



Dermoscopy of skin manifestations of behçet's disease: A first description

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Abstract

Behçet's disease is a rare vasculitic disorder and a multisystemic disease with variable systemic effects. The disease appears to involve an autoimmune response triggered by exposure to an infectious agent. Clinical criteria are; a positive result of the pathergie test, bipolar aphthoses, eye damage. Cutaneous manifestations can occur in up to 75% of patients with Behçet disease and can range from acneiform lesions, to nodules and erythema nodosum. Ocular disease has the greatest morbidity, followed by vascular disease generally from active vasculitis. Our objective is to expose the dermoscopic aspect of the skin manifestations of Behçet's disease

Keywords: behçet disease, dermoscopy, skin manifestations

Introduction

Behçet's disease is a multi systemic disease with variable systemic effects. Clinical criteria are; a positive result of the pathergie test, bipolar aphthoses, eye damage. The objective of our summary is to expose the dermoscopic aspect of the skin manifestations of Behçet's disease.

Observations

We report four patients with a history of retained Behçet disease. The average age of our sample was 20.5 years with a female sex ratio m/F=0.33, which had clinically cutaneous ulcerations, on average the number of ulcers was 3 with multiple locations trunk, legs, pubis, of varying sizes ranging from a few millimeters to 8cm for the largest, the ulcers were well limited in irregular contours sometimes to a budding depth with clean surface, sometimes topped with bleeding scabs. Dermoscopic examination of different skin lesions revealed similarities in the different patients in our sample. Central erosion was noted surrounded by a circle with a pinkish-white background with a white radiant halo, linear and central hairpin vessels, and glomerular vessels at the periphery forming an overall star or target image.

Discussion

Behçet's disease is a systemic vasculitis characterized by episodes of acute inflammation, which can affect almost all vascularized organs of the body including the skin 1. Mucocutaneous manifestations are markers of BD and their recognition may

allow diagnosis and treatment 2. The earlier the onset of manifestations, the worse the prognosis, with consequent increase in morbidity and mortality 2. Skin lesions are not specific to BD, but essential for diagnosis. The skin manifestations are; papulopustular lesions, knotty erythema, thrombophlebitis and various cutaneous and vasculitic lesions. Cutaneous ulcers affect up to 3% of patients, are recurrent, and leave scars 3. According to some authors, they are the most specific cutaneous manifestation of BD 4.

Dermoscopy of skin lesions of Behçet's disease has never been reported in the literature except for the one describing the papule lesion after the pathergie test, which is similar to those of our patients. Pathergy is defined as an altered state of the tissue response to a minimal needle puncture trauma, reflecting an exacerbated response of the innate immune system 5. Pathergy lesions are clinically manifested as erythematous papules, sometimes topped by a sterile pustule. Histopathological examination reveals inflammatory infiltration with predominance of mononuclear cells around the dermal vessels and an increase in the number of mast cells. A neutrophilic vascular reaction may also be present, resulting in leukocytoclastic vasculitis, suggesting that this infiltration is responsible for the dermoscopic image. It is suggested that central erosion corresponds to fibrinoid necrosis secondary to intravascular thrombus, hairpin vascularization to superficial and deep infiltration of perivascular inflammatory cells including lymphocytes and neutrophils and glomerular vascularization to reflection achieved by swelling of endothelial cells.



A: Erythematous symmetrical macules centered by a bilateral crust and symmetrical at the two major ones.
B: An ulceration of 8 cm from the major axis to the anterior face of the leg.
C: Two slightly infiltrated erythematous plaques with eroded surface surmounted by haemorrhagic crust on the outside of the right leg.

Fig 1: Clinical appearance

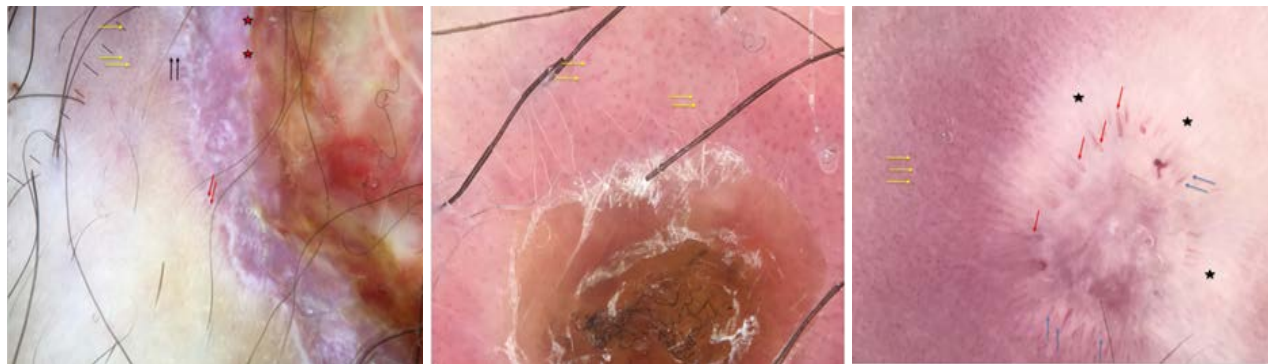


Fig 2: Dermoscopic appearance: Two different dermoscopic patterns were found; early lesions were mainly typified by a pink-white background with a white radiant halo, linear and hairpin vessels around the edges, surrounded in turn by glomerular vessels at the periphery. Appearance in a star in a rosette. In late stage lesions; ulceration, had a same appearance with linear and hairpin vessels surrounded by glomerular vessels at the periphery but with a central ulceration

Conclusion

In conclusion, reports using dermato scope in the Behçet's disease were not found. The dermoscopic study of skin lesions of Behçet's disease revealed signs that can be qualified as specific to the disease. Studies on richer series with anatomopathological and dermoscopy confrontation are necessary.

References

1. Behçet's disease: review with emphasis on dermatological aspects.
2. Zeidan MJ, Saadoun D, Garrido M, Klatzmann D, Six A, Cacoub P, *et al.* Behçet's disease physiopathology: A contemporary review. *Auto Immun Highlights.* 2016; 7:4.
3. Maria Antonieta Rios Scherrer, Vanessa Barreto Rocha. Lucas Campos Garcia Behçet's disease: review with emphasis on dermatological aspects, 2017, 7359.
4. Azizlerli G, Ozarmağan G, Ovül C, Sarica R, Mustafa SO. A new kind of skin lesion in Behçet's disease: extragenital ulcerations. *Acta Derm Venereol.* 1992; 72:286.
5. Varol A, Seifert O, Anderson CD. The skin pathergy test: innately useful? *Arch Dermatol Res.* 2010; 302:155-68.