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Subclinical Behcet's Disease

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Abstract

Behcet's Disease (BD) is a chronic inflammatory multisystem disease belongs to the Vasculitidies with small, medium and large sized vessel involvement. This disorder is characterized by Oral aphthosis, Genital aphthosis, ocular inflammation, skin lesions, vascular involvement, positive Pathergy test and HLA-B51 positivity.

When a patient is involved in Behcet's Disease (BD) but his/her disease is presented as simple oral aphthosis and in which the pathergy test and/or HLA-B51 are positive but other clinical features are obscured; it is called subclinical BD.

Two types of subclinical BD have been detected by the author yet; type I and type II.

Subclinical BD type I is a varient of BD with the combination of:

- Low frequent, minor, recurrent oral aphthosis
- Occasionally a few pseudofolliculitis lesions and
- The pathergy test and/or HLA-B51 positivity

Subclinical BD type II is a varient of BD with the combination of:

- Low frequent, minor, recurrent oral aphthosis
- Recurrent internal genital (vaginal/cervical) aphthosis and
- The pathergy test and/or HLA-B51 positivity

Indeed some parts of the cases with simple oral aphthosis are the cases with missed subclinical BD.

So, the author thinks that, the prevalence of BD may be underestimated.

Keywords: Oral Aphthosis; Genital Aphthosis; HLA-B51 Positivity; Pathergy Test; Subclinical BD Type I and II

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Introduction

Behcet's Disease (BD) is the most famous disease within countries along the ancient Silk Road as the cause of oral aphthosis. It is a chronic inflammatory multisystem disease belongs to the Vasculitidies with small, medium and large sized vessel involvement [1-3].

This disorder is characterized by [4]:

- Oral aphthosis:
- Genital aphthosis
- Ocular inflammation
- Skin lesions
- Vascular involvement
- Positive Pathergy test and
- HLA-B51 positivity

The oral aphthosis in BD is almost all recurrent, most often multiple, can be major and may be persistent. It is usually accompanied by genital aphthosis, so called Complex aphthosis [5].

The genital aphthosis in BD morphologically is similar to oral aphthosis but, it is more painful, larger and less frequent with scar except the internal genital (vaginal/cervical) aphthosis in women that is painless and without scar [6, 7].

Ocular inflammations in BD are including anterior Uveitis, posterior/Intermediate Uveitis and Retinal Vasculitis. When all of above items occur together, it is called Panuveitis [8].

Skin lesions in BD are including erythema nodosum, pseudo folliculitis, pustular lesions and dermal aphthosis [9].

Vascular features are including palpable purpura, thrombosis, aneurysm, hemorrhage, and so on [10].

Neurologic manifestations that are compatible with BD called neurobehcet and sometime it is mistaken for Multiple Sclerosis [11].

Articular involvement is more similar to SpondyloArthritis [12]. Gastrointestinal involvement of BD (Enterobehcet) is the differential diagnosis of Inflammatory Bowel Disease (IBD) [13].

Renal, cardiac and pulmonary involvements are uncommon.

In Middle East (e.g. Iran) the Pathergy test is positive in about 50% of the patients with BD. The rate of Pathergy test positivity in Eastern Asia (e.g. Japan) is the highest and it is lowest in Western countries [1].

The HLA-B5 especially B51 is positive in about 50% of the patients [4].

Main Body

When a patient is involved in Behcet's Disease (BD) but his/her disease is presented as simple oral aphthosis and in which the pathergy test and/or HLA-B51 are positive but other clinical features are obscured; it is called "subclinical Behcet's Disease".[1]

Two types of subclinical BD have been detected by the author yet; type I and type II.

Subclinical BD type I:

It is a varient of BD with the combination of:

- Low frequent, minor, recurrent oral aphthosis
- Occasionally a few pseudofolliculitis lesions and
- The pathergy test and/or HLA-B51 positivity

Subclinical BD type II:

It is a varient of BD with the combination of:

- Low frequent, minor, recurrent oral aphthosis
- Recurrent internal genital (vaginal/cervical) aphthosis and
- The pathergy test and/or HLA-B51 positivity

Subclinical BD type I:

It is a varient of BD with clinical features of low frequent, minor, recurrent oral aphthosis and occasionally a few pseudofolliculitis lesions along with pathergy test and/or HLA-B51 positivity. The pseudofolliculitis lesions have been missed by both patient and physician. The patient has not seen the a few pseudofolliculitis lesions upon his/her low back and/or buttock, so he/she does not have any complain regarding it. When the patient is visited by Rheumatologist, due to physical examination within interval phase, the lesions of skin can not be detected either. On the other hand, grossly the only clinical feature is low frequent recurrent oral aphthosis.

Indeed 3 to 6 episodes of oral aphthosis have occurred per year. In each episode one to two minor oral aphthosis with non-significant pain can be presented for a few days.

Due to pathergy test and/or HLA-B51 positivity the patient has been followed and finally the pseudofolliculitis lesions have been detected.

This type of subclinical BD can occur in both genders but men too much more than women will be involved in this type of subclinical BD.

Indeed folliculitis and pseudofolliculitis and even if every skin lesions are more important for women than men and a great percentage of men ignore their a few pseudofolliculitis.

Subclinical BD type II:

It is a varient of BD with clinical features of low frequent, minor, recurrent oral aphthosis and low frequent recurrent internal genital (vaginal/cervical) aphthosis along with the pathergy test and/or HLA-B51 positivity.

The internal genital aphthosis is painless, without any scar and it can not be seen by the patients, so the women can not detect when it does occur? The vaginal speculum examination can be negative too due to examination within interval phase. On the other hand, grossly the only clinical feature is low frequent recurrent oral aphthosis the same as the type I of subclinical BD. Due to pathergy test and/or HLA-B51 positivity the patient has been followed and finally the internal genital aphthosis has been detected.

This type of subclinical (BD) can be seen only in women.

Conclusion

The author thinks these types of subclinical BD e.g. type I and type II are common but most of them can be missed if the pathergy test is not done and HLA-B51 is not checked.

Indeed some parts of the cases with simple oral aphthosis are the cases with missed subclinical BD. So the author thinks that the prevalence of BD may be underestimated.

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[1] This definition is introduced by the author for the first time.

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