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Case Report

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ATYPICAL CUTANEOUS LESIONS IN BEHCET'S DISEASE: A CLINICO-PATHOLOGICAL CORRELATION

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ABSTRACT

A variety of cutaneous lesions may occur in Behçet's disease (BD) both at the onset and over the course of the disease. Erythema nodosum (EN) (15-78%) and Papulopustular lesions (28-96%) are the most commonly observed cutaneous lesions. We describe a case of 17 year old male with previously undiagnosed Behcet disease who presented to OPD with multiple recurrent eroded papulonodular lesions on acral areas with history of positive pathergy phenomenon. Histopathology manifestation from the lesion was suggestive of Behcet's disease. The patient was treated with oral corticosteroids and dapsone which showed a tremendous response with healing of all the lesions. To the best of our knowledge such atypical lesions of Behcet's disease have not been reported in literature so far. We suggest eroded papulonodular lesions on acral areas as observed in our patient may be included in the spectrum of the cutaneous manifestations of Behçet's disease.

KEYWORDS: Behcet, acral papulonodular.

INTRODUCTION

Behcet's disease (BD) is a chronic, recurrent, multi-system inflammatory disorder of unknown aetiology, characterised by the triad of oral ulcers (OU), genital ulcers (GU) and cutaneous lesions.[1] Skin involvement occurs in about 38-99% of BD.[2] The cutaneous lesions of the disease vary and mainly include erythema nodosum-like lesions, Papulopustular lesions (PPL), superficial thrombophlebitis, extragenital ulceration, reactivity of the skin to needle prick or injection (pathergy reaction) and other cutaneous vasculitic lesions.[2] We describe a patient with acral eroded papulonodular lesions not yet reported in literature so far.

CASE REPORT

A 17 year old male presented with chief complaint of recurrent multiple red raised painful lesions over dorsal aspect of fingers, palms and forearm for 6 months. There was history of 2 episodes of oral ulcers in past 2 months. There was history of on and off fever and pain in right ankle and knee joint for past 1 month. History of pathergy phenomenon in form of development of similar lesions at site of injection was present. There was no history of any genital ulceration, pain, redness or discharge from eyes, photophobia, gastrointestinal symptoms, neurological symptoms, or any other systemic complaints at the time of presentation. He took some injectables of unknown nature as advised by local physician. Cutaneous lesions subsided after taking injections but recurred again after stopping the treatment. On cutaneous examination multiple eroded papules and nodules of size varying from 0.1-1cm were present on dorsal aspect of distal phalanx of right thumb and right ring finger, Metacarpophalangeal joint of right index finger, middle phalanx of left ring finger, palmar aspect of left little finger and volar aspect of left forearm (fig 1). On basis of history and cutaneous findings differential diagnosis of Sweet syndrome, Behcet's disease and Erythema elevatum diutinum was kept. Pathergy test was performed with a disposable 26-gauge needle prick (needle held for 90 s in the dermis) at the flexor aspect of left forearm, approximately 2 inches below the elbow crease and read at 48 h which showed 1 mm papule. Laboratory investigations revealed raised erythrocyte sedimentation rate (28 mm/1st h) and C-reactive protein. Complete hemogram, liver and renal function test, random blood sugar, urine microscopy and stool for occult blood were normal. ELISA for HIV was nonreactive. On histopathology epidermis shows focal hyperkeratosis, parakeratosis with presence of neutrophils in keratin layers with formation of crust along with irregular acanthosis and spongiosis. Dermis show perivascular mononuclear infiltrate comprising predominantly lymphocyte admixed with few neutrophils at places. The infiltrate is seen extending into the vessel wall with exocytosis suggestive of Behcet's disease (fig 2,3,4). On basis of history of oral apthosis, cutaneous lesions and positive pathergy a diagnosis of Behcet's disease was made as per Revised international criteria for Behcet's disease.[3] The patient was treated with oral corticosteroids and dapsone which showed a tremendous response with healing of all the lesions in 2 weeks.

