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Kaposi Sarcoma in a Patient with Bullous Pemphigoid Treated with Low-Dose Corticosteroids

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Dear Editor:

Kaposi's sarcoma (KS) is a lymphatic and vascular neoplasm, that develops in patients with human herpesvirus-8 (HHV-8) infection and presents as multiple purplish papules or nodules[1]. KS is classified into four clinical types: classic, endemic, iatrogenic immunosuppression-related, and acquired immune deficiency syndrome-associated KS. The risk of KS is increased in immunosuppressed patients owing to their lack of immunity to HHV-8[1].

Bullous pemphigoid (BP) is an autoimmune blistering disorder characterized by subepidermal bullae on pruritic and urticarial plaques. BP occurs when autoantibodies develop against BP180 and BP230 and systemic corticosteroids and/or immunosuppressive agents are required[2]. Some case reports have discussed the coexistence of KS in terms of BP[2,3]; however, no case reports from Korea have been published. Herein, we describe a case of KS in a patient with BP who had received low-dose systemic corticosteroids for 25 months.

An 84-year-old female patient presented with a 4-month history of purpuric patches and nodules on the right lower leg. She had a history of hypertension and atrial fibrillation. She presented with multiple bullae and erosions on the chest for 30 months. She was diagnosed with BP based on the findings of a subepidermal blister with granulocytes in histology, C3 deposition along the basement membrane in direct immunofluo-

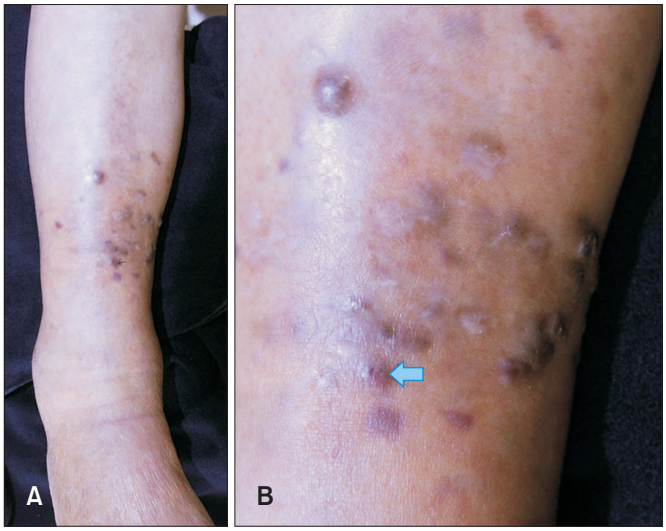

Fig. 1. (A, B) Clinical appearance of Kaposi's sarcoma skin lesions in the patient with bullous pemphigoid. Multiple violaceous patches and nodules on the right lower leg. We received the patient's consent form about publishing all photographic materials.

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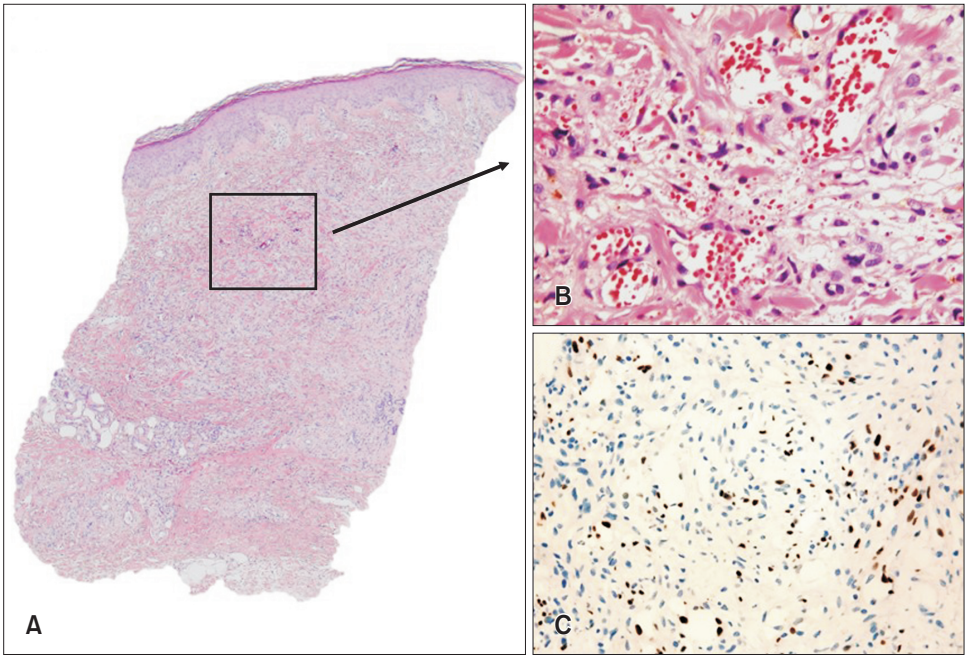

