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Case report

Corticobasal syndrome like presentation of Hashimoto encephalopathy with cortical ribboning

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ABSTRACT

Hashimoto encephalopathy (HE) and corticobasal syndrome (CBS) are heterogeneous disorders characterized by variable presentation; seizure, stroke like symptoms, myoclonus, alien limb phenomena, dystonia, limb apraxia, although extrapyramidal features and alien limb phenomena are rare in HE. Here we are reporting a 70 years female who presented with 3 months history of asymmetrical onset rapidly progressive disease associated with alien limb behaviour, myoclonic jerks, dystonia, seizure, speech impairment, asymmetrical extrapyramidal features (left > right), gait ataxia and cortical ribboning on brain imaging without visual disturbances, cognitive decline or neuropsychiatric abnormalities. EEG did not show any periodic discharges or slowing. Neuronal antibody VGKC was negative but she had euthyroid state with high titre of anti TPO antibody 590 IU/ml (normal upto 95 IU/ml).

Therefore, diagnosis of Hashimoto encephalopathy in a probable sporadic case of corticobasal syndrome was made. Patient was treated with antiepileptic and IV steroid for 5 days. At the time of discharge she was conscious and followed verbal commands.

In every case of rapidly progressive neurodegenerative disease like CBS, investigations for HE should be planned.

1. Introduction

Hashimoto encephalopathy (HE) has been initially diagnosed as either a primary psychiatric disorder or a neurodegenerative condition. High titres of thyroid autoantibodies with euthyroid state are pathognomonic for this disease. Although other laboratory findings in HE are nonspecific like increased cerebrospinal fluid protein, diffuse or focal slowing on EEG and nonspecific T2 hyper intensities on imaging [1]. High titre of anti −TPO antibodies is considered diagnostic for HE in appropriate clinical setting and it has been detected in almost 100% cases which are reported so far. Pathological findings of HE are heterogeneous with mild lymphocytic peri vascular infiltration of small vessels and gliosis in cortex, hippocampus, basal ganglia, thalamus or T cell infiltration of leptomeningeal venule [2]. Corticobasal syndrome (CBS) is also considered a heterogeneous disorder of asymmetrical rigidity, dystonia, myoclonus, cortical sensory loss or alien limb phenomenon with relatively preserved cognition in early course of the disease. Motor features like myoclonus and alien limb behaviour at disease onset are regarded characteristic for CBS but they are present in less than 50% patients [3]. Alien limb phenomenon is also reported in stroke, progressive supra nuclear palsy, Alzheimer's disease and Creutzfeldt-Jacob disease (CJD) [4].

Herein we are reporting a case of CBS like presentation of HE with cortical ribboning on brain imaging.

2. Case report

A 70 years old female presented with 3 months history of abnormal brief jerky movements in form of slippage of small objects with her left hand. After few days, she developed alien limb phenomenon in form of inability to control her left upper limb with rapidly progressive abnormal posturing. She used to say that sometimes her left upper limb would go upward and backward in midline automatically. One month after the disease onset of illness she developed abnormal patterned twisting posturing in all four extremities distally. Her symptoms progressed to inability to maintain balance in the bed as well as difficulty in walking in form of swaying on either side. Speech was explosive, effortful with

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intermingling of words(spastic dysarthria) which were hardly understandable to family members. There was no history of blurring vision, cognitive impairment, neuropsychiatric abnormalities, trauma, bowel-bladder involvement or family history.

General physical examination and examination of other systems was unremarkable. On neurological examination, patient was conscious alert following verbal commands without cognitive involvement (MMSE 24/30). Her speech was spastic with normal fundi and cranial nerve examination. Motor system revealed asymmetric rigidity (Left > Right), myoclonus, dystonia with fixed twisting posture of left hand associated with alien limb phenomenon. There were bilateral cerebellar signs. Sensory modalities were normal.

Routine investigations, including ESR and EEG were normal. Tests for neurosyphilis and HIV were also non reactive. MRI Brain revealed asymmetrical hyperintensities in fronto-occipito-parietal region (cortical ribboning) with subtle hyperintensities also present in left temporal lobe on DWI and FLAIR axial images and reduced corresponding ADC level (Fig. 1). After three days of hospitalisation she deteriorated further in form of altered sensorium subsequent to an episode of generalised tonic clonic seizure. CSF examination was normal except mild increased protein. Further work up like serum ammonia, autoimmune and vasculitis profiles(ANA, Anti-ds DNA, Anti-nucleosome, Anti-histones, Anti-Sm, Anti SS-A, Anti RO, Anti Scl-70, Anti Rib-PProtein, Anti-JO, Anti-SS-B) were also carried out. Autoimmune encephalitis work up including VGKC antibody was negative but thyroid profile revealed euthyroid state with high titre of anti TPO antibody 590 IU/ml (normal range- upto 95 IU/ml). So diagnosis of Hashimoto encephalopathy in a probable

