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FDG-PET Hypometabolism in a Rare Neurological Disorder; The Creutzfeldt-Jakob Disease, A Case Report

ABSTRACT

The Creutzfeldt-Jakob disease (CJD) is an acute onset neurological disorder with a group of symptoms, characteristic clinical and diagnostic features. As the disease progresses, mental symptoms worsen. This inevitable fatal rapidly progressive neurodegenerative disorder is thought to be caused by an abnormally folded cellular glycoprotein known as the prion protein. Many of the patients suffered from CJD in the due course lapse into a coma, the respiratory failure, heart failure, pneumonia, or other infections are generally the cause of death which is usually less than a year. Involvement of the brain is characteristically seen on Fluorine-18-fluorodeoxyglucose-positron-emission-tomography/computed-tomography-scan (F-18-FDG-PET/CTscan), and in T2, FLAIR, and DWI sequences of MRI. This case highlights a typical pathological pattern of FDG hypometabolism in F-18-FDG-PET/CTscan in the case of CJD which is rare to see in day to day clinical practice.

Key words: [18F] fluoro-2-deoxy-D-glucose, Creutzfeldt jakob disease, FDG, MRI, PET scan, Positron emission tomography

INTRODUCTION

The Creutzfeldt-Jakob disease (CJD) is an acute onset rapidly progressive and fatal neurological disorder caused by prion protein with classical clinical and imaging features. Usually, the MRI findings together with CSF and EEG changes are routinely seen in CJD symptomatic patients, which render the use of FDG-PET/CTscan non-essential for the diagnosis of CJD.[1] However, in case of a lack of complete workup for CJD (as in contraindication for MRI or difficulty in performing or interpreting EEG or lumbar puncture due to non-cooperative agitated patients), the use of FDG-PET/CTscan and the knowledge of expected FDG-PET abnormalities in CJD might be useful to help making a diagnosis also help us determining the severity of the disease which can precede the clinical symptoms as well as to exclude the other neurodegenerative disorder and paraneoplastic syndromes.

CASE REPORT

We present a case of 63-year-old male, who presented with complaints of sudden onset right upper limb pain and weakness followed by bilateral lower limb weakness with loss of coordination, imbalance, jerky movements, ataxia as well as slurring of speech which is rapidly increased in intensity over a period of few weeks. He had no significant previous medical or family history. An MRI study of the brain was performed which revealed asymmetrical T2/FLAIR hyperintensities [Figure 1a] and restricted diffusion [Figure 1b] in the left caudate and lentiform nuclei, thalamus, and left superior parasagittal frontoparietal cortex. In view of abnormal MRI findings, a FDG PET/CT scan was advised for further evaluation as well as to rule out any occult paraneoplastic etiology. A whole-body F-18-FDG-PET/CT scan was obtained after 6 hours of fasting with non-contrast CT images acquired from vertex to mid-thighs along with a dedicated brain FDG PET study as well on a Philips Trueflight PET/CT camera after 1 h of administration of intravenous 7 mCi FDG radioisotope. The PET scan revealed mild to moderate pathological FDG hypometabolism in the left medial and lateral frontal as well as temporal cortices, left-sided striatum (caudate nucleus and putamen), left thalamus, left-sided midbrain, pons, and left middle cerebellar peduncles and left occipital cortex with severe FDG hypometabolism in entire left parietal cortex and right-sided cerebellar diaschisis [Figure 2]. PET scan findings revealed a typical pattern of FDG Hypometabolism as seen in CJD. Further confirmation of diagnosis was achieved with CSF analysis which showed increased Tau protein

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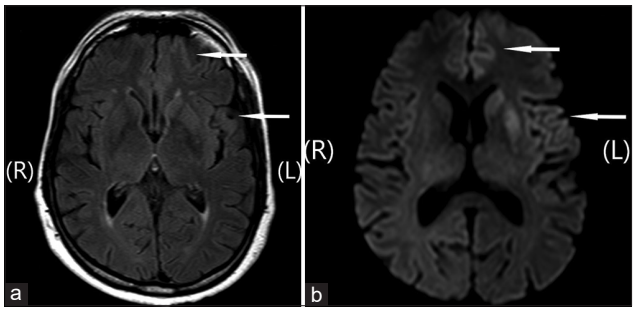

Figure 1: (a) Axial Flair MRI image of brain – shows asymmetrical hyperintesities in left (L) caudate and lentiform nuclei, thalamus and left superior parasagittal fronto-parietal cortex (arrows). (b) Axial Diffusion Weighted MRI image of brain – shows asymmetrical restricted diffusion in left (L) caudate and lentiform nuclei, thalamus and left superior parasagittal fronto-parietal cortex (arrows)

