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Case Report

Co-Occurrence of Behçet's Disease and Ankylosing Spondylitis: A Rare Case Report

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Abstract

Introduction

Behçet's Disease and Ankylosing Spondylitis are two distinct inflammatory conditions that have rarely been reported to coexist. This study reports a 41-year-old male patient with the co-occurrence of both conditions.

Case presentation

A 41-year-old male presented with chronic lower back pain for a period of 20 years. He also had left eye redness and painful oral ulcers. On physical examination, there were oral lesions and left eye redness (uveitis), and the patient's spine exhibited restricted mobility. Magnetic Resonance Imaging of the sacroiliac joint showed sacroiliacis affecting both sacroiliac joints. Hence, the patient was diagnosed as a case of mixed Behcet's disease and ankylosing spondylitis. The patient was managed with Azathioprine (Imuran) tablet (50 mg) twice daily, Colchicine tablet (1 mg) once daily, Topical steroid and Ulciguard mouth spray, Prednisolone tablet (5 mg) once daily, calcium and vitamin D3 tablet once daily, steroid eye drops, nonsteroidal anti-inflammatory drugs for 2 weeks. As a result, the patient achieved a dramatic response with all the symptoms resolved.

Conclusion

Behçet's disease and ankylosing spondylitis can coexist with overlapping presentations, making their clinical diagnosis challenging.

1. Introduction

Behçet's Disease (BD) and Ankylosing Spondylitis (AS) are two distinct chronic inflammatory diseases characterized by specific clinical presentations and multifaceted pathophysiology [1,2]. Behçet's disease, first described by Hulusi Behçet in 1937, manifests as a vasculitis syndrome affecting various-sized vessels and leads to oral and genital ulcers, uveitis, and skin lesions [3]. It most commonly affects people from the Mediterranean region to eastern Asia, with the highest prevalence observed in Turkey and Japan. The peak incidence of the disease is between twenty and forty years of age [4].

Ankylosing spondylitis, on the other hand, is a chronic systemic inflammatory condition of the axial skeleton that predominantly affects the spine and sacroiliac joints, leading to eventual fusion of the spine, known as ankylosis. Alongside joint involvement, systemic issues may arise. Males are predominant in AS, with the peak incidence occurring between the ages of 15 and 40 [1].

Though both diseases have distinct clinical and radiological features, the underlying inflammatory pathways suggest potential overlaps, with both conditions being associated with the dysregulation of T-helper cells and the production of pro-inflammatory cytokines [4]. However, the co-occurrence of BD and AS has rarely been documented in the literature, with only a handful of cases reported in medical literature to date excluding those published in non-peer-reviewed journals [5,6]. There is still ongoing debate regarding whether BD causes sacroiliitis and whether it should be classified among seronegative spondyloarthritides [1].

The present study describes a 41-year-old male patient with concurrent BD and AS presented with chronic lower back pain, painful oral ulcers, and left eye redness (uveitis).

2. Case Presentation

2.1. Patient information

A 41-year-old male presented with chronic lower back pain for a period of 20 years. He also had left eye redness and a painful oral ulcer.

2.2. Clinical findings

On physical examination, vital signs were stable. An oral lesion and left eye redness (uveitis) were observed, suggestive of BD. The patient's spine exhibited restricted mobility. All other systems, including the cardiovascular system, respiratory system, and gastrointestinal system, were normal. Additionally, the neurological examination showed normal results.

2.3. Diagnostic assessment

Routine laboratory investigations mostly showed normal findings. Erythrocyte Sedimentation Rate (ESR), viral screening, and serum vitamin D3 levels were all within normal range. Human leukocyte antigen (HLA) tissue typing of the patient revealed positive HLA-B27 and negative HLA-B51 status. Ultrasonography of the neck and echocardiography were normal. Further imaging diagnosis via a Computed Tomography scan of the chest revealed normal results, except for a bridging syndesmophyte. Magnetic Resonance Imaging of the sacroiliac joint showed sacroiliacis affecting both sacroiliac joints, suggestive of AS.

