Ann Dermatol Vol. 22, No. 1, 2010 DOI: 10.5021/ad.2010.22.1.99

CASE REPORT

A Case of Sjögren's Syndrome That Presented with Alcohol-induced Purpura

Seung-Hyun Cheong, M.D., You-Jin Han, M.D., You-Won Choi, M.D., Ki-Bum Myung, M.D., Ji-Soo Lee, M.D.[1], Hae-Young Choi, M.D.

Departments of Dermatology and [1]Rheumatology, School of Medicine, Ewha Womans University, Seoul, Korea

Sjögren syndrome (SS) is a systemic autoimmune disease that mainly affects the salivary and lacrimal glands. It may exist as a primary condition or in association with other systemic autoimmune diseases. Patients with SS usually complain of persistent dryness of the mouth and eyes and other features, including diverse general symptoms and cutaneous symptoms such as purpura. We report here on a case of 34-year-old woman who presented with purple non-blanching palpable purpura on both lower legs, and these lesions had developed soon after drinking alcohol 2 days previously. She had a 2 year history of repeatedly developing rashes in association with drinking alcohol. The physical examination showed dry eyes and dry mouth. The laboratory tests showed positivity for anti-Ro/SS-A antibody and RF and hyperimmunoglobulinemia. She was diagnosed as suffering with primary SS. Herein we report on a patient with primary SS and this patient initially presented with recurrent purpura in association with alcohol ingestion. Drinking alcohol had played a role as a possible aggravating factor for the cutaneous purpura of this patient with SS. **(Ann Dermatol 22(1) 99～101, 2010)**

-Keywords-
Alcohol-induced purpura, Primary Sjögren syndrome

INTRODUCTION

Sjögren syndrome (SS) is a systemic autoimmune disease that mainly affects the salivary and lacrimal glands. It may exist as a primary condition or in association with other systemic autoimmune diseases. These patients usually complain of persistent dryness of the mouth and eyes and other features, including diverse general symptoms and cutaneous symptoms such as purpura. Sjögren syndrome with vasculitis can be accompanied by hypergammaglobulinemic purpura of Waldenström (HGP). HGP is characterized by recurrent purpura and polyclonal hypergammaglobulinemia, positivity for RF and anti-Ro/La antibodies and an elevated ESR[1-4]. Here we report on an interesting case of primary SS with HGP that had initially presented with recurrent purpura in association with alcohol ingestion.

Received March 26, 2009, Revised July 24, 2009, Accepted for publication July 31, 2009

Reprint request to: Hae-Young Choi, M.D., Department of Dermatology, School of Medicine, Ewha Womans University, 911-1, Mok-dong, Yangcheon-gu, Seoul 158-710, Korea. Tel: 82-2-2650-5159, Fax: 82-2-2652-6925, E-mail: hychoi@ewha.ac.kr

CASE REPORT

A 34-year-old woman presented with red non-blanching palpable purpura on both lower legs and this had developed soon after drinking alcohol 2 days previously (Fig. 1). She reported that similar lesions had repeatedly developed on her lower legs for the last 2 years after consuming 1 bottle of soju (distilled liquor, 15% alcohol in 360 ml). On the past history, she had intestinal tuberculosis in 2006, which has been completely cured after her presentation to our clinic, and the rest of the medical history was not remarkable. On review of the systems, she often felt the discomfort of dry mouth, and she had frequent feelings of gravel and dryness in the eyes for several years. She didn't have any evidence of Raynaud phenomenon, joint pain, fever, headache or photosensitivity. On the laboratory examinations, the anti-Ro/SS-A antibody was positive and the serum RF and ESR were elevated. The patient showed an abnormally high immunoglobulin level with hyperimmunoglobulinemia of the serum IgG and IgA. The anti-nuclear antibody

