

# Retinal vasculitis and skin ulcer preceding Budd–Chiari syndrome in a patient with Behcet’s disease

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Behcet’s disease (BD) is a multisystem disorder with a classical triad of recurrent oral ulcers, genital ulcers, and uveitis. It involves both arterial and venous system which can lead to life-threatening complications. Both superficial and deep venous systems can be involved leading to peripheral skin lesions and devastating complications like cerebral venous thrombosis and Budd–Chiari syndrome (BCS). This report describes a case of an HLA B-52 positive BD in a 22-year-old woman who presented with retinal vasculitis and venous ulcer on the foot and later on developed ascites due to obstruction of supra-hepatic inferior vena cava (BCS). This report highlights the fact that BCS can develop in patients of BD and every ophthalmologist should be aware of this life-threatening complication while they are treating these patients, as timely diagnosis and intervention can prevent mortality.

**Key words:** Behcet’s disease, Budd–Chiari syndrome, HLA B-52, retinal vasculitis, venous ulcer

Retinal vasculitis is a common presenting feature of Behcet’s disease (BD). It is a chronic relapsing vasculitis with both ocular and systemic involvement.<sup>[1]</sup> Budd–Chiari Syndrome (BCS) is an uncommon condition caused by thrombotic or non-thrombotic hepatic venous outflow obstruction.<sup>[2]</sup> BD presenting with retinal vasculitis and subsequently developing BCS has been reported only once, almost a decade ago.<sup>[3]</sup> We report a case of BD in whom retinal vasculitis preceded BCS.

## Case Report

A 22-year-old lactating Indian woman presented with sudden painless onset diminution of vision in her left eye for 8 days. General examination revealed purpuric rashes over the

wrist [Fig. 1a] and non-pitting pedal edema with a venous ulcer on the right foot [Fig. 1b]. She had a history of multiple episodes of painful oral ulcers in the last 6 months. Her menstrual history was unremarkable, with no history of abortions.

Visual acuity was 20/20 in the right eye (RE) and 20/600 in the left eye (LE) with a relative afferent pupillary defect. Rest of the anterior segment was unremarkable in both eyes. Fundus was normal in the right eye [Fig. 2a]. The left eye had optic disc edema along with diffuse phlebitis, multiple dot hemorrhages in all quadrants, and diffuse thickening of the retina at the macula [Fig. 2b]. The general background of the fundus was pale when compared with the right eye. Fundus fluorescein angiography (FFA) of the left eye confirmed the presence of diffuse occlusive phlebitis with a filling defect in the supero-temporal vein, persisting till late phase [Fig. 2c and d]. Optical coherence tomography (OCT) of the left eye confirmed the presence of macular edema [Fig. 3b] with a neuro-sensory detachment. FFA and OCT of the right eye were within normal limits [Fig. 3a].

Her vitals were normal, total leucocyte count was 15,500 per microliter and hemoglobin was 8.5 gm/dl. Keeping in mind a predominant venous involvement, HLA typing was performed and HLA B-52 turned out to be positive. Other causes of vasculitis were ruled out as anti-nuclear antibody (ANA), anti-neutrophil cytoplasmic antibody (cANCA), and anti-phospholipid antibody (APLA) were negative.

A diagnosis of BD was made based on ocular and systemic manifestations and she was started on intravenous dexamethasone 200 mg for 3 days followed by oral prednisolone 45 mg once daily which was tapered slowly by 5 mg per week. The response to steroids was good with complete disappearance of skin lesions on the upper limb [Fig. 1c] and a significant reduction in pedal edema within 10 days of initiation of treatment [Fig. 1d]. Visual acuity improved to 20/200, macular edema had resolved completely. [Fig. 3d] The phlebitis healed, with FFA showing a significant reduction in retinal vascular leakage [Fig. 4a and b]. Visual acuity in the right eye was maintained at 20/20 until 1 month following the initial presentation when it dropped to 20/32. OCT at this visit showed focal photoreceptor outer segment loss, which was healthy in the previous visits [Fig. 3a and c]. At this point, the vasculitis in the left eye had healed completely and steroids were tapered slowly.

