Case Report

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Neuro-Behcet Disease in a Nigerian Man- A Case Report.

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ABSTRACT

Behcet disease is an inflammatory multisystemic autoimmune disease characterized by tetrad of recurrent oral aphthous ulcers, recurrent genital ulcers, uveitis and cutaneous manifestationS. Vascular and nervous system involvement are less frequent and often late feature of the disease but more likely to be associated with long term sequalae. The disease is more common amongst Asians, Middle Easterners and Southern Europe particularly along the old silk road connecting these regions, with few cases reported in blacks.
This case of neurologic Behcet disease in a young patient that presented to our hospital is being reported to heighten suspicion of this uncommon disease and to underscore its unusual presentation as a neurological disease as well as to outline management options
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Key words- Neuro-Behcet disease, Young, Black Africa

CASE HISTORY

The patient was a-39-years old-man who presented on 7th June 2021 with a 2-year history of recurrent painful oro-genital ulcers. Oral ulcers were multiple and painful, and affected the lips, buccal mucosa and the tongue. Most recent episode of oral ulcers occurred 3 months prior to presentation and resolved spontaneously after 2 weeks. Patient had no perianal ulcers or symptoms of lower gastrointestinal tract bleeding. Genital ulcers affected the scrotum, glans penis and base of the penal shaft. Ulcers usually last for 2-3 weeks before healing with scarring following usage of cocktail of over-the-counter medications. Current genital ulcers started 2-months prior to presentation and had lingered. He has had recurrent episodes of eye pain, redness, gritty sensation and hyper lacrimation in the last 2 years. Eye symptoms usually last for a week and responded to topical mydriatics and steroids prescribed at an eye clinic. No history to suggest decline in vision or diplopia. Patient also had recurrent fever and weight loss. A month prior to presentation he developed postural dizziness, vertigo, slurred speech and was noted to be apathetic and increasingly somnolent.

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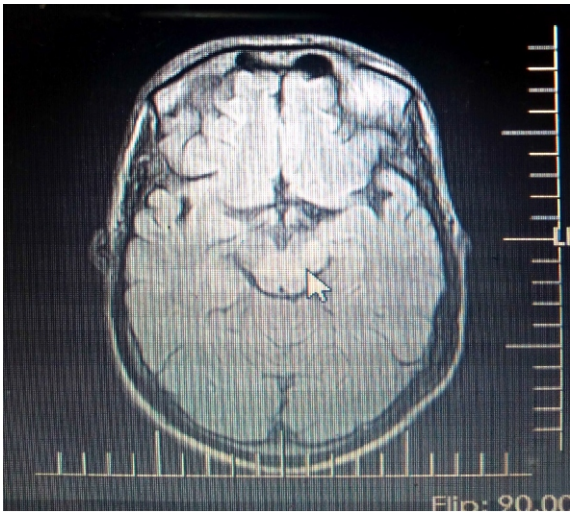

Figure 1: Magnetic resonance Image shows area of T2/FLAIR hyperintensity in the mid brain and pons

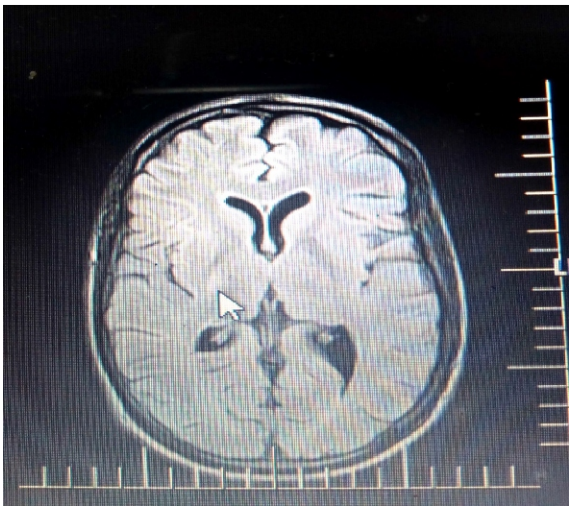

Figure 2: MRI shows T2 flair image showing area of hyper intensity in the right thalamus, with associated dilatation of the lateral ventricle

He had no prior headache, neck pain or stiffness. There was no recurrent skin eruptions or thrombophlebitis. There was no risky sexual behavior or substance use disorder.

