

Skin and Mucosa Findings of Behçet's Disease

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Mucocutaneous lesions constitute the most characteristic feature of Behçet's Disease (**BD**). As there is no specific laboratory finding of BD, the diagnosis is made based on the evaluation of clinical findings. Skin and mucosa lesions are important for the early diagnosis of the disease as they are the initial findings of the disease or occur frequently during the course of the disease. Lesions including oral ulcer, genital ulcer, papulopustular lesions and erythema nodosum are the most frequently observed cutaneous lesions in BD [1,2].

ORAL ULCERS

Recurrent oral ulcers are the most frequently observed and usually the first finding at the early period of or any time during the course of BD [3,4]. In various publications, it was reported to be seen in more than 95% of the patients [1,3,5-7]. Moreover, oral ulcer is the most frequent clinical findings of the juvenile age group, and constitutes the beginning finding of this age group. Also, during the pregnancy the prevalence of the oral ulcers increases [8]. However, a minority of patients with BD does not develop oral ulcers in the whole course of disease. Positive pathergy test and eye involvements were the most common presenting features in that cases [9].

Oral ulcers may frequently be seen in patients years before the other symptoms of BD occur. Oral ulcer is accepted as the most important diagnostic criterion in BD. According to the criteria of international working group, for the diagnosis of BD there should be minor, major or herpetiform aphthae repeating at least three times within a year [10].

Oral ulcers are frequently seen on regions such as lips, tongue, oral mucosa and soft palate [1]. Oral ulcers are less frequently seen on regions such as hard palate, dorsal surface of the tongue, gingiva, oropharynx and tonsils. Lesions start as erythematous, slightly swollen vesiculopapular lesions, and transform into round or oval ulcers with gray-yellow sole and surrounding erythematous area within 2-3 days. Pain is the most important symptoms and may cause difficulty with eating, drinking, swallowing and oral care. Local traumas may trigger the development of new lesions [1,11]. This finding may be considered as the equivalent of pathergy reaction on mucosa [2].

There are three types of aphthae observed in BD; minor, major and herpetiform. Several types of aphthae may be present at the same time. Minor aphtha is the most common type (85-99%). Minor aphthae are between 1 and 5 in count with the diameter of < 1 cm, and usually heal without scarring within 4-14 days. Usually, minor aphthae do not cause very severe pain. Major aphthous lesions are seen less frequently. They are painful with large diameter (1-3 cm) and deeper. They heal slower within 10-40 days, and usually leave scar. Major aphthae can be seen on any region of the mouth. Herpetiform aphthae are quite rare. They are usually formed of gathering of hundreds of painful ulcers with the diameter of 1-3 mm. They sometimes leave scar. Unlike minor and major aphthae, herpetiform ulcers may also occur on non-keratinized mucosa [10,12]. Studies have shown that none of the specific types of oral ulcers in BD have association with sex and disease severity [10].

While it is impossible to clinically differentiate from recurrent aphthous stomatitis, they have a characteristic feature of frequent repeating and causing generalized lesions [13]. While in some publications, it was reported that the oral ulcers in BD have some differences, there is no definite criterion to differentiate from recurrent aphthous stomatitis. In studies exploring the clinical differences between recurrent aphthous stomatitis and oral ulcers seen in BD, it was reported that oral ulcers in BD are more in number and repeat more frequently, and major oral ulcers are more common. Additionally, oral ulcers in BD are more frequently seen on oropharynx and soft palate than recurrent aphthous stomatitis. Furthermore, in BD, major ulcers were found to be significantly more frequent, more in number and take more time to heal than minor ulcers. A study comparing the oral ulcers between 1643 recurrent aphthous stomatitis patients and 3527 BD by Oh et al. has shown that major aphthae in BD are significantly more frequent, and the involvement of at least two regions of oral mucosa in BD [2,14,15].

