Coexistence of Behçet's Disease and Ankylosing Spondylitis: A Case Report

Behçet Hastalığı ve Ankilozan Spondilit Birlikteliği: Olgu Sunumu

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Abstract

Whether Behçet's disease (BD) is of the seronegative spondiloartropathy (SSpA) group has been a subject of debate for many years. On the other hand, the number of reported cases of coexisting BD and a disease of SSpA group has increased. Here we presented a patient with ankylosing spondylitis (AS) and BD to discuss the coexistence of these two diseases. *(Rheumatism 2008 23: 69-71)*

Key words: Ankylosing spondylitis, Behçet's disease

Özet

Behçet hastalığı (BH)'nın seronegatif spondiloartropati (SSpA) grubunda yer alıp almadığı uzun yıllardan beri tartışılmaktadır. Bu konu henüz tam olarak açığa kavuşmamış olsa da BH ve SSpA grubu hastalık birlikteliğini içeren olgu sunumları zaman içinde artmaktadır. Bu yazıda BH ve SSpA grubunun prototipi olan ankilozan spondilit (AS)'ye sahip bir olgu sunulmuştur. *(Romatizma 2008; 23: 69-71)*

Anahtar kelimeler: Ankilozan spondilit, Behçet hastalığı

Introduction

Behçet's disease (BD) is characterized with oral or orogenital ulcers and various systemic (eye, skin, joint, central nervous system, and blood vessels) symptoms. The basic anatomical lesion is vasculitis (1,2). Ankylosing spondylitis (AS), a prototype of seronegative spondiloartropathy (SSpA) group, is a chronic inflammatory disease of the axial skeleton primarily involving the sacroiliac joint and vertebra. The coexistence of BD and AS has been rarely reported. Whether BD is one of SSpA group and whether BD progresses with sacroiliitis development have been subjects of debate (2).

Case

A 29-year-old male patient applied with low back and neck pain of 7 years, increasing in severity in the last 3 years. The pain was inflammatory and intensified at rest and subsided with motion. The patient also suffered morning stiffness for an hour. He described recurrent oral aphtae, genital ulcers, and wide spread papulopustullary lesions of 3 years. There was no history of pain or swelling in his joints. The patient received a diagnosis of posterior uveitis and papillary edema from the ophthalmology clinic where he applied a week ago with the complaints of redness and discomfort in the eye. There was no family history of rheumatic disease. On physical examination, his spinal motions were painful and limited (the chin manubrium stern distance was 8 cm; tragus wall distance was 12 cm; finger floor distance was 34 cm; schober was 2 cm, and chest expansion was 2.5 cm). Pathergy test was negative. Complete blood count revealed normal counts of white blood cells and platelets and hemoglobin: 10.4 g/dl. Blood iron level was low, while iron binding capacity was normal, which was evaluated as chronic disease anemia. Erythrocyte sedimentation rate: 36 mm/h, C-reactive protein: 12 mg/L (normal<5), and rheumatoid factor negative. Routine biochemistry test results, immunoglubolines, and complements were within normal limits. HLA B51 was positive, while HLA B27 was negative. Anteroposterior pelvis radiography revealed stage 3 sacroiliitis on the left side, stage 2 sacroiliitis on the right side (Figure I). Lateral view of the servikal spine is seen in figure II. Considering all the findings, the patient was diagnosed as AS based on New York criteria (3) and BD based on International Study Group criteria (4). Then, the patient was started on colchicine 1 mg/day, 150 mg/day indomethacin.

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Discussion

Literature reports a few cases of BD and AS coexistence. Whether BD is one of SSpA group and whether BD progresses with sacroiliitis development have been subjects of debate (2). Oliveri et al (5) reported a 45-year-old female patient with BD and AS coexistence. In that case, cervical spine was more severely involved than the sacroiliac joints. The authors emphasised that in the coexistence of BD and AS, servical spine is more severely and earlier affected than the lumbar spine and sacroiliac joints. This might be related with more marked cervical involvement in female patients with AS. In our patient, sacroiliac joint involvement was more marked than cervical involvement. Beiran et al (6) reported a 33 -year-old male patient with painful red eye. They observed that in the period when the patient had AS clinic at the onset of the disease, anterior uveitis, an ophthalmic finding of AS, was added by pan uveitis and retinal vasculitis specific to BD when BD was also included in the clinical picture. Our patient did not suffer from anterior uveitis specific to AS, but had posterior uveitis and papillary edema associated with BD. Kallel et al (1) reported 2 cases of BD and AS coexistence. They concluded that BD and AS coexistence was rare and in BD cases with positive HLA B27, there might be a higher incidence of AS. In our case, HLA B27 was negative. In a case of Borman et al (7), a 29-year-old female patient was followed up with AS diagnosis and clinical deterioration in her condition was added by BD. Thus, the authors have emphasized in a case followed up for diagnosis of AS or BD, if the clinical progression of the disease is worse than expected, the other disease might join in the clinical picture. Similarly, in our patient, the deterioration in the clinical progression of the disease was noted in the last three years when BD developed. The 36-year-old male patient reported by Cimen et al (8) had BD of 16 years, which was added by AS in the last 1.5 years. The researchers attributed the late onset of AS to the drugs used by the patient for BD (systemic corticosteroid, NSAI, cyclophosphamide). Etaouil et al (9) reported 2 cases with BD and AS coexistence, and suggested that positive HLA B51 could be less common in those with BD alone than in those with BD and AS. However, in our patient, despite BD and AS coexistence, HLA B51 was positive, while HLA B27 was negative.

