

Ankilozan Spondilit ve Behçet Hastalığı Birlikteliği: Olgu Sunumu

A Case of Behcet's Disease and Ankylosing Spondylitis Coexistence

Rabia Terzi¹, Nese Olmez², Asuman Memis², Nazife Sefi Yurdakul³

¹*Kocaeli Derince Training and Research Hospital, Department of Physical Medicine and Rehabilitation*

²*Izmir Ataturk Training and Research Hospital, Department of Physical Medicine and Rehabilitation*

³*Izmir Ataturk Training and Research Hospital, Department of Ophthalmology*

Özet

Bu olguda ankilozan spondilit ve takibinde Behçet hastalığı kliniği gelişen 35 yaşında erkek bir olgu sunulmuştur. Olgu modifiye Newyork kriterlerine göre ankilozan spondilit tanısı almıştır. Oküler tutulumda hipopiyonlu iridosiklit ve retinal vaskülit gelişen olguda HLA-B27 pozitif, HLA-B5 negatif olarak bulunmuştur. Bu olguda ankilozan spondilit ve Behçet hastalığı bir arada gözlenmiş ve birlikteliği literatür eşliğinde tartışılmıştır.

Anahtar Kelimeler: Ankilozan spondilit, Behçet hastalığı

Kısa Başlık: Ankilozan spondilit ve Behçet hastalığı

Abstract

We presented a 35 year old case who was diagnosed as ankylosing spondylitis (AS) and developed clinical features of Behcet's disease (BD) during his follow-up. He fulfills the Modified NewYork criteria for AS. He had an ocular involvement which was diagnosed as iridocyclitis with hypopyon and retinal vasculitis. His phenotype was positive for HLA-B27 and negative for HLA-B5. In this case, a combination of AS and BD has been observed and discussed.

Key Words: Ankylosing spondylitis, Behcet's disease
Running Title: Behcet's disease and ankylosing spondylitis

İletişim Adresi:

Rabia Terzi / Kocaeli Derince Eğitim ve Araştırma Hastanesi / İbni Sina Bulvarı Derince/KOCAELİ
Tel: (0262) 3178000-8110 Mail: drrabia1@yahoo.com

Introduction:

Behcet's disease (BD) is a systemic vasculitis of unknown cause involving arteries and veins of all sizes and having recurrent mucocutaneous and frequent ocular involvement(1). Ankylosing spondylitis (AS) is the main representative of the family of spondyloarthropathies, and a chronic inflammatory joint disease of axial joints and in particular both sacroiliac joints (2).

There has been some debate as to whether BD should be included among the seronegative spondylarthritides. Back pain is far uncommon, and properly conducted studies of sacroiliac joint involvement have not demonstrated an increased prevalence in BD patients. The lack of familial association with other diseases in the seronegative spondylarthritides (SpA) and the association with HLA-B51 rather than the B27 cross-reactive group suggest that BD is not a sPA. Most importantly, the clinical features in BD are very different: there is widespread vasculitis; the genital lesion in the male usually affects the scrotum rather than the glans; urethritis is absent; nail changes are not seen; and the nature and course of eye involvement are dissimilar (1).

In literature, there are case reports about coexistence of BD and AS and a case of BD (3–6). In this report we present a case who were diagnosed as AS and during follow up developed BD. Our case meets the criteria of AS according to modified Newyork criteria and BD according to International Study Group Criteria (ISGC).

Case

Thirty-five year-old male patient admitted to our clinic with complaints of morning stiffness, low back pain, persistent right knee swelling, oral aphthous ulcerations. The first complaints of our patient started when he was 19 year-old. He had inflammatory low back pain, hip pain and treated with NSAIDs. After 6 years he was diagnosed as AS. He had right knee swelling, calf tenderness, difficulty in walking and oral aphthous ulcerations occurring 10 to 15 times a year. The patient did not give any history of genital ulceration, urethritis or preceding infection at that time. He

was started on indomethacin and sulfasalazine and corticostereoids.

When the patient was 31 year old, total hip replacement surgery was performed because of both hip joint ankylosis.

Nine years after his first symptoms the patient had ocular involvement which was diagnosed as uveitis. Patient had pain and redness of right eye. In the last year vision loss of right eye occurred. The examination at this time revealed iridocyclitis with hypopyon and retinal vasculitis. One month's later, on biomicroscopic examination there were iris pigments on right eye lens and no other signs of active iridocyclitis. Fundus fluorescein angiography showed edema of optic disc, widespread preretinal and subretinal hemorrhages. Vision was intact, there were iris pigments on right eye lens and anterior and posterior chambers were normal at 4 month's follow up.

On physical examination there were loss of lumbar lordosis, papulopustular skin lesions on chest and back, limitation of lumbar spine mobility in all three planes and right knee swelling. Sacroiliac compression tests were negative. Spinal mobility measurement were performed; Modified Schober test was 2 cm, chest expansion was 1,5 cm and occiput-wall distance was 6 cm. There was right knee effusion. Laboratory evaluation revealed sedimentation rate 23 mm/h, CRP 1,81 mg/dl, rheumatoid factor negative. HLA phenotype was positive for HLA-B27 and negative for HLA-B5.