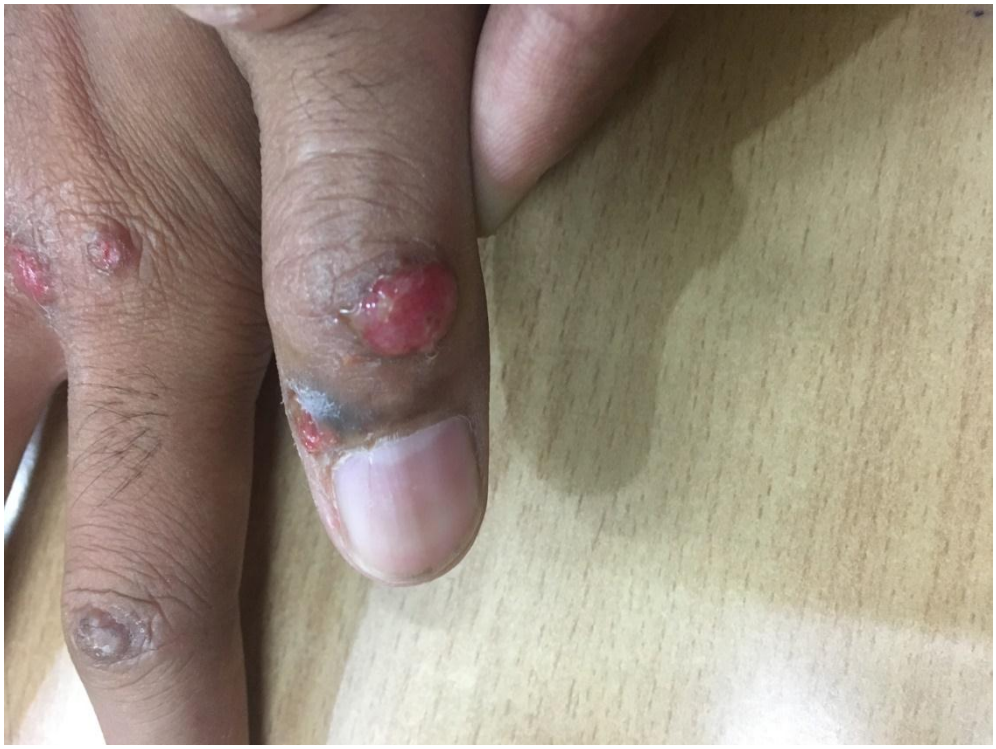

Figure 1: Multiple eroded papulonodular lesion on dorsal aspect of distal phalanx of right thumb and metacarpophangeal joint of right index finger.

