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Case Report Open Access

Generalized Pustular Psoriasis Associated with Ulcerative Colitis

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Abstract

A 38-year-old Japanese woman with steroid-dependent ulcerative colitis developed generalized pustular psoriasis. Cyclosporine improved both her cutaneous lesions and bowel condition. Recent studies show that Th17 T cells are critically involved in the pathogenesis of both generalized pustular psoriasis and inflammatory bowel disease. Although molecular mechanisms seem similar, our case is the first report showing generalized pustular psoriasis associated with inflammatory bowel disease. We believe that our case may contribute to further understanding of both diseases and give us a chance to design a suitable therapy in the case of generalized pustular psoriasis complicating inflammatory bowel disease.

Keywords: Cyclosporine A; Inflammatory Bowel Disease; pustular psoriasis; ulcerative colitis

Case Presentation

A 38-year-old Japanese woman, without history of psoriasis and other skin disorders, presented with pustules and erythematous papules on her extremities. She had been treated with corticosteroids and salazosulfapyridine for ulcerative colitis (UC) for 5 years. Because her bowel condition got worse, her doctor raised the prednisolone dose from 8 mg/day to 30 mg/day. After her bowel condition stopped worsening, he administered azathioprine and reduce her prednisolone dose to 20 mg/day. One month later, she developed the rash. As azathioprine induced drug eruption was suspected, azathioprine was discontinued and prednisolone 20mg/day was continued, then she was referred to our dermatology department. Topical corticosteroids resolved the cutaneous lesions in a week, but they relapsed one month later and worsened along with a bad bowel condition and a high fever. Sheets of erythema and pustulation spread over her extremities and trunk (Figure 1). Bacterial and fungal cultures were negative. Her blood contained elevated levels of neutrophils (7139/mm^3) and C-reactive protein (2.6 mg/dl). The biopsy of an abdominal pustule showed a spongiform pustule of Kogoj formed by large collections of neutrophils in the upper spinous and granular layers (Figure 2). Epidermal changes consisted of parakeratosis and elongation of the rete ridges. The upper dermis contained an infiltrate of lymphocytes and neutrophils, migrating from the capillaries in the papillae into the epidermis. Generalized pustular psoriasis (GPP) was diagnosed, and etretinate 30 mg/day was started but was ineffective. So cyclosporine A (CSA) (150 mg/day=3.8 mg/kg/day) was added one week later. Within a week, the skin lesions and fever subsided. Neutrophils and C-reactive protein levels decreased to within normal ranges. Etretinate was tapered and discontinued after two months. Throughout these treatments, both her skin lesions and her bowel condition improved. Prednisolone was tapered and discontinued. We have continued this CSA dose for 2 years, because her bowel condition may deteriorate if we reduce it. Pustular eruptions

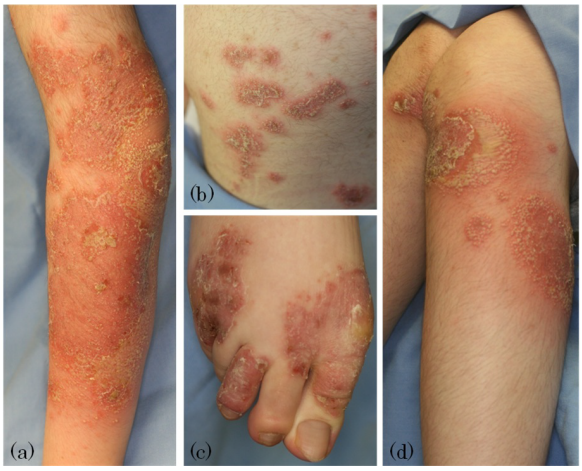

Figure 1: Clinical appearance of the skin lesions Sheets of erythema and pustulation spread over the extremities (a, c, d) and trunk (b).

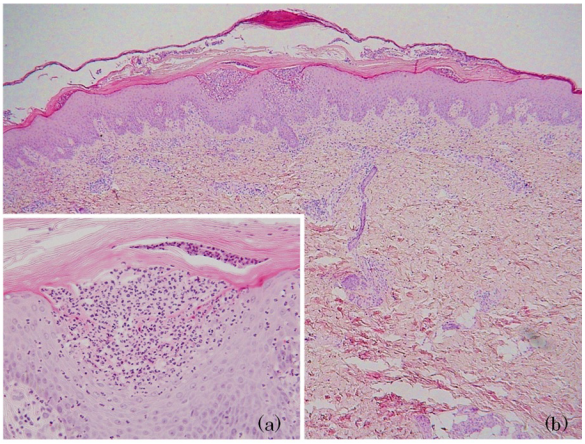

Figure 2: Pathological findings of the skin lesions
Biopsy of an abdominal pustule showed a spongiform pustule of Kogoj formed by neutrophils in the upper spinous and granular layers (a). The epidermis showed parakeratosis and elongation of the rete ridges. The upper dermis contained an infiltrate of lymphocytes and neutrophils, migrating from the capillaries into the epidermis (b).

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have not relapsed to date, and the UC symptoms are well controlled.

Discussion

GPP is a rare but severe cutaneous disease, characterized by a sudden generalized eruption of sterile pustules on a highly erythematous skin surface accompanied by a high fever [1]. Cutaneous manifestations are well-recognized complications of inflammatory bowel disease (IBD) [2]. The most common reactive skin eruptions are erythema nodosum, pyogenic gangrenosum and Sweet's syndrome [2]. An association between IBD and psoriasis has also been observed; the prevalence of psoriasis is 7-11% in IBD, compared with a general population-based prevalence of 1-2% in North America and Europe [2]. However, no reports have shown GPP accompanying IBD, except for infliximab-induced GPP in patients with IBD [3-5]. GPP is divided into two groups, one with a history of ordinary psoriasis (pso+ GPP) and the other without a history of psoriasis (pso- GPP) [1]. Systemic corticosteroids can be a precipitating factor for pso+ GPP especially at the time of withdrawal [1]. However, our patient had neither a history of cutaneous disease nor a family history of psoriasis. In addition to that, the dose of prednisolone she received when she developed GPP (20 mg/day) is much higher than her basic dose (8mg/day). Other differential diagnoses included pustular drug eruption, acute generalized exanthematous pustulosis, acute generalized pustular bacterid (Tan), pustular vasculitis and subcorneal pustular dermatosis (Sneddon-Wilkinson). Because the generalized pustular eruption appeared over one month after azathioprine discontinuation, drug eruption by azathioprine was excluded. She had taken salazosulfapyridine for five years when she suffered from generalized pustulosis, and she continues it now without cutaneous lesions, so salazosulfapyridine was excluded as well. The other differential diagnoses were excluded based on the histopathologic features. Although a relapsing course is important in the diagnosis of GPP, our case is controlled with CSA. GPP was confirmed by the clinical and pathological correlations.

In our patient, the conditions of GPP and UC seemed parallel. Both psoriasis and IBD are organ-specific autoimmune diseases which are triggered by an activated cellular immune system [6,7]. The balance between T-helper 17 cells (Th17) and regulatory T cells (T_{reg}) plays a key role. Psoriasis shares many immune-derived cytokines with IBD, including IL-17, IL-23 and TNFs. The therapeutic strategies overlap considerably. In our case, CSA controlled both GPP and UC effectively. CSA inhibits interleukin-2 production by activated T lymphocytes through a calcineurin-dependent pathway [8]. CSA is a standard treatment for GPP, as are etretinate and methotrexate, and has been used to induce remission in acute severe UC [6,8]. Thus, this case demonstrates that CSA may be a candidate treatment for this rare combination of diseases.

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