Behçet disease in dentistry

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Abstract

The present study aimed to explore the clinical manifestations of Behçet's disease in six patients using the criteria of the International Study Group for Behçet's Disease. The primary objectives were to determine the prevalence of oral aphthosis and assess potential ocular and vascular complications. For a retrospective analysis of six diagnosed Behçet's disease patients, a comprehensive clinical examination, Pathergy tests, and various biological assessments were performed. Additionally, eye examinations, slit lamp studies, and radiological investigations were carried out to evaluate ocular and vascular involvement. The results of the study reveal that oral aphthosis was present in all patients. Some individuals exhibited ocular and vascular involvement, providing insights into the diverse manifestations of the disease. The conclusions outline the importance of the early diagnosis and comprehensive assessments, that are crucial in identifying potential organ involvement in Behçet's disease, contributing to better management and improved patient outcomes.

Keywords: Behçet's disease, oral aphthosis, ocular involvement, vascular complications, comprehensive assessment

INTRODUCTION

The systemic autoimmune and inflammatory conditions often determine oral manifestations, starting from their early onset, outlining the importance of a proper oral examination as an early diagnosis measure. Behçet's disease is a rare, chronic, multi systemic disorder, progressing in episodes [1]. It has been known as to whether it is an autoimmune disease, an auto-inflammatory disease or both [2,3]. This disease is characterized by the occurrence of mucocutaneous lesions, that involve the dermis, genitalia, ocular and oral mucosa [4,5]. In severe cases, the evolution can determine severe impairment of the patient, affecting the nervous system, and involving the vascular and digestive organs [5,6,7].

The etiology of Behcet's disease is not completely known, being suggested the connection between genetic and environmental triggering factors [5]. Immunological alterations have been reported and approximately 50% of the patients exhibited an increase in the circulating immunocomplex while the disease is exacerbated [9, 10, 11]. Being described as a disease with a chronic evolution, the intensity might decrease with time, but still the complication can be life-threatening, such as myocardial infarct, thrombosis, intestine perforation and blindness [12, 13, 14]. Studies have discussed the incidence of different manifestations, among which the oral symptomatology is encountered in 95-100% patients [7,8].

In the present research, we will discuss about the current knowledge of Behçet's disease [8,9] and the management in the dental office of patients with Behçet's disease. The most important manifestation is a mouth-sex-eye triad which is characterised by oral ulcerations, genital recurrences and eye damage [15, 16, 17]. And all these types of lesions are the main clinical manifestations of Behçet's disease, but they are not always present. We can also note, among other things, skin, joint, neurological and / or vascular damage [18, 19]. Behçet's disease affects women and men [18]. To improve the prognosis, the quick detection of the disease is important when the first symptoms appear, and it is important to use the most appropriate treatment.

Focusing on the management in the dental office of patients with Behçet's disease, it is important to acknowledge the possible symptoms, the complications that a diagnosed patient could have and the potential risks related to certain interventions. This management needs the knowledge of patient treatments in order to avoid the drug interactions and to not conform to contraindicated drugs and acts [20, 21].

With a patient with recurrent mouth ulceration, the dentist must make a positive diagnosis and differential diagnosis of Behçet's disease [9, 10]. The dentist must also be able to prescribe medication for the treatment of mouth ulcerations.

Aim and objectives

The aim of this retrospective study is to identify the manifestations of Behçet's disease and assess its epidemiological aspects. By quantifying the existent symptomatology, the oral manifestations were taken into consideration as a primary sign of this disease. The objectives of this study are to determine the influence of age and gender on the prevalence and severity of Behçet's disease, to discuss the clinical criteria for the positive diagnosis of Behçet's disease and its challenges, to investigate the different clinical manifestations of Behçet's disease, including oral and genital ulcers, eye damage, skin lesions, articular damage, vascular damage, neurological damage, and cardiovascular damage, to assess the prognosis and evolution of Behçet's disease, including potential complications and mortality rates, to evaluate the current treatment options for Behçet's disease, considering symptomatic relief and disease control, to provide insights into the diagnosis of Behçet's disease in dental

practice, including the differentiation of oral lesions and to highlight the importance of complementary examinations in determining the underlying pathology of oral lesions.

