UNUSUAL PRESENTATION OF BEHCET DISEASE (NEURO-BEHCET)

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ABSTRACT

Behcet disease is a common disease and usually present by oral, genital ulcers. Can be affect multiple systems in the body. CNS can be affect but usually after present of orogenital ulcer. In our case report, the patient present with neurological manifestations before orogenital ulcers, and that is rare presentation (less than 3 percent).

Key words: Behcet disease, neurobehcet disease, Vasculitis

ملخص :مرض بهجت هو مرض معروف و منتشر ويظهر عادة بتقرحات بالفم والأعضاء التناسلية ومن الممكن أن يؤثر على أجزاء كثيرة من جسم المريض ومن ضمنها الجهاز العصبي.
لكن أعراض الجهاز العصبي تظهر عادة بعد ظهور التقرحات في الفم والأعضاء التناسلية.
في هذه الحالة المريض اشتكى من أعراض الجهاز العصبي قبل ظهور التقرحات في الفم والأعضاء التناسلية وهذا شي نادر الحدوث. (اقل من ٣ %)

INTRODUCTION

Behcet's disease is a multisystem, recurrent, inflammatory disorder.[2] It was first described by H. Behcet (1924) as a three-symptom complex comprising uveitis, oral aphthae and genital ulcerations. Later, in 1930, the Greek physician Adamantiades reported a patient with inflammatory arthritis, oral and genital ulcers, phlebitis, and iritis.[1]

Behcet disease can affect central nervous system in less than one- fifth of patient. There is, a period of approximately five to six years elapsed between the onset of the earliest non-neurologic symptoms of Behçet's disease and the appearance of neurologic symptoms or findings. Nevertheless, neurologic findings may also appear concurrently (7.5 %) or precede non-neurologic features (3 %). Twenty percent of those with neurologic findings were asymptomatic.[3,4]

DIAGNOSIS

CNS manifestation according to site of lesion, they are commonly present (>50%) by pyramidal signs, hemiparesis and headach. Some patient present by brainstem sign, fever, paraparesis, meningeal sign (aseptic meningitis), movement disorder, sensory disturbance (10-40 % of case). Less common manifestations (5% of case) like seizure, cerebellar syndrome, optic neuropathy, psychiatry disorders may present.[6] It can also present with aseptic meningitis or encephalitis, and Vasculitis. And occurs more commonly in male than female.[5]

Coarse of disease can be recurrent attacks, primary progression or secondary progression.[3]

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Neuro-Imaging: MRI

The lesion type during an acute attack is usually located in the basal ganglion region or in the brainstem, extending to the diencephalic structures, in about 30% of the cases the lesion are bilateral. In some cases a mass effect may be noted, and the centre of these lesions shows contrast enhancement. And can be small scattered lesions located in the basal ganglion region, brainstem or the internal capsule. In chronic cases, atrophy of the brainstem and enlargement of the third ventricle disproportionate to the lateral ventricles or cortical sulci can occurs.[3]

CSF examination:

Can be completely normal and can show pleocytosis and/or elevated protein content. CSFs with a high cell count, had either neutrophilic predominance or both neutrophils and lymphocytes, or lymphocytic predominance.[3]

Management:

Neuro-behcet consider as life threatening condition so we should treat the patient with intensive immunosuppressant. The combination of high dose of glucocorticoid and another immunosuppressive agent (like Azathioprine, Infliximab, and cyclosporine) is required.[7,8]

Aseptic meningitis may occur intermittently in-patient with behcet disease. In absence of other manifestation usually treated with high dose of glucocorticoid with great response.

In case of recurrent aseptic meningitis may need to use long-term maintenance therapy with another immunosuppressive agent like azathioprine.[9]

CASE REPORT

A 21-year-old male Saudi presented to hospital initially with 12 days history of headache and fever, followed by 3 days history of left sided body weakness, double vision and inability to open right eye.

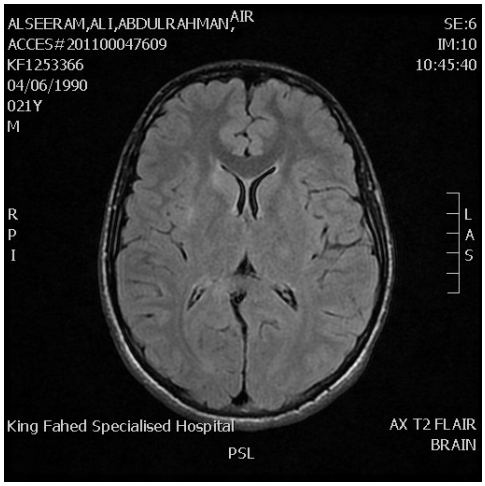


Fig 1: T2 flair (right basal ganglia hyperintense signal)

