Journal of the Turkish Academy of Dermatology
eISSN 1307-394X

Case Report

Behçet's Disease After H1N1 Vaccination

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Published:
J Turk Acad Dermatol 2011; **5 (1)**: 1151c1
This article is available from: http://www.jtad.org/2011/1/jtad1151c1.pdf
Key Words: Behçet's disease, H1N1 vaccination

Abstract

Background: Behçet's disease is a chronic, relapsing, multisystemic vasculitis characterized mainly with ulcerations of the oral and genital mucosa, ocular, articular and vascular as well as further organ involvements. The aetiology of Behçet's disease is unknown. Most widely held hypothesis of the disease pathogenesis is that an altered immune response triggered by an infectious agent or by an auto antigen in a genetically predisposed host.

We report a 26-year-old male patient with ulcers of oral and genital mucosa, which had occurred 48 hours after H1N1 vaccination. He was diagnosed as Behçet's disease with the clinical findings. Since viral and bacterial antigens can precipitate Behçet's disease via triggering immune response, we think that H1N1 vaccination may be precipitant in this case. This is the first case in the literature showing the association of Behçet's disease and H1N1 vaccination.

Introduction

Behçet's disease is a multisystemic vasculitis with classic triad of oral aphthous ulceration, genital ulceration and eye involvement [1]. The disease is most common between 20-40 years of age and though both sexes are affected equally, it has a more severe clinical course in men [2]. The aetiology of Behçet's disease is unknown. Autoimmunity, infections, heredity and environmental factors have been frequently described in the aetiopathogenesis [3]. Our patient is the first case in the literature showing the association of Behçet's disease and H1N1 vaccination while there is only one case reported to be triggered with different vaccination (typhoid vaccination) in the literature [4]. We presented this case because Behçet's disease may be related to H1N1 vaccination in this patient.

Case Report

Twenty six year-old male patient applied to our clinic 2 days ago with complaint of scrotal ulcer. Patient had H1N1 vaccination and 4 days after vaccination muscle aches, headache and fever have developed. A small inflammatory papule appeared on the scrotal area 48 hours after H1N1 vaccination and in a short time took the form of a wound with cavity. Patient had recurrent oral aphtae since 4 years with a few oral ulcers occurring every month. It had been learned that patient had been examined for Behçet's disease and was receiving colchicine treatment with recurrent aphthous stomatitis diagnosis. There was no history of a previous genital ulcer or suspicious sexual intercourse.

On dermatologic examination an ulcer was present on left lateral side of scrotum. It was sharply bordered with slightly swollen edge and was approximately 1.5 cm in diameter and the floor of ulcer was covered with necrotic tissue (**Figure 1**) and

