*Physical Medicine 2000; 3 (1) : 47-49*

**PHYSICAL MEDICINE**

# ATYPICAL BEHCET'S DISEASE WITH PERIPHERAL EROSIVE ARTHROPATHY

## PERİFERİK EROZİV ARTROPATİ İLE SEYREDEN ATİPİK BEHÇET HASTALIĞI

Pınar BORMAN MD*, Kürşat ÖZORAN MD*, Ümit SEÇKİN MD*, Serap Yücel GÖKÇEK MD**,

* Department of Physical Medicine and Rehabilitation, Numune Education and Research Hospital
** Department of Radiology, Numune Education and Research Hospital

***SUMMARY***

*Joint symptoms are well recognized features of Behcet's disease (BD) characterized as intermittent self-limiting nondestructive arthritis involving the peripheral large joints. We report a 35-year-old female patient with BD presented with erosive arthritis of right wrist and intercarpal joints and discuss the case with the review of the literature.*

**Key words :** Behcet's disease, erosive arthritis

***ÖZET***

*Behçet hastalığının eklem semptomları iyi bilinmektedir ve periferik büyük eklemleri tutan intermittant nondestrüktif artrit ile karakterizedir. Burada 35 yaşında bir kadın hastanın sağ el bileğinde ve interkarpal eklemlerinde gelişen eroziv artrit sunulmakta ve literatür gözden geçirilerek tartışılmaktadır.*

**Anahtar sözcükler :** Behçet hastalığı, eroziv artrit

### INTRODUCTION

Behçet's Disease (BD) was first described by Hulusi Behçet in 1937 as a triple symptom complex of apthous stomatitis, genital ulceration and iritis (1). Later more manifestations were described and it became a multisystem disease characterized by vasculitis (2). Joint symptoms are well recognized features of this syndrome and are usually described as a nondestructive monoarticular or symmetrical oligoarthritis which is commonly subacute and self limiting (3). Although arthropathy is a common manifestation in patients with BD, it rarely causes chronic disability or deformity (4).

Here we describe a female patient suffering from BD, associated with unilateral erosive arthritis of the wrist and finger joints.

### Case Report

A 35 year old female was referred to the Ankara Numune Education and Research hospital in March 1999 with pain and swelling of the right and left wrist, right metacarpophalangeal (MCP) joints and left knee. She had a history of painful ulceration in the mouth for 8 months and in the genital area for 2 months. Last year she has had an illness which lasted four weeks with severe muscle and joint pain, painful ulcers both in the mouth and genital area. Since then about every 2-3 months she has had recurrent attacks of painful mouth ulcers as well as the attacks of peripheral arthritis and she noticed some stiffness in the wrists. She denied any morning stiffness. She had 2 children aged 13 and 10 years old and there was no history of abortions. During her hospitalization she experienced one episode of painful oral ulceration. Physical examination revealed synovial thickening with tenderness of the left and right wrist, right MCP joints and left knee. There was a slight limitation of movement on these joints. For the evaluation of the systemic involvement, all the responsible physicians consultated the patient and no other system involvement was found. Pathergy reaction was positive and opthalmologic examination revealed normal findings. Routine biochemical test and immunoglobulins were normal. ESR was 26 mm/h and CRP was 51 mg/l (normal<5). Blood cell count revealed mild anemia with 11.2 g/l hemoglobin, white blood cell count: 7800, platelet: 356000. Repeated tests for RF, ANA and anti

DNA were negative. HLA typing were as follows: HLA-A2, HLA-B8, HLA-Bw6, HLA-Cw3, HLA-Cw7, HLA-DR13, HLA-DR17, HLA-DR52, HLA-DQ2, HLA-DQ7.

Chest radiographs and USG examination of abdomen and pelvis showed no pathologic features. X-rays of the both hands showed erosive arthritis involving the right wrist, carpometacarpal and carpal joints. There was periarticular widespread osteoporosis in the right hand and right intercarpal joint space was narrowed when compared to the left hand (Fig-1). There was no evidence of spondyloarthritis or sacroiliitis on x-rays. Computerized tomographical findings of the hands revealed narrowing of the right intercarpal and carpometacarpal joint space, demineralization and subchondral resorption of the carpal bones. There was also resorption on the metacarpal joint surface adjacent to the joint of carpometacarpal joint associated with endosteal resorption and irregularity in all phalangeal surfaces (Fig 2 and 3).

