SAGE Open Medical Case Reports
Volume 11: 1–4

DOI: 10.1177/2050313X231182237
journals.sagepub.com/home/sco
Sage

Episcleritis; and posterior uveitis misdiagnosed as orbital cellulitis in a child patient with Behçet's disease

Mohsen Jari[1], Shidrokh Nasiri[2] and Mohammadreza Ghandehari[2]

Abstract

Behçet's disease is a chronic multisystem vasculitis that presented with several manifestations. Episcleritis is a benign inflammation of the episcleral tissue and is rarely reported in Behçet's disease. We reported an 11-year-old girl who was admitted due to pain and swelling around the left eye with initial diagnosis of preseptal cellulitis. The patient had a history of recurrent oral aphthous ulcers and genital ulcers. Episcleritis and posterior uveitis were reported in ophthalmic examination and ultrasonography. Pathergy skin test and human leukocyte antigens (HLA) B5 and HLA-B51 were positive, consistent with the diagnosis of Behcet's disease. This case report emphasizes that various pathological causes may cause episcleritis and uveitis in children, one of which is rheumatic diseases, that should always be considered.

Keywords

Behçet's disease, episcleritis, uveitis, case report

Date received: 18 March 2023; accepted: 30 May 2023

Background

Behçet's disease (BD) is a chronic condition that demonstrates itself by inflammation of the veins, arteries and capillaries. Manifestations of the disease include oral aphthous ulcer, genital ulcer, pseudofolliculitis, erythema nodosum, arthritis, and ocular manifestations.[1] Ocular involvement is a rheumatologic emergency; if left untreated, it could lead to blindness. Its manifestations can show themselves as anterior uveitis, posterior uveitis, retinal vasculitis, choroidal vasculitis, optic neuritis, and iridocyclitis.[2]

Episcleritis is a rare ocular manifestation of this disease. In this report, we introduce an 11-year-old patient with episcleritis and posterior uveitis as the first manifestation of BD. To conclude, the present case report demonstrates the importance of considering episcleritis as one of the differential diagnoses in patients presenting with the initially diagnosis of preseptal cellulitis, that one of the underlying causes may be rheumatic diseases including BD.

Case presentation

An 11-year-old girl was admitted in the pediatric infectious department of Imam Hossein Children's Hospital, Isfahan University of Medical Sciences, due to pain and swelling around the left eye (Figure 1).

The patient developed redness and photophobia a week before admission, then gradually developed pain in the eye movements, swelling, and redness around the left eye. She had normal vital signs on examination. On physical examination, she had redness and swelling around the left eye. Globe movements were associated with pain and limitation. Visual acuity was 5/10 in the left and 10/10 in the right eye. Cranial nerve examination and other physical examinations were normal.

The patient was diagnosed with preseptal cellulitis and treated with clindamycin intravenous infusion. The computed tomography (CT) scan orbit was normal, and no signs of orbital cellulitis were found. After 3 days of treatment, the

[1]Department of Pediatric Rheumatology, Imam Hossein Children's Hospital, Isfahan University of Medical Sciences, Isfahan, Iran
[2]School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran

Corresponding Author:
Mohsen Jari, Department of Pediatric Rheumatology, Imam Hossein Children's Hospital, Isfahan University of Medical Sciences, Isfahan, Iran.
Email: mjari14@yahoo.com

