Case of Crohn’s disease presenting with pemphigus vulgaris: A case report

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Abstract

Pemphigus vulgaris is an autoimmune bullous disease which is characterized by erosion and bullae on the skin and mucosae. The disease is thought to occur under the influence of predisposing factors in people with genetic susceptibility. Pemphigus may coexist with other autoimmune diseases such as myasthenia gravis, lupus erythematosus, rheumatoid arthritis, and pernicious anemia as well as with neoplasias such as thymoma, carcinoma, and lymphoproliferative diseases. Crohn’s disease is a chronic inflammatory disease which involves the whole gastrointestinal mucosa from the mouth to the anus. Etiopathogenesis of this disease includes autoimmunity. A 39-year-old Turkish man presented to our polyclinic due to the erythema on the trunk and the shoulders. The patient stated that his complaints had started a month before and were accompanied by pruritus. He had a history of Crohn’s disease for 10 years. Histopathologic examination of the blister revealed pemphigus vulgaris. Physicians should be able to recognize Crohn’s disease might accompany with autoimmune skin disease. For our knowledge this is the first case with coexistence of two diseases.

Keywords: Autoimmunity
Crohn’s disease
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1. Introduction

Pemphigus vulgaris (PV) represents a group of autoimmune bullous diseases characterized by the presence of the IgGs directed against the surface of keratinocytes in the epidermis and circulation. It is reported that pemphigus might be associated with other autoimmune diseases such as myasthenia gravis, lupus erythematosus, rheumatoid arthritis, pernicious anemia, and ulcerative colitis (UC), pemphigus may also coexist with such neoplasias as thymoma, carcinoma, and lymphoproliferative diseases (Akarsu et al., 2010). Crohn’s disease (CD) is a chronic inflammatory disease which involves the digestive tract from the mouth to the anus segmentally by leaving undamaged regions and this disease exhibits the characteristic of transmural involvement, and progresses with remissions (Ozkan,
When we consider the previous manuscripts, we seldom encounter the coexistence of PV and CD (Fabbri et al., 1986). A first case with a history of CD in whose body common post-operative bullae developed is discussed here.

2. Case presentation

A 39-year-old Turkish man presented to our polyclinic due to the erythema on the trunk and the shoulders. The patient stated that his complaints had started a month before and were accompanied by pruritus. Upon learning that he had a history of CD for 10 years and upon the observation of a lesion yielding a mass image on the right lower quadrant of the abdomen in the abdominal tomography that the patient had undergone three months before, the opaque substance was injected into the bladder and his radiography were taken. As a result of the radiography, it was seen that the opaque substance had gone into the small intestine; therefore, an enterovesical fistula was considered (Fig. 3A, B). The patient underwent right hemicolectomy to include about a 20 cm portion of the terminal ileum and it was found nuclear debris and inflammatory cells in lumen by histopathological examination and CD wasn’t active at that time (Fig. 4). It was learned that he was not receiving any immunosuppressive treatment that day. Formerly he used sulfasalazine during a six years history of CD and hadn’t used any drugs for four years. There was no family, and psychosocial history. In the dermatological examination, bullae and eroded plaques were present on both shoulders and in the periumbilical region (Fig. 1A, B). There was no attribute in the routine laboratory examinations of our patient. In histopathology, suprabasal bullae formation and areas of acantholysis with a tombstone appearance were present on the epidermis and a clear mixed inflammatory cell infiltration mostly with a perivascular location was present on the superficial dermis (Fig. 2A). And also a perilesional biopsy was done for direct immunofluorescence (DIF) and DIF has demonstrated deposits of C3 and IgG stain the cell surfaces throughout the epidermis which was found in agreement with PV (Fig. 2B). PV was active at that time so systemic steroid treatment was started with consent from the patient. We couldn’t follow-up the patient because he didn’t come to our polyclinic again.

