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Behçet's disease in black skin. A retrospective study of 50 cases in Dakar.

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Abstract

Introduction: Although Behçet's disease is well-documented in Eastern populations, epidemiologic data in Sub-Saharan African population is scarce. The aim of this study was to define the epidemiologic and therapeutic aspects as well as clinical course of Behçet's disease in African black population.

Results: The study included 50 patients with Behçet's disease. The average age was 32 (18-67) years. A total of 31 patients were men and 19 were women. Two patients had a positive family history of Behçet's disease. The oral and genital aphthous lesions were present in 100% of patients. The pathergy test was positive in 16 patients (32%). Following skin conditions were observed: pseudofolliculitis in 15 patients (30%), acneiform papules in 6 patients (12%), erythema nodosum in 4 patients (8%) and leg ulcers in one patient. Ocular involvement was reported in 22 patients (44%) and joint involvement in 20 patients (40%). Neurological abnormalities were noted in 12 patients (24%). Gastrointestinal involvement with wide and deep ulcerations in the ileocecal region was observed in a patient. As treatment, a combination of oral corticosteroids and colchicine was used in 97% of our patients. Thalidomide was introduced in 3 patients and anticoagulation treatment in 19 patients. Clinical improvement was noted in 25 patients (50%), recurrence in 14 patients (28%) and 3 patients were lost to follow (6%).

Conclusion: The Behçet's disease is not uncommon in black skin and generally affects young adults. Severe aphthous ulcers of the oral cavity and genital area are the most consistent finding. (*J Dermatol Case Rep*. 2015; 9(4): 98-102)

Introduction

Behçet's disease is a multisystem, relapsing, vasculitis with mainly venous involvement. Recurrent chronic oral ulcers, ocular inflammation, genital ulcers, and skin lesions are the common manifestations.[1] Although its mortality is relatively low, it is a public health problem as eye evolvement can lead to blindness. A prompt diagnosis and treatment is the main challenge to reduce its morbidity and mortality. Given the frequency of dermatologic manifestations the place of dermatologist in the diagnosis and management is important. In Sub-Saharan Africa the Behçet's disease related data are scarce. The objectives of our study were to determine the epidemiological, clinical, therapeutic and outcome of the Behçet's disease to the dermatological clinic of the Aristide Le Dantec Hospital.

Patients and Method

This is a retrospective study of a 13-year period (2000-2013), performed at the Dermatology Department of Aristide Le Dantec Hospital, the reference center of dermatological diseases in Senegal. The medical records of all patients

with a diagnosis of Behçet's disease were collected. In all cases the diagnosis was based on the International Study Group for Behcet's disease's criteria[2] with the mandatory presence of recurrent mouth ulcers and at least two of other Behcet's disease distinctive symptoms including: recurrent genital ulcers, skin lesions, ophthalmic lesions and positive pathergy test.

Figure 1
Genital aphthous ulcers.

Figure 2
Aphthous ulcer of the tongue.

Results

All 50 Behçet's disease patients diagnosed in last 13 years were included. They were of African descent and Fitzpatrick phototype VI. The yearly incidence was 3.84. The mean age was 32 years, ranging from 18 years to 67 years. The sex ratio was 1.6 : 1 with 31 men and 19 women. A total of 23% of the patients had received antibiotics, 12%, oral corticosteroids, 9%, antinflammatory drugs, 6% colchicine and 8% oral herbal medicine before consulting in our department. The mean delayed time to consultation was 18 months. Smoking and alcohol abuse were found in 12.5% and 6.25% respectively. Two patients had a positive family history of Behçet's disease in their first degree relatives. Clinically, oral and genital ulcers was a constant finding (100%) with average of 3 recurrences per year (3-12 recurrences / year). Pathergy test was positive in 16 patients (32%), pseudofolliculitis was present in 15 patients (30%), acneiform papules in 6 patients (12%), erythema nodosum in 4 patients (8%) and leg ulcer in one (2%) patient (Fig. 1—5). Extracutaneous manifestations were dominated by ocular involvement in 22 patients (44%) (Table I), 20 patients showed joint involvement (40%) in the form of arthritis in 17 cases and monoarthritis

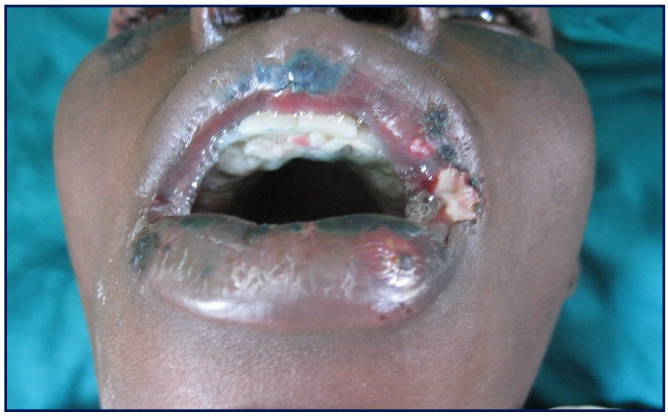

Figure 3
Oral aphthous ulcers.