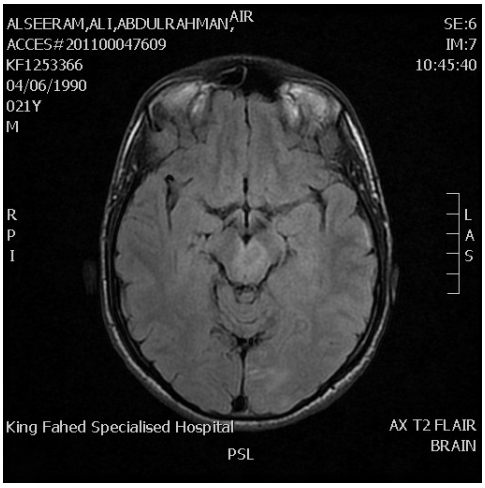


Fig 2: T2 flair (left midbrain hyperintense signal)

There was no history of oral or genital ulcer, red or painful eye and seeing halos. No skin rashes and swollen or painful joint joints.

There was no past history or family history of similar condition.

O/E: Temp: 37.8^{0}C, BP: 125/85 mmHg, RR: 19 cpm. Pulse: 90

Patient was confused and slightly slow in mentation. Had left 3rd and 4th cranial nerve palsies with left hemiparesis and bilateral hyper-reflexia. Also had mild dysmetria and ataxia over the right side, though was mobile without support. Other systems including skin and musculoskeletal systems were unremarkable, had no oro-genital ulcers, no evidence of lymphadenopathy

X-Ray Chest; unremarkable

CT brain was unremarkable.

MRI showed multiple signal change low T1 bright T2 and flair sequences at right basal ganglia, right parietal lobe, left occipital lobe and midbrain (Figure 1 and 2).

CSF-Analysis; Lumber Puncture was done opening pressure was normal, WBC 200 cell/mm3, 85% lymphocyte, 15% polymorph, protein 1.06 gm/L, sugar 2.3 mmol/L. Gram stain and AFB stains were negative, culture revealed no growth. PCR and C/S for mycobacterium was not available at the local hospital so it was send out to the central laboratory but unfortunately no result could be obtained latter on.

CBC and routine Chemistry were unremarkable. ESR and C-reactive protein were normal and ANA was negative.

Possibilities of CNS Tuberculosis, Acute Demylenating Encephalomyelitis (ADEM), Vasculitis (Behcet's disease) and Sarcoidosis were thought off and patient was started on steroids (dexamethasone 4 mg TID then decrease to 2 mg BID at discharge for 1 month then start tapering), and Anti-Tubercular drug(ATT).

Follow up; Patient improved clinically over period 2 weeks, MRI was repeated after one month, which showed regression in the size and number of lesions (Figure 3 and 4).

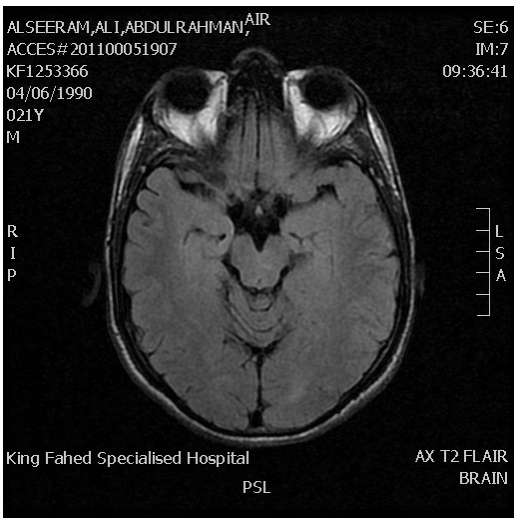


Fig 3: T2 flair (improve midbrain lesion)

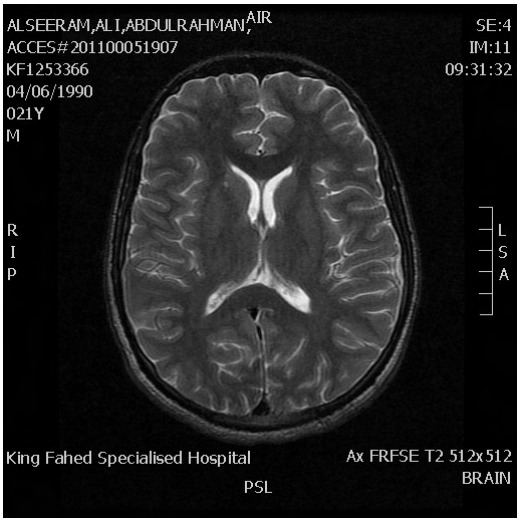


Fig 4: T2 (right basal ganglia lesion improve)

Unfortunately, patient was not compliant to medication and after 10 months had a relapse of his condition. This time patient also reported oral ulcers but no genital ulcer, no visual symptoms or skin manifestations.