3. Discussion

Immunotolerance is defined as the unresponsiveness of an organism to its own antigens or its creation of no immune response to these antigens. Autoimmunity develops when immunotolerance has been impaired and the organism has created a response to its own antigens. Cutaneous autoimmunity is characterized by the presence of autoantibodies and autoreactive T cells. PV is the prototype of antibody-related autoimmune skin diseases. The impairment of keratinocyte adhesion due to the autoantibody that has developed against desmosomal adhesion molecule Dsg-3 is defined as the most important immune step in the pathogenesis of the disease (Dogan and Atakan, 2013).

CD is seen together with the abnormal production of the proinflammatory cytokines mediated by T lymphocytes. In CD, inflammation starts on the bowel wall in a sensitive person under the influence of triggers. In normal people, diets or microbial antigens stimulate the elements of the immune system located on the bowel wall, thereby causing inflammation within physiological boundaries. This inflammation is suppressed again by the immune system and the destruction of the intestines is prevented. Starting in people with CD, this immune response is not suppressed but proceeds increasingly (Ozkan, 2003). We couldn’t see any case report of Crohn’s disease presenting with pemphigus vulgaris in literature. The
most prevalent skin finding in the inflammatory bowel disease (IBD) is erythema nodosum. Its incidence is reported as 4-6%. It is more prevalent than ulcerative colitis in CD. The IBD is present in 20-30% of the cases of pyoderma gangrenosum; however, the incidence of pyoderma gangrenosum in IBD cases is rather low. Its general incidence in the IBD is reported to be 0.6-2.2%, although being lower with CD. Pyostomatitis vegetans is more rarely seen in the IBD. It is characterized by pustules and ulcerations and seen on lips and in the buccal mucosa. Cutaneous polyarteritis nodosa is again a relatively uncommon case, and its coexistence with CD was reported in fewer than 20 cases. Acute neutrophilic dermatosis (Sweet syndrome) is characterized by painful erythematous lesions which occur suddenly and are accompanied by fever and neutrophilia, and a papule, nodule or plaque might occur. Lesions might emerge on the hands, arms, upper abdomen, nape, neck or face asymmetrically. Here is a neutrophil infiltration which is not accompanied histologically by vasculitis. Likely to be associated with systemic diseases, this syndrome might also be associated with CD; nevertheless, it is rather rare (Hatemi, 2013).

When considering the literature IBD were reported with autoimmune bullous dermatosis in 49 cases. We most frequently see the coexistence of UC and Linear IgA Dermatosis (LAD) in 20 cases as well as the coexistence of LAD and CD in 6 cases, of CD and bullous pemphigoid in 2 cases, and of CD and mucosal membrane pemphigoid in one case out of them (Shipman et al., 2012; Yamada et al., 2013).

T cells, particularly CD4 T cells are responsible for IBD. The dysregulation between regulatory T cells and CD4 T cells exists in its pathogenesis. This dysregulation causes inflammation in the intestines. Warning started major antigen is flagella antigen secreted by bacteria. In addition, the tumor necrosis factor and secretion of various cytokines such as interleukins 4 and 17 lead to inflammation, tissue damage, and the presentation of autologous antigens to the inflammatory cells in the IBD. Result of inflammation, it is thought that such antigens as BP180, BP230, and desmogleins which are responsible for autoimmune bullous dermatosis, are secreted from the colon epithelium and presented to T cells as antigens (Blumberg, 2009).

CD was observed first and autoimmune bullous dermatosis developed later in all cases in the literature. CD was present in our patient first, which was in agreement with the literature. Moreover, in a previous study, the period between CD and the development of autoimmune bullous dermatosis was reported to range from 6 weeks to 3 years on average (Blumberg, 2009).

In the study by Paige et al. (Paige et al., 1997), LAD developed in the patient with ulcerative colitis 43 years later. In a study of Harrison et al. (Harrison et al., 1996), bullous pemphigoid developed in the patient with ulcerative colitis 10 years later. The period was also 10 years in our case, which is similar to those reported previously.

In conclusion, autoimmunity is a picture which develops depending on the antibodies produced by the organism itself that perceives its own proteins as antigens for some reason. Supposing that the diseases
occurring with other autoimmune mechanisms might accompany in cases with any autoimmune disease, we intended to present the coexistence of PV in a patient with a history of CD for 10 years.

REFERENCES