On radiologic examination there were bilateral Grade 4 sacroiliitis, nonmarginal, asymmetric syndesmophytes on L1–L2, L2-L3, T12-L1. Servical and atlantoaxial radiographs were normal. Knee radiographs showed elevation of capsula possibly because of increased intrarticular synovial fluid, soft tissue swelling, osteophytosis on medial femoral tibial condyles and patella. Hand and feet radiographs showed no evidence of arthritic involvement or enthesitis.

Skin lesions were examined by a dermatologist. Microbiologic culture of papulopustular lesions on back and chest were negative. Pathergy test was negative.

Having recurrent oral aphthous lesions, iridocyclitis with hypopyon and retinal vasculitis and acneiform lesions the patient was diagnosed as BD according to ISGC. Because of recurrent ocular involvement the patient was started on azathioprine 50 mg three times a day.

Discussion

The inclusion of BD among SpA and whether sacroiliitis develops in BD are still being debated (3).

Joint involvement occurs in 45-70 % of patients with BD, usually manifesting as arthralgia and as acute or subacute arthritis. Behcet's arthritis is usually mono or oligoarthritis but can be symmetric with a potential of confusion with rheumatoid arthritis. Oligoarthritis in small or larger joints was found in 40% of a patient population followed for 1-2 years. Knees, ankles, wrists and tendon entheses may be involved in a recurring but seldom destructive episodic process. Involvement of distal interphalangeal joints, spine and sacroiliac joints is very uncommon (7).

There are several studies about the prevalence of sacroiliitis and AS in BD patients. Maghoury et al studied the prevalence of sacroiliitis in a group of patients with BD. Two out of 27 (7,4 %) BD patients had grade 2 sacroiliitis and, 1 equivocal sacroiliitis (8).

Taarit et al retrospectively reviewed the medical records of 309 patients with joint manifestations. Sacroiliitis was observed in 6% cases and 2 cases were found to have AS (8). Dilsen et al reported prevalence of sacroiliitis and AS in BD. Among 331 BD cases, 34% had sacroiliitis and 10% had AS in their series(9).

Oliveri et al studied 20 BD and 20 controls and observed sacroiliitis in 6 out of 20 patients and 1 of controls (10). Oliveri et al also reported a case of coexistence of BD and AS with advanced cervical involvement (11).

Borman et al reported a 29 year-old woman who was diagnosed as AS and during follow up developed BD with severe inflammatory joint involvement. HLA phenotype was negative for

HLA-B51 and positive for HLA-B27 (5). Cimen et al reported a case of clinically occult coexistence of AS and BD and revelation of AS after a long time of BD diagnose (12). Etaouil et al reported 2 cases of AS and BD in combination. In both patients HLA-B27 was positive and HLA-B51 was negative (4).

Signs of AS were sought in 11 patients suffering from BD by Dubost et al. Three patients had definite AS and 2 had phenotype B27. They stated that this combination is rare but it is felt to be legitimate to seek the other disease when one of these 2 conditions is diagnosed (6) .

Yazici et al reported only a single case of AS in 184 BD patients in the Turkish population (13). Yazici et al reported that the high observer variation in interpreting a film of the anteroposterior (AP) view of the pelvis for sacroiliitis maybe a major cause of reported sacroiliitis in Behçet's disease (14).

Chang et al performed a study to clarify whether BD could be classified into the SpA complex. The prevalence of definitive sacroiliitis in BD group and SpA was 46.4% and 5.2% respectively. The patterns of eye involvement were different between these two groups. They concluded that BD could not be classified into the SpA complex (15).

In our case the patient was initially diagnosed as AS. He had oral aphthous ulcerations recurring 10–15 times a year but had no other signs of BD for years. First ocular involvement which was diagnosed as uveitis occurred 9 years after the beginning of his musculoskeletal symptoms. He had vision loss of right eye in the last year and diagnosed as iridocyclitis with hypopyon and retinal vasculitis. He was then diagnosed as BD according to ISGC with oral aphthous ulcerations, ocular involvement and acneiform lesions on his chest and back.

Our patient had uveitis and iridocyclitis with hypopyon and retinal vasculitis in different times. Beiran et al reported a case of a young man suffering from both ankylosing spondylitis and BD in whom the character of the ocular involvement changed according to the predominant disease at a given time. When the clinical picture was one of

AS, only anterior uveitis was observed, while the clinical picture of BD occurred with panuveitis and retinal vasculitis (16).

Our patient's phenotype was positive for HLA-B27 and negative for HLA-B5 which is consistent with HLA phenotypes of BD associated AS cases in literature.

Overall, nearly 90% of SpA patients are positive for HLA-B27 and 60–80 % of BD patients are positive for HLA-B5 (17, 18). However patients with both BD and SpA are far less likely to be HLA-B5 positive than those with BD alone (19). Furthermore, HLA-B27 does not seem more common in BD than seen in the normal healthy population (18). However the prevalence of HLA-

B27 is extremely high among patients with both BD and SpA (6,17).

Hamza et al suggested that BD may increase the risk of SpA among patients with the HLA-B27 antigen (19). Kallel et al suggested that HLA-B5 negativity in BD patients may increase the risk of AS (18).

As a result whether the combination of BD and AS in our patient is a co-existence or if there is a casual relationship remains to be determined. Patients with rheumatologic diseases should be followed for long time and new symptoms should alert the physician for reviewing the diagnosis and coexistent diseases.

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