Two months later she presented with ascites and was referred to a physician for the same, where she was diagnosed to have BCS with an obstruction in the supra-hepatic Inferior Vena Cava (IVC) [Fig. 4c]. She had been advised surgery for relieving the IVC obstruction.

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| Quick Response Code:  | Website:<br>www.ijo.in          |
|  | DOI:<br>10.4103/ijo.IJO_1338_20 |

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Received: 06-May-2020

Revision: 20-May-2020

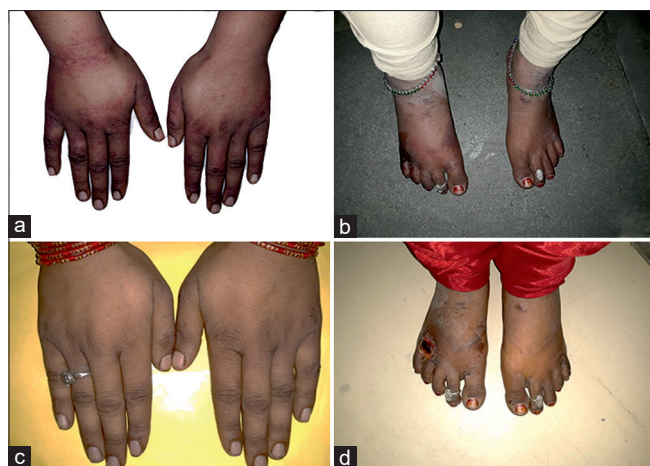
Accepted: 03-Jun-2020

Published: 20-Aug-2020

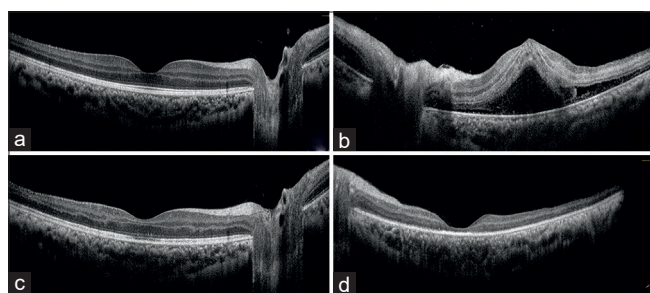
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**Cite this article as:** Bhatia P, Sen A, Kohli GM, Shenoy P. Retinal vasculitis and skin ulcer preceding Budd–Chiari syndrome in a patient with Behcet’s disease. Indian J Ophthalmol 2020;68:2015-7.



**Figure 1:** Pre and post treatment images of hands and feet. (a) Purpuric rashes over the wrist. (b) Non-pitting pedal oedema with venous ulcer on right foot. (c) Purpuric rashes over the wrist have disappeared after initiating systemic steroids. (d) Pedal oedema has decreased



**Figure 3:** OCT at presentation and follow-up. (a) Normal OCT of right eye at presentation. (b) OCT of left eye showing macular oedema with a large neurosensory detachment. (c) OCT of right eye at one month showing photoreceptor outer-segment loss at the fovea. (d) OCT showing photoreceptor loss and resolved macular oedema

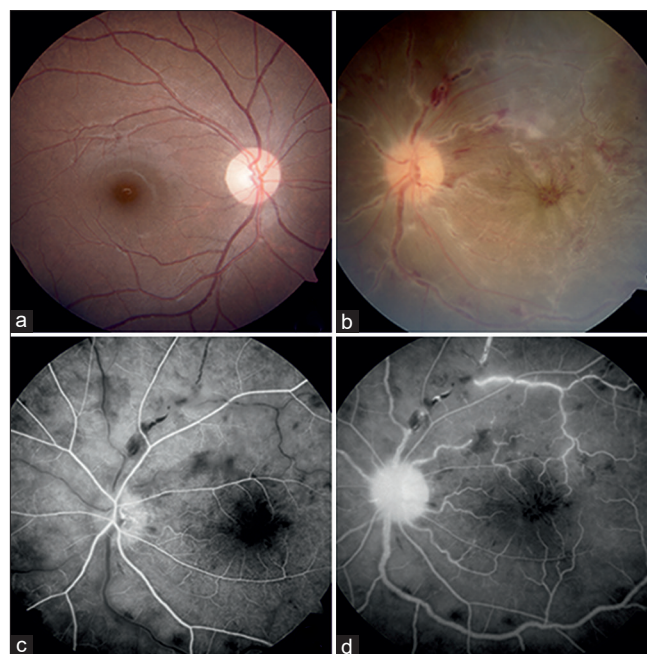
## Discussion

The clinical diagnostic criteria for BD according to the International Study Group guidelines requires the presence of recurrent oral ulcers (major criteria) and at least two of the four minor criteria which includes: recurrent genital ulcerations, uveitis, skin lesions, and a positive pathergy test.<sup>[1]</sup>