MATERIAL AND METHODS

In this study, we retrospectively analysed six patients who exhibited various manifestations of Behçet's disease. The diagnosis of Behçet's disease was confirmed based on the criteria set forth by the International Study Group for Behçet's Disease, which specifically required the presence of oral aphthosis.

Each of the six patients underwent a comprehensive clinical examination, which included conducting the Pathergy test, a non-specific hypersensitivity test used to assess skin reactivity. Additionally, a thorough biological assessment was performed, encompassing several parameters such as the sedimentation rate, complete blood count, C-reactive protein levels, prothrombin level, and activated partial thromboplastin time. These tests were crucial in evaluating the patients' overall health status and identifying potential underlying systemic inflammation.

Furthermore, the patients' ocular health was meticulously evaluated through eye examinations, including fundoscopy, which involves examining the back of the eye to detect any abnormalities. A slit lamp study, which is a specialised microscope, was also utilised to obtain a detailed view of the anterior eye structures, aiding in the identification of any ocular complications associated with Behçet's disease.

As with any retrospective study, there may be limitations, such as incomplete data or potential biases. However, by adhering to the International Study Group's established criteria, we ensured a standardised and reliable approach to diagnosis.

The comprehensive nature of the clinical assessments in this study provides valuable insights into the diverse manifestations and potential organ involvement in Behçet's disease. These findings can contribute to the existing knowledge base on the disease, potentially leading to improved diagnostic approaches and treatment strategies in the future.

RESULTS

Six patients with manifestations of Behçet's disease were included in this retrospective study. The diagnosis was confirmed based on the criteria established by the International Study Group on Behçet's Disease, which required the presence of oral aphthosis. The average age of the diagnosis of Behcet disease was 41 years old. In this study were included people between the ages of 18 and 50, the first sign being given by a 28-year-old person (*Table 1*).

Table 1. Characteristics of the included subjects

Patient	Age (years)	Gender	Duration of Disease (months)
P1	**32**	**Female**	**18**
P2	**45**	**Male**	**24**
P3	**28**	**Female**	**12**
P4	**39**	**Male**	**36**
P5	**50**	**Female**	**48**
P6	**34**	**Male**	**15**

The study participants consisted of three females and three males, with ages ranging from 28 to 50 years. The duration of the disease varied between 12 to 48 months. Besides the demographic details of the patients, the clinical examination provided valuable information regarding the progress of the disease. The oral mucosa, the performance of the Pathergy test and the presence of cutaneous and ocular manifestations was assessed in order to provide a complete clinical description (Table 2).

Table 2. Clinical Characteristics of Behçet's Disease in the included group

Patient	Oral Aphthosis	Pathergy Test	Eye Involvement	Other Manifestations
P1	Yes	Positive	Uveitis	Arthritis, Cutaneous Lesions
P2	Yes	Negative	Conjunctivitis	Arthritis, Genital Ulcers
P3	Yes	Positive	Retinal Vasculitis	Arthritis, Oral Ulcers
P4	Yes	Positive	Uveitis, Optic Neuritis	Arthritis, Gastrointestinal Symptoms
P5	Yes	Negative	Uveitis, Retinal Vasculitis	Arthritis, Neurological Symptoms
P6	Yes	Positive	Conjunctivitis, Uveitis	Arthritis, Vascular Involvement

All six patients met the criteria for oral aphthosis, which was the primary diagnostic requirement. The Pathergy test was positive in four patients and negative in two. Eye involvement was observed in all patients, with uveitis being the most common manifestation. Additionally, other manifestations, such as arthritis, cutaneous lesions, genital ulcers, and gastrointestinal symptoms, were observed in various patients.