Only 1-3% of the patients with recurrent oral ulcers develop BD [8]. As recurrent oral aphthae, especially in children, might be an indicator of BD, they should not be ignored. However, there is no way to foresee whether a patient with recurrent oral aphthae will develop BD or not [14]. The lack of a valid laboratory test to rule out BD may cause problems in diagnosis at the early period of the disease in which the oral ulcers are the only findings [16]. Bang et al., in their prospective study including 67 patients with only recurrent oral aphthae, demonstrated only 35 patients

(52.2%) developed BD with a mean of 7.7 years after the onset of recurrent oral ulcers. Also, they reported that the frequent repeating of oral ulcers is an indicating signal for the development of BD [16].

Histopathologically, non-specific changes such as lymphocyte, macrophage and neutrophil infiltration are seen on the base of the ulcer. In severe inflammation, leukocytoclastic or lymphocytic vasculitis may also be seen [15].

GENITAL ULCERS

It is one of the three original symptoms identified by Dr. Hulusi Behçet. It is one of the most important symptoms of BD, and seen in 51-97% of the patients [5,11]. It is observed as the initial symptom in 6% of the juvenile age group [8]. It is the second most common skin finding after oral aphthae [2]. Genital ulcers are show a striking similarity to oral ulcers in appearance and prognosis. They are deeper than oral ulcers, heal within 10-30 days and tend to leave scar. Mat et al. observed that approximately two third of the genital ulcers in BD leave scar, and the rate of scar formation is based on the size of the ulcer. Ulcers on labia minor and small ulcers usually don't leave scar [17]. In suspected BD, even if there is no active genital ulcer, scars from previous lesions should be examined. In men, they are most frequently seen in scrotal region. Approximately 90% of the lesions are seen in this region. Penis and other regions are rarely involved in men [2,11]. Epididymitis is also common in men. However, urethritis is not observed in BD. This characteristic may help the differentiation from Reiter's Syndrome [6]. In women, labium major is the most commonly involved region. Labia minor, vaginal mucosa and rarely cervix may also be affected. Ulcers have also been reported to occur on urethral orifice [1]. Genital ulcer is more frequently seen in Caucasian race [5,18]. Studies have shown that patients with BD who had genital ulcer experience the other findings more frequently, but eye involvement is less frequent [19].

Histopathological examination of the genital ulcer shows similar findings to oral ulcers [2].

The genital ulcer seen in BD should be differentiated from venereal diseases such as syphilis, chancroid and herpes simplex, fix drug eruption, erythema multiforme, erosive lichen planus, and autoimmune bullous dermatoses [15].

PAPULOPUSTULAR LESIONS

Papulopustular lesions are the most common mucocutaneous lesions seen in BD. While the prevalence of papulopustular lesions in patients with BD varies greatly between countries, it is between 28 and 96% [12,20]. They are characterized with folliculitis or acne-like pustulae. Papulopustular lesions in BD is also termed as Behçet's pustulosis, folliculitis, acneiform eruption and pseudofolliculitis. They usually start as papule with erythematous base, and transform into pustule within 24 to 48 hours. Papulopustular lesions are usually seen on the body and extremities, and on the face [20].

Papulopustular lesions are an important diagnostic criterion for dermatologists. According to international working group, the sensitivity of this lesions is 70%, and the specificity is 76% [20].

The formation mechanisms of the papulopustular lesions in BD are yet to be known completely. A study has shown that granulysin positive cytotoxic T-lymphocytes play an important role in the pathogenetic mechanism of the acneiform lesions. In another study, the prevalence of papulopustular lesions was found to be significantly increased in HLA-B51 positive BD patients with C438T polymorphism on *SUMO4* gene. Moreover, HLA-B5109 subtype was found to be observed less frequently in BD patients with papulopustular lesions [20-23]. According to some research; the frequency of papulopustular lesions is higher in men than women, and the prognosis in men is worse. In other studies, it was demonstrated that androgens may play a role in the formation of papulopustular lesion and disease activity [20,24].

Papulopustular lesions in BD are mostly known as sterile pustulae. There are some opinions regarding that microbiological examination of the pustulae in BD and in acne vulgaris may provide a clue for the differential diagnosis of these two diseases. In a study, mostly *Staphylococcus aureus* and *Prevotella spp.* were detected in the pustulae of BD patients, and *coagulase-negative staphylococci* were detected in the pustulae of acne patients. However, it is not known that these microbiologically detected microorganisms have a role in the disease pathogenesis [4,20,25].