Examination revealed a chronically ill-looking and wasted young patient with bilateral injected conjunctivae. He had multiple aphthous ulcers over the lips, buccal mucosae, tongue and hard palate. Also had multiple penile shaft and scrotal ulcers that were non discharging. He had bilateral inguinal lymph node enlargement, largest measuring 2cm by 1.5cm

Blood pressure was 110/70mmHg, His heart sounds are normal 1st and 2nd. On neurological examination the patient was conscious, but had slurred speech and was slow witted, Cranial nerves examination was normal. Motor examination showed grade 4/5 power in muscle groups. The patient was ataxic and had unsteady gait. Other systemic examination findings were unremarkable. Eye review by an ophthalmologist showed features of recurrent granulomatous anterior uveitis. A diagnosis of Behcet's syndrome with CNS involvement was entertained

Results of investigations revealed- PCV-35%, TWBC-6.53 X 10^9/L, N-66.0%, L-30.6%, platelet count was 379 x 10^9/L. Erythrocyte sedimentation rate was markedly elevated at 140mm/Hr. He had normal urea and electrolytes level, antinuclear antibody test was negative, VDRL was non-reactive, Hepatitis B, Hepatitis C and HIV screenings were all negative. Brain MRI showed multiple areas of T2/FLAIR hyperintensities in the right thalamus, mid brain and pons in keeping with thalamic and pontine infarcts. The MRI finding of the patient is shown in figures 1 and 2.

Patient had pulse methyl prednisolone 1 gram daily for 3 days and was commenced concomitantly on tabs azathioprine 50mg 12 hourly, Tabs Colchicine 0.5mg 12hourly, Tabs Prednisolone 30mg daily, in

addition to topical steroid and mydriatic eye drops. He showed remarkable and progressive improvement over the course of 3 weeks hospital stay as oro-genital ulcers healed without sequalae, hypophonia improved and patient was able to stand and ambulate with minimal to no support.

He was subsequently discharged home at 4th week and currently on follow up at both rheumatology, neurology and ophthalmology clinics with sustained clinical improvement.

DISCUSSION

Behcet's syndrome is an autoimmune disease characterized by a tetrad of recurrent oro-genital aphthous ulcers, uveitis, skin manifestations and arteriovenous vasculitis. [1] The disease is more common and severe in Mediterranean region and south east Asia along the old silk road, [1-3], with cases rarely reported amongst black Africans. [4,5] Age commonly affected is between 20-40 years. The index patient fits into the age profile of Behcet's syndrome as he presented at the age of 39 years, with 1st symptoms occurring 2 years prior to presentation. The patient had prior symptoms of recurrent oro-genital ulcers and uveitis. Oral ulcers are the most common symptoms of Behcet's disease occurring in greater than 90% of patients. [1, 2] In 70% of patients, recurrent oral aphthous ulcer is the presenting symptom of the disease. [3] Oral ulcers tend to precede other symptoms of Behcet disease by several years.[1-3] The occurrence of genital ulcers in this patients is in keeping with the pattern of presentation of Behcet's disease as it is the 2nd most common symptom. Genital ulcers affect 80-90% of patients. (1,2) They occur more frequently in male patients, are more painful and deeper and more likely to leave residual scars compared to genital ulcers in female patients. [1-3]

The patient also had eye involvement at the onset of disease and was noted to have granulomatous anterior uveitis at slit-lamp examination. Ocular disease has been reported to occur in approximately 50% of patients with Behcet's disease, mostly in male patients, and can present with features of anterior uveitis, posterior uveitis or a pan uveitis. The temporal relationship between Oculo-Behcet disease and other manifestation of Behcet syndrome varies, with most cases of eye disease occurring years after onset of Behcet disease. The occurrence of ocular involvement at the onset of Behcet disease, as is the case in this patient, has been reported in less than 20% of cases of Oculo-Behcet disease. Symptoms of nervous system involvement (slurred speech, apathy, ataxia, vertigo, dizziness, gait abnormality and somnolence) commenced 2 years into the disease, prompting the patient's presentation for medical care. MRI done at point of care showed vasculitic infarct in the thalamus, mid brain and pons. The finding of multiple infarcts involving the thalamus, mid brain and pons explains the presentation of the patient with neuropsychiatric manifestations

Central nervous system (CNS) disease occurs less frequently in patients with Behcet's syndrome, compared to aphthous ulcers and cutaneous manifestation. Neuro-Behcet disease affects approximately 5-30% of patients,[7] and often portends poor prognosis [7,8] There is often a time lag of 1-7 years between the development of oral ulcers and neurological manifestation in patients with Behcet's syndrome. [9] Neurological involvement in Behcet's syndrome is more frequent and severe in males compared to female patients. The disease predominantly affects the brain parenchyma, presenting as brain stem lesion in over half of the patients. Isolated pyramidal signs, hemispheric disease and spinal cord involvement, are also seen in patients with brain

parenchymal neuro-behcet disease. Other features reported include- aseptic meningitis, seizures, neuropsychiatric symptoms such as hallucination and personality changes (apathy, withdrawal, disinhibition), pseudotumor cerebri, cranial nerve palsies, pyramidal, extrapyramidal and cerebellar symptoms.[8,9]

Non-parenchymal brain disease is due to dural sinus thrombosis and presents with features of raised intracranial pressure. [8,9] Dural venous thrombosis is reported to be more common in patients of pediatrics age group compared to parenchymal disease. [10] Unlike other vasculitis, peripheral nerve disease is uncommon in Behcet's syndrome.[11,12]

Underlying pathophysiologic mechanism of neuro-Bechet disease include- autoimmune induced inflammation (meningoencephalitis), lymphocytic infiltration, vasculitis and thrombosis.

Mortality in Neuro-Behcet disease could be as high as 41%. [9] Adverse prognostic factors include- parenchymal brain disease, elevated cerebrospinal fluid proteins and/or pleocytosis, brain stem involvement, primary or secondary progression of neurologic disease and relapses during treatment.[8]

The patient was managed with pulse IV methylprednisolone in addition to tabs azathioprine with progressive improvement in symptoms. As patient speech improved, ataxia and postural dizziness reduced and he was able to commence graded ambulation without support before discharge 4 weeks into admission

Treatment of CNS involvement in Behcet's disease is dependent on operative mechanism-and often entail usage of steroids and disease modifying antirheumatic drugs (DMARDs) such as azathioprine, cyclophosphamide, mycophenolate mofetil, chlorambucil etc. The index patient responded to a combination of IV methylprednisolone and oral azathioprine. Azathioprine is the most commonly used DMARD in patients with severe manifestation of Behcet's disease because of its predictable and low side effect profile. [12] Recent studies have shown the advantage and short- and long-term superiority of immune biologics (anti-TNFs) over conventional disease modifying antirheumatic drugs such as cyclophosphamide and azathioprine in the management of severe manifestation of Behcet's syndrome, including CNS disease. [13-15] The option of using immune biologic was explored in the patient but shelved due to lack of finance

.CONCLUSION

Behcet's syndrome though rare amongst blacks, do occur. Presentation with CNS involvement occurs late in disease and is associated with severe morbidity. Management with pulse methylprednisolone and maintenance therapy with Azathioprine was effective in control of vasculitic CNS disease in the index case

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