Fig. 2. (A) Scanning view of microscopy image showing diffuse dermal angiomatosis (H&E, ×10), (B) subtle slit-like vascular spaces and extravasated red blood cells (H&E, ×400), (C) positive immunohistochemistry staining for HHV-8 (×200).

rescence analysis, and IgG deposition on the epidermal side of salt-split skin at a 1:40 dilution in indirect immunofluorescence analysis. Initially, 8 mg/day oral methylprednisolone was administered for three weeks, which was tapered after disease control. No other immunosuppressive agents had been administered. She maintained complete remission of BP on 1~6 mg/day oral methylprednisolone for 24 months.

Physical examination revealed a dozen well-demarcated violaceous macules, patches, and nodules on the anterior aspect of the right lower leg (Fig. 1). Laboratory results showed no previous exposure to human immunodeficiency virus (HIV). Punch biopsy specimen of the skin was obtained from one of the nodules. Histopathologic examination revealed numerous vessels forming slit-like spaces, erythrocyte extravasation, and vessels protruding into other vessels (Fig. 2A, B). The sample tested positive for HHV-8 (Fig. 2C), D2-40, CD31, and CD34 by immunohistochemical staining. Based on these results, KS was confirmed. The patient was referred to a different hospital for radiation therapy.

The development of KS after immunosuppression for a bullous disease was first reported in 1970[4], and since then, approximately 25 such cases have been reported. The most commonly implicated agents are prednisone and azathioprine, with administration of mycophenolate mofetil being reported as a rare cause[5]. These drugs are considered to be involved in the reactivation of HHV-8[5]. In a retrospective study of 1,362 non-HIV KS patients, 14 (1.0%) patients were diagnosed with bullous disease[2]. Although it is still unknown whether this concurrence is associated or incidental, the incidence of BP in KS patients is high considering the very low incidence of autoimmune bullous diseases[2].

Here, we reported that KS developed in a BP patient despite the administration of low-dose systemic corticosteroids. The onset of KS in our patient may be attributed to immunosuppression after corticosteroid therapy. Future studies are required to confirm whether an associated mechanism exists between KS and BP.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis Secondary to Human Immunodeficiency Virus Infection: A Case Report

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Dear Editor:

Vasculitides are multifactorial and often autoimmune conditions characterized by necrosis of the blood vessels induced by infiltrated leukocytes and inflammation, causing tissue injury. Various viral infections including hepatitis B virus, hepatitis C virus, cytomegalovirus, Epstein–Barr virus, and human immunodeficiency virus (HIV) are considered common causes inducing secondary vasculitides[1].

A 29-year-old male presented with erythematous patches and papules with erosions on his chest and lower legs (Fig. 1A~C). The patient was recently diagnosed with inflammatory bowel disease. Despite treatment with 5-aminosalicylic acid, the patient had recurrent mucus and/or bloody stool, with a weight loss of 5 kg during the recent 3 months. Furthermore, skin eruptions progressed into multiple atrophic whitish scar-like patches (Fig. 1D) with focal reticulated livedo reticularis-like lesions and multiple erythematous nodules evolving into necrosis, suggestive of vasculitis (Fig. 1E, F). A biopsy from the lower leg showed typical leukocytoclastic vasculitis with eosinophils involving the upper dermal blood vessels without any granulomatous lesions (Fig. 2). Direct immunofluorescence was negative. Laboratory tests showed mild leukopenia (2,800/μl), peripheral eosinophilia (14.4% of white blood cells), and thrombocytopenia (88,000/μl) with elevated erythrocyte sedimentation rate (41 mm/h). Myeloperoxidase anti-neutrophil cytoplasmic antibody (ANCA), anti-cardiolipin immunoglobulin M, and lupus anticoagulant were also detected. Under the

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