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Recurrent Oral and Scrotal Inflammatory Lesions in A Young Man: First Case Report of Behcet's Disease in Gambia

Asmell Ramos Cabrera[1], Yanet Correa Lopez[2], Hector Oscar Victoria Barzaga[3], Lamin Drammeh[4]

[1] HOD Internal Medicine Department, American International University West Africa

[2] Mariana Grajales Faculty of Health Sciences.

[3] Carlos J Finlay Faculty of Health Sciences.

[4] Researcher of MRC. London school of hygiene and tropical medicine

ABSTRACT

Behcet's disease (BD) is a rare condition, considered as a variable vessel vasculitis with multi-system involvement, characterized by the triad of recurrent mouth and genital ulcers with eye involvement, also may involve joints, skin, central nervous system, and gastrointestinal tract with important heterogeneity among patients regarding demographic features and organ manifestations. BD has a worldwide distribution. However, it is observed commonly among populations living along the historic Silk Road.The etiology of the disease remains unknown, but the most widely held hypothesis of disease pathogenesis is that of a profound inflammatory response triggered by an infectious agent in a genetically susceptible host. Number of reported cases is inexplicably low in sub-Saharan region and especially lowest in West Africa where only few cases have been reported. The aim of this presentation is to announce the first case description of BD in Gambia as well as, to sensitize physicians, Internal Medicine residents and general practitioners in our sub region with the pattern of the disease, as only way to reduce the diagnosis time frame.

KEYWORDS: Behcet's disease, canker sore, scrotal ulcer. The Gambia.

ARTICLE DETAILS

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1. INTRODUCTION

Behçet's disease (BD) is a multisystem disorder named after the Turkish dermatologist, Hulusi Behcet (1889–1948), who recognized and reported in 1937 a triad of symptoms consisting of recurrent eye inflammation, oral ulcers, and genital ulcers. [1-3] The disease had also been alluded to in descriptions as far back as Hippocrates. [3] BD is particularly common in the Far East and the Mediterranean basin, and is frequently noted between the 30th and 45th degree latitudes in Asian and European populations.[3] corresponding to the Old Silk Road, an ancient trading route stretching between the Mediterranean, the Middle East and the Far East. In contrast, this disorder is uncommon in the American continents, Oceania, and sub-Saharan Africa. [3-5]. in sub-Saharan Africa, the prevalence of BD is not known. Few cases have been reported elsewhere [6–8]. Reports in black African migrants done in London and Paris showed that BD rarely encountered among individuals of central and West African descent. [9, 10]

CASE PRESENTATION

MJ, Male, aged 42 years, during the last 5 years has been in prison, being frequently referred to the hospital for medical attention due to the presence of recurrent mouth ulcers Fig [1],

typically painful and associated with difficulty eating, also has been suffering painless genital ulcers, on the scrotum, multiple in number, tender with no discharge and without scares Fig [2], additionally regular episodes of arthritis, this one currently affecting the right knee, it is swollen, hot, painful to touch and to movement, associated with difficulty walking Fig [3], others complains are mild abdominal pain, located in the suprapubic region and vomits but no diarrhea.

System Review: Intermittent headache and low-grade fever last 4 days. No history of drug intake. During the last two years has been admitted on several occasions on account of similar symptoms with small variations in the severity, with remission after antibiotics, NSAID and steroids without major problems; during all hospitalizations always reported as chief complaints

oral and genital ulcers. Six times HIV and VDRL tests were requested all of them negatives.