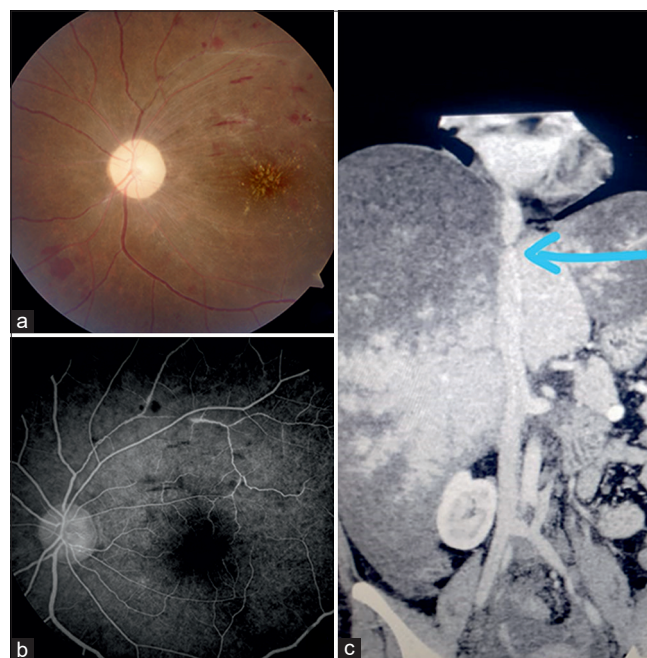
In the present case, the diagnosis of BD was relatively straightforward with presence of recurrent oral ulcers, retinal vasculitis, and papulopustular skin lesion. Our clinical diagnosis was supported by a positive test for HLA B-52 antigen.

There have been a few reports of concomitant BCS and BD.<sup>[4]</sup> However, in most of these cases the patients presented initially with BCS, the causality of which was later established to BD.<sup>[4]</sup> BCS developing subsequent to ocular BD is a rare scenario with only one case reported so far, wherein the authors noted the development of ascites due to BCS within a week of being diagnosed as BD.<sup>[3]</sup>

HLA B-51 and B-52 are split antigens of HLA B-5. Although HLA B-52 has been more commonly found in association with Takayasu's arteritis, it has also been reported with BD.<sup>[5,6]</sup> BD can have multisystem involvement and needs long-term immunosuppression not only to preserve vision, but also to avoid life-threatening complications.<sup>[1,7]</sup> In the



**Figure 2:** Fundus photograph and FFA at presentation. (a) Fundus photograph of right eye at presentation. (b) Fundus photograph of left eye at presentation showing vasculitis, disc edema and macular edema. (c) Early phase fluorescein-angiogram of left eye. (d) Late phase fluorescein-angiogram of left eye showing occlusive phlebitis and disc leak



**Figure 4:** (a) Fundus photograph showing healed phlebitis with resolution of disc oedema and macular oedema. (b) FFA showing healed retinal vasculitis with dye filling up in the supero temporal arcade. (c) CT scan showing supra-hepatic IVC Obstruction (Blue arrow)

present case initial immunosuppression was achieved with pulse steroid therapy and subsequently by oral steroids. Immunosuppression with azathioprine was initially planned but was deferred in view of ascites and deranged liver function. During follow-up visits, loss of photoreceptor outer segments

in the right eye was noted, the reason for which remains undetermined. This could possibly represent a subclinical vasculitis in the RE, that we could not assess angiographically due to her systemic condition.

## Conclusion

What makes this case noteworthy is the fact that our patient presenting with BD subsequently developed systemic complications of BCS with IVC involvement. BCS is a life-threatening complication of BD and every ophthalmologist should be aware of this entity while treating these patients.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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