Bullous and Pustular Rheumatoid Vasculitis: Treatment by Plasma Exchange

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A case of rheumatoid vasculitis (RV) with bullous and pustular lesions is reported. These skin manifestations represent very unusual symptoms of RV. A dramatic clinical improvement was obtained by plasma exchange and immunosuppressive therapy. *Key words: Vasculitis; Rheumatoid; Arthritis; Bullae; Pustules; Plasma exchanges.* (Received April 14, 1983.)

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Skin lesions resulting from vasculitis in the course of rheumatoid arthritis (RA) include digital micro-infarcts, nodules, urticarial-like plaques, ulceration and, more rarely, bullae (7). A case of localized pustular vasculitis in RA has recently been reported. The authors suggested that immune complexes may play a role in the pathogenesis of pustular vasculitis (6).

We report here a case of rheumatoid vasculitis (RV) with bullous and pustular lesions which was successfully treated by plasma exchange (PE) and immunosuppressive therapy (IST).

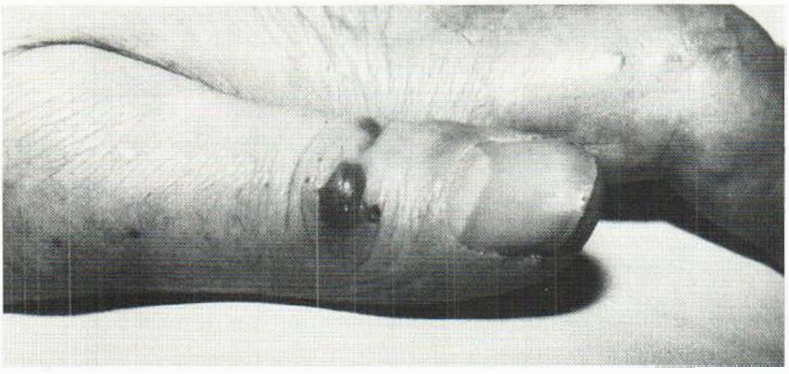

Fig. 1. Bullae on a finger.

CASE REPORT

A 57-year-old Caucasian woman had had RA for 3 years. At that time, the rheumatoid factor titre was within the normal range. Previous treatment of her articular manifestations included gold, D. Penicillamine and non-steroidal anti-inflammatory drugs, sulfones and even steroids.

For 6 months, she developed urticaria and small cutaneous ulcerations on the fingers. She was then referred to the Department of Dermatology.

Physical examination of the skin revealed 1) urticaria with recurrent swellings of the skin. In a few instances, oedema of the skin and subcutaneous tissues was severe simulating thrombophlebitis. All these lesions, however, disappeared within a few hours. 2) Small nail-fold and haemorrhagic finger pulp infarcts. 3) Small subcutaneous nodules. 4) Vesicules pustules and bullae, 5 mm to 1 cm in diameter, occasionally haemorrhagic on the dorsum of the fingers and hands.

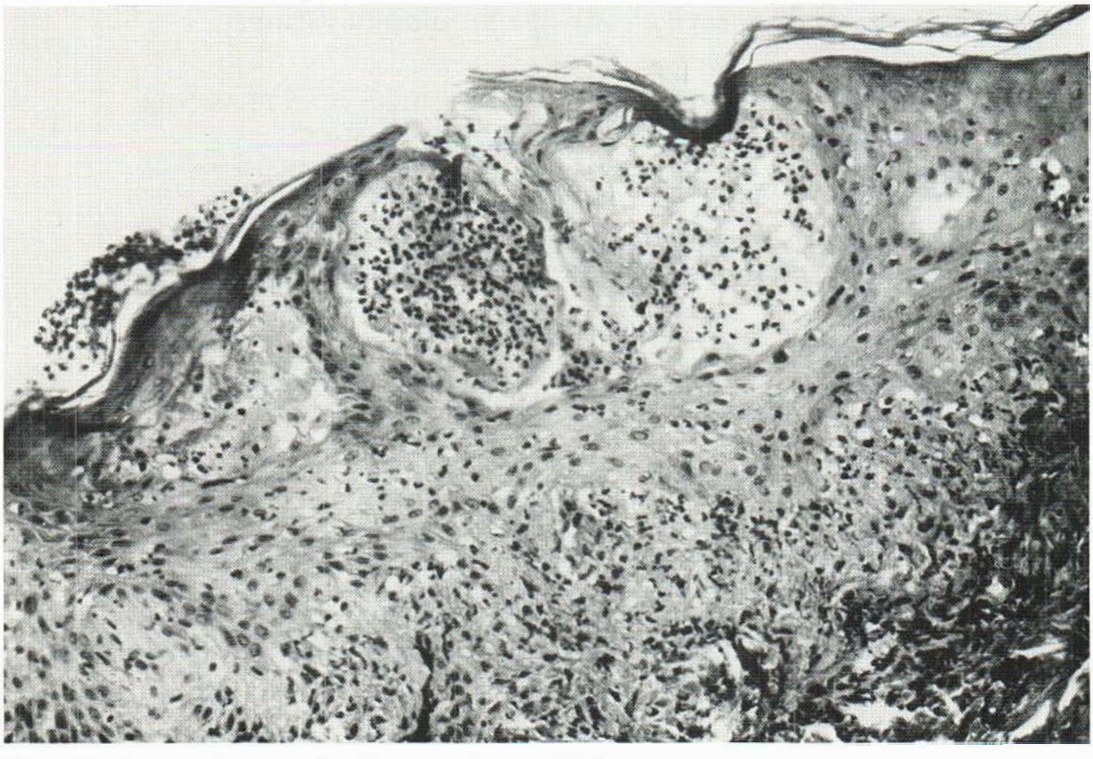

Fig. 2. Kogoj's spongiform pustule with underlying vasculitis in the upper dermis.

