

# A Rare Presentation of Behçet Ulcer on the Vaginal Cuff: Case Report

## Vajinal Cuff'da İzlenen Nadir Bir Behçet Ülseri Olgusu

Betül YORGUNLAR,<sup>a</sup>  
Suna ÖZDEMİR,<sup>a</sup>  
Lebriz Hale AKTÜN TAMER,<sup>a</sup>  
Mustafa ACET<sup>a</sup>

<sup>a</sup>Department of Gynaecology and Obstetrics, Medipol University Faculty of Medicine, İstanbul

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Yazışma Adresi/Correspondence:  
Betül YORGUNLAR  
Medipol University Faculty of Medicine,  
Department of Gynaecology  
and Obstetrics, İstanbul,  
TÜRKİYE/TURKEY  
jindr\_betul@yahoo.com

**ABSTRACT** Behçet Syndrome (BS) is a rare chronic, inflammatory, recurrent and multisystemic disease that is characterised by vasculitis throughout the body. BS is an autoimmune disease that affects the neurological and gastrointestinal systems that is characterized by reoccurring oral and genital aphthosis, arthritis, eye and cutaneous lesions. In BS, genital lesions have been reported at rates of 60-90% in different series. Lesions are observed most commonly on the vulva labium majora, followed by the vagina and uterine cervix in descending order. However, no previous study has reported a lesion on the vaginal cuff of a patient who has undergone total hysterectomy. In this study, for the first time, we defined a rare vaginal cuff ulcer in a patient previously diagnosed with BS. This case study will be the first in literature and contribute to it by revealing the appearance of a genital ulcer on the vaginal cuff caused by BS. Moreover, we think that it should be considered in the differential diagnosis of vaginal lesions.

**Key Words:** Behçet syndrome; ulcer; vagina

**ÖZET** Behçet sendromu (BS) nadir görülen, vaskülit ile karakterize kronik, inflamatuvar, tekrarlayan, multisistemik bir hastalıktır. BS, tekrarlayan oral ve genital aftlar, artrit, göz lezyonları, kutanöz lezyonlar, gastrointestinal sistem ve merkezi sinir sistemi tutulumuyla karakterize otoimmün bir hastalıktır. Behçet sendromunda genital tutulum farklı serilerde %60-90 olarak rapor edilmiştir. Genital bölgede en sık tutulum vulvada labium majuslarda görülmektedir. Daha sonra azalan sıklıkta vajen ve servikte rapor edilmiştir. Fakat şu ana kadar histerektomize bir hastada vajinal cuffta herhangi bir tutulum bildirilmemiştir. Biz bu makalede daha önceden BS olduğu bilinen hastada ilk defa görülen nadir bir vajinal cuff ülseri tanımladık. Bu olgu BS'ye bağlı genital ülserin vaginal cuff da görülmesi bakımından literatürde ilk olup bu anlamda literatüre katkı sağlayacağını ve vajinal lezyonların ayırıcı tanısında göz önüne alınması gerektiğini düşünüyoruz.

**Anahtar Kelimeler:** Behçet sendromu; ülser; vajina

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Behçet Syndrome (BS) has been defined as a multisystemic disease affecting the skin, joints, peripheral vessels and nervous system that can also affect all other tissues, first defined by Hulusi Behçet, MD, a Turkish dermatologist. The current criteria for BS diagnosis were determined by the principles that were defined and proposed by the "International Study Group for Behçet Disease" in 1990. According to these criteria, BS diagnosis can be made in the presence of oral ulceration and two of the following; genital ulceration, typical eye lesions (uveitis), typical skin le-

sions and a positive pathergy skin test.<sup>1,2</sup> Although BS is seen on all over the world, it is more common in countries along the historical silkroad including Turkey, the Mediterranean, Middle Asia, Asia and Japan.<sup>3-6</sup>

Genital ulcers, like oral ulcers, commonly appear on the skin and mucosa of the genital tract. In women, these lesions commonly develop on the vulva and less frequently on the vagina or cervix.<sup>5</sup> However, at present, no Behçet's ulcer has been reported on the vaginal cuff.

## CASE HISTORY

A 47 year old female patient applied to our clinic with the complaints of vaginal discharge and groin pain for the past one week. In her medical history, she had one normal vaginal delivery and total abdominal hysterectomy operation for myoma uteri 7 years ago. Moreover, the patient had been taking colchicine since diagnosed with BS 25 years ago. In addition, when oral and genital sores developed, she applied topical corticosteroid cream for treatment. The patient had not been suffering from genital ulcers for 3 years. Furthermore, the patient had had an attack of uveitis 8 years ago and made a full recovery with medical treatment. Additionally, the patient was being followed by cardiovascular surgeons for thromboflebitis affecting the lower extremities. In the patients gynaecological examination, the perineum and vulva were normal; with speculum examination, there were two ulcers of 1 cm and 0.5 cm in size observed on the vaginal cuff and smear was taken (Figure 1). The uterine cavity could not be observed by ultrasonography (USG) due to the hysterectomy and the ovaries were bilaterally atrophic. As topical treatment, betamethasone valerate cream was prescribed. The smear results were normal. The patient was called for a control examination 3 weeks after the first admission. In her control examination, there was no genital ulcer or scar observed (Figure 2).