2.4. Therapeutic intervention

After diagnosing the patient with a case of mixed BD and AS, the patient was managed with the following interventions: Azathioprine (Imuran) tablet (50 mg) twice daily, Colchicine tablet (1 mg) once daily, Topical steroid and Ulciguard mouth spray, Prednisolone tablet (5 mg) once daily, calcium and vitamin D3 tablet once daily, steroid eye drops, nonsteroidal anti-inflammatory drugs (NSAID) for 2 weeks.

2.5. Follow-up and Outcome

As a result of the treatment regimen, the patient achieved a dramatic response with all the symptoms resolved. The patient continued on his treatment plan with close follow-up every 3 months.

4. Discussion

The rare concomitant occurrence of BD and AS observed in this case is rarely reported in the literature, which offers an opportunity to delve deeper into the complexities of autoimmune and inflammatory disorders. Such cases not only challenge clinical diagnostic acumen but also provide insights into potential overlapping pathophysiologies.

Behçet's disease predominantly manifests in populations from the Mediterranean region to eastern Asia. Turkey exhibits the highest prevalence of Behçet's disease, with 420 cases per 100,000 population. The overall prevalence of BD is 15.2 per 100,000 and does not differ by gender. Prevalence rates among Jewish, Arab, and Druze populations were 8.6, 26.2, and 146.4 per 100,000, respectively [7]. Meanwhile, AS has a worldwide distribution, albeit with a noted male predominance. The exact prevalence varies, occurring in 0.1-1.4% of the general population [8]. Our case was a 41-year-old male. Eser et al.'s study reported a 29-year-old male [9], and Olivieri et al.'s study reported a 45-year-old female with these coexisting diseases [10].

Clinical manifestations of BD include uveitis, genital ulcers, and recurrent oral aphthous ulcers [11]. Among the systemic symptoms, eye disease presents the highest morbidity, followed by vascular diseases, often due to active vasculitis [12]. Meanwhile, AS is characterized by abnormal ossification and ankylosis, primarily affecting the sacroiliac joints and axial bone. Additional clinical manifestations include peripheral joint pain, enthesitis, and extra-articular organ involvement [13]. Clinically distinguishing between BD and AS can be challenging, given the overlap in some symptoms. While sacroiliitis is a hallmark of AS, its association with BD remains contentious, with debates ongoing regarding BD's classification among seronegative spondyloarthritides [5]. The current case was presented with chronic lower back pain for the past 20 years. Similar cases of chronic lower back pain have been presented, with one case showing additional swelling and discomfort in both knees. Similar to our case, this patient had left eye redness and a painful oral ulcer [14].

During physical examination, the current patient's spine exhibited restricted mobility. In contrast to our case report, other reports indicated minimal yet painless spinal motion. Throughout our case finding and clinical examination, the patient was vitally stable, with normal GIT, respiratory, and cardiovascular systems. Our patient was characterized by a painful oral ulcer and left eye uveitis, similar to some other cases with posterior uveitis and oral ulcer [1,15].

At the heart of both diseases lies a dysregulated immune system. The fact that both diseases manifest dysregulation of T-helper cells and increased pro-inflammatory cytokine production indicates potential genetic predispositions that can be triggered by environmental or unknown factors. Infections, gut microbiota alterations, or even dietary factors could play roles in triggering or exacerbating these diseases [5]. Understanding these triggers, especially in cases with overlapping conditions, could offer insights into prevention or early intervention strategies. The HLA-B27 positivity in the current patient is a well-established marker for AS, while the absence of HLA-B51 negates one of the common genetic associations with BD [16,17]. In most reported patients with coexisting BD and AS, HLA-B27 and HLA-B51 are positive [1,14]. However, a case by Koçyiğit et al. had HLA-B5 positivity and HLA-B27 negativity [15]. This divergent genetic presentation in a patient with both conditions points to possible other genetic or epigenetic factors at play. In the current case, magnetic resonance imaging scans revealed sacroiliitis in both joints, consistent with the modified New York Criteria for Ankylosing Spondylitis diagnosis [18]. However, both ultrasound and computed tomography scans only revealed normal findings in this case.

