Atypical clinical presentation of lichen planus bullous in a systemic sclerosis patient

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We report the case of a 54-year-old woman with progressive systemic sclerosis who presented erythematous papules diagnosed in biopsy as lichen planus bullous (LPB).

Systemic sclerosis (SSc) is an autoimmune connective tissue disease characterized by specific antibodies, vascular abnormalities with progressive damage of blood vessels and diffuse fibrosis leading to their failure. Systemic sclerosis coexisting with systemic lupus erythematosus, rheumatoid arthritis, dermatomyositis, acquired vitiligo, Sjögren syndrome or chronic hepatitis are well documented [1].

Lichen planus (LP) is a common chronic autoimmune disease associated with immunological dysfunction. Both antigen-specific and non-specific mechanisms may be involved in the pathogenesis [2]. Several factors including stress, genetics, systemic diseases, hepatitis viruses and drugs were implicated as causative agents [3]. Clinical presentation of LP varies resulting in 20 subtypes within the disease. The bullous group was divided into LPB and LP pemphigoides (LPP), which are distinguished by clinical, histological and immunological characteristic features.

Cases of co-existing autoimmune skin disorders were described many times suggesting that one autoimmune disease may induce another.

A female patient aged 54 years suffering from SSc since 2005 was admitted to the Department of Dermatology, Medical University of Lodz. The patient complained of hand and finger joint pain, diarrhea, and dysphagia. Before developing SSc symptoms the patient was diagnosed with gastroesophageal reflux and Barrett's esophagus. The patient has been under dermatological control since 2005 due to SSc and received vasodilators and vitamin E.

In December 2011, erythematous papules accompanied by itching appeared on the forearms (Figure 1). Dermatological examination revealed flat-topped elevated papules, which were reddish-purple and Wickham's striae on lesion surfaces. Blisters were not found. In the mucous membrane of the oral cavity, lacy streaks along buccal occlusion line were observed.

Laboratory results (blood count, liver and kidney function, erythrocyte sedimentation rate, C-reactive protein, protein electrophoresis, urine test) were within normal range. The echocardiographic research revealed a slight disorder in mobility of the left ventricle walls, slight mitral and tricuspid incompetence and trace of pericardial effusion. Scintigraphic study of esophageal motility showed slow transit in the lower part. Chest X-ray and spirometry showed no pulmonary change. Results of serum sample testing for HCV antibodies and antigen HBs were negative. No casual or other provocative factors for LP were detected.

The examination of the skin biopsy revealed subcorneal bulla typical of LPB, which in clinical presentation was not observed either within papules or on the uninvolved skin. On histopathological examination LPB with increased thickness of corn and granular layers, a "saw tooth" pattern of epidermal hyperplasia, dermal-epidermal separation and a band-like lymphocytic infiltration were found (Figure 2). Indirect immunofluorescence (IIF) for anti-BMZ antibodies was negative. Direct immunofluorescence (DIF) revealed deposits of IgG (++) along basement membrane k IgG (++) against hyaline bodies (Figure 3). The autoantibody to the NC16A domain of BP180 was negative.

During hospitalization general treatment was instituted – vasodilators (intravenous infusion of 40,000

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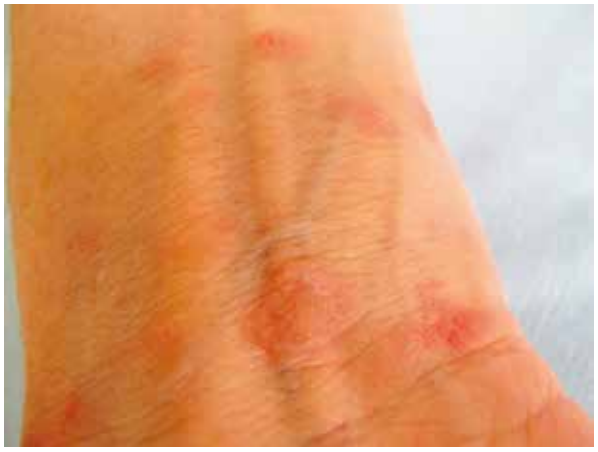

Figure 1. Clinical picture of flat-topped elevated papules with Wickham's striae

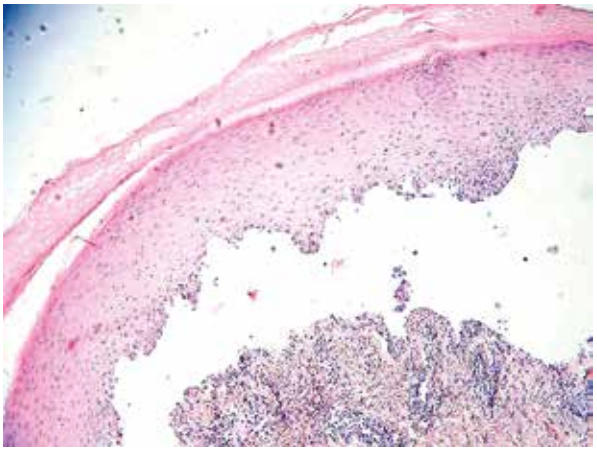

Figure 2. Histopathology examination revealed: subcorneal bulla as well as increased thickness of corn and granular layers, a "saw tooth" pattern of epidermal hyperplasia, dermal-epidermal separation, and band-like lymphocytic infiltration

