The Journal of International Medical Sciences Academy
2023; (July-September); Vol 36; No. 3

CASE REPORT

Cortical Ribbon Sign in Osmotic Demyelination Syndrome

P. R. Sowmini, K. Muralidharan, Kondapally Swamy, S. Kamatchi, S. Sakthi Velayutham, K. Malcolm Jeyaraj, R. Viveka Saravanan, K. Mugundhan

Abstract

Osmotic demyelination syndrome (ODS) is a demyelinating disorder due to a marked change in osmolarity. Common site of involvement is central pons. The extrapontine lesions are seen in the neostriatum, thalamus, internal capsule, midbrain, and cerebellum. Cortical involvement is very rare in ODS, accounting for only 5-10% and this case report highlights the rarity of radiological involvement involving cerebral cortex in a cortical ribboning pattern which occurred even before the involvement of pons.

Department of Neurology, Govt. Stanley Medical College, Chennai - 600001, Tamilnadu, India

Corresponding Author: Dr. K. Mugundhan, Head of the Department of Neurology, Govt. Stanley Medical College, Chennai – 600 001, Tamilnadu, India Email: mugundhan69@gmail.com

Received: 22th July 2022

Accepted: 01th February 2023

How to Cite this Article: Sowmini PR, Muralidharan K, Swamy K, Kamatchi S, Velayutham SS, Jeyaraj KM, Saravanan RV, Mugundhan K. Cortical Ribbon Sign In Osmotic Demyelination Syndrome. J Int Med Sci Acad 2023;36(3):325-326.

Access this article online : www.imsaonline.com

Introduction

Osmotic demyelination syndrome (ODS) is a demyelinating disorder due to a marked change in osmolarity. Common site of involvement is central pons. The extrapontine lesions are seen in the neostriatum, thalamus, internal capsule, midbrain, and cerebellum. Cortical involvement is very rare in ODS, accounting for only 5-10% and this case report highlights the rarity of radiological involvement involving cerebral cortex in a cortical ribboning pattern which occurred even before the involvement of pons.

Case Report

50-year lady presented with weakness of all four limbs and altered sensorium of three weeks duration. She was asymptomatic until 6 weeks back when she developed recurrent vomiting and headache when she was found to have borderline elevated BP, for which she was advised a low-salt diet. She was treated with IV fluids for persistent vomiting. Three weeks later, she developed one episode of generalized seizures, for which she was hospitalized. The next day, she developed weakness in all four limbs and became hyporesponsive thereafter.

On admission, she was stuporous. Rigidity and paucity of movements were noted in all four limbs. One week later, she became conscious but was aphasic and apathetic. A diagnosis of akinetic rigid syndrome was made.

Her routine blood investigations including blood sugar and electrolytes were normal. MRI Brain showed bilateral symmetrical T2/FLAIR hyperintensities with diffusion restriction in putamen, caudate nucleus (Fig-1), bilateral cerebellar hemisphere, superior frontal cortex, bilateral medial temporal lobe, left insular cortex, bilateral ventrolateral thalami and bilateral posterior temporal, parietal and perirolandic cortices (Fig. 2). Cortical ribboning pattern was noticed. Brain stem showed normal signal in all the sequences.

Keeping the pattern of cortical ribbon, differential diagnoses being considered were extrapontine myelinolysis, hypoxic-ischemic encephalopathy, hypoglycemia, hyperammonemia, autoimmune-encephalitis, paraneoplastic encephalitis, infectious encephalitis and Creutzfeldt–Jakob disease (CJD)[2]. There was no history of hypoxia and her blood sugar was normal. Diagnosis of CJD was least considered as it was not fitting into clinical criteria. She was treated with IV Methylprednisolone in view of autoimmune encephalitis.

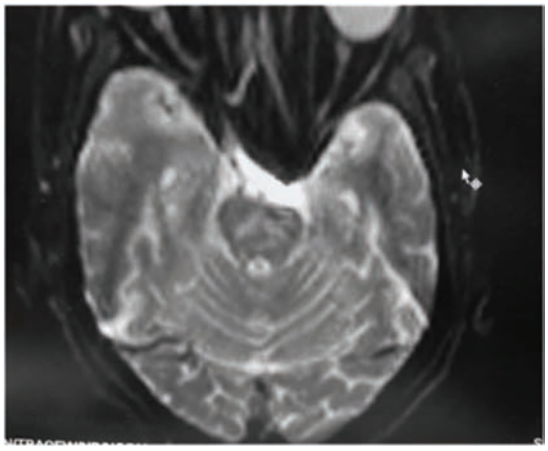

Figure 1. MRI Brain (FLAIR) showing hyperintensity in bilateral putamen and caudate nucleus