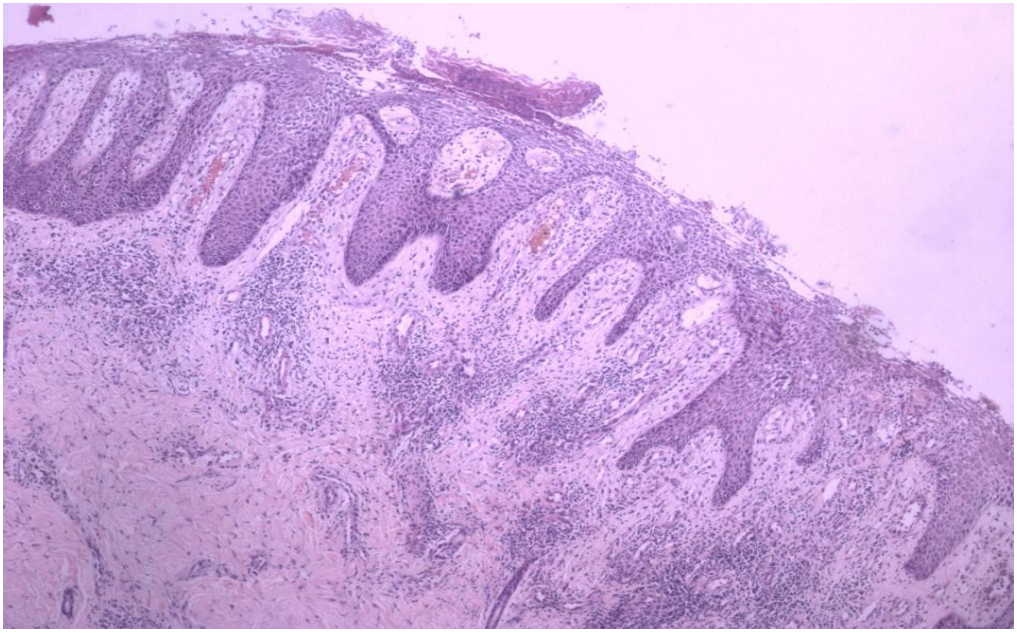

Figure 2: Histopathology (4x magnification) showing focal hyperkeratosis, parakeratosis irregular acanthosis and spongiosis in epidermis. Dermis show perivascular mononuclear infiltrate comprising predominantly lymphocyte admixed with few neutrophils. The infiltrate is seen extending into the vessel wall with exocytosis.

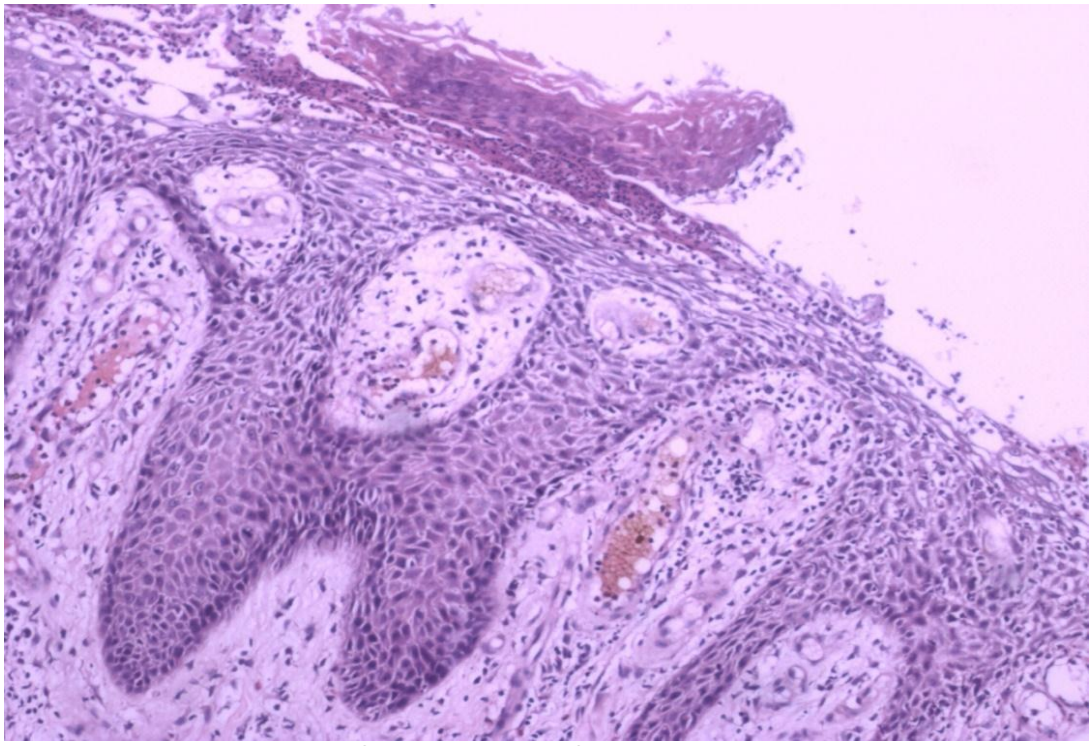

Figure 3: Histopathology (10X magnification) showing focal hyperkeratosis, parakeratosis with presence of neutrophils in keratin layers with formation of crust along with irregular acanthosis and spongiosis in epidermis.