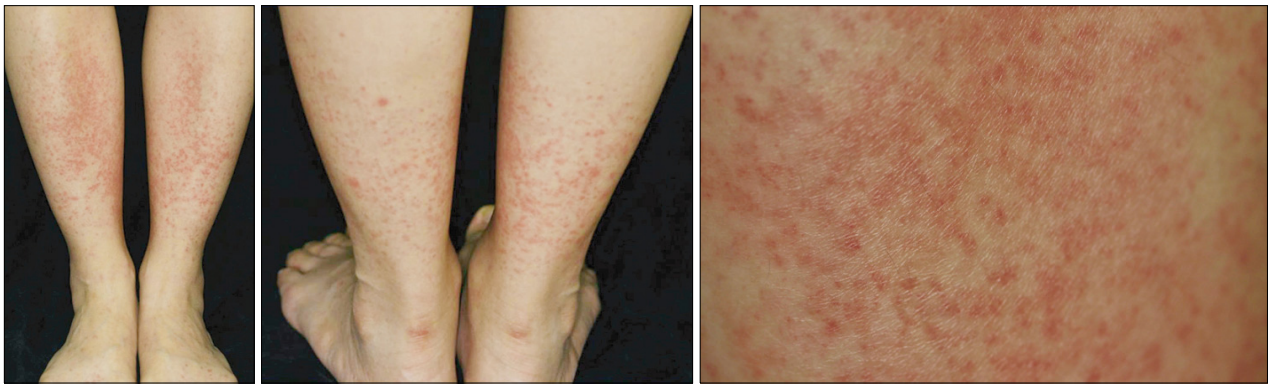

Fig. 1. Red non-blanching palpable purpura on both of her lower legs.

(ANA) showed a high titer of 1 : 640 with a speckled pattern, but she was negative for anti-DNA antibody, anti-neutrophilc cytoplasmic antibody (ANCA), anti-La/SS-B antibody and anti-Sm/RNP antibody. The salivary gland scan using technecium 99 showed decreased uptake of radiotracer on both parotid glands and both submandibular glands. The Schirmer blotting paper test showed a decreased amount of resting lacrimal fluid of her eyes. Skin biopsy of the purpuric lesion was not performed because she refused it. On the basis of these findings, the diagnosis was primary SS. According to the history of recurrent purpura with polyclonal hypergammaglobulinemia, positive RF and an elevated ESR, she was also diagnosed to have HGP in association with SS. Pilocarpine was prescribed for treatment to relieve the discomfort of her dry eyes and mouth.

DISCUSSION

Sjögren syndrome (SS) is a chronic autoimmune disease that primarily affects the salivary and lacrimal glands. It is characterized histologically by a lymphocytic infiltration of the affected glands and it is clinically characterized by ocular and oral dryness. SS is often found in association with other autoimmune disorders such as rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), dermatomyositis and scleroderma, and these cases are termed secondary SS. In the absence of any other associated autoimmune disease, this condition is classified as primary SS.

To fulfill the diagnostic criteria (the American-European Consensus Group revised criteria), patients must have objective signs of dryness on physical examination and laboratory confirmation of an autoimmune process as evidenced by positive autoantibody to Ro/SS-A, La/SS-B or both or a characteristic lip biopsy[5].

There are other features that are not included in the diagnostic criteria but that can be seen in SS, including diverse general symptoms and cutaneous symptoms. Among the various extraglandular manifestations, Ramos-Casals et al.[6] reported that cutaneous involvement was detected in 16% of the patients with primary SS, with cutaneous vasculitis being the most frequent process, and patients with cutaneous vasculitis had a higher prevalence of extraglandular manifestations such as articular involvement, peripheral neuropathy, Raynaud phenomenon and renal involvement and a higher frequency of serologic features such as an elevated ESR, hypergammaglobulinemia and positivity for RF, ANA and anti-Ro/SS-A antibody, as compared with the control group of patients without vasculitis. Those authors mentioned that most of the patients with vasculitis could be considered to have hypergammaglobulinemic purpura of Waldenström (HGP). HGP is a syndrome that was first described in 1943, and it includes recurrent purpura, hypergammaglobulinemia, positivity for RF and anti-Ro/La antibodies and an elevated ESR[1]. The purpura usually manifest on the lower part of the body after several aggravating events such as prolonged standing. HGP can be primary or associated with other connective tissue diseases, including SS[1-4].

Our patient had peculiar history of repeated development of the purpura in association with drinking alcohol. Alcohol is known to have a profound effect on inflammation and immunity by involving cytokines[7]. Therefore, it can cause allergic-type reactions and induce purpuric lesions[8,9]. In addition, Chua et al.[10] reported an interesting case of cutaneous IgA-associated vasculitis that was induced by alcohol. The patient had a 3-year history of intermittent rash on his lower legs after consuming 2 pints of beer. Those authors performed an alcohol provocation test with 10% sterile alcohol and the serial skin biopsies. The skin biopsy showed IgA, C3 and fibrin deposition in the vessel walls. Chua et al.[10] have shown that alcohol can induce a cutaneous vasculitis with IgA and C3 deposition

in a patient with an elevated IgA serum concentration. They also stated that other patients with elevated serum IgA levels might develop similar pathology after consuming alcohol.

As for our case, she had primary SS accompanied by HGP, so she showed repeated development of the purpura, which may have been triggered by alcohol. Recurrent cutaneous purpura induced by alcohol ingestion has not been reported for patients with primary SS and HGP. Therefore, it is an interesting case of primary SS with HGP, and the patient initially presented with recurrent purpura in association with alcohol ingestion. This implicated that alcohol could have played a role as a possible aggravating factor of the recurrent purpura in this patient who suffered from SS with HGP.

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