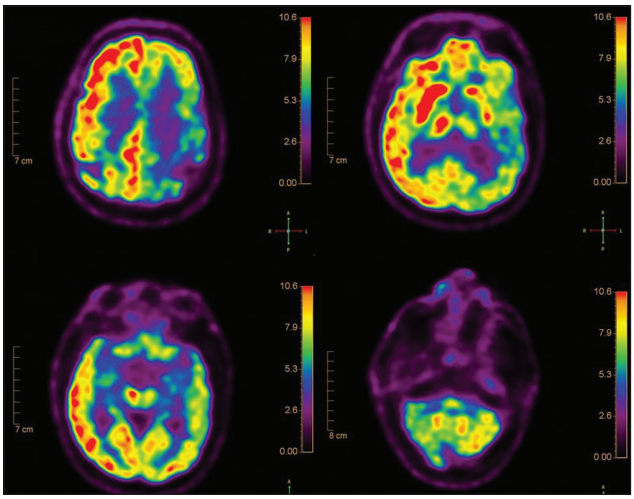

Figure 2: Sequential Axial brain FDG-PET scan images – are showing pathological mild to moderate hypometabolism in left frontal cortex and left half of the anterior and posterior cingulate gyrus, severe hypometabolism in left parietal cortex, with mild to moderate hypometabolism in left straitum and left thalamus, left temporal and occipital cortices, left sided mid brain and pons as well as left occipital lobe with right cerebellar diaschisis

and neuron-specific enolase, with normal CSF cell counts, electrolytes, and no evidence of pathogenic microorganisms on gram stain or atypical cells on cytology. CSF glucose was also within normal range. EEG revealed three-phase sharp wave complexes. Complete blood count, urine routine microscopy, ANA blot, serum electrolytes were unremarkable. No abnormal metabolic activity or structural lesion was found in the rest of the PET/CT scan to suggest a concomitant paraneoplastic process.

DISCUSSION

A study on the CJD patients by using [18F] fluoro-2-deoxy-D-glucose positron emission tomography (FDG-PET) scan, analyzed nine patients through PET statistical parametric mapping (SPM), which revealed lateralized FDG-PET hypometabolism in the medial and lateral parts of the frontal and parietal cortices.2,3 In that study, FDG-PET proved more sensitive than MRI for detecting cortical abnormalities.[2]

In clinical CJD subtypes, few case reports have shown FDG-PET hypometabolism involving cortical areas related to the clinical signs. The pyramidal or extrapyramidal signs observed in CJD are one of the main clinical features required for the diagnosis of sporadic CJD, and the lateralized brain FDG PET hypometabolism in CJD patients is probably led to an estimation of the involved brain areas which corresponds to the clinical signs reflecting genuine clinical and FDG-PET correlation. In most of the CJD patients left-lateralized brain hypometabolism is seen thus explaining the left predominant brain areas involved in the clinical subgroup analyses.[2,3]

Signs frequently encountered in the CJD include pyramidal and extrapyramidal (rigidity, bradykinesia, and limb dystonia), as well as limb dystonia, sensory loss, alien limb, apraxia, akinetic mutism, myoclonus, dementia, ocular movement disorders, and visual signs.[4] Sometimes, one or more of these clinical symptoms are predominant and considered as clinical CJD subtypes, for example, the Heidenhain variant when prominent visual signs are present; or the corticobasal syndrome subtype in the presence of movement disorder and higher cortical dysfunction such as cortical sensory loss, apraxia.[1,5]

In particular, the lateral frontal and mesial parietal hypometabolism is generally found in CJD patients, however, hypometabolism can also be observed in the occipital cortex in CJD patients with visual signs, and hypometabolism in the middle cerebellar peduncles and pons in CJD patients with ataxia, or hypometabolism in the lateral parietal and prerolandic cortex in CJD patients with corticobasal syndrome. Larger patient numbers are needed to further analyze the relationship between clinical presentation and FDG-PET metabolism.

CONCLUSION

CJD reveals a typical pathological hypometabolism pattern on FDG-PET scan which not only helps in making a diagnosis but can also predict its severity based on the cortical involvement and prognosis of patient. However, due to the combined high sensitivity and specificity of electroencephalogram abnormalities, MRI changes as well as abnormal protein detection in the cerebrospinal fluid, the additional diagnostic value of FDG-PET for CJD is probably limited but ineluctable.

To the best of our knowledge, a systematical analysis of the relationship between clinical presentation and FDG-PET hypometabolism in CJD patients has never been reported in an Indian scenario.

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