

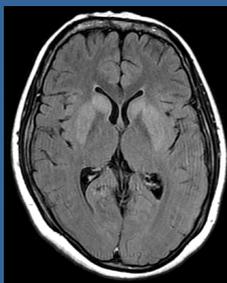
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## Background

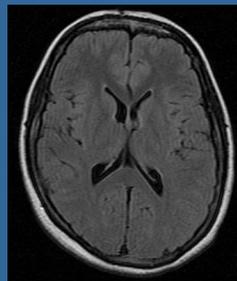
- Central Pontine Myelinolysis (CPM)/ Extrapontine Myelinolysis (EPM) is usually associated with rapid correction of hyponatremia or rarely hypernatremia. Alcoholism and chronic nutritional deficiency are considered other causes of CPM/EPM.
- CPM/EPM is pathologically defined as areas of symmetric demyelination in the center of basis pontine, as well as extrapontine locations such as basal ganglia, thalamus and cerebellum. EPM is found in ~10% of patients in CPM but also can be found in isolation.
- Various pathophysiological mechanisms have been postulated to be involved in CPM/EPM including oligodendroglial dehydration resulting in myelinolysis and necrosis, oligodendroglial apoptosis secondary to metabolic stress imposed on the cells that attempt to prevent osmotic dehydration, and astroglial response to osmotic myelinolysis leading to accumulation of myelinotoxic substances.
- Various reports on the prognosis of CPM/EPM exists, ranging from a poor prognosis with high mortality rate to more favorable outcome in recent reports.
- The best therapeutic management after development of symptoms of CPM/EPM is unknown and most strategies are limited to case reports. There are a few reported cases in the literature of IVIG administration in the literature of IVIG administration in CPM/EPM, all of them associated with favorable outcome.
- We report 2 cases of EPM with significant improvement in their clinical signs and symptoms after IVIG therapy.

## Case 1

- 20 year-old female who presented to the emergency room with 2 weeks history of nausea and vomiting. Her serum sodium increased from 100 to 126 mmol/L within 24 hours of admission.
- Nine days after initial presentation, she developed dysarthria, dysphagia and spasms in the hands. She was also reported by her family to be more somnolent during the day and sometimes laughing without reason.
- Brain MRI revealed T2 hyperintensities in the caudate and putamen bilaterally consistent with EPM.



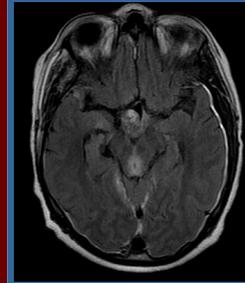
Case 1: Hyperintensities in caudate and putamen bilaterally (T2 FLAIR)



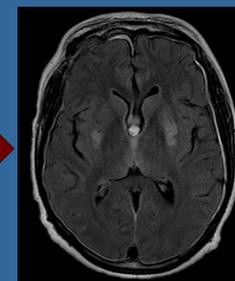
Case 1: Resolution of the hyperintensities in caudate and putamen 8 months after presentation. (T2 FLAIR)

## Case 2

- 18 year-old male with craniopharyngioma, and secondary panhypopituitarism who was admitted to the hospital with dehydration and hypernatremia.
- His serum sodium changed from 167 to 157 mmol/L within the first 12 hours. Three days later he became weak in all the extremities, along with dysarthria and lethargy necessitating intubation.
- Brain MRI revealed increased T2 FLAIR signals in caudate, putamen, thalami, periaqueductal region and cerebellar hemispheres consistent with EPM.



Case 2: Hyperintensities in periaqueductal region (T2 FLAIR)



Case 2: Hyperintensities in caudate and putamen (T2 FLAIR)

## Results

- Both of the cases received a 5-day course of IVIG (0.4 g/kg/day).
- The first case demonstrated significant improvement of her symptoms one day after completion of IVIG. Her dysphagia, dysarthria and strength almost completely returned to baseline during the next few weeks.
- Our second case also showed improvement in his mental status and increased strength in extremities 2 days after completion of therapy. The clinical improvement continued during the next few weeks until he reached his baseline.

## Conclusion

- CPM/EPM is a devastating demyelinating condition with little therapeutic intervention after symptom onset.
- Immunological mechanisms may be involved in the pathogenesis of CPM/EPM, hence immune modulating treatments such as IVIG maybe a promising therapeutic option. Further studies and observations are required to determine the role of immune modulating treatment in CPM/EPM.

*Authors have nothing to disclose*

## Selected References

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