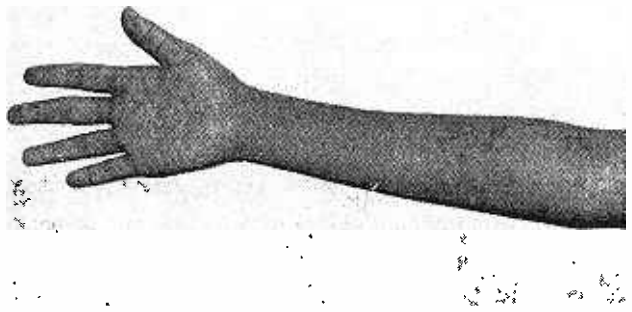

Fig. 1. Right forearm and hand: petechiae and ecchymoses over radial half of flexor surface of forearm and palm.

(4) normal radial and ulnar pulses on both sides.

The platelet count was normal at 175,000/cu. mm., and serum for cryoglobulins was negative.

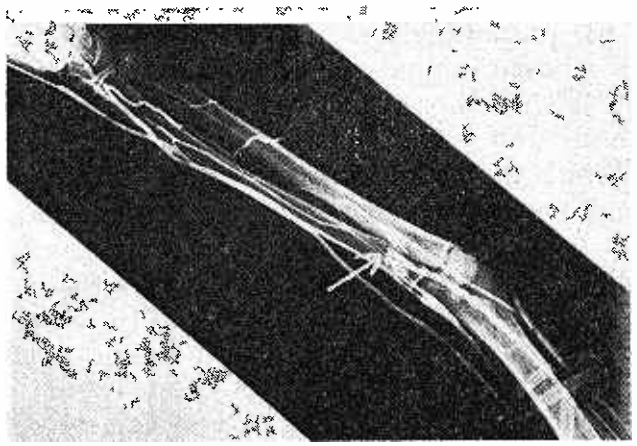

Fig. 2. Venogram: thrombosis of median veins of right forearm (arrowed).

A venogram was done on 29.11.73 (see Fig. 2). The veins were compressed above the right elbow for the procedure. The basilic and cephalic veins were visualised and appeared normal. The median vein of the forearm which is usually present was not seen but instead the proximal stumps at the cubital fossa were demonstrated, suggesting the deeper vein or veins in the forearm were blocked.

The patient was started on anti-coagulant therapy on 26.11.73, with i.v. Heparin and Warfarin Sodium. The painful swelling subsided after four days, and the petechiae faded after eight days.

On follow-up, the right hand and forearm have remained free from symptoms, and she was generally well on maintenance prednisolone of 10 to 15 mg. daily.

Case 2

L.C.C., a 34-year old female Chinese was first seen in 1964 with thrombocytopenic purpura. Haematological investigations at that time showed a haemoglobin of 8·1 g.%, WBC of 3,800/cu.mm., platelet count of 5,000/cu.mm., and a reticulocyte count of 4%. Blood for LE cells and Coomb's test was negative. Bone marrow aspiration revealed a hypoplastic marrow with few megakaryocytes. She was managed as a case of hypoplastic anaemia and given blood and platelet transfusions, testosterone, and prednisolone.

She did fairly well on follow-up. In January 1972 she presented with fever, jaundice, and passage of dark urine. Investigations during this admission showed a peripheral pancytopenia with 3·6% reticulocytes. Ham's test was positive. There was haemosiderin in the urine. Serum iron was 65 ug.%, TIBC 294 ug.%. The consultant haematologist (Dr. S.B. Kwa) was of the opinion that the patient has Paroxysmal Nocturnal Haemoglobinuria. She was given iron and folic acid but no prednisolone.

Fig. 3. Right thigh: clustered petechiae over anterior surface.

On 21.11.73 she developed sudden onset of pain, and progressive swelling of her left leg, associated with red spots over her left leg and thigh.

Local examination, on admission, revealed:-

(1) her left lower limb to be larger and paler than her right,

(2) several petechiae scattered over her left leg and thigh anteriorly (see Fig. 3),

(3) her left leg was warmer,

(4) Homan's sign was positive,

(5) equal femoral, dorsalis pedis, and posterior tibial arterial pulses.

The patient was in good general condition, and there were no petechiae elsewhere in the body. The haemoglobin was 9·8 g.%, WBC 6,200/cu.mm., and platelet count 50,000/cu.mm. Urine again showed haemosiderin.

She was anticoagulated with Warfarin Sodium.

