Behçet Disease in a Child: A Rare Disorder with an Unusual Complication and Favorable Outcome

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A 6-year-old boy presented to us with a history of painful oral lesions involving the tongue, oral mucosa, and buccal mucosa for 1 month. On day 5 of the illness, he started having fever and developed a pustular lesion over right forearm. For these complaints, he was hospitalized elsewhere and treated with intravenous antimicrobials and acyclovir but did not respond. Lesions continued to progress and he developed an ulcer at the site of venipuncture. He had a history of recurrent painful oral ulcerations in the past (every 4–6 weeks, each episode lasting 7–10 days, since the age of 3 years with increasing frequency), for which he was being given topical medications. There was no history of genital ulcers, abdominal pain, diarrhea, headache, arthritis, or ocular complaints. On examination, he was febrile and had multiple oral ulcers involving the tongue, buccal mucosa, labial mucosa, and hard palate [Figure 1a]. The lesions were shallow surrounded by a narrow rim of erythema and most were covered with a greyish membrane. There was an ulcerative lesion on the ventral aspect of the right forearm [Figure 1b] and papulopustular lesions over the right elbow and left lateral malleolus. Systemic examination including ophthalmological examination was unremarkable. Investigations revealed hemoglobin (Hb) 10.8 g/dl, white blood cell (WBC) count 3800/µl (P_{50}, L_{36}, M_{10}, E_4), and platelet count 303 × 10^3/µl. Pathergy test was positive. Antinuclear antibody (ANA), done by indirect immunofluorescence using Hep2 cells, was negative. Fecal calprotectin was normal and human leukocyte antigen B51 (HLA B51) was negative. Skin biopsy was performed which revealed leukocytoclastic vasculitis [Figure 2]. The child started having high-grade fever during the hospital stay. Hb, total leukocyte count, and platelets progressively declined (Hb 9.7 g/dl, WBC 3400/µl, platelets 229 × 10^3/µl). Serum transaminases were mildly elevated, aspartate amino transferase (AST) 86 U/l (Normal 7–55 U/l) and alanine amino transferase (ALT) 74 U/l (Normal 8–48 U/l). Considering a possibility of macrophage activation syndrome (MAS), bone marrow aspiration and biopsy were performed, which revealed hemophagocytosis. Serum triglycerides and fibrinogen were normal. He was treated with intravenous immunoglobulins (2 g/kg) to which he responded. Fever subsided, oral ulcers, however, persisted. In view of recurrent oral ulcers, positive pathergy test, and skin biopsy showing leukocytoclastic vasculitis, a possibility of Behçet's disease was considered and oral

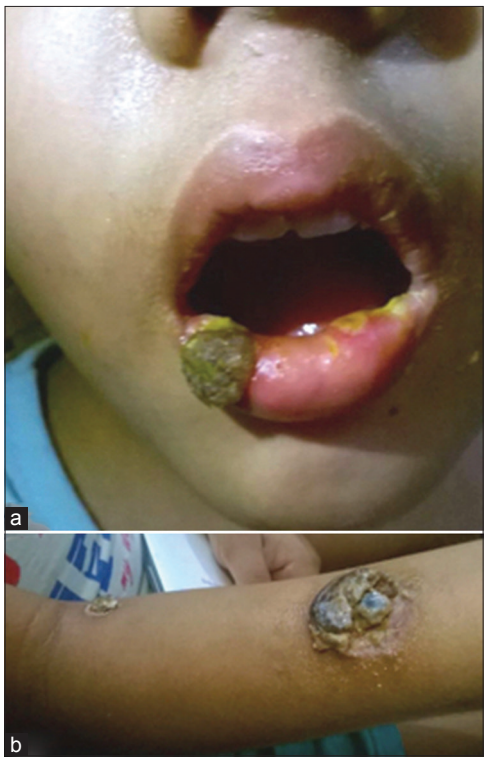

Figure 1: (a) Aphthae on lower labial mucosa. An aphtha with overlying thick crust is present on the right side. (b) Ulcers with overlying thick crust on the forearm

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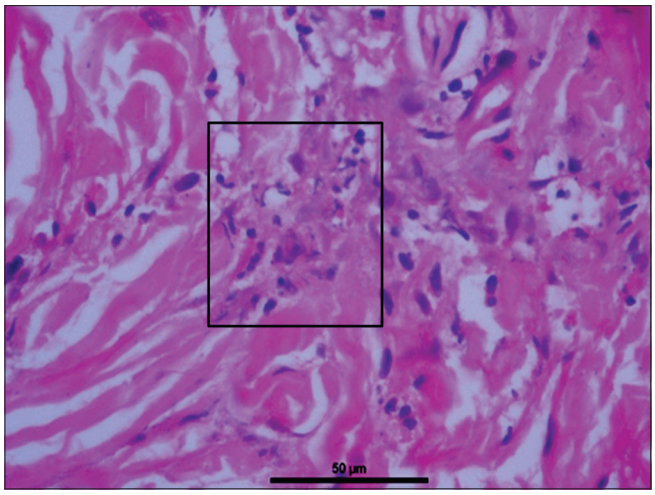

