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Are We Missing Behçet's Disease?

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Abstract | **Case Report**

Behçet's disease is already famous with classical triad of recurrent oral ulcers, genital ulcers and uveitis. It is also known for its multisystemic involvement. However, an accidental detection of this disease is unusual. We will report here the story of a gentleman who had presented with urinary tract infection and was found to have underlying Behçet's disease for years. He had peripheral neuropathy as the only neurological manifestation without involvement of other neuro-axis as commonly seen in Neuro-Behçet's disease. He also lacked prominent systemic involvement. This incident underscores the necessity of careful clinical examination, keeping suspicion high for this disease especially for late presenters as we may miss and expose patients to unnecessary sufferings and even to risk of serious complications. This case also shows the importance of inter-departmental coordination while approaching unusual cases or common cases with unusual presentation.

Keywords: Behçet's disease, neuro behcet's syndrome, late-onset Behçet's disease.

INTRODUCTION

Neurological manifestation varies widely in Behçet's disease; 5-23% of cases present with it as the first symptom [1]. Neuro-Behçet's disease may present with more common parenchymal (intra-axial) form, less common extra-axial form like venous sinus thrombosis or a combination of these two [2]. Isolated peripheral nervous system involvement in neuro-Behçet's disease is very rare [2].

Here, we will present a case who presented with lower urinary tract symptoms and was incidentally diagnosed to have Behçet's disease.

CASE

Sixty eight year old gentleman, without known comorbidities except smoking had presented to us with dysuria and whitish urethral discharge associated with generalized weakness for fifteen days. He was apparently well before this episode, except complain of pain and swelling of hands, knees and ankles for last three years without early morning stiffness. He also had decreased hearing bilaterally for same duration. He recollected that he has developed blackish discolouration of both lower limbs specifically below knees for last one year and recurrent multiple oral ulcers for an indefinite period (Figure 1 & 2).

Fig-1: Nodular Lesions over leg which are healing up

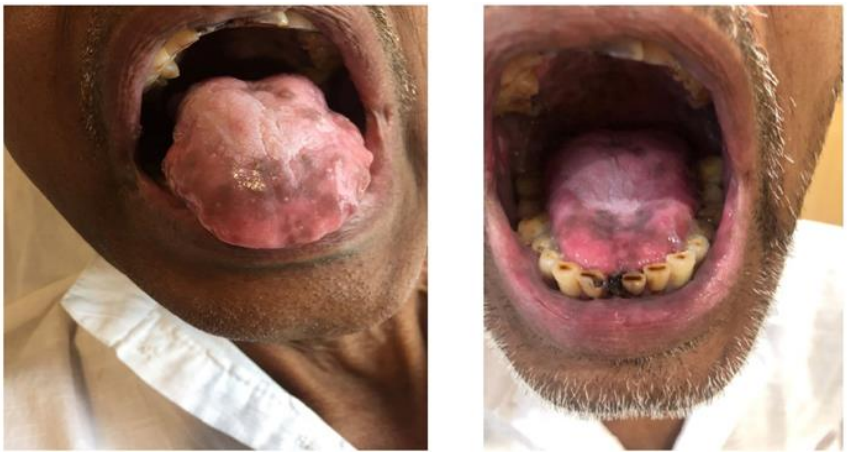

Fig-2: Hyperpigmented scar marks of multiple oral ulcrs

He had attended doctors of multiple speciality and got symptomatic relief from these symptoms during exacerbations.

On further query, he gave history of recurrent genital ulcers since last two to three years. His general examination revealed old hyperpigmented scar marks of multiple oral and genital ulcers along with few active oral aphthous ulcers. There was evidence of multiple acneiform lesions along with papulopustular to nodular skin lesions over forearm and elbows. A detailed history of tender nodular lesions over leg and healing up of those lesions after 2-3 weeks with hyperpigmentation was suggestive of erythema nodosum.

These type of skin lesions with evidence of recurrent oral and genital ulcerations matched the diagnostic criteria of the Behçet's disease according to "International study group for Behçet's disease" [3]. Though pathergy test was negative, genetic testing came positive for HLA B5 supporting our suspicion to be true.

Neurological examination revealed normal higher mental function with decreased hearing bilaterally, right more than left. His hearing impairment was later confirmed with pure tone audiometry, where moderate sensorineural on left side and profound mixed hearing loss was evident on the right. X ray revealed sclerotic mastoids.