Fig 1. Mouth Ulcers

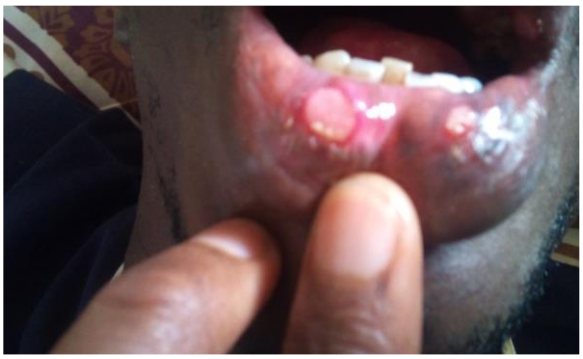
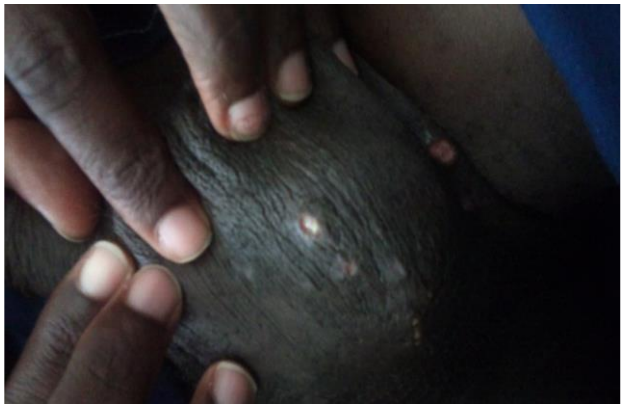
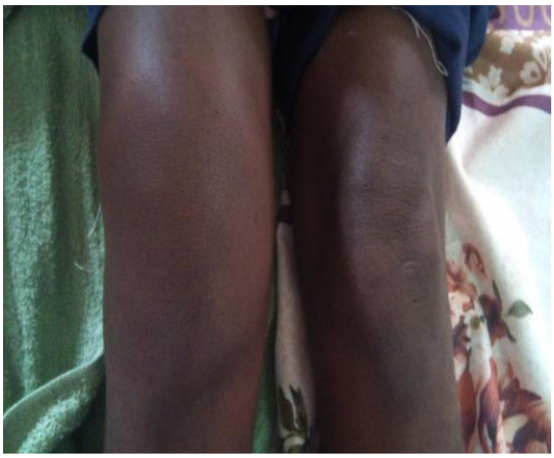


Fig 2. Scrotal ulcers



Right knee swollen Fig 3.



Past medical history no relevant information about previous illness like rheumatoid arthritis, rheumatic fever, or Gout. No STD reported. Nevertheless, recurrent episodes of axial pain and major joints inflammation commented during prior admissions. No drugs history, several events of ¨allergic¨ to intramuscular injection reported.

On examination:

General-not pale, a cyanosed, anicteric, no finger clubbing or oedema, febrile to touch (38.30 C). Cardiovascular system: pulse 100 beats/min, BP 106/77mmhg, s1 and s2 heard, normal, no murmurs, Respiratory system: RR 22cycles/min, vesicular breath sounds heard. Abdomen: full, moves with respiration, mild tenderness in the suprapubic region, no rebound tenderness, no guarding, no palpable organomegaly. Central nervous system: conscious and oriented, no neurological deficit. Anterior uveitis in the left eye.

Skin and mucous exam showed multiple oral ulcerations under the tongue, one of them is well circumscribed lesion of 2 cm, another on the lower lips measuring 4cm, tender, and no active discharge. Similar ulcerations on the scrotums, multiple in number measuring 1- 2cm, mild tenderness, no active discharge. At the right gluteal area, scar as a result of "penicillin allergy" is seen. Pseudo folliculitis barbae.

Musculoskeletal: swollen right knee, tender, fluctuant, overlying skin is normal, warm to touch, with mild limitation of extension and flexion movements.

