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Case Report

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Henoch-Schonlein Purpura in A Patient with Behcet's Disease Presenting with Penile Ulcer

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Abstract

Background: Behçet's disease is a chronic, relapsing, multi-systemic inflammatory process characterized by a triad of oral ulcers, genital ulcers and ocular inflammation.

Case report: Seventeen-years-old male patient who has been suffering recurrent oral aphthous lesions for one year presented with complaints of general fever, arthralgia, and rash on his legs which started ten days ago. He had different sizes of purpuras and two painful lesion of erythema nodosum at his both legs. A 4-5 cm necrotic wound through corpus to glans penis was observed at physical examination. His acute phase proteins were elevated; the pathergy test was positive with normal eyes examination. Skin biopsy revealed leukocytoclastic vasculitis with the deposition of Ig A and C3. These biopsy findings were compatible with a diagnosis of Henoch-Schönlein purpura.

Conclusion: Although Bechet's disease is rare in childhood, it should be considered in differential diagnosis of children who have recurrent aphthous lesions. Since adolescents may be shy to mention about their genital ulcers during anamnesis, all systems particularly genital system should be examined carefully. In addition, HSP should be kept in mind in patients with Bechet's disease who have palpable purpura.

Keywords: Atypical Penis Involvement; Behcet's Disease; Henoch-Schönlein Purpura

Introduction

Behcet's Disease (BD) was first described in 1937 by Professor Ordinarius Dr. Hulusi Behcet as a triple complex of recurrent oral aphthae, genital ulcers and hypopyon-iridocyclitis [1]. Although its etiology and pathogenesis is not fully known, widely accepted hypothesis dictates the disease is caused by an intense immune-mediated inflammatory response that is triggered by environmental factors (frequently infectious agents) in genetically susceptible individuals. Geographical distribution of the disease condenses along the countries situated along the historical "Silk Road" [2]. Genital ulcers are mostly reported on scrotum in men. In this paper, we present an adolescent with BD who has an atypical genital involvement and Henoch-Schönlein Purpura (HSP).

Case Report

Seventeen-years-old male patient who previously has been suffering recurrent oral aphthous lesions for one year presented with complaints of general fever, arthralgia, and rash on his legs which started ten days ago. In his physical examination, BW: 50 kg (3-10P), height: 174 cm (50-75P), body temperature: 38.40C, blood pressure:100/74 mmHg, there were 3-4 oral aphthae on his oral mucosa. There were purpuric lesions at varying sizes and two painful lesions resembling erythema nodosum at lower extremities. At first, patient refused the genital examination, but after a short and effective persuasive speech; he agreed to be physically examined. A big necrotic lesion on the tip of penis extending to glans penis was observed but there was no lesion on scrotum (figure 1).

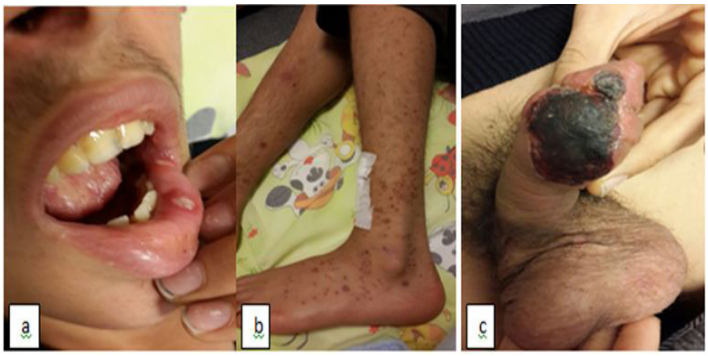

Figure 1: (a) Recurrent oral ulcers in the mouth **(b)** Purpuric rash and erythema nodosum at lower extremities **(c)** Big necrotic lesion on the tip of penis extending to glans penis.