Figure 2: High-power photomicrograph of the skin biopsy showing destruction of the dermal capillaries as indicated by swollen endothelial cells and neutrophil infiltration with nuclear debris which has been highlighted by a black box. (hematoxylin and eosin stain, ×500)

prednisolone was started in tapering doses (initial dose 0.5 mg/kg) along with colchicine at a dose of 1 mg/day in two divided doses (0.06 mg/kg/day). Prednisolone was stopped in a month and he continues to be on colchicine and has remained asymptomatic for more than 1 year. He is on regular follow-up. There has been a recurrence with minor aphthae and skin lesions have healed with scarring [Figure 3a and b].

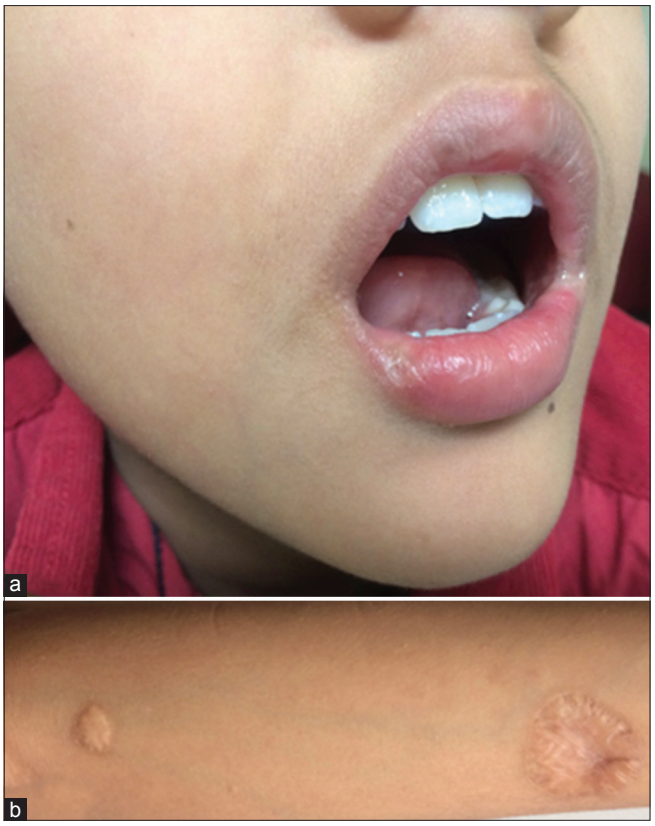

Figure 3: (a) At 1-year of follow-up, completely healed lesions with scarring on lip. (b) At 1 year of follow-up, lesions have healed with scarring

Behçet's disease is a rare childhood vasculitic disorder classified as variable vessel vasculitis.[1] Recurrent oral ulceration is the most common and sometimes the only presenting feature.[2] Pathergy test and HLA B51 do not find a place in the recent classification criteria.[3] Leukocytoclastic vasculitis has been described as a histopathological feature of cutaneous manifestation of Behçet's disease.[4] MAS has been described with a multitude of childhood rheumatological diseases, the most common being systemic juvenile idiopathic arthritis, systemic lupus erythematosus, and Kawasaki disease. It is an exaggerated inflammatory response resulting in cytokine storm which occurs due to immune dysregulation, aberrant activation of T-lymphocytes, and macrophages. Delay in recognition and treatment is fatal. Diagnosis of MAS is based on a combination of clinical features and laboratory findings in the form of decreasing blood cell lines, low serum fibrinogen, transaminitis, elevated serum ferritin, and triglycerides. Treatment consists of immune-modulation with corticosteroids, cyclosporine, and etoposide. Refractory cases may require interleukin-1 blockade.[5] MAS has not been commonly described in association with Behçet's disease. A single case report in an adult who developed Epstein Barr virus associated hemophagocytic lymphohistiocytosis while on follow-up is available.[6] Alpsoy *et al*. reported that serum from Behçet's disease patients caused proinflammatory activation of macrophages.[7] MAS, though not commonly reported in patients with Behçet's disease, must be considered in clinically appropriate settings. Delay in recognition and treatment can be fatal.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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