CNS: No focal motor or sensory disorder, left eye anterior uveitis (iridocyclitis) GCS: 15/15

Investigations:

- Chest x ray: Normal
- X ray of the Rt knee: Normal
- Pelvic and Hips X ray: Normal
- Hb=14.3 g/ L
- WCC: 11.5 x 109 l
- RDT: Malaria negative
- VDRL: Non-reactive
- HIV: Negative
- Hepatitis B and C: Negative
- Urinalysis:
  - ✓ Protein 1+
  - ✓ PH= 9
  - ✓ SG=1.020
- ESR: 85 Mm/H
- CRP: 40 Mg/L
- Urea creatinine and electrolytes: Normal
- Urinary Culture: Negative
- Blood Culture: Negative
- Pathergy test: It was performed using 2 subcutaneous punctures with a blunt sterile needle to one arm and two subcutaneous punctures with pointed needle into the other arm simultaneously. It was read 48 hours later, and the result showed a sterile erythematous papule of more than 4 mm. [11,12]

1.
2. **2. DISCUSSION**

The diagnostic approach to a patient with multiple clinical problems has always been a challenge demanding a holistic approach, multi-disciplinary team, a variety of numerous, complex, and expensive studies.In general, individuals with Behcet's disease frequently report low self-esteem, negative body image, anxiety, fear and tension, the chronic and debilitating nature of the condition makes it imperative that clinicians improve in the entire spectrum from diagnosis to treatment; considering the plethora of probable diagnoses, such as sexually transmitted illnesses, mucocutaneus parasitosis,

systemic vacuities, inflammatory bowel diseases or rheumatological sicknesses and even cancer. [2,4,6,13,14] Bearing in mind this wild spectrum, we proceeded to carry out the differential diagnosis based on: Clinical pattern of symptoms and signs, age, sex, race, ethnicity, geographic zone, and laboratory/ Radiological studies. The case in question, association between recurrent oral ulcers and genital ulcers are so strong that the recommendation is simple "Everyone who has mouth sores three times within a 12-month period should be consider as potentially BD patient, additionally anterior uveitis plus non-erosive arthritis of major joints plus history of ¨skin allergic reaction¨ after intramuscular injection remained BD until proven otherwise. Oral ulcers are the hallmark manifestation," [14-19] and lead to the diagnosis approach, nevertheless the biggest challenge is not to perform diagnosis per se, but on the contrary, the main problem to face lies in excluding a varied group of entities that may at some point present a similar clinical pattern. We consider that due to his stay in prison as a young and sexually active person, they contributed the medical personnel who previously assessed him considered the diagnostic stereotype, involuntarily causing a delay in diagnosis, affecting his quality of life and especially his affective sphere, leading to continuous cycles of depression during the last 2 years prior to the BD diagnosis being made. Six negative tests for HIV, Syphilis and Hepatitis B it made imperative to consider the probability of a systemic disease, patient with ambiguous symptoms which encompasses multisystem involvement is a daily challenge for internist and requires a detailed interview, deep medical examination, and appropriate use of clinical method and further engagement of a multidisciplinary team to ensure holistic approach with the aim of trying to explain the plethora of symptoms and signs just only through a single disease. The usually mono-articular involvement with non-erosive arthritis and taking the axial axis, with negative rheumatoid factor, could include spondyloarthropathies as part of the differential diagnosis, radiography of the pelvis and lumbosacral spine were negative, and clinically did not satisfy the Rome and ASAS criteria, the absence of psoriasis, chronic diarrhea and STD, excluded psoriatic arthritis, that related to idiopathic ulcerative colitis and although crohn's disease can have canker sores and could be seen in BD patients, the absence of symptoms (except minor lower abdominal pain) of the lower and upper segment of the digestive tract ruled out the entity. Uveitis, it is not specific and can be found in diseases such as ankylosing spondylitis, Crohn's disease, Reiter's syndrome, Juvenile chronic arthritis, or Rheumatoid arthritis; all previously were ruled out before; however, HLA 27 study isn't available in Gambia and could be helpful if we consider the strong association between Behcet morbus and Axial Spondylarthrosis. [20-25]. additionally increased values of inflammatory markers like erythrocyte sedimentary rate and C-reactive protein in spite of the ambiguous specificity, confirmed an active inflammatory response. [26] Cards are on the table let's play! The presence of mild fever should prompt to ruled out infection, but as recurrent, self-limited condition, with spontaneous remission with or without antibiotic, furthermore repeated negative cultures for blood, sputum, and urine over the last two years, moreover, prolongation of the clinical picture over time (more than 2 years) has had supported to excluded infectious. Notwithstanding the chronicity, paradoxically the patient was looking in good shape also the impact in his hemoglobin value, total proteins count, liver and renal function had been nil and typically our patient use to recover well after few weeks without any sequelae. Therefore, cancer is clinically unlikely. Several conditions can present with oral aphthous ulcers [27-29], necessitating a thorough workup to narrow the differential diagnose. Physical examination should be used to screen for trauma secondary to dental appliances, widespread vesiculobullous eruptions, and signs of hormone imbalance, as well as to look for stigmata of systemic disease implication. Blood work up should be used to rule out hematologic, sepsis, nutritional deficiencies and autoimmune or hereditary conditions. The differential diagnosis for oral ulcerations includes several entities see table [1] below.

