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Interesting 3T MRI findings in a case of sporadic Creutzfeldt-Jakob disease

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ABSTRACT

A 36 year old lady with possible sporadic Creutzfeldt-Jakob disease which manifested with rapidly progressive cognitive decline and abnormal behaviour, pyramidal, extrapyramidal signs and akinetic mutism. Scalp EEG findings show diffuse slowing of background activity of delta range. MRI brain revealed high signal intensity in bilateral caudate nuclei and putamen on T2WI, FLAIR sequence and DWI. The role of MRI in diagnosis of the CJD is discussed.

INTRODUCTION

CJD is one of the transmissible spongiform encephalopathies, a rare but important group of diseases, affecting humans and other animals characterized by fatal neurologic illness, unusual neuro-pathologic changes and unconventional transmissible causal agent. In his ground-breaking work for which he got a Nobel prize, Stanley Prusiner proposed that the transmissible agent in CJD was a protein which catalyses the conversion of normal native protein to isomeric form[1]. Clinically it is characterized by rapidly progressive dementia, myoclonus, ataxia and akinetic mutism. Diagnosis often rests on EEG and 14-3-3 protein in CSF. However, some recent studies have reported signal abnormalities in the deeper gray matter nuclei which are characteristically observed in the disease. We report a case of possible CJD with characteristic MR findings.

CASE REPORT

A 36 year old lady, housewife, manifested acute onset of abnormal movements of all 4 limbs. The illness was gradually progressive and was dominated by cognitive decline and loss of social inhibition, abnormal behaviour, irrelevant talk, inappropriate smile. During the next 2-3 months, she developed unsteady gait, was muttering to self and became incontinent, unresponsive and subsequently became bed bound. At admission she was mute and used to respond with inappropriate smile. On examination vitals were stable, grasp reflex (+), palmar reflex (+), generalized rigidity, deep tendon reflex (+++), plantar normal. On investigation the patient was found to have normal hemogram, KFT, LFT, blood sugar, peripheral blood smear (including acanthocytes). Thyroid function test (including serum thyroglobulin antibodies, thyroid peroxidase antibodies), HIV, cerebrospinal fluid (CSF) analysis and X-ray

