the well-known distribution of the EGFR protein in skin sections, and because the anti-Neu antibody, C-18, does not stain smooth muscle cells (data not shown).

There are known several antibodies, such as the anti-α smooth muscle actin antibody (6), the anti-smooth muscle myosin antibody (7) and the anti-desmin antibody (7), which can be used as a differentiation marker of smooth muscle cells. Although exactly which epitope antibody C-20 detects in smooth muscle cells is not clear so far, this antibody may be useful in identifying smooth muscle cells in various situations. We are now undertaking efforts to reveal to which antigen antibody C-20 reacts in smooth muscle cells.

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A Case of Pigmented Purpuric Eruption Associated with Hereditary Spherocytosis

Sir,

Pigmented purpuric eruptions are chronic conditions of unknown aetiology which comprise a group of clinical patterns of erythrocyte extravasation due to pericapillary inflammation. Progressive pigmented purpuric dermatosis (Schamberg's disease), purpura annularis telangiectodes (Majocchi's disease), pigmented purpuric lichenoid dermatosis (Gougerot and Blum's disease) and lichen aureus are the subtypes of pigmented purpuras.

Here we present a case of pigmented purpuric eruption associated with hereditary spherocytosis.

CASE REPORT

A 20-year-old man presented with the complaints of fatigue, nausea, vomiting and cutaneous eruptions. General physical examination revealed splenomegaly and icteric scleras. On dermatologic evaluation, there were red, annular and linear macules 0.5 – 3 cm across with minute telangiectases and pinhead-sized petechiaes located on the extremities and abdominal region symmetrically (Fig. 1). The lesions had started three years previously, and were characterized by waxing and waning periods. He had no drug history.

Histopathologic examination of the lesional skin biopsy specimen showed dilated capillaries surrounded by mononuclear inflammatory infiltrate and extravasation of erythrocytes in the upper dermis.

Complete blood count revealed a hemoglobin of 11.9 gm, hematocrit of 32.2% and white blood count of 6600/mm^3. Mean corpuscular hemoglobin concentration was elevated. Osmotic fragility was abnormal (0.60 – 0.45%; control: 0.45 – 0.35%), and reticulocyte count was found to be increased (8%). Total bilirubin was 2.9 mg/dl (normal: 0.2 – 1.0 mg/dl) and direct bilirubin was 0.4 mg/dl (normal: 0.1 – 0.3 mg/dl). There were spherocytes in the peripheral blood smear. Cryoglobulins and cryofibrinogens were found to be negative. Serum protein electrophoresis was within normal limits.

Hematologists advised splenectomy, but he did not consent. Because he had no complaints, we did not give any medication for the skin lesions. The patient was seen three months later. He still had skin eruption, but it was less severe. After that, he did not attend regular examinations.

DISCUSSION

Pigmented purpuric dermatoses, pigmented purpura and purpura pigmentosa chronica are the synonyms of pigmented purpuric eruptions which are preferred by some authors. Pigmented purpuric eruptions run a chronic, recurrent course in most patients. Their clinical features show considerable overlap with typical diseases (Schamberg's disease, Majocchi's disease, Gougerot and Blum's disease and lichen aureus). All are similar histologically. Extravasation of erythrocytes due to pericapillary inflammation and perivascular lymphocytic infiltration occurs.

Pigmented purpuric eruptions may be considered a purely local cutaneous inflammation characterized by lymphocytic vasculitis. Data available suggest that the vascular damage and extravasation of erythrocytes are secondary to a localized cell-mediated immunologic reaction. The fact that predominantly CD4+ T-cell infiltration occurs in lesional skin (1, 2) supports this view.

The aetiology of pigmented purpuric eruption is unknown. Some conditions, including gravity and increased venous pressure, some drugs (thiamine propyldisulfide, chlordiazepoxide)

(3), and step aerobics (4) have been blamed. The first one is possibly an important factor in most patients.

The lesions of pigmented purpuric eruption are not related to any blood dyscrasias. However, the research of Vignale & Rizzo (5) has had interesting results. They studied hemostasis parameters in 10 patients with pigmented purpura, with the following results: reactional thrombocytosis, increased levels of platelet circulating aggregates, increased response of platelet aggregation to different agonists, delayed activation of the contact system, decrease of the activity of the fibrinolysis activators, and diminished function of the HMWK ("High Molecular Weight Kininogen") coagulation fraction.