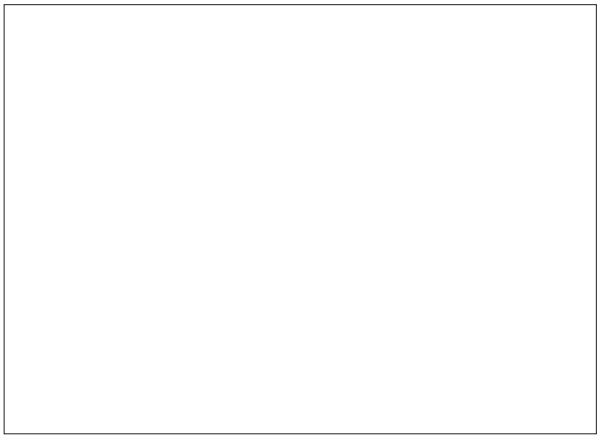


**Fig-1:** The x-rays of the both hands showed erosive arthritis involving the right wrist, carpometacarpal and carpal joints associated with periarticular widespread osteoporosis in the right hand and right intercarpal joint space was narrowed when compared to the left hand.

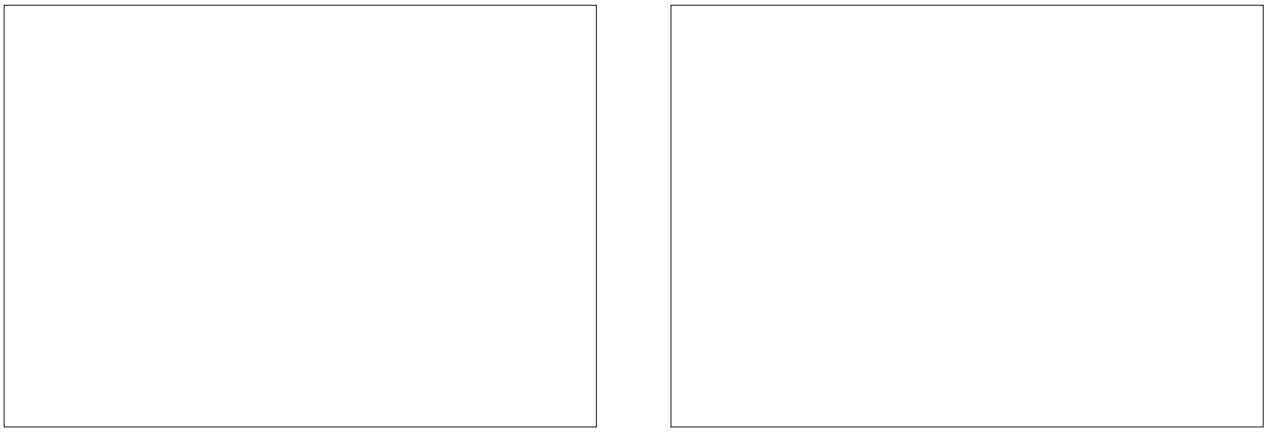
She was treated with colchium (0.5 mg) twice daily and indomethacin 150 mg/daily.

## DISCUSSION

Our patient met the criteria of International Study Group for the diagnosis of BD (2). She presented oral apthosis, genital ulcers and pathergy test positivity with features of arthritis in the hands and feet.

The joint manifestations of BD are observed in the majority of the patients with an incidence ranging between 10-55% (4). It is well known that arthritis in BD is usually non-deforming monoarticular or symmetrical oligoartricular commonly subacute and self-limiting (1-4). Like other manifestations of BD, the difference in the occurrence of joint involvement may be influenced in part by the origin of the series.

Radiological erosive changes are reported to be in 1% of Japanese patients with BD (5). In series of Vernan-Roberts (6), 2 patients had radiological erosive changes in the hip and manibriosternal joints respectively. Shimizu (7) reported erosive changes of MTP joint and interphalangeal joints of the feet in 2 patients. Yurdakul et al (3) reported erosive changes in 5 of



**Figs 2 and 3:** Computerized tomography of the hands revealed narrowing of right intercarpal and carpometacarpal joint space, demineralization and subchondral resorption of carpal bones. There was also resorption on the metacarpal joint surface adjent to the joint of carpometacarpal joint associated with endosteal resorption and irregularity in all phalangeal surfaces.