It is long-known that papulopustular lesions in BD patients significantly accompanied by arthritis. Recently, it was observed that in patients with acne-arthritis cluster, enthesitis rate was also found to be increased. Moreover, acne-arthritis cluster was found to be significantly increased among familial cases [26].

Papulopustular lesions in BD and acne vulgaris lesions may be confused easily. The differentiation of the papulopustular lesion in BD from acne vulgaris is especially important in young patients with papulopustular lesions yet to be diagnosed with BD. Papulopustular lesions in BD are not always follicular, however acne lesions are always follicular. Some authors recommended that only non-follicular lesions which have histopathologically leukocytoclastic vasculitis or neutrophils vascular reaction should be considered to be associated with BD [10]. However, Kutlubay et al., in their study, have shown that papulopustular lesions seen in BD do not have much different clinical and histopathological characteristic from the lesions in individuals with acne vulgaris [27,28].

ERYTHEMA NODOSUM-LIKE LESIONS

Erythema nodosum-like lesions are one of the non-specific skin symptoms of BD. To evaluate this symptom in favor of BD, it should not be associated with any other reason. Erythema nodosum-like lesions are common in BD, and observed approximately in 45% of the patients. They are frequently seen in women. Typical clinical presentation is characterized with bilateral, pretibial, painful and warm erythematous nodules. They can also be localized on face, neck, forearm and

thighs. Lesions do not ulcerate, and heal within 2-3 weeks without scarring, or sometimes leave pigment [1].

While they are clinically similar to classic erythema nodosum secondary to other systemic diseases, they are differentiated by their microscopic characteristics. Differently from classic erythema nodosum lesions, the main histopathological characteristic of these lesions is vasculitis or vascular reaction [1]. However, nodular vasculitis should be considered for differential diagnosis. Demirkesen et al., in their study comparing the histopathological characteristics of erythema nodosum, nodular vasculitis, and erythema nodosum-like lesions in BD, described the erythema nodosum-like lesions as neutrophilic vascular reaction accompanied by subcutaneous tissue changes. They reported that as septal panniculitis and lymphocyte-rich infiltration are observed, and vasculitis and other vascular changes are not observed in erythema nodosum, it is substantially different from erythema nodosum-like lesions in a histopathological manner, however, histological characteristics of erythema nodosum-like lesions and nodular vasculitis are not different [1,29]. Misago et al. reported that approximately 27% of the 26 patients (7 of the 26 lesions) with erythema nodosum-like lesions in BD had septal panniculitis without vasculitis in the clinicohistopathological examination and share complete similarity with classic erythema nodosum. Additionally, vasculitis findings were observed in most patients (19 of 26 lesions). Consequently, they reported that erythema nodosum-like lesions in BD patients indicate mild prognosis, and in BD with erythema nodosum-like lesions with severe vasculitis especially accompanied by phlebitis, gastrointestinal system involvement is more common [30].

SUPERFICIAL THROMBOPHLEBITIS

BD is a systemic vasculitis, and affects various types and sizes of veins. The most common involvement of BD is in venous system, and clinically it mostly presents as superficial thrombophlebitis [1]. While superficial thrombophlebitis is rare, it is a characteristic lesion of BD [5]. The incidence of thrombophlebitis is between 27 to 47% [31]. It is more common in male patients. It is characterized with linear arrangement of many, sensitive erythematous subcutaneous nodules involving a vein trace usually on lower extremities and rarely on upper extremities [1]. Thrombus in the vein might be felt by palpation [2]. Thrombotic vein develops sclerosis over time. As it may involve many segments of the venous system at the same time, the place of the nodules may change within days [1]. The most commonly involved vein is vena saphena magna. They might be confused with erythema nodosum-like lesions. It is difficult to differentiate erythema nodosum-like lesions from superficial thrombophlebitis lesions by unaided eye [32]. High-resolution ultrasonography may help to differentiate these two entities. Superficial thrombophlebitis appears as hypoechoic nodules on ultrasonography, and erythema nodosum-like lesions appear as hyperechoic nodules [10].