Patient was confused, dysartheric, had right side complete and left partial ptosis, pupil 3mm bilateral reactive to light. Fundus examination was normal also had right 6th nerve palsy. Had left hemiparesis (4/5 weakness). Sensations were normal, Gait

was ataxic. No evidence of uveitis or retinitis. Chest, CVS and abdominal examination was unremarkable

INVESTIGATION

CBC, Chemistry was unremarkable. ESR: 15mm 1st hour, ANA was Negative and complements C3 and C4 were normal. Serology for HSV 1 & II, Brucellosis, VDRL and HIV was negative. PPD test and skin Pethargy tests were normal.

X-Ray Chest; unremarkable

MRI Brain and cervical Spine:
Multiple T1 hypointense, T2 and flair hyperintense lesion were seen involving brainstem, thalamus, Cerebellar peduncle and left occipital lobe and basal ganglia. Some of the lesions were partially enhanced with GAD contrast. Cervical spine was unremarkable (Figure 5 and 6).

Cerebrospinal fluid (CSF):
Opening pressure was Normal. WBC 60 cells/cmm, 40% were polymorphs and 56% lymphocyte. Proteins; was 1 gm/ L.

Gram & AFB stains and culture were negative.HLA-B52, Mycobacterium culture and PCR were sent to central lab and result came negative.

In Differential Diagnosis this time suspicion of Neuro Behcet's disease was very high so again started on high dose of steroids (methylprednisolone 1g for 5 days followed by prednisolone 50 mg and steroid sparing drug Azathioprine 50 mg for 1 week then increase to 100 mg daily.

Hospital Course and Follow up:
Patient again showed clinical recovery in about 2 weeks. He was discharged on oral prednisilone 50 mg daily then tapering dose with calcium, vitamin D supplementation and Azathioprine 100 mg daily.

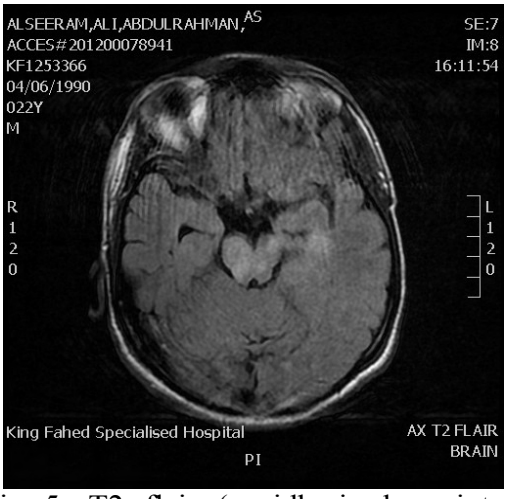
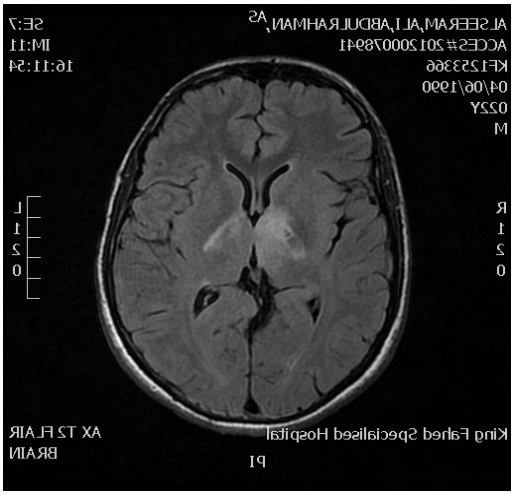


Fig 5: T2 flair (midbrain hyperintense lesion)


Fig 6: T2 flair (basal ganglia hyperintense lesion)

DISCUSSION

Our patient had a diagnostic challenge at 1st presentation, as he presented with brainstem, basal ganglia deficit and alteration of sensorium, fever and headache of subacute to chronic course. It was suggestive of subacute multifocal disseminated and diffuse involvement of CNS with progressive course. Initially diagnosis of Encephalitis, including ADEM (Acute disseminating encephalomyelitis), were thought of. Suspicion of CNS-

Tuberculosis could not be ruled out, because of the clinical subacute presentation with headache, fever. Neuroimaging, CSF findings were suggestive and PCR and culture for Mycobacterium were not available at our hospital. However patient was started on High dose IV methyl prednisolone for 5days with anti-tubercular (ATT) and anti-viral coverage, followed by oral steroids (dexamethasone) tapering dose.

Possibility of Vasculitis including neurobehcet's disease was remote at this stage, as there were no other symptoms and signs suggestive or fulfilling diagnostic criteria. Anyhow, our patient was lucky enough to be started on steroids without delay, though for treatment of ADEM and as adjunct with ATT. He was saved from devastating consequences of Neuro Behcet's disease and made a good clinical recovery.

The relapse of his condition due to non compliance to medications, the history of oral ulcer and pattern of lesions on MRI raised our index of suspicion further towards Vasculitis particularly Neuro Behcet's disease. Furthermore, the anti-tubercular workup and serology for connective tissue disease and infections was negative.

CONCLUSION

Neuro-Behcet can be devastating if left untreated, so it is very important to have high index of suspicion and this rare possibility should be considered in the differential diagnosis of such cases.

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