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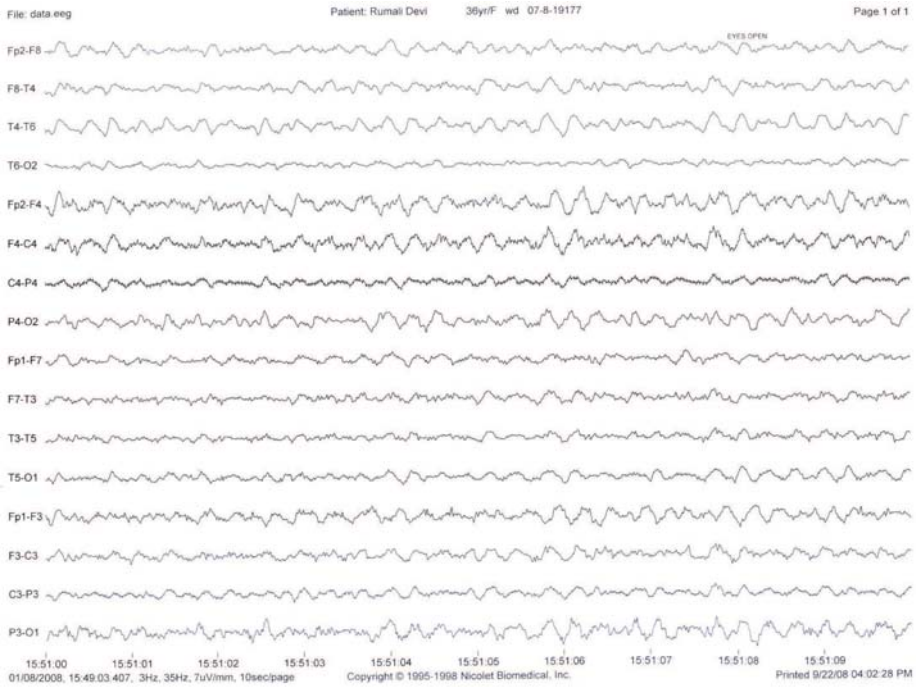
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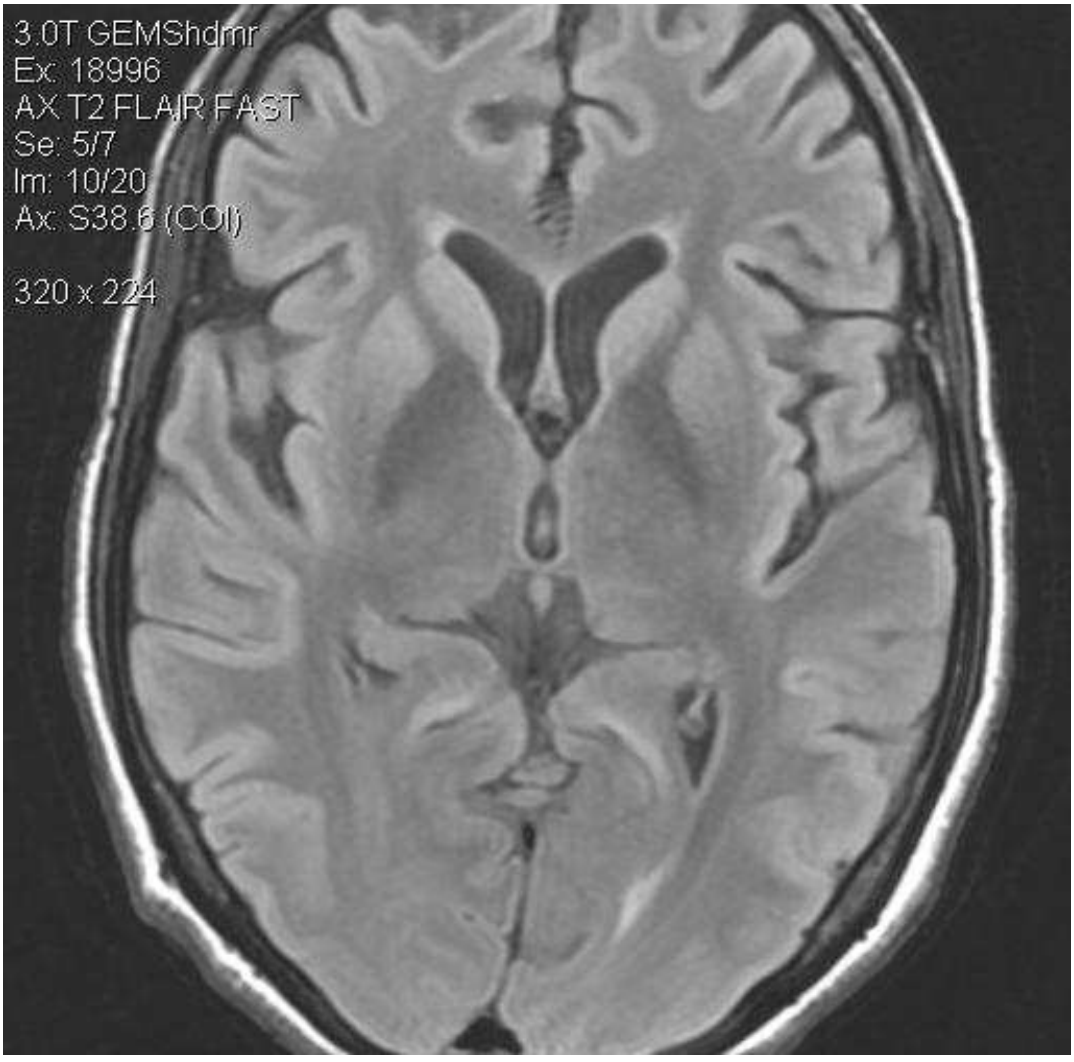
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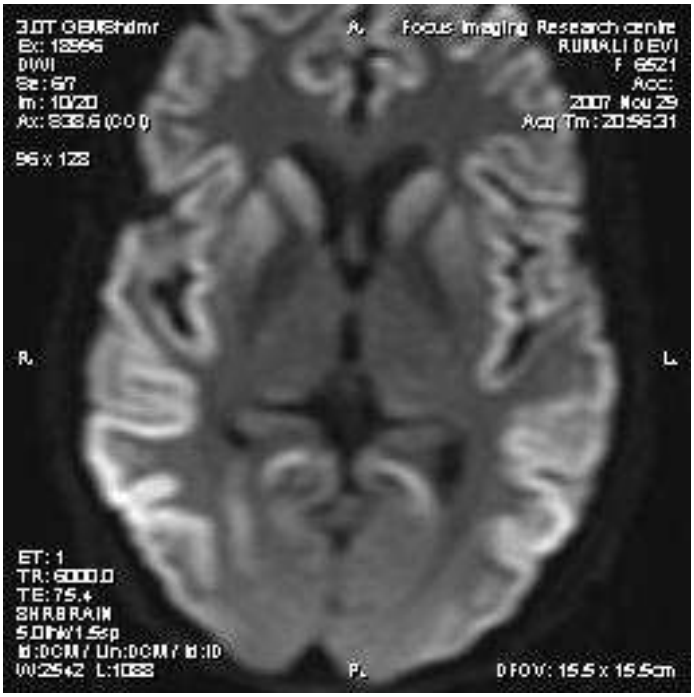
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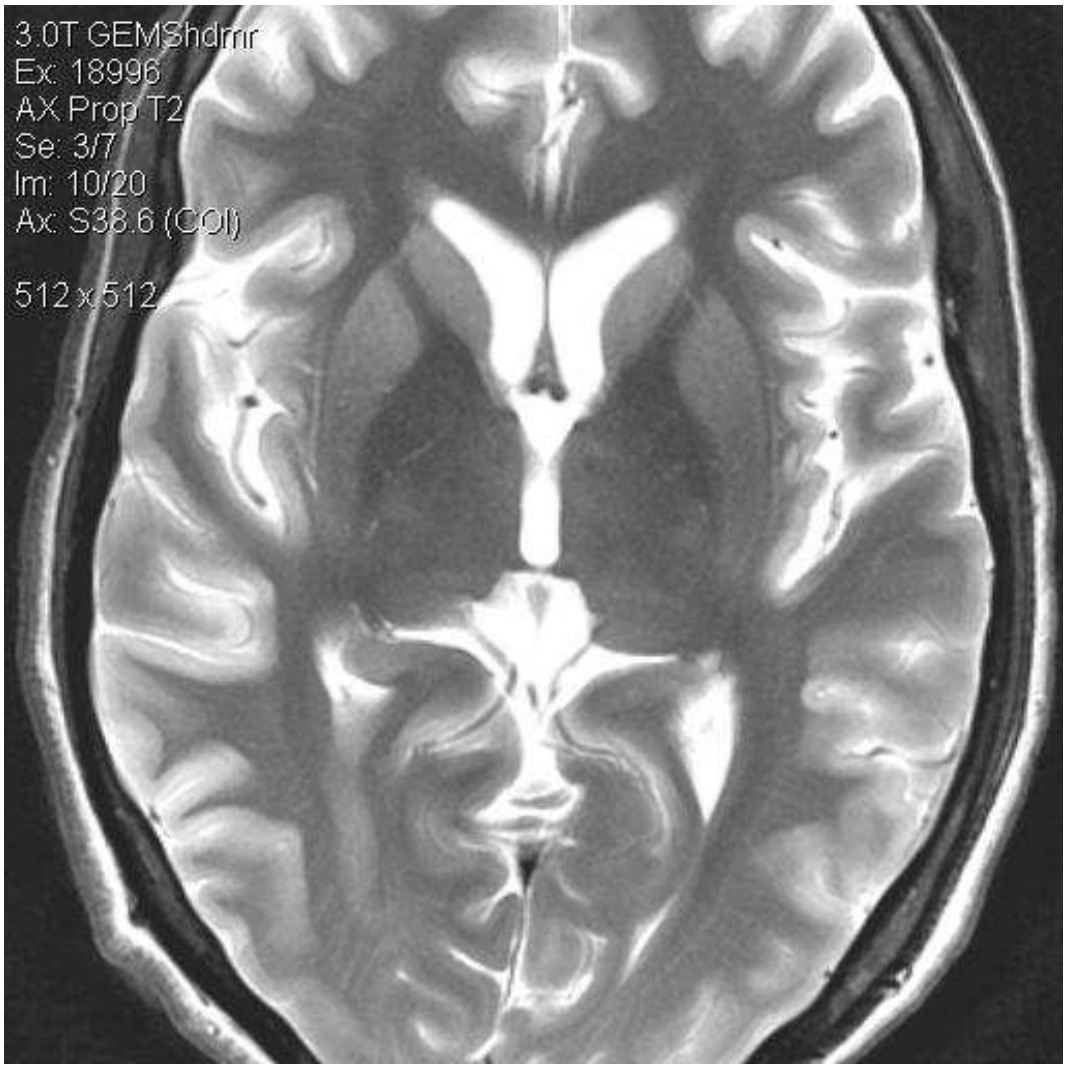
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chest were normal. Scalp EEG findings show diffuse slowing of background activity of delta range (Fig. 1). MRI brain revealed subtle hyperintensity in bilateral caudate nuclei and putamen on T2WI (Fig. 2.). This was confirmed on fluid attenuated inversion recovery (FLAIR) sequences (Fig. 3). Diffusion weighted images highlighted bilateral symmetric hyperintense signals in the caudate, putamen with similar hyperintense regions in the cortex seen as cortical ribbonning (Fig 4).

