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-
sessions and a positive pathergy skin test.\textsuperscript{1,2} Although BS is seen all over the world, it is more common in countries along the historical silkroad including Turkey, the Mediterranean, Middle Asia, Asia and Japan.\textsuperscript{3-6}

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.\textsuperscript{5} 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 thrombophlebitis 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 ulcerations.\textsuperscript{7} Other criteria include genital ulcers, erythema nodosum, papulopustular rashes, eye lesions and pathergy skin test positivity.\textsuperscript{4,5} Recent studies with large series have determined an equal incidence of BS in both males and females.\textsuperscript{6} The same study reported that symptoms including erythema nodosum and genital aphts were more commonly seen in females.\textsuperscript{6}

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. In BS, genital lesions have been shown to occur at a rate of 60-90%. 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. 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. About 2% of patients with inflammatory bowel disease will develop pyoderma. In vaginal cuff cellulitis, the vaginal surgical margin is erythematous, edematous, and hyperemic early following surgery, and there are purulent vaginal secretion.

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±0.9 weeks.

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. The recovery time for these lesions has been reported as being 10-30 days in various studies. The size and depth of the lesion and the presence of bacterial infection are important in scar formation. On the other hand, a recent study suggested that ulcers of the labia minora recover without scarring like oral lesions. 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. Published 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. 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. In addition, genital lesions may cause urinary and walking difficulties. 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. 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. Colchicine can be used as a secondline alternative. A recent study has shown that colchicine reduces the occurrence of genital ulcer. 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