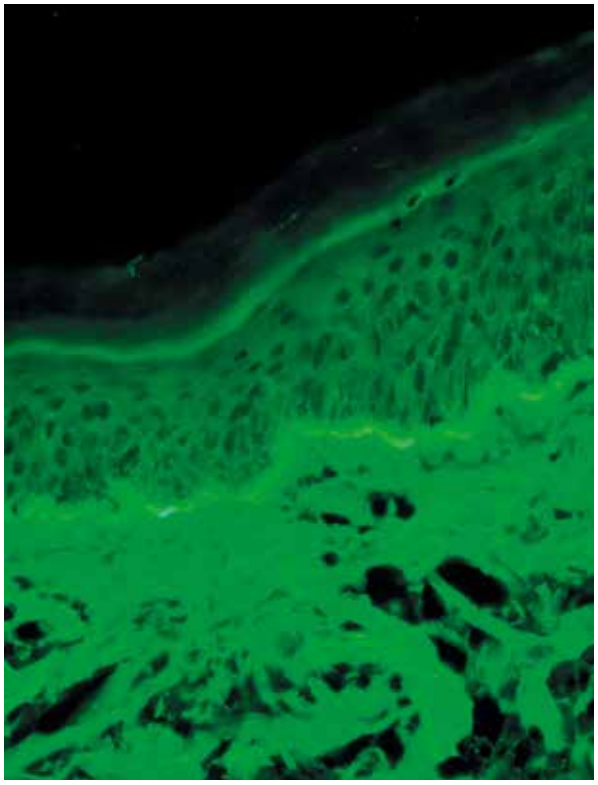

Figure 3. Direct immunofluorescence (DIF) revealed deposits of IgG (++) along basement membrane and IgG (++) against hyaline bodies

dextran and pentoxyphilline). For topical treatment, corticosteroid ointment was ordered – clobetasol; soon, the papule eruption disappeared. No new changes were observed.

Bullous subtypes of LP were first described in 1892 by Kaposi [4]. Two distinct forms of LP with bullae were defined – LPP and LPB. Before immunofluorescence studies were introduced, the differential diagnosis between LPB and LPP had been based on clinical presentation and histopathological changes.

On dermatological examination, LPB blisters are observed only on papules, placed mainly on palms and feet. Histological changes include subcorneal blisters together with typical changes for lichen planus. The course of LPB is thought to be milder than that of LPP [5]. In LPB, dissemination of blisters is observed only for a short period of time.

Clinical presentation of LPP is different with tense, subepidermal bullae on seemingly healthy skin coexisting with violaceous LP-like lesions. According to Murphy and Cronin, only very rarely blisters may be localized within the lichen planus area [6]. Lichenus planus pemphigoides develops at all ages but the age of LPP onset is lower compared with LP bullous [7] and affects more men than women (males : females 3 : 2) [8]. On histological examination, which differentiates LPP from LPB, the lack of characteristic futures for LP, apart from subcorneal blisters, is prominent and the subepidermal bulla may be not distinguishable from bullous pemphigoids (BP).

Importantly, in LPP, linear deposits of IgG and C3 along the basal membrane zone on immunofluorescence are observed in more 50%. Importantly, in LPP, IgG and C3 deposits are located on both the roof of the blister whereas in LPB only at the floor. Interestingly, LPP sera were found to react with antigens similar to BP, such as BP180 and BP230, as well as a unique band at 200 kDa [9]. It is an open question whether LPP is a type of LP or co-existence of LP and pemphigoid.

In our case, immunofluorescence on a sample taken from the papule area showed only IgG deposits along basal membrane. Although the diagnosis of LPB was

based on the hypothesis that immunoglobulin deposits on the dermal-epidermal junction may be due to immunological disorders observed in SSc. In the clinical picture of our patient, bullae within lichen papules were not found on the skin without lesions. The subject literature describes cases when papule dissemination preceded appearance of bullae by a few weeks or even months.

Topical or systemic corticosteroid medications are used in management. Low doses of corticosteroids with acitretin or dapsone may also be used. PUVA-therapy is another effective method. Successful treatment with adalimumab in resistant lichen planus was also reported [10].

Patients usually respond well to a short-term treatment.

Patients with an autoimmune disease are at a higher risk of developing a second autoimmune disorder, and their co-existence is more frequent than expected only by chance. The presented case of our patient with SSc in whom LPB symptoms developed confirms the observations described above.

Conflict of interest

The authors declare no conflict of interest.

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