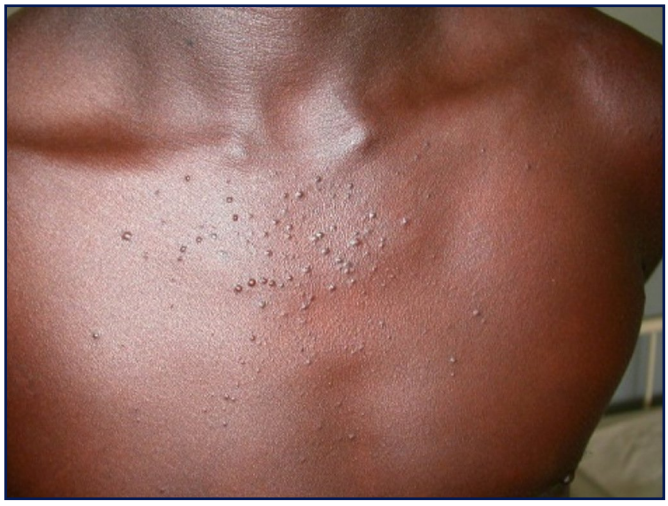

Figure 4
Acneiform papules.

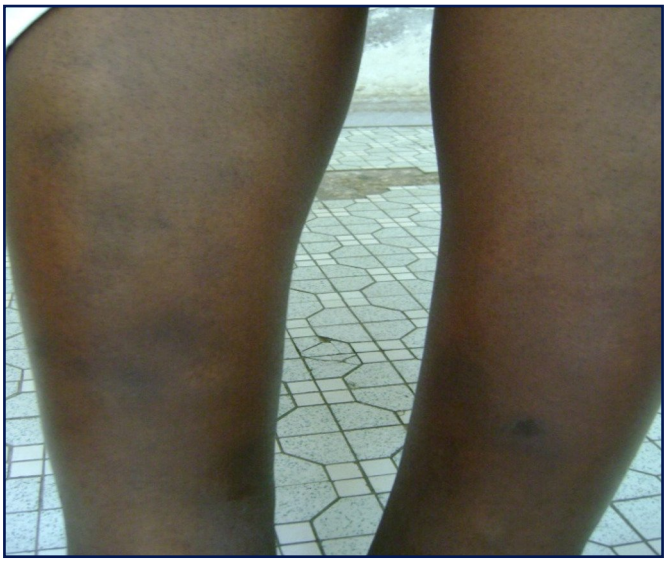

Figure 5
Erythema nodosum.

in 3 cases. Neurological abnormalities were present in 12 patients (24%) with most the common manifestation being severe and treatment refractory headache in 8 patients, and intracranial hypertension, seizure one and polyneuropathy one case. Vascular disorder in 9 patients (18%) mainly consisted of venous thrombosis. Intestinal involvement was found in patient with colonoscopy consisted of wide deep ulcers in ileocecal region. Cutaneous histopathology performed in 3 patients and revealed vasculitis with complements deposits and inflammatory infiltrates. A number of 16 patients showed anemia, among them 10 patients had hypochromic microcytic and 6 patients normocytic normochromic anemia. In treatment, a combination of oral corticosteroids and colchicine was used in 97% of our patients. Thalidomide was prescribed in 3 patients. Anticoagulant therapy as antiplatelet agent was prescribed in 10 patients and low molecular weight heparin in 9 patients. Local treatments included antiseptic and local anesthetics. Clinical improvement was noted in 25 patients (50%), recurrences in 14 patients (28%) and we had 3 cases of lost to follow (6%). As complication blindness was noted in 7 patients (14%), infectious and gastrointestinal abnormalities in 6 patients (12%). No death was reported.

Discussion

This study shows that Behçet's disease is not a rare condition in Senegalese black population. In Sub-Saharan Africa, data on Behçet's disease is scarce. In North America, the prevalence of Behçet's disease was recently estimated at 5.2 cases / 100,000 people by the Olmstead committee in Minnesota.[3] A recent study in Martinique estimated a prevalence of Behçet's disease to 7 / 100,000.[4] The highest prevalence is observed in Turkey ranging from 19.6 to 420 / 100,000.[5,6,7,8]

In our series, this hospital prevalence is fairly representative as many patients consult in other department such as internal medicine depending on the primary manifestation of their disease.

Behçet's disease is more common in young adults with a peak incidence between 20 and 30 years. It is very rare in ages of older than 60 years. In children few cases have been reported. In our series, the mean was 32, similar to other series reported in literature.[5,6] The average age of 27.5 years was reported in series by Hamza[9] and 30 reported by Benamour.[10]

Our sex ratio of 1.63 which shows a male predominance what confirms the results of many other studies.[7,8,11] However, in Japanese as well as Anglo-Saxon series a female predominance is frequently reported.[12]

Most of our patients initially received an inappropriate treatment due to misdiagnosis, a consequence of the lack of knowledge on Behçet's disease among the general practitioners. A proportion of 8% of our patients had used an oral herbal medicine what can be explained by socio-cultural factors and availability of traditional healers in Senegalese community. A 18-month delay before diagnosis was noted in most African series.[13] Although some of our patients consulted earlier because of the severity of oral-genital ulceration disturbing feeding and sexual activity.

