(3) Recurrent cutaneous suppuration complicating acne with paroxysmal nocturnal haemoglobinuria (PNH)

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D.I.T., male, aged 25 years.

History. In August, 1967, aplastic anaemia was diagnosed after 3 months of dyspnoea, epistaxis and spontaneous bruising. He was treated with prednisolone 20 mg daily, and 1 month later, when aged 15, he developed acne vulgaris on the face, back and chest. Steroids were discontinued after 9 months. The pancytopenia persisted although the abnormal bleeding tendency improved. March, 1970. Deep vein thrombosis of the right leg.

FIGURE 1. Diffuse ulcerative suppuration on acne area of shoulders.
June, 1970. An acute crisis with pancytopenia and reticulocytosis was diagnosed as PNH. He had more severe acne on the face and neck, with acne conglobata on the back and suppurative hidradenitis in the left axilla and the groins. Large irregular necrotic ulcers developed on both calves, healing slowly to leave scars.

April, 1971. Further ulceration of both calves occurred following a haemolytic crisis, and rapidly developing areas of suppuration appeared in the acne tissues of his face and back (Fig. 1), with some resemblance to pyoderma gangrenosum. A sudden left hemiparesis and hemianaesthesia developed, from which he recovered; it was assumed to be due to cortical vein thrombosis.

Two weeks later he developed abdominal pain with vomiting. Inferior veno-caval obstruction was confirmed by venography. He was anticoagulated with Warfarin. The leg ulcers improved slowly. He was discharged home on anticoagulants.

Between January and July, 1972, three blood transfusions were given for severe anaemia. There was
recurrent ulceration of the calves. Further severe suppurative ulcers again developed in the acne on the shoulders and face (Fig. 2). These began with nodular haemorrhagic areas, spreading rapidly and ulcerating, healing slowly with marked scarring. The lesions were self-limiting and were un-

![Biopsy of the shoulder showing suppurative and perivascular infiltration with fibrin deposits in small blood vessels (x65).](image)

affected in their course by topical steroids or systemic antibiotics.

August, 1972. Left hemiplegia due to cortical vein thrombosis.


April, 1973. Recurrent ulcerative suppuration on acne affected areas of cheeks and shoulders, but healing within a few weeks.


Immunological investigations. Immunoglobulins normal. Direct Coombs test negative. Total serum complement level 720 units/ml (normal range). Polymorph function tests. (a) Phagocytosis; polymorph preparations ingested test particles normally, the average proportion of phagocytosing cells being 79°° (normal limits 65-89°°). (b) Nitro-blue tetrzaolium test; 9°° of the patient's cells reduced this dyestuff (within normal limits). Mantoux test; strongly positive at 1:1000 dilution.

X-rays. Inferior venocavogram showed complete occlusion of the iliac veins and the inferior vena cava. The upper level of the occlusion was not demonstrated. Collateral circulation was seen on the anterior abdominal wall, lateral abdominal wall and retro-peritoneally, connecting with the azygos veins and the internal mammary veins.

IVP. Bilateral equal excretion was seen. No radiographic evidence of renal vein thrombosis was seen.

Organisms cultured from suppurations included beta haemolytic streptococcus (Lancefield Group C), Staphylococcus aureus (coagulase positive), S. albus (coagulase negative).

Histology (Fig. 3). Ulcer on shoulder; acute suppurative and perivascular inflammation and involvement of the small vessels with fibrin deposits.

Discussion. The three essential features here are PNH 'recurrent leg ulcerations' and acne complicated by suppurations resembling pyoderma gangrenosum. PNH may be preceded by a period of marrow aplasia or hypoplasia and pancytopenia, as we observed in our own patient (Dacie, 1967). Venous thrombosis is a frequent complication of PNH and thrombo-embolism is a major cause of death. Leg ulceration has been noted only when the patient has been severely anaemic, and anaemia, venous insufficiency and thrombosis in the legs may be contributory factors here. Conglobate acne dates from adolescence at the age of 15, but the type of suppuration with burrowing pyoderma is not typical of acne conglobata, nor is it associated with haemolytic crises or severe anaemia, and it is self-limiting and uninfluenced by treatment. Cutaneous haemorrhagic necrosis has been observed in patients receiving coumarin anti-coagulants (Meyler & Herxheimer, 1972), but in these cases, ulceration is preceded by erythema and petechiae. Our patient, however, continued to develop supplicative lesions during a short period in which he was not receiving Warfarin. The clinical findings and investigations provide no evidence for the commoner causes of pyoderma gangrenosum; it was decided not to subject the patient to further barium studies because of his poor general condition. Tests so far performed do not suggest an abnormality of cellular or humoral immunity. The clinical progression of the lesions from induration and erythema to ulceration, together with the histology, suggest that there may be a vascular aetiology, but whether vessel occlusion from fibrin deposition is due to a cutaneous vasculitis, is secondary to inflammation of neighbouring tissue, or has resulted from intravascular coagulation associated with PNH is not clear.

REFERENCES