There was evidence of distal more than proximal weakness of lower limbs (MRC grade 4/5 to 4+/5) with decreased deep tendon reflexes. Graded sensory loss in lower limbs with clinical features of small and large fibre involvement was evident. Romberg test was positive and tandem was impaired. His upper limb examination was within normal limit

Nerve conduction study was corroborative, revealing mildly asymmetric sensorimotor axonopathy (sensory more than motor) of lower limbs and unremarkable upper limb finding.

The routine blood tests were normal except reversed albumin and globulin ratio with mildly raised ESR. So, we performed a serum protein electrophoresis to rule out plasma cell dyscrasia as a possible etiology of the peripheral neuropathy and it proved to be negative. A detailed laboratory tests were done; all other immunological (ANA, RA, Anti CCP antibody etc.) and serological (including HIV, VDRL) markers were found to be negative.

Keeping in mind the multisystem involvement of Behçet's disease, a detailed cardiac, gastrointestinal workup was done and ophthalmological consultation was taken. However, there was no evidence of other systemic involvement. A doppler study of lower limbs was performed and venous or arterial thrombosis was ruled out.

On evaluation of his presenting complain, urine routine examination revealed twenty pus cells and culture sensitivity was negative. His lower urinary tract symptoms improved with a course of antibiotic.

He was started with a course of steroid, colchicine and azathioprine along with neurorehabilitation for his neurological symptom. With this regimen his symptoms responded very well.

DISCUSSION

The common neurological symptoms among patients with Behçet's disease are headache, upper motor neuron related weakness, brainstem and cerebellar involvement; cognitive disorders and behavioural abnormality [4].

Our patient presented with urinary symptoms which never returned after treatment of urinary tract infection. It is less likely due to Behçet's disease where bladder involvement occurs more commonly in early age [5]; more so, as it completely reversed in such a short duration.

Peripheral nervous system involvement is very rare, detected mostly in patients who presents with other significant neurological symptoms [4]. Our patient had peripheral neuropathy as the only prominent neurological finding. Neuropathy in Behçet's disease is also variable including sensorimotor polyneuropathy, mononeuritis multiplex and even Guillain– Barré syndrome [6].

In the study done by Akbulut *et al*., amongst electrophysiologic studies done on 63 patients with Behcet's disease, frequency of subclinical neuropathy was found in 14.2% of patients with sensory involvement, noted to be much more common [7]. In their study, Birol *et al*., concluded that patients who exhibited electrophysiological peripheral neuropathy had much longer disease duration [6]. In our case, disease duration is not exactly known; clinical and electrophysiological finding revealed evidence of quite advanced involvement of peripheral nerves (we ruled out other possible etiologies). It is possible that his disease remained subclinical for a long time.

Our case had sensorimotor axonopathy matching electrophysiological finding in other case series. He had predominant lower limb involvement which matched with predominant findings of Birol *et al.*, [6].

MRI brain in our case revealed old ischaemic foci in corona radiata, central pons with periventricular ischaemic changes. In our case, we are not sure whether the vascular insults were secondary to Behçet's disease as he had other vascular risk factors and site of stroke was also not classical.

Though studies revealed common linkage between ocular involvement and genetic linkage [8], our patient inspite of having HLA B5 positivity, had an unremarkable ocular finding.

He had bilateral hearing impairment predominantly sensorineural type agreeable with other case series. In the study done by Kemal O *et al*., pure tone audiometry detected a sensorineural hearing loss in 10 of the 29 patients of Behcet's disease (34.5%) in comparison to healthy controls, a statistically significant difference underscoring the requirement of routine auditory testing in patients with Behçet's disease [9].

Our patient is a late presenter with unusual type of neurological manifestation i.e peripheral neuropathy without any other neuroaxis involvement. The multiple white matter changes and brainstem vascular insults were not classical of vasculitis secondary to Behçet's and would be better attributable to small vessel disease secondary to his background risk factors. His arterial and venous angiogram were also within normal limit ruling out Behçet's disease induced non parenchymal vascular pathology.

CONCLUSION

Our case had a very indolent course and disease came to notice when he presented with dysuria secondary to urinary tract infection. It justifies the need for close follow up of patients with fewer than three criteria of the Behçet's disease, keeping suspicion high with multi-departmental approach especially in late presenters as this disease causes morbidity and serious complications.

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