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Behçet's Disease Beyond the Triad: Unusual and Atypical Presentations

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Abstract

Behçet's disease (BD) is a chronic, multisystem, autoinflammatory disorder classically characterized by recurrent oral ulcers, genital ulcers, and uveitis. However, atypical presentations including vascular thrombosis, pulmonary artery aneurysms, gastrointestinal lesions, and neurological involvement can precede the classic triad, making early recognition challenging. We report three unusual cases: (1) a 38-year-old male with multisite vascular thrombosis, (2) a 39-year-old female with pulmonary artery aneurysms, and (3) a 30-year-old male with parenchymal neuro-Behçet's disease. All cases required multidisciplinary management and early immunosuppressive therapy to prevent life-threatening complications. These cases underscore the importance of high clinical suspicion and thorough investigation in patients presenting with atypical symptoms suggestive of BD.

Keywords: Behçet's Disease, Vascular Thrombosis, Pulmonary Artery Aneurysm, Neuro-Behçet, Atypical Presentation, Immunosuppression

Introduction

Behçet's disease (BD) is a variable vessel vasculitis affecting arteries and veins of all sizes. Initially described in 1937 by Hulusi Behçet, BD is classically defined by recurrent oral aphthae, genital ulcers, and uveitis. Atypical presentations such as vascular thrombosis, pulmonary aneurysms, gastrointestinal lesions, and neurological involvement are increasingly recognized. Early identification of these unusual manifestations is essential to prevent life-threatening complications.

Materials and Methods

This study is a descriptive case series of three patients diagnosed with BD with atypical presentations at Suhar Hospital, Oman. Detailed clinical histories, physical examinations, laboratory investigations, imaging studies, and treatment regimens were collected. Diagnoses were made according to the International Criteria for Behçet's Disease. Patient consent was obtained for publication.

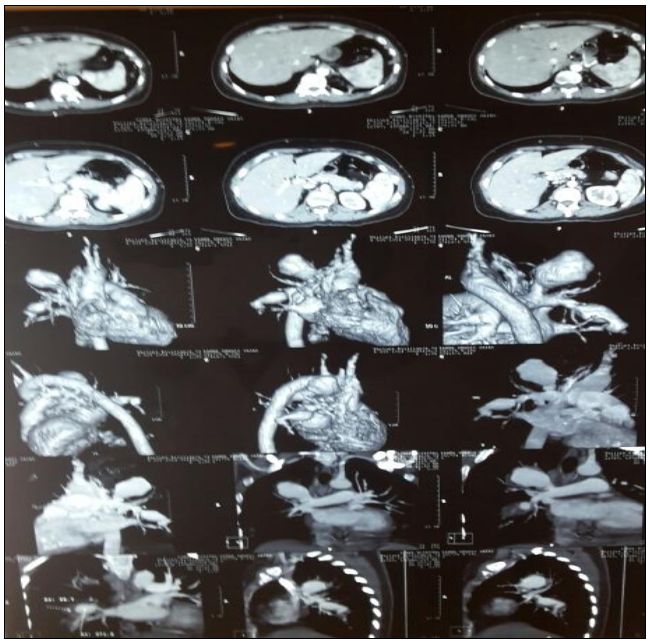
Results and Discussion

Case 1: Vascular Involvement Without Classic Triad

- Patient: 38-year-old male with history of idiopathic intracranial hypertension, seizures, and post-cholecystectomy.
- Presentation: Fever, weight loss, oral aphthous ulcers (no genital or ocular involvement).
- Investigations: Pan-CT showed IVC thrombosis, segmental pulmonary emboli, dilated subclavian/axillary veins, focal sigmoid thickening. Lab work-up unremarkable except high CRP/ESR; HLA-B51 positive.
- Management: IV methylprednisolone × 3 days, oral prednisolone taper, colchicine, azathioprine, warfarin 9 months.
- Outcome: Complete recovery; asymptomatic on colchicine PRN.

Case 2: Isolated Pulmonary Artery Aneurysms

- Patient: 39-year-old female with asthma and psychiatric history.
- Presentation: Fever, cough, hemoptysis, night sweats, weight loss; history of recurrent oral/genital ulcers.
- Investigations: CT chest revealed two large right pulmonary artery aneurysms with mural thrombosis; infectious and ANCA work-up negative. She had history of recurrent mouth and genital ulcers. She had positive HLA B51.



- Management: IV cyclophosphamide; anticoagulation withheld due to bleeding risk; cardiothoracic surgery not indicated.
- Follow-up: Lost to follow-up after transfer to tertiary care.

Case 3: Parenchymal Neuro-Behçet's Disease

- Patient: 30-year-old male with BD since 2016, chronic inactive hepatitis B, poor medication compliance.
- Presentation: Severe headache, diplopia, blurred vision; left sixth cranial nerve palsy.
- Investigations: MRI: parenchymal brainstem lesions; CSF: lymphocytic pleocytosis; infectious work-up negative. Multiple Sclerosis ruled out.
- Management: IV methylprednisolone × 5 days, maintenance prednisolone taper, azathioprine, colchicine; anti-TNF deferred; HBV prophylaxis with entecavir, latent TB prophylaxis with INH.
- Outcome: Dramatic improvement; follow-up MRI showed complete recovery.

Discussion

- Immunosuppression is central to BD management, especially in vascular and neurological involvement. Anticoagulation is not routinely recommended unless pulmonary artery aneurysms ruled out and anticoagulation are advised in certain cases according to the last EULAR guidelines.
- Unusual BD presentations often precede the classic triad, requiring high clinical suspicion.
- Multidisciplinary management including rheumatology, neurology, pulmonology, cardiology, and ophthalmology is essential.

Conclusion

Behçet's disease may present with atypical manifestations beyond the classic triad. Awareness of these presentations, early diagnosis, and timely immunosuppressive therapy are crucial to prevent severe complications and improve patient outcomes.

References

1. International Team for the Revision of the International Criteria for Behçet's Disease. J Eur Acad Dermatol Venereol. 2014; 28:338-347.
2. Hatemi G, *et al*. 2018 EULAR recommendations for the management of Behçet's disease. Ann Rheum Dis. 2018; 77:808-818.
3. Sakane T, *et al*. Behçet's disease. N Engl J Med. 1999; 341:1284-1291.
4. Hamuryudan V, *et al*. Pulmonary arterial aneurysms in Behçet's disease: Clinical features and outcome. Arthritis Rheum. 2004; 50:1391-1397.
5. Al-Araji A, Kidd DP. Neuro-Behçet's disease: Epidemiology, clinical characteristics, and management. Lancet Neurol. 2009; 8:192-204.