

MRI in Osmotic Demyelination: The “Mexican Hat” Sign

O Islam, G Dhillon

Citation

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Abstract

Osmotic demyelination syndrome (ODS) is often both a difficult clinical and radiological diagnosis. This article documents classic imaging features of OSD with magnetic resonance imaging examination. Features of diffusion weighted imaging in our case of OSD is also presented.

CASE DISCUSSION

A 58-year-old male had been recently discharged from a Florida intensive care unit following a six day stay for treatment of hyponatremia, jaundice, and abnormal liver function tests. Four days later he presented to the local emergency department with agitation, delirium and altered level of consciousness. Physical examination demonstrated a jaundiced, afebrile, normotensive patient without cardiovascular, respiratory, abdominal or neurological abnormalities. Initial blood work showed a normal complete blood count and electrolytes with minor elevation of total bilirubin and alkaline phosphatase. Within 48 hours he developed acute oropharyngeal dysphagia. Unenhanced computed tomography (CT) examination revealed focal hypoattenuation in the mid and dorsal pons. [Figure 1] A follow-up magnetic resonance imaging (MRI) study demonstrated abnormal low T1 and high T2 mexican hat shaped signal within the central pons with sparing of the corticospinal tracts ventrolaterally. [Figure 1] Abnormal increased T2 signal was also demonstrated in the thalami and putamen bilaterally. [Figure 1] Restricted diffusion was noted in these corresponding regions with diffusion weighted imaging (DWI). [Figure 2] Clinical and radiological findings were compatible with central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM).

Figure 1

Figure 1: A. Unenhanced CT demonstrates a large central hypoattenuating focus in the central pons. B,C. Corresponding axial T1 & T2 weighted MRI sequences demonstrate a mexican hat shaped signal abnormality in the basal pons with sparing of the tegmental and ventrolateral tissues (CPM). D. Coronal FLAIR sequence demonstrates increased signal in the hippocampi, lateral putamen and thalami bilaterally (EPM).

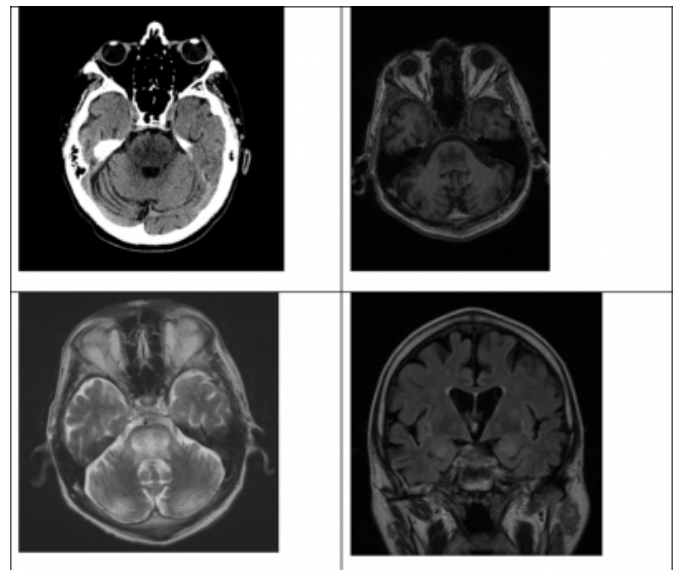
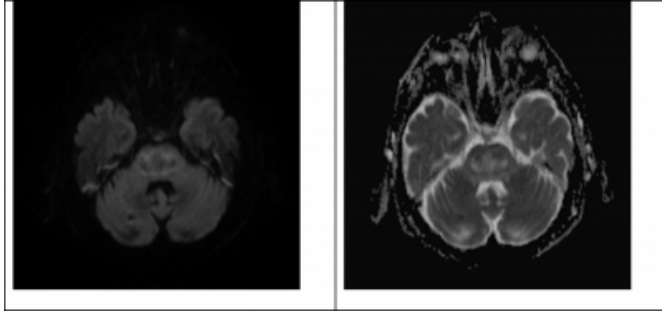


Figure 2

Figure 2: A&B The Mexican Hat Sign. A. DWI demonstrates low signal intensity in the basal pons representing increased water diffusivity with a faint surrounding rim of higher DWI signal representing infarction. B. Apparent diffusion coefficient map confirms the DWI findings of increased water diffusivity central to a surrounding thin rim of restricted water motion in the base of the pons.