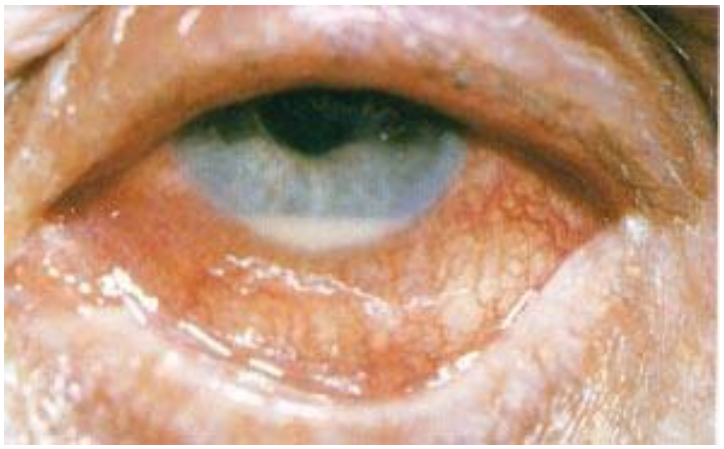

Figure 1. Ophtalmic symptomatology in Behcet's disease

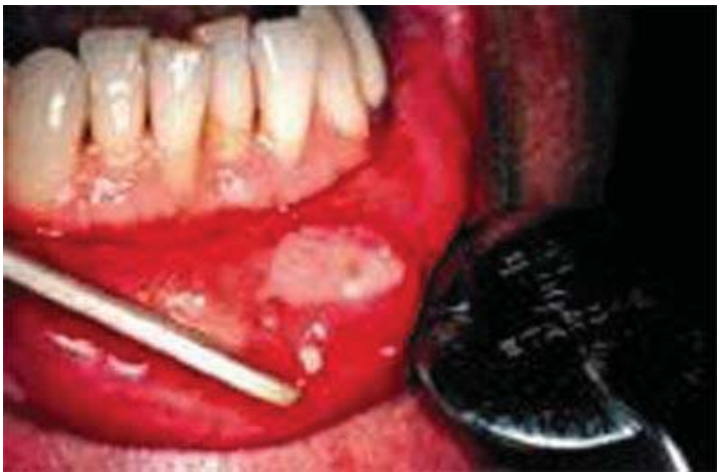

Figure 2. Oral aphtosis in Behcet's disease

The laboratory investigations revealed elevated levels of C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) in all patients, indicating active inflammation (Figure 1). However, the prothrombin level and activated partial thromboplastin time were within normal ranges.

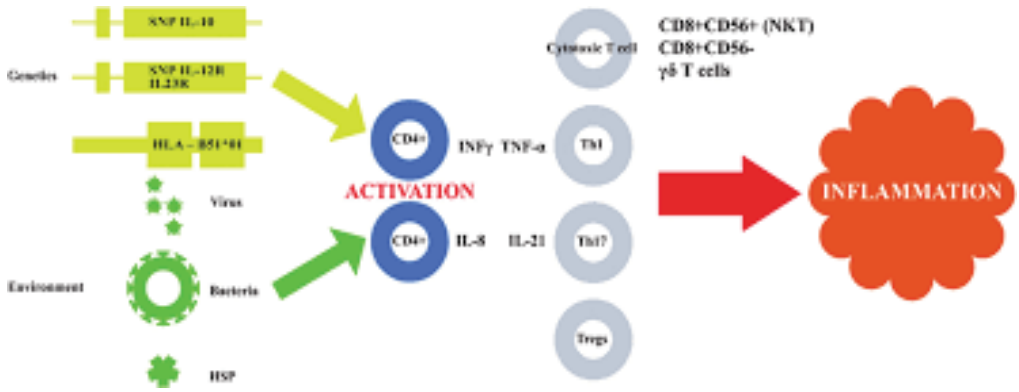

Figure 3. Inflammation process

The radiological examinations, including chest and sacroiliac radiography, revealed no abnormalities in four patients. However, two patients showed signs of vascular involvement, and further examinations with ultrasound Doppler and angio-CT confirmed the presence of vasculitis.

