Multiple autoimmune syndrome

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ABSTRACT

We report a case in which the presence of dermatological autoimmune conditions, vitiligo and alopecia areata, led to the diagnosis of a systemic autoimmune disease, ulcerative colitis.

Key Words: Cutaneous autoimmune diseases, Ulcerative colitis, Multiple autoimmune syndrome

INTRODUCTION

The combination of at least three autoimmune diseases in the same patient has been defined as multiple autoimmune syndrome (MAS).[1-3] In this unusual condition, dermatological autoimmune diseases and especially vitiligo have an important place.[4] Ulcerative colitis (UC) is a chronic inflammatory bowel disease (IBD) of unknown etiology. Familial or genetic, infectious, immunologic, and psychological factors have been implicated. Abnormalities of both humoral and cell-mediated immunity have been described. Patients with a clustering of autoimmune diseases may help to delineate the pathogenesis of ulcerative colitis.

CASE REPORT

A 28-year-old Indian woman presented with hypopigmented macules in December 1999. Within one month they became depigmented. The lesions were extensive and involved the scalp, right palm, front and back of the chest and abdomen. She was diagnosed as having vitiligo and treated initially with a topical steroid and later with PUVA therapy. She also had alopecia areata on the scalp.

In early January 2001, the patient presented with acute abdominal pain and blood stained stools. There was no response to antibacterial and antiparasitic treatment. Since the patient already had two autoimmune diseases, the possibility of a third one was considered and a diagnosis of IBD was entertained. Colonoscopy showed pancolitis. A biopsy confirmed the diagnosis of ulcerative colitis. She had a hectic course in hospital. Methylprednisolone had to be given in large doses. She is now in remission and is on a maintenance dose of 5-aminosalicylic acid.

DISCUSSION

Disorders of autoimmune pathogenesis occur with increased frequency in patients with a history of another autoimmune disease. The tendency to develop another disease occurs in about 25% of these patients. The definition of multiple autoimmune syndrome is based on 91 reported cases of such associations in the literature. MAS can be classified into three groups according to the prevalence of their associations with one another.[2] Type 1 comprises myasthenia gravis, thymoma, polymyositis and giant cell myocarditis. Type 2 includes Sjögren's syndrome, rheumatoid arthritis (RA), primary biliary cirrhosis (PBC), scleroderma and autoimmune thyroid disease. Type 3 groups together autoimmune thyroid disease, myasthenia and/or

thymoma, Sjögren's syndrome, pernicious anemia, idiopathic thrombocytopenic purpura (ITP), Addison's disease, insulin-dependent diabetes, vitiligo, autoimmune hemolytic anemia (AIHA), systemic lupus erythematosus (SLE) and dermatitis herpetiformis. For this group, HLA-B8 and/or -DR3 or -DR5 seems to be an important factor.

Other conditions found in various combinations in MAS are: pemphigus and autoimmune thyroid disease in type 1 MAS; chronic active hepatitis (CAH), SLE, pemphigus, bullous pemphigoid, AIHA, ITP, alopecia areata and Addison's disease in type 2 MAS; and acquired primary hypogonadism, hypophysitis, RA, PBC, relapsing polychondritis, multiple sclerosis, CAH, ulcerative colitis, and scleroderma in type 3 MAS. This classification helps to detect a new condition liable to appear in a patient who has had two previous autoimmune diseases. It provides a basis for analysis of the pathophysiological mechanisms of autoimmunity.

The pathogenesis of multiple autoimmune disorders is not known. Environmental triggers in a genetically susceptible individual are believed to cause disorders of immune regulation. In animal experiments multiple autoantibodies have been shown following cytomegalovirus infection. Multiple autoantibodies can be found in a patient and some of the specific mono- or polyclonal autoantibodies may be multiple organ reactive.

Autoimmune phenomena may be prominent in inflammatory bowel disease. Ulcerative colitis, in particular, exhibits a high incidence of associated autoimmune diseases, including hypothyroidism, primary sclerosing cholangitis, vitiligo, and alopecia areata.[5] Jewell et al found that at least one autoimmune disorder was present in 9% of patients with UC, 2% with Crohn's disease and 2% in controls.[6] These results provide further indirect evidence of the involvement of autoimmune mechanisms in the pathogenesis of ulcerative colitis. The pathogenetic mechanism might be organ-specific cellular antigen(s) shared by colon and extracolonic organs.

In conclusion, the presence of one autoimmune disease should alert one to watch for another one. The occurrence of multiple autoimmune phenomena in this case indicates the need for continued surveillance for the development of new autoimmune disease in predisposed patients.

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