## DISCUSSION

Mucosal ulcerations are among the main findings of BS and are characterized by recurrent oral ul-



FIGURE 1: Two ulcers of 1 cm and 0,5 cm on the vaginal cuff.



FIGURE 2: No genital ulcer or scar after treatment.

cerations.<sup>7</sup> Other criteria include genital ulcers, erythema nodosum, papulopustular rashes, eye lesions and pathergy skin test positivity.<sup>4,5</sup> Recent studies with large series have determined an equal incidence of BS in both males and females.<sup>6</sup> The same study reported that symptoms including erythema nodosum and genital aphthae were more commonly seen in females.<sup>6</sup>

Genital ulcers are the second most common finding in BS, and like the oral ulcers, they are usually located on the mucosa and skin of the genital tract. The most common location of genital ulcers is the labia majora of the vulva; however they may be

located less often on the vagina or cervix.<sup>5</sup> In BS, genital lesions have been shown to occur at a rate of 60-90%.<sup>8</sup> The ulcer of the present case was detected on vaginal cuff. To our knowledge, there are no data in the literature about the Behçet's ulcer of vaginal cuff. In a previous case report, vaginal ulceration was described in a patient with ulcerative colitis after abdominal hysterectomy.<sup>9</sup> Histopathology was found as nonspecific chronic granulomatous inflammation on biopsy at the first time, the patient was diagnosed as pyoderma gangrenosum because recovery was determined after local and systemic steroid treatment. However, BS was diagnosed in the patient after recurrent vaginal ulceration.

In addition to pyoderma gangrenosum, granuloma, cellulitis of the vaginal vault, BS should be considered in the differential diagnosis of the vaginal ulcerations. About half of the pyoderma gangrenosum are associated with underlying systemic conditions, such as inflammatory bowel disease, arthritis, and haematological malignancies.<sup>10</sup> About 2% of patients with inflammatory bowel disease will develop pyoderma.<sup>11</sup> In vaginal cuff cellulitis, the vaginal surgical margin is erythematous, edematous, and hyperemic early following surgery, and there are purulent vaginal secretion.<sup>12</sup>

In another one of our studies, the incidence of genital lesions (scar + ulcer) in BS was found to be 74.3%, with the labia majora being the most commonly affected region at 55.1%, followed by labia minora, pubic area and perineal area. The duration for ulcer recovery was  $2.6 \pm 0.9$  weeks.<sup>13</sup>

The genital lesions are usually circular or oval and look like they have been punctured with a staple. In contrast to oral ulcers, genital lesions recover with scar formation that is specific to BS.<sup>3</sup> The recovery time for these lesions has been reported as being 10-30 days in various studies.<sup>8,14</sup> The size and depth of the lesion and the presence of bacterial infection are important in scar formation.<sup>15</sup> On the other hand, a recent study suggested that ulcers of the labia minora recover without scarring like oral lesions.<sup>16</sup> This suggestion could be due to the difficulties in differing mucosal scar tissue on the labia minora with the naked eye from scar-free normal mucosa. Pub-

lished data regarding vaginal or cervical lesion recovery is insufficient. In our case, we noted that recovery was without scarring like in oral lesions. This may be due to the structural characteristics of the vaginal mucosa.

Genital lesions are usually painful however they may rarely be asymptomatic and asymptomatic females have no symptoms.<sup>16</sup> Active ulcerated lesions and scars may cause gynaecological complications in female patients with BS. The present case admitted to our clinic with vaginal discharge. For instance, deep ulcers located in the vagina may result in the formation of fistulas to the bladder and urethra.<sup>17</sup> In addition, genital lesions may cause urinary and walking difficulties.<sup>18</sup> Moreover, large genital ulcers may recover with scar tissue that may cause dyspareunia and affect the quality of life. Although there are a large number of studies regarding oral and systemic symptoms in BS, studies relating to genital ulcers, the formation of scar tissue and other gynaecological symptoms are limited in literature.

Recurrent painful genital ulcers may decrease the quality of life of the patients. Topical anaesthetics, dapsone, sucralfate, granulocyte colony-stimulating factor, cyclosporine, topical and intralesional corticosteroids have been used either alone or in combination, having varying degrees of supporting evidence in the treatment of genital ulcers in BS.<sup>19-23</sup> For the topical treatment of genital ulcer and cutaneous lesions, corticosteroid and antiseptic creams can be applied for a short period of time (7 days). Topical sucralfate reduces the healing duration and pain of genital ulcer.<sup>20</sup> Colchicine can be used as a secondline alternative. A recent study has shown that colchicine reduces the occurrence of genital ulcer.<sup>23</sup> Similarly, we applied local steroid treatment to our case, and then no scar or ulcer formation occurred.