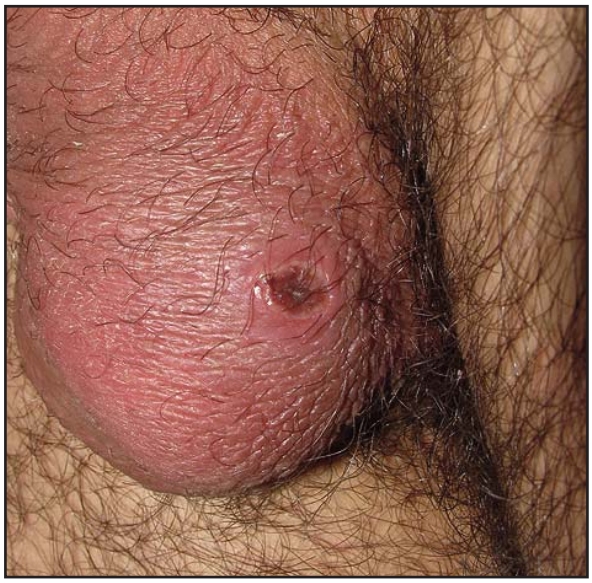

Figure 1. Genital ulceration on the scrotum of the patient

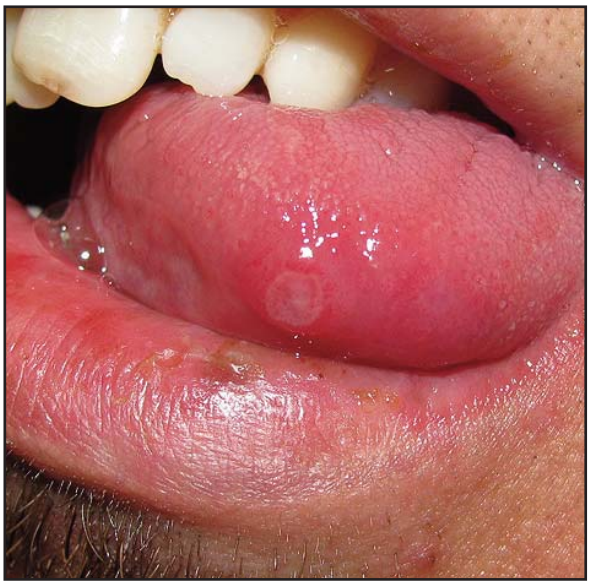

Figure 2. Minor oral aphtae on the tip of the tongue of the patient

on the tip of the tongue there was an aphthae which was 5 mm in diameter (**Figure 2**). On legs and buttocks there were scattered erythematous papules and pustules 1-2 mm in diameter. The pathergy test was performed on front arm and was 1 positive after 48 hours (**Figure 3**). Eye examination was normal and there was no pathology on systemic examination. Family history was of no significance.

Routine laboratory studies were made and complete blood count, biochemistry panel, complete urine examination, erythrocyte sedimentation rate, TPHA, VDRL-RPR, hepatitis markers and chest X-ray were within normal limits. CRP level was high with 19 U/L normal range: 0-5). Gram staining of ulcer samples was negative for bacteria and leukocytes. On wound culture coagulase positive staphylococci were isolated and were considered to be contamination.

Based on *International Study Group Criteria* patient was diagnosed as Behçet's disease with four signs

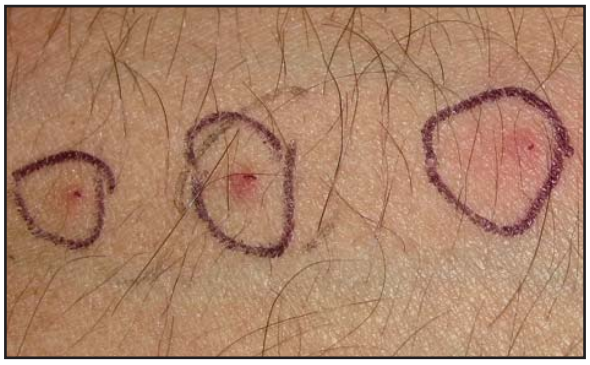

Figure 3. Positive pathergy reaction

[**5**]. Treatment with colchicine tablets (0.5 mg three times in a day) and topical corticosteroid for genital ulcer (Prednisolone + iodochlorhydroxiquine) were started. The patient's genital ulcer began to heal after 2 weeks, leaving with the atrophic scar.

Discussion

Behçet's disease is a chronic, relapsing, multisystemic disorder characterized by recurrent oral and genital ulcers, ocular lesions, skin manifestations, arthritis, intestinal, vascular and neurological involvement [**3**]. The highest prevalence has been reported in Turkey as 8-37/10.000 [**6**].

Behçet's disease is considered to be a vasculitis triggered by immunological mechanisms, but pathogenesis could not be fully elucidated [**3**]. For today, the most emphasised hypothesis is that Behçet's disease is an irregular immune response in genetically predisposed individuals against environmental antigens such as viral, bacterial, etc., and / or autoantigens such as heat shock proteins [**2,7**].

Genital ulceration, oral ulceration, acneiform eruptions and 1+ positive pathergy test of the patient fulfil the criteria of International Study Group for Behçet's Disease. The occurence of Behçet's disease 48 hours after vaccination in the case implicate vaccination as a possible trigger although the timing of vac-

cination before the onset of Behçet's disease may also just be a coincidence. The relation to vaccination with Behçet's disease is not clear whether it is a causal association or a mere coincidence.

Though reports are present implicating vaccination in occurrence of autoimmune and vasculitic diseases strong associations have not been demonstrated between autoimmunity and vaccination except in a few cases [**8**]. Development of *Henoch-Schönlein* purpura (HSP), leukocytoclastic vasculitis, ANCA-associated vasculitis, giant cell arteritis following influenza and meningitis C vaccination have been reported [**9, 10, 11, 12, 13, 14**]. Moreover HSP and Churg-Strauss vasculitis were reported after hepatitis B and pneumococcal vaccinations [**11, 15, 16**]. Behçet's disease is also a systemic vasculitis.

Only one case of Behçet's disease triggered by vaccination has been reported previously. *Molloy* et al reported a case of Behçet's disease occurring 4 days after the third typhoid vaccination [**4**].

Although it is not clear how vaccinations trigger autoimmune diseases several mechanisms including molecular mimicry, polyclonal activation and the induction of systemic or local proinflammatory states have been proposed [**8, 12**].

Onset of Behçet's disease 48 hours after H1N1 vaccination make us consider that vaccination can be added to the list of potential triggering causes.

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