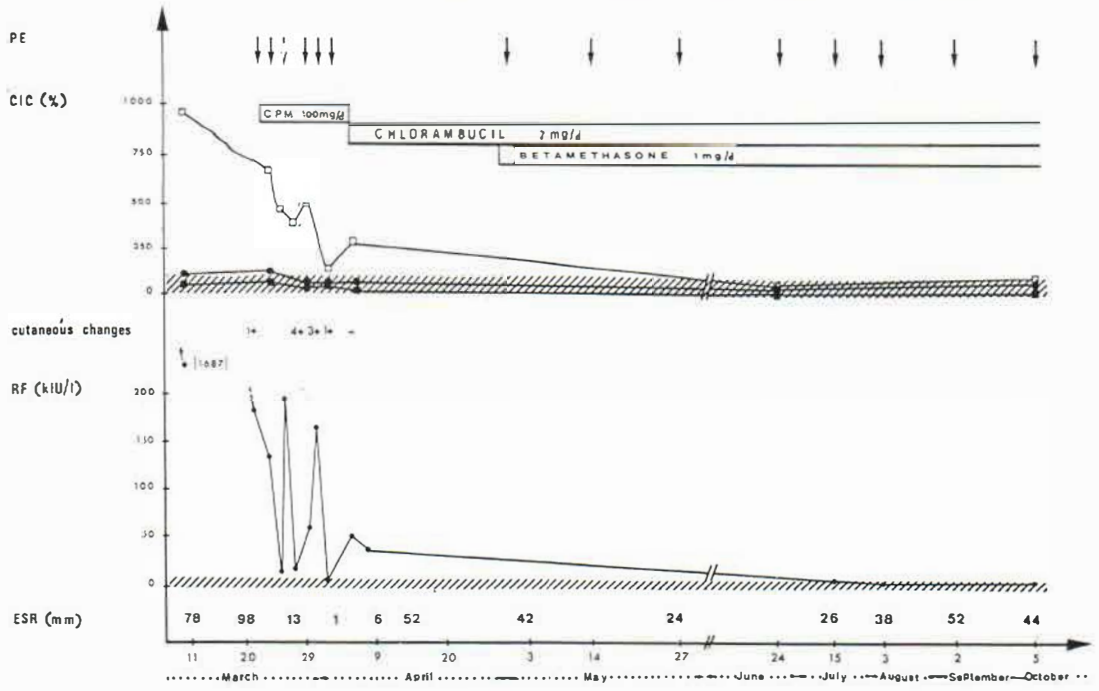

Fig. 3. Immune complexes and rheumatoid factor levels during the course of PE and IST therapy. PE: plasma immune complexes. □, CIC IgG (%); ●, CIC C4 (%); ■, CIC IgM (%). RF: rheumatoid factor assayed by RF-EIA. ESR: erythrocyte sedimentation rate. ▩, Normal range.

There was no livedo, no cutaneous ulceration or gangrene of the extremities. Other physical findings included a polyneuropathy.

Microscopic examination of a biopsy specimen taken from a pustule on a finger showed a spongiform pustule with underlying leukocytoclasic vasculitis in the upper dermis.

Laboratory evaluations demonstrated an elevated ESR and an elevated rheumatoid factor titre (1687 KUI/l) (5). Circulating immune complexes (CIC) of the IgG type were also elevated (3). Autoantibody tests and cryoglobulins proved negative. Cultures from the pustules were sterile. Other results—including complete blood cell counts, liver and renal function tests— were within normal limits. Complement levels (CH50, C3) were slightly depressed.

It was decided to treat this patient with PE and additional immunosuppressive therapy. PE consisted in plasma filtration (PLASMAFLO ASAHI Hi05), discontinuous-flow centrifugation (HAEMONETICS PEX 60) and continuous-flow centrifugation (IBM 29.97). 1.5 of the patient's plasma mass was removed and simultaneously replaced by a 4% diluted albumin solution. PE were performed three times per week for 2 weeks. This frequency was then gradually lengthened to 1 per month. IST was associated including cyclophosphamide (100 mg/day) for 13 days then chlorambucil (4 mg/day) and betamethasone (1 mg/day). Apart from shivering, no incidents or accidents occurred.

After three PE, an exacerbation of the cutaneous lesions was observed. Relief of the articular pain and improvement of the skin manifestations was observed after the sixth PE. Maintenance treatment, comprising one PE per month, chlorambucil (2 mg/day) and betamethasone (1 mg/day), completely controlled the articular disease and the cutaneous manifestations. The clinical improvement was well correlated with a dramatic reduction in CIC levels and rheumatoid factor titres.

DISCUSSION

The bullous lesions of our patient (Fig. 1) represent a very unusual manifestation of RV (4).

The pustular lesions showed the typical picture of Kogoj's spongiform pustule (Fig. 2),

as in the case reported by Miyachi et al. (6). These 2 cases suggest that RV should be included in the list of aetiological factors for pustular vasculitis.

The dramatic clinical improvement of these lesions during PE and IST was probably related to the anti-inflammatory effects of IST, but also to the reduction in the concentration of CIC. This suggests the involvement of CIC, at least to some extent, in the production of bullous and pustular skin lesions.

In our case, PE combined with immunosuppressive therapy appeared to be an excellent treatment for the cutaneous and articular lesions of RV (12). However, the ultimate value of such a treatment must await further studies.

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