The treatment approach in this patient, integrating therapies for both BD and AS, resulted in symptom resolution. Azathioprine and Colchicine are conventionally used for BD, with their anti-inflammatory properties proving beneficial [19]. NSAIDs are the first line of treatment for AS, providing symptomatic relief from inflammation and pain [20]. The addition of calcium and vitamin D3 supplements is essential for patients on long-term corticosteroids to counteract bone demineralization. In another reported case, Infliximab treatment resulted in substantial pain alleviation and symptom improvement [15]. In a study by Chang and colleagues, a 28-year-old man underwent treatment with NSAIDs, low-dose methotrexate, and sulfasalazine for three years, while our case report involved the use of analgesics and painkillers with no improvement. Their patient stopped using narcotics and experienced a recurrence of symptoms, including knee swelling and a scrotal sore [1].

5. Conclusion

Although rare, the concomitant occurrence of BD and AS can still happen and are associated with some overlapping symptoms. This underscores the complex interplay of genetic, environmental, and immunological factors in the pathogenesis of autoimmune and inflammatory diseases. Clinicians should be vigilant to the potential overlap of symptoms and ensure a comprehensive approach to diagnosis and management.

Declarations

Conflicts of interest: The author(s) have no conflicts of interest to disclose.

Ethical approval: Not applicable.

Patient consent (participation and publication)**:** Written informed consent was obtained from the patient for the publication of the present case report and any accompanying images.

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Authors' contributions: SMG participated in data collection, designed the study. AAQ, SMG participated in preparing the manuscript. HSM, AKG, KFHH and JIH critically revised the manuscript, literature review. AAQ, KFHH confirmed the authenticity of the data. All authors approved the final version of the manuscript.