In light of these data, the authors suggested that cutaneous pathology seen in pigmented purpuric eruptions may be related to hematologic alterations of the platelet-HMWK-endothelial cells-kinins-coagulation-fibrinolysis system.

Nevertheless, to our knowledge, there is still no case of pigmented purpuric eruption associated with a hematologic disorder in the literature, except our patient.

Effective and established therapeutic modalities are lacking in cases of pigmented purpura. Topical and systemic steroids may have some benefit. PUVA was found to be effective in two cases (6). Okada et al. (7) treated a case of purpura pigmentosa chronica unresponsive to conventional therapies (topical corticosteroids, anti-inflammatory agents, etc.) with oral cyclosporin A. High therapeutic effectiveness of cyclosporin A in this case supports the hypothesis that CD4+ T-cells play an important role in the pathogenesis of pigmented purpura (1, 2). However, because of its potential side effects, it is not reasonable to recommend cyclosporin A in all cases. It may be regarded as an alternative approach in patients with widespread lesions who are distressed by their condition. However, it should be kept in mind that a therapeutic approach is not required in most patients with pigmented purpuric eruption.

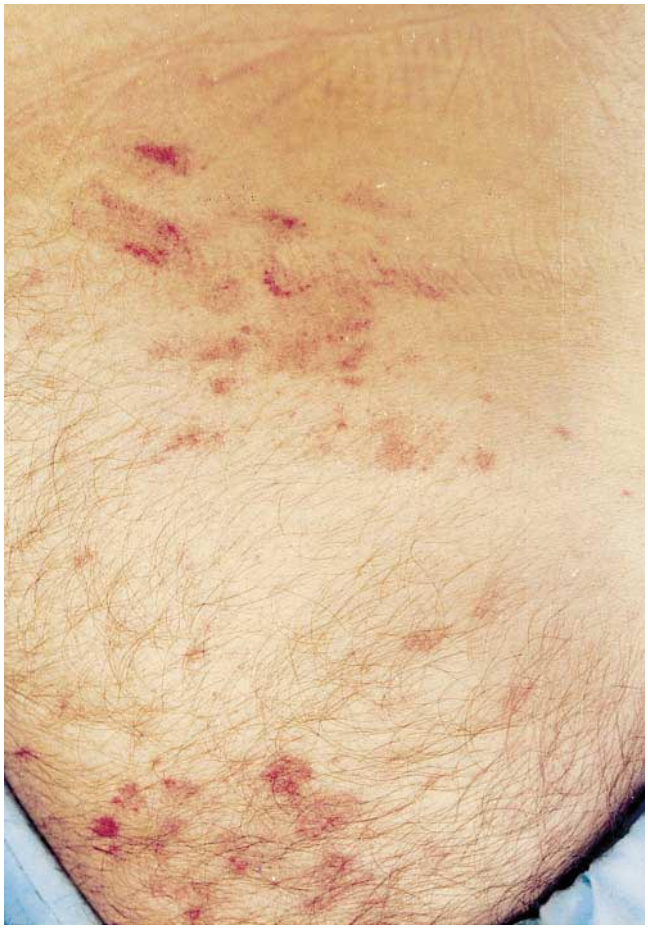

Fig. 1. Red, annular and linear macular lesions with petechias on the right side of the abdomen.

Hereditary spherocytosis is a familial hemolytic disorder characterized by anaemia, intermittent jaundice, splenomegaly, and responsiveness to splenectomy. It is transmitted as an autosomal dominant trait, but may remain clinically silent for long periods. Chronic leg ulceration and pigmentation resulting from healed ulcers are the unusual cutaneous complications of hereditary spherocytosis (8).

Our case had no ulcer or pigmentation on the ankles. We did not observe any correlation between hemolytic episodes and the activation of cutaneous eruptions. To our knowledge, this is the first case of pigmented purpuric eruption associated with a hematologic disorder. However, whether there is a causal relationship between pigmented purpuric eruption and hereditary spherocytosis remains unclear.

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