DISCUSSION

According to the WHO criteria for diagnosis of sporadic and iatrogenic CJD, our patient had possible CJD. In accordance to these criteria, identification of 14-3-3 in CSF is an accurate test to detect sporadic CJD. This however could not be carried out due to lack of availability. Clinical criteria (history, EEG and CSF analysis) in cases of CJD are not 100% accurate as put forth by Zerr et al[2], which is why so much interest has been generated in MR imaging as a means to further improve diagnostic accuracy noninvasively. The characteristic histopathologic features of CJD are spongiform degeneration, astrocytic gliosis and neuronal loss associated with deposition of prion protein[3]. The morphologic diagnostic feature is spongiform degeneration of grey matter, which is characterized by individual and clustered vacuoles in neuronal and glial processes. The lesions are widespread in the cerebral cortex, striatum, thalami and grey matter structures in the brain stem[3]. These histopathologic features correspond to MR findings. Earlier used only to exclude other illnesses, MR images describe characteristic, usually symmetrical, hyperintensity of the caudate head and putamen in 67-79% of sCJD cases with varying specificity by Finkenstaedt et al[4]. These basal ganglionic changes have been shown on T2WI, fluid attenuated inversion recovery (FLAIR) and diffusion weighted images (DWI). Shiga et al noted that diffusion weighted imaging is considerably more sensitive (92%) to these changes than FLAIR (41-59%) and T2W (36-50%)[5]. Cortical restriction on DWI was found to persist for several weeks potentially distinguishing the changes from infarction. In addition, in the early stages asymmetric changes are more prevalent than previously thought and this has important implications in interpretation of MR images performed early in cases of rapidly progressive dementia[5,6]. MR also plays an important role in the diagnosis of variant CJD, Variant CJD is characterized by younger age at time of onset, longer duration of disease and a clinical picture of nonspecific sensory and psychiatric symptoms leading to late diagnosis. EEG and CSF 14-3-3 has low specificity in vCJD than sCJD[7]. Here MR shows a characteristic distribution of symmetric hyperintensity in the pulvinar of thalamus best appreciated on axial image. The degree of hyperintensity is greater than anterior putamen and is known as 'Pulvinar Sign' of vCJD. Additional involvement of dorsomedial thalamic nucleus results in double hockey-stick appearance along with pulvinar changes. The other notable MRI finding is periaqueductal grey high signal without any cerebral atrophy[8].

From the imaging perspective the MRI plays an important role not only in the diagnosis of CJD but also in excluding the other possible causes of rapidly progressive dementia and diffusion weighted imaging offers a useful way as an early diagnostic marker of CJD

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