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References

1. Chang HK, Cho EH, Kim JU, Herr H. A case of coexisting Behcet's disease and ankylosing spondylitis. The Korean Journal of Internal Medicine. 2000;15(1):93. doi:10.3904/kjim.2000.15.1.93
2. Shorsh A. Mohammed, Fahmi H. Kakamad, Soran H. Tahir, Marwan N. Hassan, Miran Najmadden Hassan, Berun A. Abdalla, et al. May-Thurner syndrome as the presenting symptom of Behcet's disease; a rare case report. *Barw Medical Journal*, 1(1). doi:10.58742/bmj.v1i1.17
3. Tahira SH, Kakamad FH, Hussein DM, Mohammed SA, Rshid RJ, Salih KM, et al. Behcet's disease presenting with focal fibrosing mediastinitis and cava thrombosis; a case report with a review of the literature. Barw Medical Journal. 2023;1(2). DOI: N/A
4. Habibagahi M, Habibagahi Z, Saidmardani SM, Sadeghian F. No definite association between human parvovirus B19 infection and Behcet disease. Iranian journal of medical sciences. 2015;40(6):493. DOI: N/A
5. Aktas Cetin E, Cosan F, Kucuksezer UC, Bilgic S, Cagatay Y, Gul A, et al. Behçet's disease: immunological relevance with arthritis of ankylosing spondylitis. Rheumatology international. 2013; 33:733-41. doi:10.1007/s00296-012-2446-9
6. Aso S. Muhialdeen, Jaafar Omer Ahmed, Hiwa O. Baba, Ismael Y. Abdullah, Hemn Ali Hassan, Kayhan A. Najar, et al. Kscien's List; A New Strategy to Discourage Predatory Journals and Publishers (Second Version). Barw Medical Journal. 2023;1(1). doi:10.58742/bmj.v1i1.14.
7. Krause I, Yankevich A, Fraser A, Rosner I, Mader R, Zisman D, et al. Prevalence and clinical aspects of Behcet's disease in the north of Israel. Clinical rheumatology. 2007; 26:555-60. doi:10.1007/s10067-006-0349-4
8. Dean LE, Jones GT, MacDonald AG, Downham C, Sturrock RD, Macfarlane GJ. Global prevalence of ankylosing spondylitis. Rheumatology. 2014;53(4):650-7. doi:10.1093/rheumatology/ket387
9. Eser F, Bodur H, Seçkin Ü. Coexistence of Behçet's disease and ankylosing spondylitis: A case report. Archives of Rheumatology. 2008;23(2):069-71. DOI: N/A
10. Olivieri IG, Gemignani GA, Busoni FR, Pecori FA, Camerini EM, Trippi DO, et al. Ankylosing spondylitis with predominant involvement of the cervical spine in a woman with Behçet's syndrome. Annals of the rheumatic diseases. 1988;47(9):780-3. doi:10.1136/ard.47.9.780
11. Yazici Y, Hatemi G, Bodaghi B, Cheon JH, Suzuki N, Ambrose N, et al. Behçet syndrome. Nature Reviews Disease Primers. 2021;7(1):67. doi:10.1038/s41572-021-00301-1
12. Demirkesen C, Tüzüner N, Mat C, Senocak M, Büyükbabani N, Tüzün Y, et al. Clinicopathologic evaluation of nodular cutaneous lesions of Behçet syndrome. American journal of clinical pathology. 2001;116(3):341-6. doi:10.1309/GCTH-0060-55K8-XCTT
13. Zhu W, He X, Cheng K, Zhang L, Chen D, Wang X, et al. Ankylosing spondylitis: etiology, pathogenesis, and treatments. Bone research. 2019;7(1):22. doi:10.1038/s41413-019-0057-8
14. Celik C, Halil UÇ, UÇKUN AÇ, ALEMDAROĞLU E. Coexistence of Behçet's disease and ankylosing spondylitis. Archives of Rheumatology. 2011;26(3):271-2. doi:10.5606/tjr.2011.045
15. Koçyiğit H, Turan Y, Bayram K, Gurgan A, Deveci H, Güvenç A. Coexistence of behçet's disease and ankylosing spondylitis: a case report. Archives of Rheumatology. 2010;25(4):217-20. doi:10.5152/tjr.2010.32
16. Akassou A, Bakri Y. Does HLA-B27 status influence ankylosing spondylitis phenotype?. Clinical Medicine Insights: Arthritis and Musculoskeletal Disorders. 2018; 11:1179544117751627. doi:10.1177/1179544117751627

17. Burillo-Sanz S, Montes-Cano MA, García-Lozano JR, Olivas-Martínez I, Ortego-Centeno N, García-Hernández FJ, et al. Behçet's disease and genetic interactions between HLA-B* 51 and variants in genes of autoinflammatory syndromes. Scientific Reports. 2019 Feb 26;9(1):2777. doi:10.1038/s41598-019-39113-5
18. Kardeş SI, Karagülle MI, Karagülle MZ, ERDOĞAN N. Coexistence of Behçet's disease and ankylosing spondylitis: Case report and review of the literature. Acta Med Mediterr. 2016; 32:947. doi:10.19193/0393-6384_2016_4_114
19. Alibaz-Oner F, Direskeneli H. Advances in the Treatment of Behcet's Disease. Current rheumatology reports. 2021;23(6):47. doi:10.1007/s11926-021-01011-z
20. Fan M, Liu J, Zhao B, Wu X, Li X, Gu J. Indirect comparison of NSAIDs for ankylosing spondylitis: Network meta-analysis of randomized, double-blinded, controlled trials. Experimental and Therapeutic Medicine. 2020;19(4):3031-41. doi:10.3892/etm.2020.8564