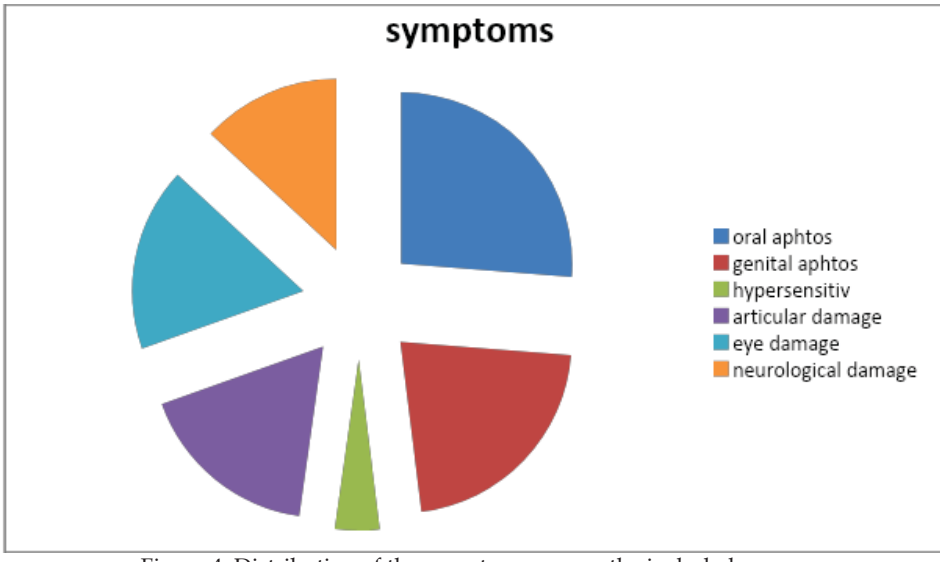

Figure 4. Distribution of the symptoms among the included group

In conclusion, this retrospective study of 6 patients with manifestations of Behçet's disease demonstrated the importance of oral aphthosis as a diagnostic criterion (Figure 4). Eye involvement, particularly uveitis, was a common feature, and some patients presented with systemic manifestations, including arthritis and vasculitis.

DISCUSSIONS

In the discussions section, the study sheds light on important aspects of Behçet's disease without repeating detailed data from previous sections. The historical significance of the disease is emphasised, with records of its existence dating back to ancient times, as described by Hippocrates. This historical background provides valuable insights into the long-standing impact and prevalence of the disease in different regions.

The epidemiological data highlights variations in Behçet's disease prevalence across different geographic areas, suggesting the influence of environmental factors. The study also reveals how age and sex can affect disease manifestation, with men showing more complications than women in certain regions.

Regarding the disease's cause, the study emphasises the complex interplay of genetic and environmental factors, leading to abnormal inflammatory responses and immune dysfunctions. These findings are consistent with previous research, supporting the notion of multiple factors contributing to the disease's onset.

The study emphasises the challenges in definitively diagnosing Behçet's disease due to the lack of specific biological tests. Clinical evaluation remains crucial, with recurrent oral lesions being important indicators. Dentists play a vital role in recognizing oral aphthosis as an initial symptom, highlighting the need for vigilance in identifying potential cases. Various clinical manifestations are discussed, including oral and genital aphthosis, eye damage, skin lesions, and damage to the articular, vascular, and nervous systems. Eye involvement is particularly common and early detection is crucial to prevent severe complications.

Treatment options are outlined, emphasising tailored therapeutic approaches based on disease severity and organ involvement. Corticosteroids are fundamental, while other immunosuppressants are considered for severe cases. The study acknowledges the limitations and potential adverse effects of certain treatments.

