Abstract:
Creutzfeldt-Jakob Disease (CJD) is a rare transmissible spongiform encephalopathy caused by proteineous infectious particle called PrP(prion). Among subtypes CJD most common is sporadic CJD(s CJD) and less common is variant CJD(v CJD). MRI is indispensable in making diagnosis and ruling out other conditions. Bilateral symmetrical abnormal high signal intensity in the posterior thalamus is the pulvinar sign and in dorsomedial thalamic nuclei pulvinar is the hockey-stick sign. In the appropriate clinical context the MRI identification of pulvinar hockey stick sign is a useful non-invasive test for the diagnosis of vCJD ( upto 90% sensitivity) and can avoid tonsillar biopsy. The pulvinar sign was originally defined simply as hyperintensity of the pulvinar. In young patients, the normal basal ganglia are relatively hyperintense compared to old and rarely in sCJD patients can have pulvinar sign and this could be mistaken for vCJD. Hence the pulvinar sign is redefined as hyperintensity of the pulvinar relative to the signal intensity of the anterior putamen. Hyperintensity of all deep grey-matter nuclei, with the pulvinar less bright than the anterior putamen and caudate is a feature of sCJD. Here we describe a case of sCJD with pulvinar hockey stick sign mimicking vCJD. Our patient had pulvinar hyperintensity less bright than putamen, cortical ribboning and age of onset in fifth decade which were typically suggestive of sCJD. So far vCJD has not been reported in our country.
Keyword : CJD, Pulvinar sign, hockey stick sign
INTRODUCTION:
Creutzfeldt-Jakob Disease (CJD) is a rare transmissible spongiform encephalopathy caused by proteinaceous infectious particle called PrP (prion). A number of CJD subtypes are recognized. The most common is sporadic CJD (sCJD). Clinically the disease is characterized by a rapidly-progressive dementia culminating in an akinetic mute state. Neurological features seen during the illness reflect widespread neuronal damage, and include myoclonus, cerebellar ataxia, pyramidal signs, extrapyramidal signs and cortical blindness. Death ensues usually within about 5 months of symptom onset. There is no known effective treatment. Definitive diagnosis can be made only with autopsy and brain biopsy. So WHO proposed a criteria for diagnosing possible and probable CJD. MRI is helpful to rule out other differential diagnosis as well as to support diagnosis of CJD.

CASE REPORT:
58 year old female, house wife with no previous co-morbid illness came with history of acute onset of memory decline progressing rapidly over 3 months. Patient was apparently normal 3 months back, and of daily living like washing clothes, cooking for family members, cleaning the house and taking care of all house hold needs. Initially patient’s relatives noticed that she had forgetfulness and repeatedly asked same questions over and over again. She started misplacing objects, couldn’t cook properly and had difficulty in money handling. Patient also had recurrent falls and unsteadiness of gait. Her symptoms progressed rapidly over next 2 months to present state where she is socially withdrawn, apathetic, could not identify relatives and also had way finding difficulty. Later she also developed frequent sudden jerky movement of limbs. On examination patient was conscious but had poor attention and detailed lobar functions could not be done. Her MMSE was only 7. She was in an akinetic mute state. She could move all 4 limbs and had no pyramidal or extra pyramidal signs. She had an ataxic gait and stimulus sensitive myoclonus to sound. Since patient had myoclonus and rapidly progressing dementia CJD was suspected. Her routine blood investigations, thyroid profile, liver function tests were normal. Electroencephalogram showed periodic sharp wave complexes at 1 per second. CSF analysis for 14-3-3 could not be done due to financial constraints. Diagnosis of probable CJD was made with help of magnetic resonance imaging (MRI).

DISCUSSION:
CJD is a transmissible spongiform encephalopathies that affects many different animal species and are characterized by a progressive fatal neurologic course and the presence of spongiform change, neuronal loss, astrocytosis, and deposition of partially protease-resistant prion protein in brain. This abnormal protein is thought to be the infective agent and is termed a “prion”. A number of CJD subtypes are recognized.
The most common is sporadic CJD (sCJD), which is found worldwide and has an incidence of about one per million annually. In 1996, a new clinicopathologically distinct form was described as variant CJD (vCJD) and most cases have been reported from the United Kingdom. MRI is indispensable in making diagnosis and ruling out other conditions. Bilateral symmetrical abnormal high signal intensity in the posterior thalamus is the pulvinar sign and in dorsomedial thalamic nuclei and pulvinar is the “hockey-stick” sign. Characteristic findings in sCJD on MRI are best seen on diffusion-weighted images (DWI) and to a lesser extent on fluid-attenuated inversion recovery (FLAIR) images and are 91% sensitive and 95% specific for the diagnosis of sCJD. These findings include hyperintense signals within the cortical ribbon, basal ganglia, and thalamus. In sCJD, cortical ribboning is the most common MRI finding, followed by basal ganglia hyperintensities. MRI features of vCJD were medial thalamic and periaqueductal grey matter high signal, and the notable absence of cerebral atrophy. Present indications are that FLAIR sequences are most likely to show the abnormality. The pulvinar sign was originally defined simply as hyperintensity of the pulvinar. In young patients, the normal basal ganglia are relatively hyperintense compared to old and rarely in sCJD patients can have pulvinar sign and this could be mistaken for vCJD. Hence the pulvinar sign is redefined as hyperintensity of the pulvinar relative to the signal intensity of the anterior (4)putamen.

Here we describe a case of sCJD with pulvinar/hockey stick sign mimicking vCJD. Hyperintensity of all deep gray-matter nuclei, with the pulvinar less bright than the anterior putamen and caudate is a feature of sCJD which is useful to differentiate it from vCJD. Our patient had a similar picture. In addition to pulvinar sign, lack of cerebral atrophy and younger onset favours vCJD. Further studies are needed to assess the radiologic-neuropathologic correlates in vCJD and sCJD.

CONCLUSION:
In the appropriate clinical context the MRI identification of pulvinar sign is a useful non-invasive test for the diagnosis of vCJD and can avoid tonsillar biopsy. But it can rarely occur in sCJD like in our case and it should not be misdiagnosed as vCJD.

REFERENCES: