Subcorneal Pustular Dermatosis Accompanied by Seronegative Arthritis

Hiroshi Nagai\textsuperscript{1,2} and Susumu Harada\textsuperscript{1}

\textsuperscript{1}Department of Dermatology, Sanda City Hospital, Hyogo, and \textsuperscript{2}Division of Dermatology, Clinical Molecular Medicine, Faculty of Medicine, Kobe University Graduate School of Medicine, 7-5-1 Kusunoki-cho, Chuo-ku, Kobe 650-0017, Japan. E-mail: hnagai@med.kobe-u.ac.jp

Accepted May 3, 2002.

Sir,

Subcorneal pustular dermatosis (SPD) is a rare disorder characterized by a chronic benign and relapsing vesicopustular eruption that generally affects the trunk, particularly at flexural sites (1). Despite marked improvements in investigative techniques, the pathogenesis is still controversial. In some patients, SPD has been described in association with monoclonal gammopathies (2) and multiple myeloma (3). Furthermore, in addition to other neutrophilic dermatoses (Sweet’s syndrome, pyoderma gangrenosum and erythema elevatum diutinum), SPD has been reported to be associated with inflammatory diseases such as rheumatoid arthritis (RA) (4) and Crohn’s disease (5). Here we describe a patient with a long history of SPD with arthritis that was non-typical RA. We found only one report in the literature similar to our case. SPD accompanied by seronegative arthritis is extremely rare.

CASE REPORT

A 33-year-old woman presented with a 20-year history of skin eruption and joint manifestation. When she was 13 years old, she developed a symmetrical rash beginning on the extensor surface of both knees. Skin lesions increased in number and were disseminated over almost her entire body, except for her head. Joint manifestations with occasional swelling and dull pain began in elbows, knees, wrists and fingers, almost at the same time as the occurrence of the skin lesions. Thereafter, the skin lesions and joint manifestations repeatedly appeared and disappeared. On examination, flaccid pustules and vesicles were present on her trunk and extremities. Nikolsky’s sign was negative. There were no mucosal lesions. Examination of the joints revealed mild tenderness and swelling of some joints including elbows, knees, wrists and hands. In particular, swelling and tenderness of the right third proximal and distal interphalangeal joint were marked.

Skin biopsy histology showed subcorneal pustules composed of neutrophils, without acantholysis. A direct immunofluorescence study was negative. Bacterial cultures of swabs from skin were also negative. The findings were consistent with SPD, but not with IgA pemphigus. A full blood count and biochemical screening were within the normal ranges. Rheumatoid factor, antinuclear antibody and anti-DNA antibody were negative. Serum IgG level was 2,520 mg/dl (normal range, 870–1,700), IgA 420 mg/dl (normal range, 110–410), IgM 297 mg/dl (normal range, 46–260). On the basis of a diagnosis of SPD, the patient was treated with 75 mg dapsone, daily. Skin lesions and arthralgia responded promptly to dapsone, and resolved completely within 5 days. At present the patient’s condition is good, with few skin lesions and very little arthralgia under daily treatment with 25 mg dapsone.

DISCUSSION

In the literature 5 cases of SPD with seropositive arthritis have been reported and 3 cases of SPD with seronegative arthritis (4, 6). However, as Butt & Burge (4) pointed out, the diagnosis is uncertain in 3 of these cases: one patient with seropositive arthritis who died after developing a generalized pustular rash may have had pustular psoriasis; the clinical features in 2 cases with seronegative arthritis, which occurred in reaction to infection and settled in 2 weeks, suggest pustular bacteroid (7, 8). Thus, we suggest that this is the second fully confirmed report of SPD accompanied by seronegative arthritis. The other fully confirmed case (9) was similar to our case regarding the following points: 1) onset was in childhood; 2) eruptions and arthralgia began at almost the same time; 3) skin lesions and arthralgia responded promptly and were resolved by treatment with dapsone.

