Subcorneal pustular dermatosis associated with rheumatoid arthritis and raised IgA: simultaneous remission of skin and joint involvements with dapsone treatment

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Abstract

A 44 year old white woman with rheumatoid arthritis for 19 years developed subcorneal pustular dermatosis. She had increased polyclonal IgA and IgA rheumatoid factor. After 4 months' treatment with dapsone 100 mg daily the patient had neither skin lesions nor active joint disease.

Subcorneal pustular dermatosis (SPD) is a rare pustulous disease often unaccompanied by other systemic diseases. In some patients SPD has been described in association with benign monoclonal gammopathies and multiple myeloma.¹ Among the M components, IgA appears unusually often.¹ In a few other cases SPD has been associated with inflammatory diseases, such as rheumatoid arthritis (RA),²⁻⁴ systemic lupus erythematosus,⁵ and unclassified arthritis.⁶⁻⁸ Little is known about the immunological status of these patients except that they had no M component. We report a further observation of SPD associated with RA. The patient had an increased polyclonal IgA and IgA rheumatoid factor. Both RA and SPD were remarkably improved by dapsone treatment.

Case report

A white woman aged 44 with RA for 19 years had had a symmetrical erosive polyarthritis, which responded poorly to gold salts, D-penicillamine, hydroxychloroquine, and, finally, low dose steroids. Rheumatoid factor had first been detected eight years after onset (latex test: 1/1280, Waaler-Rose test: 1/256). At that time antinuclear antibodies had not been found. In July 1986 she complained of a pruritus soon followed by a bullous rash in the back. One month later the skin disease continued and the patient was admitted to hospital. On admission the skin lesions involved the trunk, flexural areas, and the proximal part of the limbs (fig 1). There were no lesions on the face, scalp, palms, and soles, nor on the mucous membranes. The eruption, which spread in a centrifugal gyrate pattern, consisted of flaccid pustulous elements, 5 to 10 mm in size, located mainly at the periphery. Pustules were sterile and direct immunofluorescence showed no immunoglobulin deposits. Histological analysis of a recent lesion showed subcorneal unilocular pustules, mainly filled with neutrophils (fig 2). All these features appeared characteristic of SPD. At the same time the RA flared with active synovitis of several joints, including the left knee and right ankle, with a morning

stiffness of one hour. The patient also had fixed deformities of hands and feet, and atloido-axoidal dislocation. Biological examinations showed haemoglobin 120 g/l; leucocytes $4.6 \times 10^9/l$, polymorphonuclear leucocytes 72%, lymphocytes 14%, platelets $213 \times 10^9/l$, erythrocyte sedimentation rate 52 mm/1st h, latex 1/80, antinuclear antibodies 1/640, without anti-double-stranded DNA (*Crihidia* L), raised gammaglobulins with an IgA of 8.10 g/l, IgG 22.9 g/l, IgM 1.77 g/l. The records mentioned an increased IgA eight years earlier (11.3 g/l). No M component or monoclonal light chain was detected on immunoelectrophoresis of blood and urine. IgA rheumatoid factor was found at 9.7 U/ml by an enzyme linked immunosorbent assay (ELISA) (normal <1.36 U/ml). It had not been looked for previously. The HLA groups were A2, A10, B12, B16, DR2, DR4. Dapsone (100 mg daily) was given and the pruritus disappeared in 48 hours; no new eruption was seen thereafter. The recovery of the skin lesions was complete by day 12. At that time the synovitis had improved markedly. After four months' treatment with dapsone the patient had neither skin lesions nor active joint disease.

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Figure 1: Typical distribution of the lesions (groins and abdomen). Note annular and circinate patterns, and left knee swelling.

Figure 2: Histological findings (high power view): subcorneal pustule filled with polymorphonuclear leucocytes.

Discussion

Subcorneal pustular dermatosis was described in 1956 by Sneddon and Wilkinson, who separated SPD from other, unclassified, pustular eruptions.⁹ It is a rare disease characterised by flaccid pustules, which have a tendency to coalesce, forming annular and circinate patterns. The hallmark of SPD is a pustule, which is found exclusively in the subcornea and filled with polymorphonuclear leucocytes.

Usually, an immunofluorescence study of the skin is negative, but in rare and atypical cases intercellular, intrapustular, or subcorneal depositions of IgA have been reported, the significance of which is unknown.¹ Mild acantholysis may be found in older lesions. This finding, in association with IgA deposits in the epidermis, has occasionally suggested diagnosis of superficial pemphigus.¹ In our case the clinical and histological features were characteristic of SPD, immunofluorescence was negative, and D-penicillamine was excluded as a possible cause of the dermatosis as it had been withdrawn several years before onset.

This observation is similar to the description of Weiner,⁴ who reported the first case of SPD associated with RA. Our case provides some additional considerations which may be relevant to the wider issue of dapsone treatment in RA.

When SPD is associated with joint diseases the skin lesions and the articular involvement usually follow the same course.^{4 5 7} When arthritis precedes SPD the skin lesions develop

with a flare of synovitis.^{4 5} Both improve concomitantly with dapsone treatment,^{4 5} as reported in our patient. Dapsone, however, is not considered to be a major treatment of RA in general. One study rated it less efficient than hydroxychloroquine.¹⁰

Our patient had IgA abnormality—that is increased blood concentrations and IgA rheumatoid factor, which extends the IgA-SPD relation to patients with SPD-RA. The precise role of IgA in SPD has been widely discussed,¹ and the significance of IgA rheumatoid factor in RA is not fully understood.¹¹ It is noteworthy that pyoderma gangrenosum, as well as SPD, has been described in association with RA.¹² Moreover, case reports of associated pyoderma gangrenosum, SPD, and IgA paraproteinaemia have been published.¹³ Finally pyoderma gangrenosum and SPD belong to a wide spectrum of neutrophilic dermatosis, closely related by their similar associated diseases. In our patient the raised IgA preceded SPD by several years as is probably the case for IgA M components in other patients. The IgA might have promoted SPD, rather than the other way round. Similar IgA abnormalities might have been overlooked in SPD associated with arthritis as most case reports mention only the absence of M component. If this is the case IgA abnormalities might have some predictive value for a good response to dapsone. Subcorneal pustular dermatosis associated arthritis and possibly RA in general might prove more responsive to dapsone in the presence of an increased IgA or IgA rheumatoid factor.

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