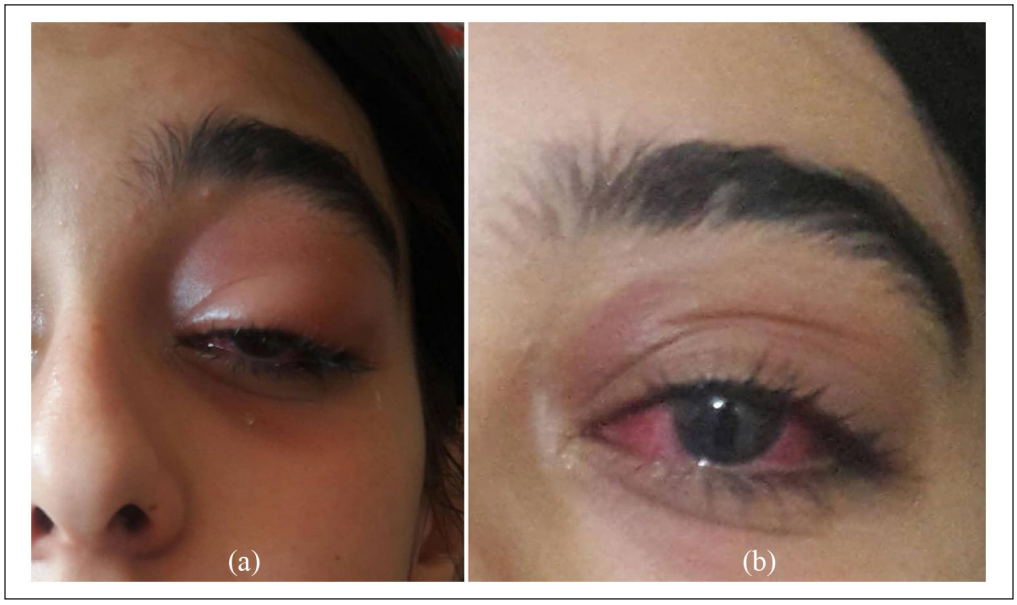

Figure 1. Patient images: (a) Swelling and erythema around the left eye which first was misdiagnosed as cellulitis. (b) Magnified image of the left eye demonstrating Swelling and erythema of the scleral tissue.

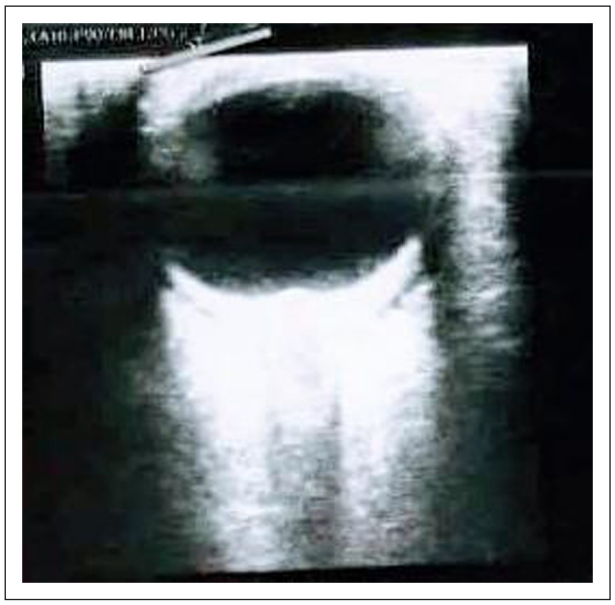

Figure 2. Sonographic image of the left eye. The thickness of the wall of the globe in the posterior pole is increased (2.4 mm), and the appearance of scleral edema with Classic view of T- sign is seen, which confirms episcleritis.

patient's condition did not improve. Therefore, an ophthalmology consultation was done for the patient

In ophthalmology consultation, posterior uveitis (diffuse choroiditis) was reported on the left eye with an ophthalmic examination. In the ultrasonography of the left eye, the thickness of the wall of the globe in the posterior pole was increased (2.4 mm), and the appearance of scleral edema with Classic view of T- sign Was seen, which confirmed episcleritis (Figure 2). Episcleritis and posterior uveitis were reported, and rheumatology consultation was recommended. Finally, the patient was transferred to the pediatric rheumatology department. A full history was taken there. The patient had a history of recurrent oral aphthous ulcers (5–6 times a year), genital ulcers, and chronic and vague abdominal pain. The results of laboratory tests including complete blood count, erythrocyte sedimentation rate, C-reactive protein, and serum electrolytes were normal. Serology tests for cytomegalovirus, toxoplasmosis, parvovirus B19, tuberculosis, and rheumatologic tests including angiotensin-converting enzyme, antinuclear antibody, and antiphospholipid antibodies were negative. The patient did not meet the laboratory and clinical criteria for lupus and granulomatosis with polyangiitis (GPA).

Both human leukocyte antigen (HLA) B5 and HLA-B51 tests were positive. The result of the pathergy skin test was positive (Figure 3).

According to the criteria of the International Study Group for Behçet Disease, the patient was recognized as a case of BD-related episcleritis and posterior uveitis, and the treatment started by prescribing pulse methylprednisolone and intravenous infliximab. Treatment was continued with oral prednisolone 2 mg/kg/d, azathioprine 2 mg/kg/d, and colchicine 1 mg/d.

After a 1-month follow-up, the patient had no symptoms and ocular exam through slit lamp and funduscopy was normal. Infusion of infliximab 8 mg/kg was given, and oral prednisolone 1 mg/kg/d with azathioprine and colchicine was continued. Three months after discharge, the patient did not have any complaints about aphthous ulcers and GI system visual acuity of both eyes was 10/10, and ocular exam was normal.