Table. 1

Drug Induce:
Fixed Drug eruption
Linear IgA bullous dermatosis
Drug-induced bullous pemphigoid
Drug-induced pemphigus
Stevens-Johnson syndrome and TEN
Autoimmune diseases:
Crohn's (orofacial granulomatosis)
Behcet's, Celiac, systemic lupus
Lichen planus
Linear IgA, Bullous dermatosis
Wegener's granulomatosis.
Trauma:
Dental appliances
Necrotizing sialo metaplasia.
Hematologic:
Anemia, neutropenia
hyper eosinophilic syndrome
Fever syndromes:
Cyclic neutropenia
PFAPA (periodic fever, aphthous stomatitis, pharyngitis, cervical adenitis)
Sweet syndrome
Familial Mediterranean fever
➢ hyperimmunoglobulinemia D Vesiculobullous disorders:
Pemphigus vulgaris
linear IgA disease.

However, most of the above entities are skin limited disorders, without involving eyes, joints, or CNS. Other conditions are associated to trauma, linked to toxics, medications and drugs related. Differentials diagnosis also include Blau syndrome,

Majeed's and PAPA syndrome all of them are rare disorders commonly seen during childhood usually before the age of 4, inducing osteomyelitis with erosive and mutilates arthritis, largely affects skin, joints, and eyes with granulomatous dermatitis. Familial Mediterranean fever (MEFV) is associated to fever, abdominal pain, chest pain, which can make it hard to breathe deeply, painful and swollen joints, usually in the knees, ankles and hips, arthritis, aphthous changes in lips and/or oral mucosa, erythematous rash on the legs, especially below knee and muscle aches, mainly affecting ethnic groups living at Mediterranean based and is characterized by recurrent, self-limited episodes of fever and serositis. [30,31] a monogenic auto inflammatory disease secondary to MEFV gene mutations in the chromosome 16p13. The diagnosis of Familial Mediterranean fever (FMF). The case in question was not fitting the Tel-Hashomer diagnosis criteria. table [2]

Table 2.

Tel-Hashomer diagnosis criteria
Major criteria
Recurrent episodes of fever accompanied by serosal inflammation (of the peritoneum, synovium or pleura)
Primary AA type amyloidosis
Good response to treatment with colchicine
Minor criteria
Recurrent episodes of fever
Erythema resembling that seen in erysipelas
Family history of FMF in a first-degree relative
Two or more major symptoms or one major plus two minor symptoms.

¨Gluteal skin lesion¨ was pointed in context, and Pathergy test requested, becoming positive after 48 hrs. Concerning the result, is more frequent in males than females and bears no relation to the severity of the disease in addition patients with pathergies are prone to have vascular involvement. Nonetheless, other diseases may induce pathergy such as: pyoderma gangrenosum, sweet's syndrome and it has been described in patients with chronic myeloid leukemia. [11,12] Being a man with involvement of large joints arthritis and pain of the axial skeleton with negative rheumatoid factor led us to a well-identified group of seronegative arthropathies. The absence of intestinal manifestations such as severe, chronic or recurrent abdominal pain or diarrhea clinically excluded inflammatory bowel disease.