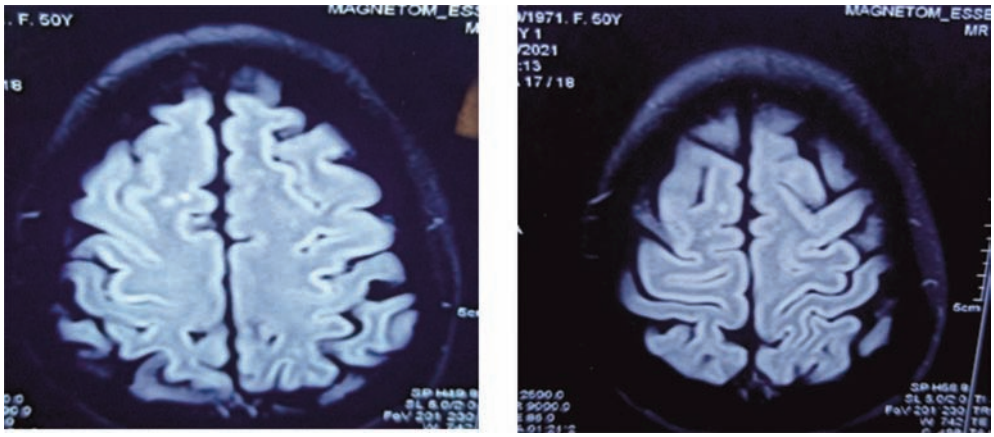

Figure 2 MRI Brain (FLAIR) showing cortical ribbon pattern in bilateral cortical areas

Serum ammonia level was normal (40mcg/dL). Workup for infectious encephalitis, autoimmune, and paraneoplastic encephalitis did not yield anything. There were no spikes sharp waves or periodic complexes in EEG. A repeat MRI done one week later showed T2 Hyperintensity in the central pons, suggestive of Osmotic demyelination syndrome (Fig 3). She was treated with supportive measures and three weeks later she was discharged with normal sensorium and she went walking with support.

The primary risk factor associated with Osmotic Demyelination Syndrome (ODS) is the rapid correction of hyponatremia. Additional risk factors include malnutrition, chronic alcoholism, primary adrenal insufficiency, prolonged use of diuretics, hypokalemia, hyperglycemia, fluid resuscitation, hemodialysis, and liver transplant [3]. However, in this particular case, there was no evidence of hyponatremia or a history of alcoholism, liver disease, severe electrolyte imbalances, or significant medical conditions. The excessive administration of intravenous fluids for vomiting, coupled with a low salt diet, likely contributed to the development of osmotic demyelination.

MRI findings in Osmotic Demyelination Syndrome (ODS) typically show abnormal hyperintensity affecting both the pons and extrapontine regions, such as the basal ganglia, thalami, and cerebral white matter. Extrapontine Myelinolysis (EPM) commonly involves the caudate nucleus, lentiform nucleus, and thalami, while atypical sites include grey matter, white matter, corpus callosum, splenium, cerebellum, hippocampus, and external capsule. However, in this case, involvement of the basal ganglia, thalamus, and cerebral cortex preceded that of the pons [3]. This temporal progression supports the hypothesis proposed by Babanrao SA et al., suggesting that EPM may precede pontine myelinolysis. Additionally, the timing of lesion appearance on MRI may be delayed, and repeat imaging at 10–14 days may reveal lesions not initially detected, as observed in this case [4].

Conclusion

In half of the cases of Osmotic Demyelination Syndrome (ODS), symptoms present with only pontine lesions, while 30% exhibit both pontine and extrapontine lesions. Isolated extrapontine lesions are less common, occurring in just 20% of ODS cases, which complicates diagnosis [5]. This case underscores the uncommon occurrence of cortical ribbon signs in ODS, found in only 5-10% of cases. It emphasizes the importance of considering Extrapontine Myelinolysis (EPM) as a potential diagnosis when patients are at risk of ODS and their imaging indicates neurological damage in various brain regions, as ODS-related damage can extend beyond the pons [5].

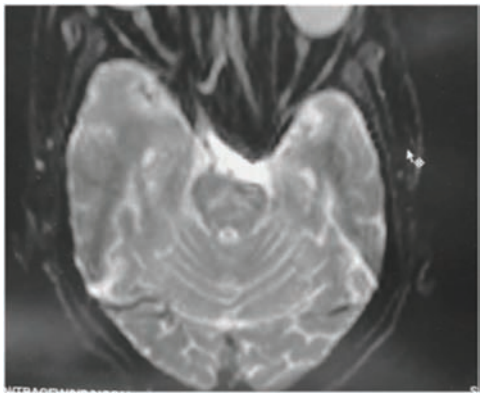

Figure 3. Repeat MRI Brain (T2) showing hyperintensity in the central pons

Conflict of Interest:	All authors declare no COI
Ethics:	There is no ethical violation as it is based on voluntary anonymous interviews
Funding:	No external funding
Guarantor:	Dr. K. Mugundha, will act as guarantor of this article on behalf of all co-authors.

Reference

1. *Chua GC, Sitoh YY, Lim CC, Chua HC, Ng PY. MRI Findings in Osmotic Myelinolysis. ClinRadiol. 2002;57:800*
2. *Miller GM, Baker HL, Okazaki H, Whisnant JP. Central pontine myelinolysis and its imitators: MR findings. Radiology. 1988;168:795–802*
3. *Calakos N, Fischbein N, Baringer JR, Jay C. Cortical MRI findings associated with rapid correction of hyponatremia. Neurology. 2000;55:1048–1051*
4. *Babanrao SA, Prahladan A, Kalidos K, Ramachandran K. Osmotic myelinolysis: Does extrapontine myelinolysis precede central pontine myelinolysis? Report of two cases and review of literature. Indian J Radiol Imaging. 2015;25(2):177-83.*
5. *Y Tatewaki, K Kato, Y Tanabe, S Takahashi. MRI findings of corticosubcortical lesions in osmotic myelinolysis: report of two cases. Br J Radiology. 2012;85(1012):e087-e090*