47 patients. The erosive changes were in the joints of the feet, temporomandibular joint and cystic changes in the shaft of middle phalanx were also reported in one patient. Kim et al (4) investigated the characteristics of arthropathy in BD and reported marginal erosion, joint space narrowing in the feet of only two patients with BD, although the incidence of the arthropathy was higher in Korean patients with BD. Jawad (8) indicated erosive arthritis in the wrist and carpi of a patient bilaterally with repeated attacks of synovitis in the same joints and suggested that the repeated attacks of synovitis led to an erosive and destructive arthritis resembling the radiological appearance of RA, which it may be confused. Kötter (9) et al also reported a female patient with erosive arthritis of the fifth toe joints associated with posterior uveitis, who was treated successfully with interferon therapy. As seen above, the erosive arthritis were rarely reported in the literature and the majority of the patients had erosive arthritis in the joints of the feet. Our patient had episodic mild synovitis affecting the small joints of the right hand accompanied by the development of erosions in several small joints of the right hand. Tissue typing was positive for HLA-B5 which has been associated with the arthropathy of BD (10). However she was also homozygous for HLA-DR4 which is associated with erosions in other inflammatory joint disorders.

The onset of BD is usually after puberty between the second and fourth decades (2,3). In a recent report by Krause et al (11), a significant correlation between the age at onset of BD and several aspects of the disease (like disease onset at an older age) is positively correlated with increased disease spectrum. The older the patients were at the disease onset the more organ system were eventually involved. In our patient the disease onset was on the third decade which can assumed as an early period. The lack of multiple system involvement in our patient may be explained by this reason.

The patients' positivity for HLA-B5(B51) are reported to be more susceptible to eye, joint and neurological complications (10). The clinical situation and tissue subtyping of our patient confirms the fact that HLA-B 5 is more frequent among the patients with joint involvement as an integral part of this entity.

In conclusion we described a patient with atypical BD presented with peripheral arthropathy and in whom erosive arthritis was the main symptom for many months. BD is common in Turkey and we suggest that this condition should be considered in the differential diagnosis of patients with asymmetrical oligoarthritis in the upper extremity associated with radiologic erosive changes and questions for the symptoms of BD should be asked for the accurate diagnosis and appropriate therapy in order to avoid irreversible tissue injuries.

**REFERENCES**


1. O'Duffy JD. Behcet's disease. Curr Opinion Rheumatol 1994;6:39-43.
2. The International Study Group for Behcet's Disease. Evaluation of diagnostic (classification) criteria in Behcet disease-towards internationally agreed criteria. Br J Rheumatol 1992;31:299-308.
3. Yurdakul S, Yazıcı H, Tüzün Y, Pazarlı H, Yalçın B, et al. The arthrtitis of Behçet's disease: a prospective study. Ann Rheum Dis 1983;42:505-515.
4. Kim HA, Choi KW, Song YW. Arthropathy in Behçet's disease. Scand J Rheumatol 1997;26:125-9.
5. Oshima Y, Shimizu T, Yokohan R, Matsumota T, Kano K, Kagami T, et al. Clinical studies on Behcet's syndrome. Rheum Dis 1963;22:36-44.
6. Vernon Roberts B, Barnes CG, Revell PA. Synovial pathology in Behçet's syndrome. Ann Rheum Dis 1978;37:139-145.
7. Shimuzu T, Ehrlick GE,Inaba G, Hayashi K. Behcet's disease. Semin Arthritis Rheum 1979;8:223-60.
8. Jawad ASM, Goodwill CJ. Behçet's disease with erosive arthritis. Ann Rheum Dis 1986;45:961-962.
9. Kötter I, Dürk H, Eckstein A, Zierhut M, Fierlbeck G, Saal JG. Erosive arthritis and posterior uveitis in Behcet's disease: treatment with ( interferon and interferon. Clin Exper Rheumatol 1996;14:313-315.
10. Yazıcı H, Tüzün Y, Pazarlı H, Yakın B, Yurdakul S, Müftüoğlu A. The combined use of HLA-B5 and the pathergy test as diagnostic marker of Behçet disease in Turkey. J Rheumatol 1980;7:206-10.
11. Krause I, Uziel Y, Guedj D, Mukamel M, Molad Y, Amit M, Weinberger A. Mode of presentation and multisystem involvement in Behcet's disease: The influence of sex and age of disease onset. J Rheumatol 1998;25:1566-9.