In his laboratory examination, complete blood count was normal. Erythrocyte Sedimentation Rate (ESR) was 52 mm/hr, CRP level was 14.5mg/dL (0-0.5). His biochemical parameters, urinalysis and coagulation tests were normal. C3 and C4 levels were normal. VDRL-RPR and Treponema Pallidum Hemagglutination (TPHA) tests were negative. Additionally, ANA, p-ANCA, c-ANCA were negative. Pathergy test was positive, and ocular examination was normal. HLA B51 was positive. Considering his findings, oral aphthous lesions, penile ulcer, erythema nodosum lesions in addition to positive pathergy test; he was diagnosed to have Behcet's disease based on the diagnostic criteria of International Behcet study group [2]. Palpable purpura on both lower limbs led us to perform a skin biopsy for the differential diagnosis of vasculitis. Skin biopsy revealed leukocytoclastic vasculitis with the deposition of Ig A and C3. These biopsy findings were compatible with a diagnosis of HSP. Based on palpable purpura on lower limbs and typically leukocytoclastic vasculitis with predominant IgA. He was diagnosed as HSP according to EULAR/PRINTO/PRES criteria [3]. He was started on colchicine 1 mg/kg/day oral prednisone 30mg twice a day, with resolution of his symptoms and a decrease in ESR and CRP. Local treatment was administered to the lesion on the tip of penis.

Discussion

Behcet's disease is generally observed in young adults aged between 20 and 40 years old but can rarely be seen in children. The disease has a more severe course in young adults, males and in far eastern races [2,4]. Our case was an adolescent, which is a rare period for development of this disease [5]. Almost all patients have recurrent oral ulcers and stomatitis that tend to be multiple in number. Lesions are not seen at outer parts of the lip. Oral ulcers that recur three times maximum in a year are important for diagnosis. They generally heal on their own in 1-3 weeks [6]. Our case had oral aphthae that occurred 4-5 times during the last one year. The genital ulcers appear to be the second frequent manifestation in the disease's cases. Ulcers are similar to oral aphthae in appearance and are generally painful. Genital ulcers are 90% in scrotum in males and in vulva in females [7]. Lesions on the penis are rarely encountered during the course of BD. Although our case did not mention about his genital lesion in his initial anamnesis, considering his other findings, genital examination was insisted, and a painful lesion covered with crust at tip of the penis, which was 4-5 cm in size and extended towards glans penis was observed. Our case genital examination was insisted, and a painful lesion covered with crust at tip of the penis was observed. There is no diagnostic laboratory test for Behcet's disease; therefore, diagnosis is made upon clinical findings. We diagnosed our case according to criteria of international Behcet's study group [2]. He had palpable purpuric rash resembling HSP and the histologic examination of the skin lesion confirmed cutaneous leukocytoclastic vasculitis with IgA-containing immune deposits. Henoch-Schönlein purpura is a systemic small vasculitis involving the skin, kidney, joints and gastrointestinal tract. There are limited papers reporting the coexistence of BD and HSP [8]. Given the rarity of Behçet's associated HSP, this coexistence can be explained by chance. However, since cytokines are important elements in the pathogenetic process of both of these diseases, an upregulation in the genes of certain cytokines or receptors which because enhanced inflammation may be an alternative explanation. Treatment of the disease is generally empirical and targets suppression of inflammation. Our patient had his acute phase reactants normalized and his clinical findings improved during follow-up. Treatment of the disease is generally empirical and targets suppression of inflammation. Treatment depends on findings and severity of organ involvement. In our case, prednisolone and colchicine were initiated and dose adjustments were made according to findings during follow up. Our patient had his acute phase reactants normalized and his clinical findings improved during follow-up.

Conclusion

Although Behcet's disease is rare in childhood, it should be considered in differential diagnosis of children who have recurrent aphthous lesions. Since adolescents may be shy to mention about their genital ulcers during anamnesis, all systems particularly genital system should be examined carefully. In addition, HSP should be kept in mind in patients with Behçet's disease who have palpable purpura.

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