The study underscores the complexity of Behçet's disease prognosis, with early care playing a pivotal role in patient outcomes. Prompt treatment is essential to avoid irreversible damage and improve the quality of life for affected individuals.

The special section on Behçet disease in dentistry highlights the challenges faced by dentists in diagnosing the disease. Recognition of recurrent oral lesions and differentiation from other oral pathologies is crucial, emphasising the need for collaboration between dental and medical professionals for early diagnosis and management.

Overall, the study contributes valuable information to the understanding of Behçet's disease, offering insights into its historical context, epidemiology, etiopathogenesis, clinical manifestations, treatment approaches, and prognosis. These findings have implications for future research, aiming to explore more effective diagnostic methods, treatment modalities, and management strategies to improve patient outcomes.

Recommendations of EULAR (European League Against Rheumatism):

1. All patients with involvement of the posterior segment of the eye should be placed under general corticosteroid therapy and azathioprine

2. If the patient has a loss of visual acuity greater than 2/10 and / or retinal damage (vacuitis or maculopathy), cyclosporine A or infliximа should be combined with corticosteroids and azathioprine (the alternative being interferon-α)

3. There is no evidence on the treatment of large vessel involvement. For thrombosis venous immunosuppressant's (azathioprine, cyclophosphamide, cyclosporine); for pulmonary or arterial aneurysms corticosteroids are recommended and cyclophos

4. There is also no solid evidence for the use of anticoagulants, antiaggregants and fibrinolytics in thrombosis and venous or arterial lesions

5. There is no evidence for the treatment of intestinal manifestations of MB. Can be used sulfasalazine, corticosteroids, azathioprine, anti-TNF, and thalidomide before surgery, except emergency

6. Colchicines allow arthritis control in the majority of cases.

7. There is no controlled data that can guide the treatment of neuro-Behçet. For corticosteroids, interferon-α, cyclophosphamide, azathioprine, methotrexate and anti-TNFs. For cerebral venous thrombosis, the corticosteroids are recommended.

8. Cyclosporin A is contraindicated (unless expressly indicated for uveitis) in patients having an attack of the central nervous system.

9. The decision to treat mucocutaneous manifestations depends on their perceived severity. patient and doctor. In the front line for aphtosis and acne form lesions, it is recommended to use topical treatments. Colchicines is the treatment of erythema knotty. Azathioprine, interferon and anti-TNF drugs should be discussed in severe cases.

CONCLUSIONS

Behçet's disease remains a challenging and mysterious condition, with its definitive cause still unknown. Experts believe that a combination of genetic and environmental factors contributes to its development. Notably, both men and women can be affected by this disease, but the way it presents may differ between genders, leading to more complications in men [3, 4, 5].

Despite extensive research, there is no specific biological test for a certain positive diagnosis of Behçet's disease. Hence, clinical evaluation plays a vital role, focusing on identifying oral and genital aphthosis and eye damage as the main manifestations [6, 7]. Dentists play a critical role in early detection as oral aphthosis often serves as the initial symptom [9, 10]. Therefore, dentists need to be vigilant and inquire about other potential indicators of Behçet's disease. In some instances, dentists may also find themselves treating

patients with this condition, underscoring the importance of understanding effective management strategies to prevent complications.

The prognosis of Behçet's disease relies heavily on early detection and regular treatment. Patients must be well-informed about their condition and treatment options to effectively manage the disease and improve long-term outcomes.

In conclusion, Behçet's disease poses several challenges due to its uncertain cause and the lack of a definitive diagnostic test. Dentists' involvement is crucial in recognizing oral aphthosis and initiating appropriate actions. Educating patients about their condition and treatment is essential for better prognosis and overall well-being. Ongoing research is necessary to unravel the complexities of this disease and develop more targeted therapies.

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