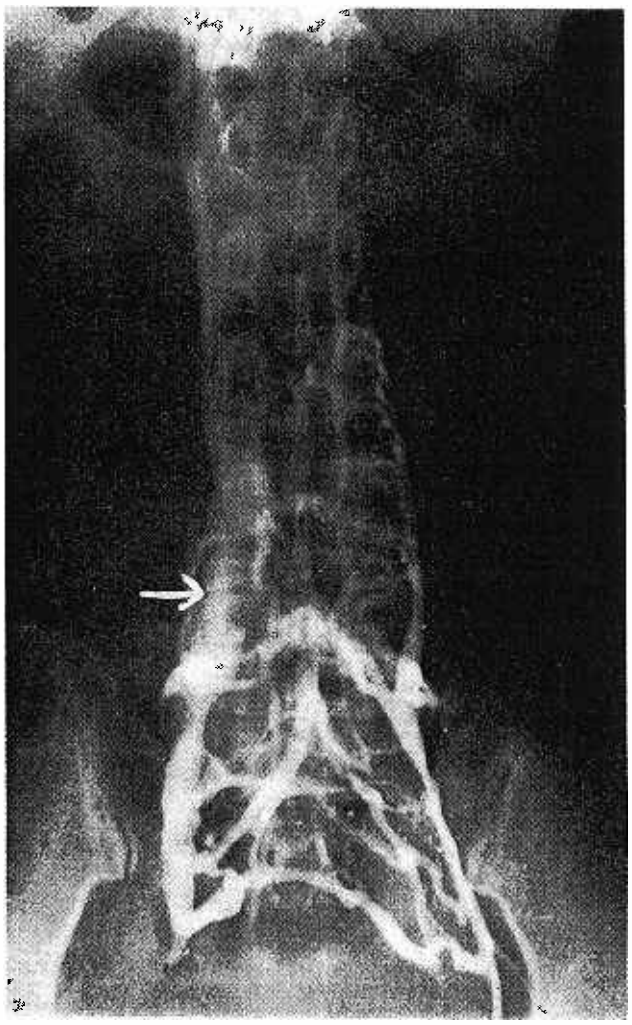

Fig. 4. Venogram: thrombosis of left iliac veins. Arrow indicates right common iliac vein; left iliac veins replaced by collaterals.

A venogram was performed on 14.12.73. An injection was made into the left external iliac vein. This showed well developed collaterals from the left to the right side (see Fig. 4). The left common iliac vein was obliterated and replaced by collaterals.

The purpura over her left lower limb faded after 8 days. The swelling of her left leg gradually subsided and her limb was back to normal size by the time of her discharge on 17.12.73.

She has remained well on follow-up.

DISCUSSION

The factors responsible for the venous thrombosis in these 2 cases will not be discussed, suffice it to say that thrombotic complications are known to occur in Systemic Lupus Erythematosus, Paroxysmal Nocturnal Haemoglobinuria, and corticosteroid therapy.

The mechanisms involved in the production of the purpuric eruption in an area distal to a thrombosed vein is primarily mechanical (Allen, 1962). Thrombosis leads to an increase in intravenous and intracapillary pressure. The level to which the venous pressure rises, depends on the extent and acuteness of the thrombosis as well as on the presence or absence of venous collateral channels. If the pressure is raised high enough to overcome the resistance of the capillary endothelium, breaks occur at various points in the capillary network with extravasation of red blood cells in the dermis and clinical purpura. There are other examples of purpura due to raised intravascular pressure in the absence of any other disease. Crops of petechiae may occur on the relatively loose tissue of the face and neck after prolonged coughing or fits.

Hypoxia, resulting from venous stasis is also probably involved in the pathogenesis. Oxygen deficiency acts by impairing the integrity of the capillary endothelium.

In Case 1, prednisolone therapy may be an additional factor, as corticosteroids are known to increase capillary fragility by causing changes in the surrounding connective tissue (Shuster, 1960); while in Case 2, the platelet count of 50,000/cu.mm. may be contributory. The mechanisms whereby platelet deficiency may be associated with purpura are complex (Marcus, 1969) although it is unusual for spontaneous purpura to develop with platelet counts above 50,000/cu.mm.

The segmental nature of the purpura depends on the distribution of the venous tributaries,

and will just be as variable. The pattern of purpura assisted in the diagnosis of Case 1 which was initially thought to be due to "vasculitis" (arteritis).

Segmental purpura, when it occurs at all, is seen *early* in the course of the thrombosis, probably within hours as the maximum rise in intracapillary pressure will be during the acute phase of the occlusion. Purpura was present in the first day of symptoms of both these cases.

The exact incidence of segmental purpura is not known. Awareness of this physical sign will, if present, lead to increased confidence in the clinical diagnosis of acute deep venous thrombosis.

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