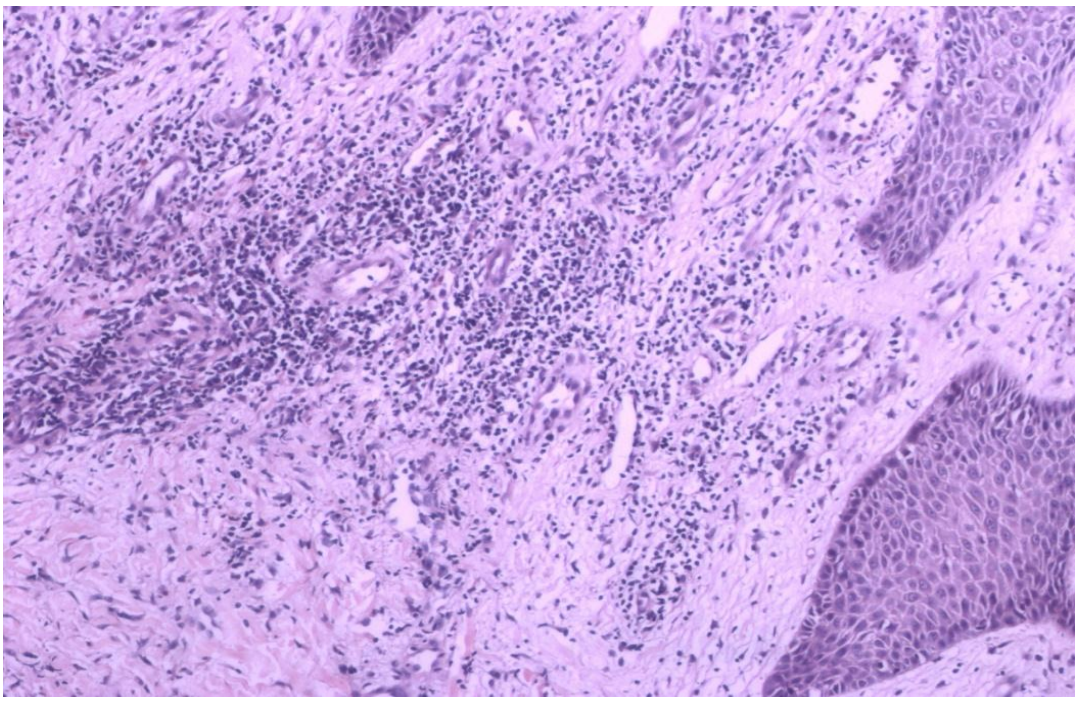

Figure 4: Histopathology(10X magnification) showing perivascular mononuclear infiltrate comprising predominantly lymphocyte admixed with few neutrophils. Infiltrate seen extending into the vessel wall with exocytosis.

DISCUSSION

Behçet's disease (BD) is a chronic, relapsing, multi-systemic, inflammatory process with the clinical features of mucocutaneous lesions and ocular, vascular, articular, gastrointestinal, neurologic, urogenital, pulmonary and cardiac involvement.[4] BD has been regarded as a Th1 type autoimmune disease, because of the association with human leukocyte antigen-B51 and hyperreactivity against streptococcal antigen.[5] Oral Ulcers (92-100%), Genital Ulcer (57-93%), cutaneous lesions (38-99%) and ocular lesions (29-100%), together with arthropathy (16-

84%) are the most frequent clinical features of the disease.

Usually Oral ulcers are the presenting feature of Behcet disease in the majority of the patients worldwide (47-86%) but in our patient cutaneous lesions were present as onset lesions which is quite rare.[4] The cutaneous lesions of the disease vary and mainly include erythema nodosum-like lesions, Papulopustular lesions, superficial thrombophlebitis, extragenital ulceration, reactivity of the skin to needle prick or injection (pathergy reaction) and other cutaneous vasculitic lesions.[4]Other cutaneous vasculitic lesions include Sweet's syndrome-like, pyoderma gangrenosum-like, erythema multiforme-like lesions, polyarteritis –like lesions, palpable purpura, subungual infarctions, hemorrhagic bullae, furuncles and abscesses.[4,6] Cantini et al and king et al have reported pernio like cutaneous lesions and purpuric papulonodular lesions on acral areas respectively.[7,8] In our patient eroded papulonodular lesions were present on acral areas which have not been described in literature so far. The eroded papulonodular lesions seen in our patient may be attributed to vasculitic changes which were also reported in the histopathology of the lesions. We suggest that eroded papulonodular lesions on acral areas as observed in our patient may be included in the spectrum of the cutaneous manifestations of Behçet's disease.

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