Chang et al (10) evaluated 58 cases with BD, 56 cases with SSpA, and 3 cases with AS and BD clinic and conducted intergroup comparisons including the control group. They found that none of the BD patients had HLA B27, while sacroiliitis or enthesitis had slightly increased in severity in those with BD than in the controls. In the patients with BD, positive HLA B51 was significantly higher than in those with a disease of SSpA group; thus, supporting the hypothesis that BD is not of SSpA group. In 2 of the 3 cases with clinical features of BD and AS, HLA B51 and B27 were positive; however, clinically, one of the patients was diagnosed with BD and AS coexistence, while the other was more like AS. In the third patient, despite positive HLA B27, the clinical features were suggestive of BD. Kotevoglu et al (11) detected late onset AS in 2 of the 135 BD patients. In their study, AS showed an unusual progression, with amyloidosis and renal failure in the first case, which are not common findings in AS, and positive HLA B51 and negative HLA B27 in the second case, which are also not common in AS.

In conclusion, although BD and AS coexistence is a rare entity, the number of cases reports about coexisting BD and AS, a prototype of SSpA group, has been increasing. In the light of the relevant literature, rather than speculating whether BD belongs to SSpA group, evaluating coexistence of BD with a disease from this group (particularly AS) seems more feasible.

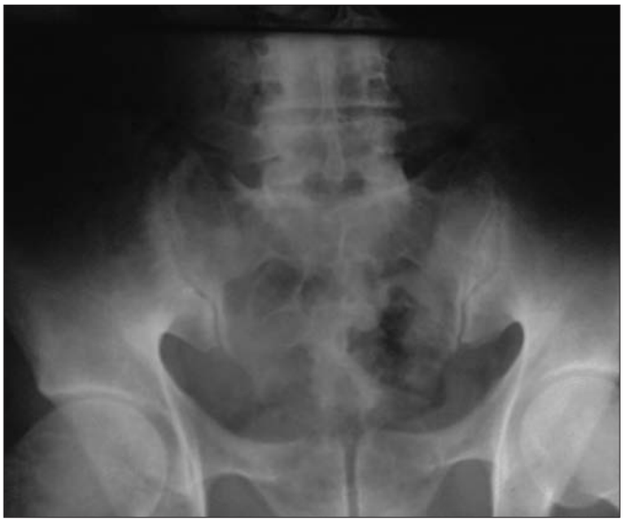

Figure 1. Anteroposterior pelvis radiography: Stage 3 croiliitis on the left side and stage 2 sacroiliitis on the right side

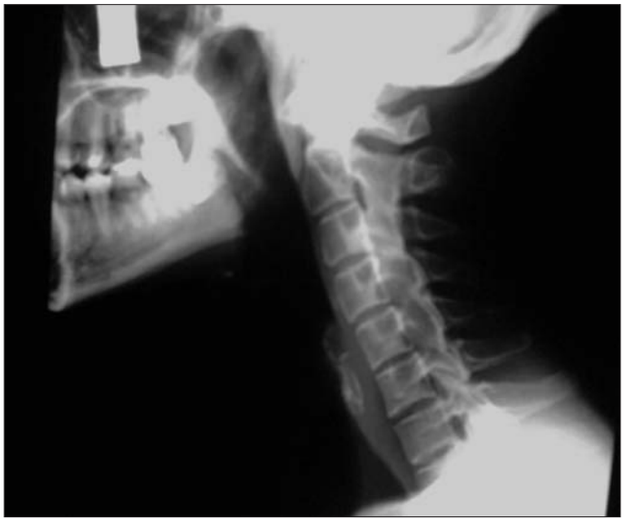

Figure 2. Lateral cervical radiography: Straightening of cervical lordosis

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