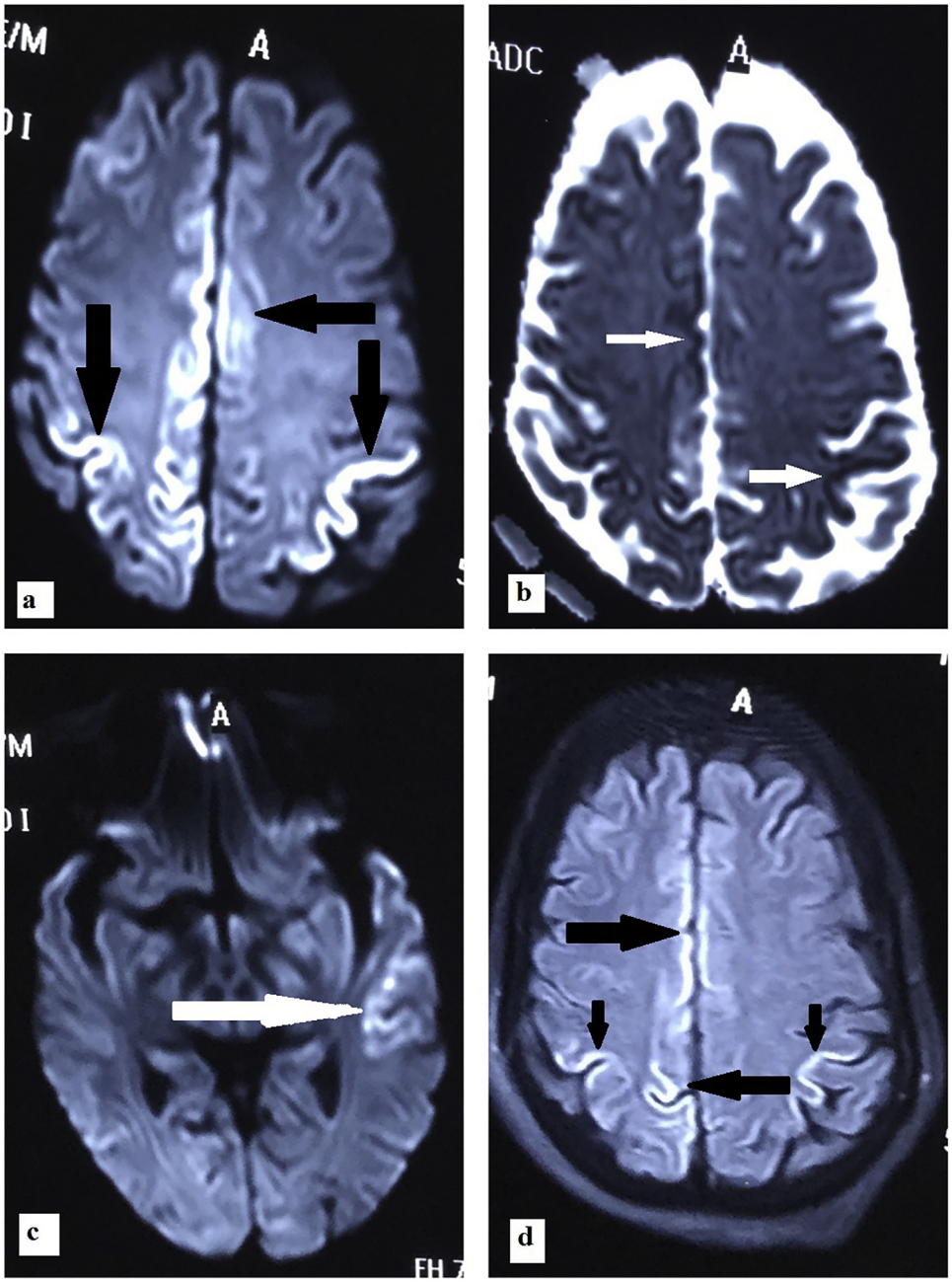

Fig. 1. MRI Brain revealed bilateral asymmetrical hyperintensities in fronto-occipito-parietal region (cortical ribboning), in left temporal lobe on DWI and FLAIR axial image, (a,c,d) and reduced corresponding ADC level (b).

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