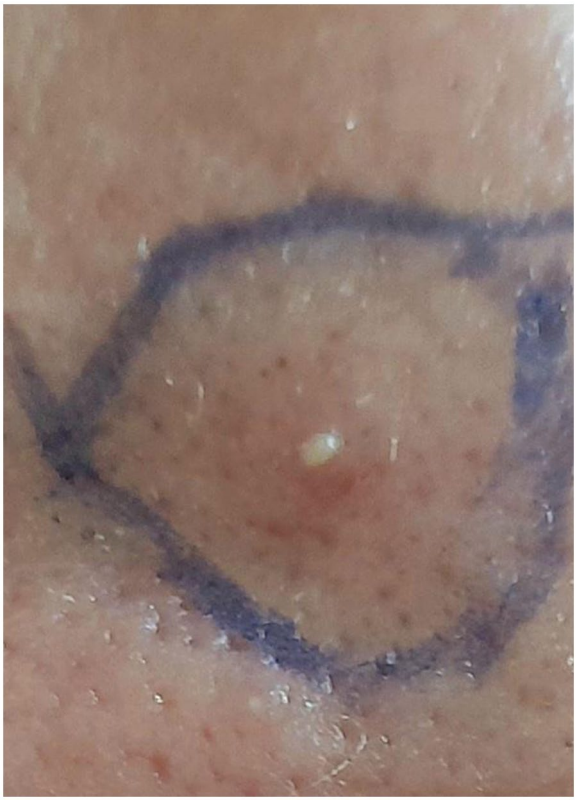

Figure 3. Positive pathergy test.

Discussion

Behçet's disease (BD) is a chronic multisystem vasculitis that develops by both endogenous and exogenous factors. This disease manifests itself with mucocutaneous, gastrointestinal, musculoskeletal, neurological, and ophthalmological lesions.[1]

BD etiology is yet to be unknown but genetic, viral, and allergic factors have been suggested. Immune complexes, aberration of T cell subsets, complement abnormalities and abnormal polymorph function have been seen. Autoantibodies are rare but have been reported against mucosal antigen. Anticardiolipin antibodies have been suggested to be associated with retinal vascular disease.[2,3]

Genetics bases of BD demonstrate in different geographic areas; there are some HLA typing that are strongly related to BD. For example, HLA-A28 and HLA-B12 are elevated in American people. Increased HLA is found in Turkish and Japanese patients.[3] In this case, we found positive HLA-B5 and HLA-B51.

Diagnosis in children is often delayed due to the different course of the disease from that of adults.[4] Children-BD have significantly fewer genital ulcers, yet they have more mild gastrointestinal symptoms, central nervous system involvement, and arthralgia.[5] One of the manifestations of BD is ocular involvement. In all, 30%–70% of patients suffer from this presentation which is said to be the most common in young patients but associated with a worse prognosis. Ocular involvement emerging at the onset of the disease is rare.[6]

Bilateral panuveitis is more common than iridocyclitis, keratitis, optic neuritis, and episcleritis.[7,8] Uveitis is a common finding and is almost pathognomonic of BD when it occurs in association with retinal branch vein occlusion.[9] Although episcleritis itself is unlikely to perforate the globe, this entity is painful and can, as in this case, be a sign of underlying serious disease. The exact mechanism for episcleritis remains a mystery. This presentation generally heals without treatment.[10] There is only one case report about episcleritis in pediatrics BD that concluded episcleritis and papillitis must be added to unusual complications of this disease.[11]

Episcleritis can occur with rheumatoid arthritis, juvenile idiopathic arthritis, reactive arthritis, systemic lupus erythematosus, inflammatory bowel disease, vasculitis, leukemia, Herpes simplex and zoster, Yersina infections, Lyme disease, and tuberculosis.[12–23] We ruled out these causes in this patient. An important point to note in our study was misdiagnosing the patient with preseptal cellulitis due to the first manifestations of the patient which were swelling and redness around the left eye.

Conclusions

To conclude, the authors suggest that in patients with the diagnosis of episcleritis, we must consider rheumatic diseases as one of the causes and BD as one of the differential diagnoses. Ignoring this scenario may lead to loss of a timely diagnosis and treatment; leaving patients with further risk of advancing to more complications and severe illness.

Acknowledgements

The authors are thankful to the patient who accepted to publish her case report.

Authors' contributions

M J: conception of the work and final edit, Sh N and M GH: have drafted the work.

Availability of data and material

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) disclosed receipt of the following financial support for the research, authorship, and/or publication of this article: This study is funded by Isfahan University of Medical Sciences, grant number 33007.

Ethical approval and consent to participate

We restate that institutional approval is not required to publish the case.

Informed consent

*Written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article.