A recent population-based study in the Paris metropolitan area on North African and Asian ancestry population emphasized the importance of genetic factors in Behcet's disease.[14] However we found only two familial cases. The frequency of the familial form varies between 2 and 18% depending on the population.[14] Familial forms seem to be more severe than sporadic and they are strongly associated with HLA B51. Familial occurrence is more common among Korean (15.4%) than Chinese and Japanese (2.2-2.6%) as well as in Arab, Israeli or Turkish populations (2-18.2%), which are more than in European origin patients (4.5%).[15]

Table I: Ophthalmologic manifestations in patients with Behcet's diseases.

Ophthalmologic manifestations		Number of cases
Uveitis	Anterior	8/50 (16%)
	Posterior	3/50 (6%)
	Intermediate	3/50 (6%)
	Panuveitis	4/50 (8%)
Retinal vasculitis		6/50 (12%)
Papilledema		1/50 (2%)

Table II: Frequency of aphthous ulcers by series.

Mucosal lesions	Our series	Hamzaoui [16]	Benamour [10]
Oral aphtous ulcers	100%	100%	94%
Genital aphtous ulcers	96%	87,5%	70%

Clinically, all of our patients showed oral aphthous ulcers, almost always associated with genital ulcers except one case who presented oral ulceration without genital ulcer. She was special regarding the disease severity as she had an average of 7 recurrences per year. These results are consistent with most of the series (Table II) which show a frequency of 90-100% for oral aphthous ulcer[16,10] presenting at the beginning and in all stages of the disease. One feature of the Behçet's disease in the tropical regions is the clinical resemblance of its oral and genital sores to the ulcers due to sexually transmitted infections such as chancroid, lymphogranuloma venereum and herpes simplex virus. It could partly explain the diagnosis delay observed in some patients. Exaggerated reaction in puncture sites was observed in 80% of cases in the Benamour's series[10] contrary to Anglo-Saxon series where it is rarely found. In our series, the pathergy test performed in 30 patients and was positive in 33% of patients. It was less frequently positive compared to 80% reported in the literature.[10]

Pseudofolliculitis reported in 31% of our series less than 56% what was observed by study Filali-Ansari.[17] They had ubiquitous locations with a preference on the back, the legs and the buttocks. Erythema nodosum which is more commonly reported in Asian studies was found only in 4% of our patients. The lesions were most commonly on the legs and occasionally in other body parts including the face.[18] Acneiform papules were noted in 8% of patients, which is close to the results of a Tunisian series by Hamzaoui with a frequency of 7.9%.[16]

Ocular manifestations, with over 40% were the most comment extra-cutaneous abnormality in our patients. The frequency of ocular involvement is variously assessed in different studies depending on the inclusion criteria: it ranges from 29% to 100%.[9,10] Uveitis was the most common ocular manifestations and found in 18 cases. In Japan Behcet's disease is responsible of 13 to 15% of endogenous causes of uveitis. In France, it is the cause of 2.4% of uveitis according Bodaghi[19] and 1.7% according to Nguyen.[20]

According to a study in Monastir University Hospital by Khairallah,[21] Behcet's disease is responsible for 13.5% and 34.6% of all posterior uveitis, and panuveitis respectively. The joint involvement is the second most important extra-cutaneous manifestations, reported in 42% of our patients. The frequency of joint involvement is comparable to other series with a range between 45-70%.[22,21,23,24] The neurological manifestations of Behçet's disease are highly variable. They were present in 25% of patients and dominated by chronic headaches. It is the most frequent neurological manifestation of Behçet's disease. It is reported in 62.2% of patients in a series by Saips *et al*.[25] A British study[26] reported a higher prevalence of 83.3%. Vascular involvement was noted in 8 (17%) of our patients. The incidence of vascular disease varies from one population to another ranging from 6-38% depending on the series.[9,16,27,28 29,30] Deep vein thrombosis is the most common, as it is documented by Tunisian, French and Moroccan studies (31%, 35% and 36% respectively).[28,29,30] Hamzaoui[16] reported a dissemination of venous thrombosis to all venous territories which confirm the ubiquitous nature and venous tropism of the Behçet's disease. Regarding treatment, our series showed a good response to treatment with a recovery rate of over 50%. Janati[31] and Cochereau-Massins[32] reported a frequency of blindness of 12% and 16% respectively, similar to our findings.

Conclusion

Behçet's disease is not rare in black population and mainly affects young adults. Recurrent oral and genital aphthous ulcers are the main symptoms of Behçet's disease. Because of the variety of dermatological manifestations, a good knowledge of this condition by dermatologist is mandatory to prevent visceral and ocular involvement.

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