DISCUSSION

CPM was first described in 1959 by Adams et al. as a symmetric, demyelinating focus most prominent in the central pons.¹ Although the original population studied was four chronic alcoholic and malnourished patients, CPM is a now a well recognized condition which can be seen in patients with cirrhotic liver disease, organ transplants, severe burns, Addison’s disease, and electrolyte disturbances. Rapid correction of prolonged hyponatremia is the most important risk factor for the development of CPM.² Associated symmetrical extrapontine lesions are well documented.³ The most common locations include the basal ganglia, midbrain, thalami, and cerebellum. Classic clinical symptoms associated with CPM include seizures, pseudobulbar palsies, spastic quadriparesis, and altered level of consciousness leading to coma or death without medical intervention. Less commonly, CPM may be suspected with only mild neurological symptoms. The term “osmotic demyelination syndrome” (ODS) more appropriately describes this rare condition.

The pathogenesis of ODS relates to osmotic injury to the endothelium resulting in the release of myelinotoxic factors and/or the production of edema.^{2,4} Post-mortem histologic examinations demonstrate symmetric demyelination of the basis pontis spreading centrifugally from the median raphe with relative sparing of neurons, axis cylinders and blood vessels, except in the center of the lesion where the disease process is most intense.^{1,3,5} The number of oligodendrocytes is notably decreased with associated myelin-laden phagocytoses and mild astrocytic gliosis. An inflammatory

reaction is conspicuously absent differentiating CPM from pontine infarction and inflammatory demyelinating diseases.⁶ The classic description of CPM notes sparing of the tegmentum and corticospinal tracts (CSTs).³ The pathogenesis of this selective pattern of demyelination is poorly understood and not present in all cases.

MRI is the imaging modality of choice in evaluating ODS. Well described findings include focal symmetric, trident or mexican hat shaped, high signal in the basal pons on T2-weighted and fluid attenuation inversion recovery (FLAIR) sequences with corresponding decreased T1-weighted signal.^{3,7} Demyelination sparing of the CSTs results in bilateral ventral paramedian foci of normal signal providing the mexican hat sign. Associated symmetric signal abnormality may or may not also be present in the aforementioned extrapontine locations. It is not uncommon for MRI to be negative in patients with suspected ODS up to 10 days after the onset of clinical symptoms. Therefore, the diagnosis of ODS cannot be ruled out in the setting of normal MRI. DWI is a relatively new MRI technique sensitive to the motion of water. Recent authors have documented restricted diffusion associated with the lesions in ODS as well as during the early clinical phase when conventional MRI imaging may be negative.^{7,8,9} Ruzek et al. demonstrated restricted diffusion within 24 hours of onset of tetraplegia as the first imaging manifestation of ODS in the setting of otherwise normal conventional MRI sequences.⁸ DWI and apparent diffusion coefficient (ADC) values are very useful in distinguishing conditions which may be confused with ODS e.g. brain tumors, encephalomyelitis, multiple sclerosis.¹⁰

CONCLUSION

ODS can occur in a variety of clinical settings with variable and often severe neurological symptoms. Early diagnosis with DWI may demonstrate changes of ODS sooner in the course of the disease allowing for earlier medical intervention and possibly a more favorable clinical outcome. DWI should be performed in all suspected cases as identification of the “Mexican Hat” sign can serve as an early imaging marker of ODS.

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Author Information

Omar Islam, MD, FRCP

Department of Diagnostic Imaging Kingston General Hospital Queen’s University Kingston, Ontario Canada

Gurmohan Dhillon

Department of Diagnostic Imaging Kingston General Hospital Queen’s University Kingston, Ontario Canada