Considering the recurrent association of ocular manifestations, scrotal ulcers, and allergy reactions at previous intramuscular injection sites, they lead us to consider behcet's disease as our working diagnosis. There are no pathognomonic laboratory tests to diagnose BD, and as such, the diagnosis is based on clinical criteria. Clinical manifestations of BD are heterogeneous and may involve virtually all organ systems. Not only the frequency but also the type of organ lesions seems to differ between regions. [32-34].These differences could be related to genetic and environmental influences. Our patient's symptoms and signs were not different of the literature reporting the typical triage of recurrent oral ulcer (85%), genital ulcer (4.6%), arthritis (1.7%) and uveitis (8.4%). in addition, back pain (1.7%). [3, 5, 7, 15, 18] Gastrointestinal and neurological symptoms tend to be more prevalent among patients in the U.S. and northern Europe than those in Istanbul and other areas in the Silk Road region. Mean age of the referred patients was 37.4 years in serial studies is similar to our patient 42 years. Average time from the onset of the first symptoms to the final diagnosis in the reviewed publications ranges between 2.5 and 5 years. The diagnosis was made after almost 2 years from the beginning of the symptoms as interesting element. Certainly, literature reviewed reported 55% female and family history also consider ethnicity as relevant element to support the diagnosis however what is relevant in our case report is that there isn't any connection with geographic or ethnicity associated to BD nevertheless reported around the world with or without genetic or familiar connection with ¨Silk Road geographic area. [36-38]. Though, related to age, sex and time of diagnosis is not uniform, the fact exists wide variations between findings reported in different latitudes and have been showing a lot of disparities between male and female patterns as well as the clinical pattern of joints, neurological or gastro-intestinal diseases. [7-10].The truth is that there is still much to understand in relation to the BD and in summary after multiple differential diagnosis, our case satisfied the- The International Clinical Criteria for Behcet's Disease. [39-42] classification Table [3]:

The International Clinical Criteria for Behcet's Disease. Classification states patients must present with:
Recurrent oral ulcerations (aphthous or herpetiform) at least three times in one year.
Additionally, patients must present any two of the following:
✓ Recurrent genital ulcerations
✓ Eye lesions (uveitis or retinal vasculitis) observed by an ophthalmologist
✓ Skin lesions (erythema nodosum, pseudo folliculitis, papulopustular lesions, acneiform nodules) found in adult patients not being treated with corticosteroids
✓ Positive "pathergy test" read by a physician within 24-48 hours of testing

CONCLUSIONS

The rare and complex Behcet's disease is often misdiagnosed or undiagnosed, leading to significant and unnecessary morbidity; as a result of is not a commonly known condition, a lot of physicians in many areas of medicine do not know about BD, oral and genital components can affect daily living, socializing and other associated quality of life parameters. [43] Clinicians

who are familiar with BD are concerned about its nature; because it is a very complex diagnosis that relies on the exclusion of a constellation of diseases. [44-46] In summary, Behcet's disease continues to be a challenge in daily clinical practice, especially for young practitioners who are not familiar with multisystem diseases, with a wide range of differential diagnoses and require an interdisciplinary approach. The problem is aggravated in daily practice in developing countries where the precariousness of public health institutions with chronic lack of diagnostic facilities and highly qualified human resources prevent or further delay the diagnosis of the disease with the consequent appearance of complications, either physical, psychosocial or both. It is in this context, and it is the objective of the presentation of this case, firstly, to remind the younger generations about EB and its peculiar symptomatic association and, secondly, and as importantly, to demonstrate the validity of the use of the clinical method in the approach rational to the patient, in public institutions with scarce technical and human resources where the internist specialist could develop the holistic vision that characterizes our specialty.

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