Based on the laboratory data, radiographic findings and clinical course, the arthritis in our case was non-typical RA. We speculated that seronegative arthritis may have been caused mainly by aseptic infiltration of neutrophils into the joints as well as into the epidermis, since the arthralgia promptly disappeared after treatment with dapsone. On the other hand, in cases of SPD accompanied by seropositive arthritis, arthritis generally precedes skin eruptions (4, 10). Thus, the immune conditions in RA may contribute to the onset of SPD. Concerning the response of arthritis to dapsone treatment in the literature, some cases responded (10), but others did not (4). Since dapsone is not a standard medicine for RA, it seems reasonable that seropositive arthritis should not respond to dapsone and the reason that some seropositive arthritis responded to dapsone has still to be investigated. One explanation could be that dapsone diminishes the additive symptoms of arthritis due to aseptic infiltration of neutrophils as well as seronegative arthritis, but not the symptoms of RA itself.
We consider that in our case the seronegative arthritis completely differed from RA in terms of the pathogenesis. Although in previous reports both have often been lumped together as arthritis, we suggest that a distinction should be made between seronegative arthritis and RA.

REFERENCES

1. Sneddon IB, Wilkinson DS. Subcorneal pustular dermatosis. Br J Dermatol 1956; 68: 385–394.
2. Kasha EE, Epinette WW. Subcorneal pustular dermatosis (Sneddon-Wilkinson disease) in association with monoclonal IgA gammopathy: a report and review of the literature. J Am Acad Dermatol 1988; 19: 854–858.
3. Atukorala DN, Joshi RK, Abanmi A, Jeha MT. Subcorneal pustular dermatosis and IgA myeloma. Dermatology 1993; 187: 124–126.
4. Butt A, Burge SM. Sneddon-Wilkinson disease in association with rheumatoid arthritis. Br J Dermatol 1995; 132: 313–315.
5. Delaporte E, Colombel JF, Nguyen-Mailfer C, Piette F, Cortot A, Bergoend H. Subcorneal pustular dermatosis in patients with Crohn’s disease. Acta Derm Venereol 1992; 72: 301–302.
6. Lutz ME, Daoud MS, McEvoy MT, Gibson LE. Subcorneal pustular dermatosis: a clinical study of ten patients. Cutis 1998; 61: 203–208.
7. Lin RY, Schwartz RA, Lamb W. Subcorneal pustular dermatosis with polyarthritis. Cutis 1986; 37: 123–126.
8. Undar L, Goze F, Hah MM. Subcorneal pustular dermatosis with seronegative polyarthritis. Cutis 1988; 42: 229–232.
9. Olsen TG, Wright RC, Lester AI. Subcorneal pustular dermatosis and crippling arthritis. Arch Dermatol 1979; 115: 185–188.
10. Roger H, Thevenet JP, Souteyrand P, Sauvezie B. Subcorneal pustular dermatosis associated with rheumatoid arthritis and raised IgA: simultaneous remission of skin and joint involvements with dapsone treatment. Ann Rheum Dis 1990; 49: 190–191.

BOOK REVIEW

Textbook of Contact Dermatitis. 3rd edition. R. J. G. Rycroft, T. Menné, P. J. Frosch and J.-P. Lepoittevin, eds. Berlin: Springer Verlag, 2001.

This is the third and well-motivated edition of this textbook within less than 10 years. Since the second edition, the book has grown and now includes 46 chapters written by world-leading authors in their respective fields. The increased number of chapters reflects the expansion in both clinical and research interest in contact dermatitis. The book, which is up to date and covers all aspects of contact dermatitis, starts with a historical review and then continues with three sections describing the basic mechanisms of allergic and irritant contact dermatitis, molecular aspects, pathology including epidemiology and dermatotoxicology, including skin penetration and predictive testing. The last three parts of the book cover clinical aspects of contact dermatitis, diagnostic testing and, finally, extensive coverage of allergens related to specific exposure. In addition, prevention, legislation and computers in the management of contact dermatitis are discussed. The final chapter comprises a comprehensive list of patch test concentrations and vehicles for testing contact allergens.

This edition of the Textbook of Contact Dermatitis is highly recommended for reading but it is also an outstanding reference textbook for all persons active in the field of contact dermatitis.

Magnus Lindberg,
Department of Medicine, Occupational and Environmental Dermatology,
Karolinska Institutet and Stockholm County Council,
Stockholm, Sweden