ORCID iD

Mohsen Jari https://orcid.org/0000-0001-7001-6794

References

1. International Team for the Revision of the International Criteria for Behçet's Disease (ITR-ICBD). The International Criteria for Behçet's Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria. *J Eur Acad Dermatol Venereol* 2014; 28(3): 338–347.
2. Jari M, Mohammadi T and Taheri E. Bilateral retrobulbar optic neuritis as the first manifestation of neuro-Behçet disease. *Case Rep Rheumatol* 2020; 2020: 8834399.
3. Bulur I and Onder M. Behçet disease: new aspects. *Clin Dermatol* 2017; 35: 421–434.
4. Citirik M, Berker N, Songur MS, et al. Ocular findings in childhood-onset Behçet disease. *J AAPOS* 2009; 13(4): 391–395.
5. Krause I, Uziel Y, Guedj D, et al. Childhood Behçet's disease: clinical features and comparison with adult-onset disease. *Rheumatology* 1999; 38(5): 457–462.
6. Yildiz M, Koker O, Adrovic A, et al. Pediatric Behçet's disease—clinical aspects and current concepts. *Eur J Rheumatol* 2020; 7(1): 38–47.
7. Mendes D, Correia M, Barbedo M, et al. Behçet's disease—a contemporary review. *J Autoimmun* 2009; 32(3–4): 178–188.
8. Tugal-Tutkun I, Onal S, Altan-Yaycioglu R, et al. Uveitis in Behçet disease: an analysis of 880 patients. *Am J Ophthalmol* 2004; 138(3): 373–380.
9. James DG, Graham E and Hamblin A. Immunology of multisystem ocular disease. *Surv Ophthalmol* 1985; 30(3): 155–167.
10. McCluskey P and Powell RJ. The eye in systemic inflammatory diseases. *Lancet* 2004; 364(9451): 2125–2133.
11. Parentin F, Lepore L, Rabach I, et al. Paediatric Behçet's disease presenting with recurrent papillitis and episcleritis: a case report. *J Med Case Rep* 2011; 5(1): 81.
12. Promelle V, Goeb V and Gueudry J. Rheumatoid arthritis associated episcleritis and scleritis: an update on treatment perspectives. *J Clin Med* 2021; 10(10): 2118.
13. Misra A and Edelsten C. Orbital varices diagnosed as episcleritis in a child with juvenile idiopathic arthritis. *J Pediatr* 2005; 146(4): 574.
14. Zuber TJ, Pearlstein MV, Hwang CJ, et al. Periorbital swelling and episcleritis may be a sign of cutaneous lupus erythematosus. *Clin Case Rep* 2019; 7(7): 1422–1425.
15. Taniguchi Y, Nishikawa H, Yoshida T, et al. Expanding the spectrum of reactive arthritis (ReA): classic ReA and infection-related arthritis including poststreptococcal ReA, Poncet's disease, and iBCG-induced ReA. *Rheumatol Int* 2021; 41(8): 1387–1398.
16. Schuettenberg SP. Nodular scleritis, episcleritis and anterior uveitis as ocular complications of Crohn's disease. *J Am Optom Assoc* 1991; 62(5): 377–381.
17. Watanabe E, Tanaka A, Sugawara H, et al. Unclassified vasculitis with episcleritis, thrombophlebitis, deep vein thrombosis, pulmonary vasculitis, and intracranial vasculitis: an autopsy case report. *Am J Case Rep* 2019; 20: 886–895.
18. Tojima H, Kinoshita T, Kasahara Y, et al. [A case of Wegener's granulomatosis with episcleritis, mediastinal lesions, and pleural effusion and without serum anti-neutrophil cytoplasmic antibodies]. *Nihon Kyobu Shikkan Gakkai Zasshi* 1994; 32(1): 62–67.
19. Oono S, Kurimoto T, Oku H, et al. Adult T-cell leukemia presenting with episcleritis and secondary glaucoma. *Jpn J Ophthalmol* 2009; 53(1): 70–71.
20. Hoshing A, Seth A and Subramaniam A. Presumed herpes simplex virus ulcerative nodular episcleritis treated by topical acyclovir alone. *Oman J Ophthalmol* 2022; 15(3): 360–362.
21. Ahvonen P and Dickhoff K. Uveitis, episcleritis and conjunctivitis associated with Yersinia infection. *Acta Ophthalmol Suppl* 1974; 123: 209–212.
22. Aldebasi TM, Alasiri AA, Alnahdi MA, et al. Tubercular episcleritis: a review of literature. *Middle East Afr J Ophthalmol* 2022; 29(1): 51–55.
23. Flach AJ and Lavoie PE. Episcleritis, conjunctivitis, and keratitis as ocular manifestations of Lyme disease. *Ophthalmology* 1990; 97(8): 973–975.