NEUROLOGICAL MANIFESTATIONS OF BEHCET'S DISEASE: A CASE REPORT

*Dawodu CO *Adeleye OA

*Department of medicine
Lagos State University College of Medicine [LASUCOM]
Ikeja, Lagos.

Correspondence to:
Dawodu CO

Email: brainattack2003@yahoo.com
Tel: +2348033522435

Abstract

Behcet's disease causes inflammation of blood vessels but it is of unknown aetiology. When it involves the nervous system, it may present with benign headaches, aseptic meningitis, meningoencephalitis, cranial nerve palsies, seizures, personality changes, and cerebral venous thrombosis.

We present the case of A.G is a 26-year old male, was a student in a Nigerian tertiary institution and resided in Lagos and Ibadan. He presented with a 2-year history of progressive and recurrent headaches, genital and mouth ulcers and weakness of the left limbs.

Physical examination revealed a right handed, conscious, but apathetic and abulic young adult. All meningeal signs were positive. He had bilateral 6^{th} nerve palsies, upper motor neuron (UMN) left 7^{th} nerve palsy and a left partial ptosis. He had a bulbar speech with a cerebellar dysarthric twist and a grade 3 spastic left hemiparesis. However, his sensory system was intact and his presenting vital signs were within normal range. His brain computed tomogram confirmed areas of multiple infarcts, and a number of negative tests ruled out other possible differential diagnosis. He was treated for Behcet's disease and had a dramatic positive response to antiplatelets and steroids.

He was adequately stabilized on an out-patient basis, when under aunt's care in Lagos. He, however, died following his relocation to Ibadan. An autopsy could not be done.

KEYWORDS: *Behcet's Disease, Neurological manifestations*

Introduction

Behcet's disease, also called Adamantiades is a disease of unknown aetiology.[1] It is common in the Middle East, Asia, and Japan were it has a male predilection.[2] Behcet's disease tends to develop in the second decade of life but the mean age at onset is 27 years in males and 24.5 years in females.[3] The causative factors/triggers are unknown. However familial occurrence has been reported particularly in the Middle East.[4] Evidence also points to HLA-B51 positivity being associated with a worse prognosis.[5] Infectious agents, such as herpes simplex virus type I, parvovirus B19, hepatitis C, and streptococcus species may also be associations.[6] Most symptoms of the disease are caused by inflammation of a diversity of blood vessels including veins as well as arteries. Common symptoms are mouth sores, genital sores, inflammation on the inside of the eye, and skin problems. 10-30% of cases have neurological manifestations.[7] Behcet's disease involving the nervous system may present with benign headaches, aseptic meningitis, meningoencephalitis, cranial nerve palsies, seizures, personality changes, and cerebral venous thrombosis.[7] The International Study Group for Behcet's Disease proposed a series of diagnostic criteria.[8] In order to make the diagnosis of Behcet's disease, a patient must have recurrent oral ulcerations and at least two of the following: recurrent genital ulceration, eye lesions, skin lesions, and/or a positive pathergy test. The latter is not specific for the disease.

Case Presentation

Mr A.G was a 26-year old male student in a Nigerian tertiary institution residing in Lagos and Ibadan. He presented with a 2-year history of recurrent headaches, genital and mouth ulcers and weakness of the left limbs. The headaches were described as generalized, intermittent, with no periodicity. He gave a negative history of vomiting and photophobia. Genital and mouth ulcers were associated with intermittent fever and weight loss but no diarrhea.

Salient negatives include; head injury, seizures, cough, chronic cough contact, previous transfusion with blood or blood products, family history of the above and promiscuous sexual activity. Mum was late and dad was unemployed. Prior to this presentation, he had been reviewed once in a tertiary hospital in Ibadan, but his dad insisted on voluntary discharge and subsequently continued his son's management with prayers. Patient deteriorated badly, before presenting to Lagos State University Teaching Hospital (LASUTH), through a paternal aunt's

effort.

His clinical examination revealed a right-handed, conscious, but apathetic and abulic young man. He had bilateral 6th nerve palsies, UMN left 7th nerve palsy and a left partial ptosis. He had a bulbar speech with a cerebellar dysarthric twist. Nuchal rigidity, Kernig's and Brudzinski were all present as well as a grade 3 spastic left hemiparesis.

The sensory system was intact and so were the vital signs within normal range. He was investigated for the following differential diagnosis; Bechet's disease, Tuberculous meningitis, Leukodystrophy, and Biswanger's disease.

Discussion

Local documented reports of Behcet's Disease were almost non-existent, more so for neuro-Behcet's. Literature on blacks and Behcet's disease have been outside the African continent. These reports include ocular manifestations in a couple of patients domiciled in London; [9] and another case report in a black patient domiciled in America.[10]

An extensive case report of neuro-Behcet's in a 20-year old patient dwelt more on the neuropsychiatric manifestations observed in the patient. [11]

Diagnosing Behcet's disease is difficult because no specific test confirms it, more so for its multi-systemic involvement and its different clinical presentations. As expected our patient had presented at different hospitals and the disease was advanced before it was finally diagnosed. We did not do a Pathergy test, since it was not specific and patient had minute skin manifestations when he presented.

Systemic steroids have been reported to be effective, especially for ophthalmologic and neurological manifestations of the disease.[12]

The importance of emotional support and understanding was negligible in our patient. Disease specific social security benefits, which might have been of help to our patient are not yet available locally. We tried to rule out our differentials of likely predominant white matter involvement with the available laboratory services. The latter two differentials had been nullified also by patient's age. When treatment was commenced for the first differential, response was not observed after 15 days. A lapse on our management was that we did not seek the expertise of an ophthalmologist to identify possible complications related to eye inflammation. International

Study Group for Behcet's Disease is a study group that proposed the criteria for the purpose of conducting research on the disease.[8]

In the International Study Group for Behcet's Disease proposed diagnostic criteria, neurological manifestations were not included.[8]

Conclusion

Behcet's disease does occur in all races. There is need for increased awareness by clinicians in Africa in order to avoid a misdiagnosis of this disease. The international Bechet's study group may improve possible criteria by including neuro-Behcet's presentations.

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