In conclusion, the variety of lesions may be observed in the vaginal and vulvar regions in females. However, the approach to these lesions differs depending on the etiological agent. BS should be considered in the differential diagnosis of lesions located in the genital region of patients even though they may be from an area where the incidence of BS is low.

## REFERENCES

1. Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. *Lancet* 1990;335(8697):1078-80.
2. O'Neill TW, Rigby AS, Silman AJ, Barnes C. Validation of the International Study Group criteria for Behçet's disease. *Br J Rheumatol* 1994;33(2):115-7.
3. Evereklioglu C. Current concepts in the etiology and treatment of Behçet disease. *Surv Ophthalmol* 2005;50(4):297-350.
4. Kaklamani VG, Vaiopoulos G, Kaklamanis PG. Behçet's disease. *Semin Arthritis Rheum* 1998;27(4):197-217.
5. Pickering M, Haskard DO. Behçet's syndrome. *J R Col Physicians Lond* 2000;34(2):169-77.
6. Tursen U, Gurler A, Boyvat A. Evaluation of clinical findings according to sex in 2313 Turkish patients with Behçet's disease. *Int J Dermatol* 2003;42(5):346-51.
7. Evaluation of diagnostic ('classification') criteria in Behçet's disease--towards internationally agreed criteria. The International Study Group for Behçet's disease. *Br J Rheumatol* 1992;31(5):299-308.
8. Zouboulis CC. Epidemiology of Adamantides-Behçet's disease. *Ann Med Interne (Paris)* 1999;150(6):488-98.
9. Lilford RJ, Tindall VR, Batchelor AG. Post-surgical pyoderma gangrenosum of the vaginal vault associated with ulcerative colitis and Behçet's disease; a case report. *Eur J Obstet Gynecol Reprod Biol* 1989;31(1):93-4.
10. 10-Bennett ML, Jackson JM, Jorizzo JL, Fleischer AB Jr, White WL, Callen JP. Pyoderma gangrenosum. A comparison of typical and atypical forms with an emphasis on time to remission. Case review of 86 patients from 2 institutions. *Medicine (Baltimore)* 2000;79(1):37-46.
11. Callen JP. Pyoderma gangrenosum. *Lancet* 1998;351(9102):581-5.
12. Hemsell DL. Infection after hysterectomy. *Infect Dis Obstet Gynecol* 1997;5(1):52-6.
13. Ozdemir S, Ozdemir M, Celik C, Balevi A, Toy H, Kaniş U. Evaluation of patients with Behçet's disease by cervical cytology and colposcopic examination. *Arch Gynecol Obstet* 2012;285(5):1363-8.
14. Zouboulis CC. Genitoanal lesions in Adamantides--Behçet's disease. *J Eur Acad Dermatol Venereol* 1997;9(Suppl 1):106-7.
15. Singer AJ, McClain SA. Persistent wound infection delays epidermal maturation and increases scarring in thermal burns. *Wound Repair Regen* 2002;10(6):372-7.
16. Ghate JV, Jorizzo JL. Behçet's disease and complex aphthosis. *J Am Acad Dermatol* 1999;40(1):1-18.
17. Al-Otaibi LM, Porter SR, Poate TW. Behçet's disease: a review. *J Dent Res* 2005;84(3):209-22.
18. Kontogiannis V, Powell RJ. Behçet's disease. *Postgrad Med J* 2000;76(900):629-37.
19. Lin P, Liang G. Behçet disease: recommendation for clinical management of mucocutaneous lesions. *J Clin Rheumatol* 2006;12(6):282-6.
20. Alpsoy E, Er H, Durusoy C, Yilmaz E. The use of sucralfate suspension in the treatment of oral and genital ulceration of Behçet disease: a randomized, placebo-controlled, double-blind study. *Arch Dermatol* 1999;135(5):529-32.
21. Bacanlı A, Yerebakan Dicle O, Parmaksizoglu B, Yilmaz E, Alpsoy E. Topical granulocyte colony-stimulated factor for the treatment of oral and genital ulcers of patients with Behçet's disease. *J Eur Acad Dermatol Venereol* 2006;20(8):931-5.
22. Avci O, Gürler N, Güneş AT. Efficacy of cyclosporine on mucocutaneous manifestations of Behçet's disease. *J Am Acad Dermatol* 1997;36(5 Pt 1):796-7.
23. Yurdakul S, Mat C, Tüzün Y, Ozyazgan Y, Hamuryudan V, Uysal O, et al. A double-blind trial of colchicine in Behçet's syndrome. *Arthritis Rheum* 2001;44(11):2686-92.