While superficial thrombophlebitis is a symptom detected during dermatological examination, it indicates vascular involvement, and it is closely associated with deep vein thrombosis and, in

central nervous system, dural sinus thrombosis [2,10]. Patients with superficial thrombophlebitis, therefore, should be monitored closely for vascular involvement [2]. Gürler et al., in their study analyzed 2147 BD patients, and observed 0.5% of the patients had thrombophlebitis as initial symptom [33]. The presence of superficial thrombophlebitis is accepted as an indicator of more severe disease. Coskun et al. emphasized that in patients with superficial thrombophlebitis, physicians should be alert for visceral organ involvement at a later time [31].

EXTRA-GENITAL SKIN ULCERS

While they are rarely seen in BD patients, they are the most characteristic and specific skin lesions. They are seen in 3-6% of BD patients. Clinically, they are similar to oral aphthae. Extragenital skin ulcers are recurrent, and usually heal with scarring. They are painful, yellowish and necrotizing ulcerated lesions. Lesions may be seen around the genital area and inner parts of thighs. They may also be seen on various regions including perianal skin, legs, breasts, axillary regions, interdigital regions and neck. [5,10,34]. In a study, vasculitis findings were observed on lesions in histopathological examination [34].

SWEET-LIKE SKIN LESIONS

Sweet-like lesions are rarely associated with BD. Sweet-like lesions are characterized with painful, erythematous nodule and plaques accompanied by fever and leukocytosis. Lesions are usually localized on face, neck and extremities [1].

While Sweet-like skin lesions may occur in BD patients as skin sign, some patients may also have coexistent Sweet's Syndrome and BD in clinical and histological examination [13,35]. It is highly difficulty to differentiate Sweet's Syndrome associated with BD, and Sweet-like lesions seen in BD. The main difference between these two conditions is the difference in HLA types which are responsible from the diseases. While HLA B51 is common in BD patients, HLA B54 is mostly positive in Sweet's Syndrome [35]. Oral and genital ulcers are rarer in Sweet's Syndrome. Joint and ocular involvement pattern might be different in Sweet's Syndrome than BD. While asymmetrical polyarthralgia, conjunctivitis and episcleritis are more frequent in Sweet's Syndrome, monoarticular or polyarticular pattern and panuveitis are more typical in BD. Also, fever is not common in BD [1].

Patch type neutrophilic infiltration is present in the histopathological examination of Sweet-like skin lesions. Leukocytoclasia and extravasated erythrocytes are common. There is dense cell infiltration on dermis consisting of lymphocytes, histiocytes and neutrophils. Vasculitis signs may also be detected in some patients [2,4].

BEHÇET'S CELLULITIS

It may be confused with Sweet's syndrome. It is a painful, large, erythematous-edematous lesion. It is usually observed on lower extremities. Clinically, it may also be confused with

infectious cellulitis and superficial thrombophlebitis. Histopathologically, vasculitis is present rather than a neutrophilic dermatosis seen in Sweet's Syndrome [5].

LEG ULCERS

Leg ulcers in BD may be caused by vasculitis or deep vein thrombosis. They have clinically chronic progression, and are resistant to treatment [10].

PYODERMA GANGRENOSUM

Pyoderma gangrenosum-like lesions are very rare in BD patients. They are painful, and expand and transform into superficial ulcerated lesions. They are usually localized on lower extremities and thighs [5].

OTHER CUTANEOUS LESIONS

The spectrum of the cutaneous lesions has been expanded by case reports. Other skin lesions such as pernio-like cutaneous lesions, acral purpuric papulonodular lesions, subungual infarctions, bullous necrotizing vasculitis, Henoch-Schönlein purpura, neutrophilic eccrine hidradenitis, Raynaud phenomenon, kaposi's sarcoma, hidradenitis suppurativa, furuncles, hemorrhagic bullae abscesses and polyarteritis nodosa like lesions are less common but may occur [10,12,36-43].

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