Cavitating Osmotic Demyelination with an Unusual Pontine Appearance

Dear Editor,

Osmotic demyelination syndrome (ODS), classically associated with rapid correction of hyponatremia, can lead to permanent sequelae.[1] Increasingly, risk factors other than hyponatremia and its rapid correction have been identified, and these include severe hypernatremia, hyperglycemia, hypokalemia, chronic alcoholism, chronic kidney disease, hepatocellular dysfunction, hypophosphatemia, etc.[2] The classical radiological feature in ODS is a triangular signal change in the central pons, with sparing of pontine tegmentum and ventrolateral pons,[1] which has been variously named as "Mercedes-Benz sign," "trident" sign, "piglet sign," and "butterfly" sign.[3] We report an unusual case of severe ODS with pontine cavitation, involving non-classical brain areas, and good improvement, in a patient with a combination of severe hypernatremia and hyperglycemia.

A 31-year-old woman presented to the emergency services with altered mental status for three days. She had a history of intrauterine death in the seventh month of pregnancy, just before symptom onset. The offspring was delivered vaginally at an outside center. She had not undergone any antenatal care during this pregnancy. She did not have any history of alcohol use or liver disease, or prior comorbidities. At presentation, she was intubated in view of poor sensorium (Glasgow coma scale 8/15). Her blood pressure was 70/50 mm Hg, and her pulse rate was 124 per minute. She appeared dehydrated. No features of papilledema or meningeal irritation were observed. Fundus examination was normal. No focal neurological deficits were present, and deep tendon reflexes were elicited normally. Plantar response was flexor bilaterally. Her random blood sugar (891 mg/dL) and corrected serum sodium levels (171 mEql/L, normal 135-145 mEq/L) were highly elevated. Urine analysis showed the presence of ketonuria (5.5 mmol/L, normal <0.6 mmol/L). Her arterial blood gas showed metabolic acidosis. The renal function test was suggestive of pre-renal azotemia. Both sugar and sodium levels were normalized over the next 3-4 days, and she

recovered to normal sensorium. At this point, she was observed to have flaccid quadriparesis with power in upper and lower limbs being Medical Research Council (MRC) grade 2/5. Magnetic resonance imaging (MRI) brain showed multiple T2-weighted and Fluid Attenuated Inversion Recovery (FLAIR) hyperintensities, predominantly affecting the belly of pons, bilateral putamen, thalami, and external capsules. Diffuse hyperintensities were seen in both cerebellar hemispheres, along with folial enhancement [Figure 1]. Cerebrospinal fluid (CSF) analysis showed acellular CSF, with normal glucose (69.9 mg/dL, corresponding blood glucose 100 mg/dL) and elevated protein (76 mg/dL, normal 15–45 mg/dL). CSF analysis for infective workup (Gram stain, bacterial culture, fungal smear, smear for acid-fast bacilli, VDRL) and malignant cytology was non-revelatory. The radiological findings were suggestive of severe and extensive ODS with both central and extrapontine myelinolysis. The patient gradually improved to independent ambulation over the next three months, with mild spasticity of limbs (modified Ashworth scale 1). Repeat MRI after four months displayed partial resolution, but the development of cavitation was noted in the pons, giving rise to an appearance of swimming goggles [Figure 2].

ODS is associated with rapid electrolyte-induced osmotic shifts classically involving serum sodium but has also been reported in ketotic and non-ketotic hyperglycemia.[4] Occasionally, as in the present case, both may co-exist.[5-7] Several atypical radiological features were noted in our patient, including the presence of thalamic hyperintensities, involvement of the external capsule and cerebellar white matter, along with folial enhancement. Additionally, the double-barreled appearance of the pons at follow-up was reminiscent of "swimming goggles," which has not been reported thus far. The presence of a "double-hit" in the form of a combination of severe hyperglycemia and hypernatremia may have contributed to the severity of the radiological findings. Another possibility considered in the setting of pregnancy was posterior fossa posterior reversible encephalopathy syndrome (PRES). The presence of extensive edema in the posterior fossa may have supported this consideration. However, the clinical composite at presentation of a normotensive patient without headache and/or visual complaints, and the setting of severe electrolyte disturbance made PRES highly unlikely.

This case demonstrates that severe hyperglycemia and hypernatremia can lead to extensive radiological osmotic demyelination, which may reverse over time. We also report the novel radiological pontine "goggles" appearance in association with ODS, with the caveat that the utility of this radiological sign needs to be further assessed in larger radiological cohorts of patients, as cavitation may possibly also be seen in association with pontine infarction or severe demyelination.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients

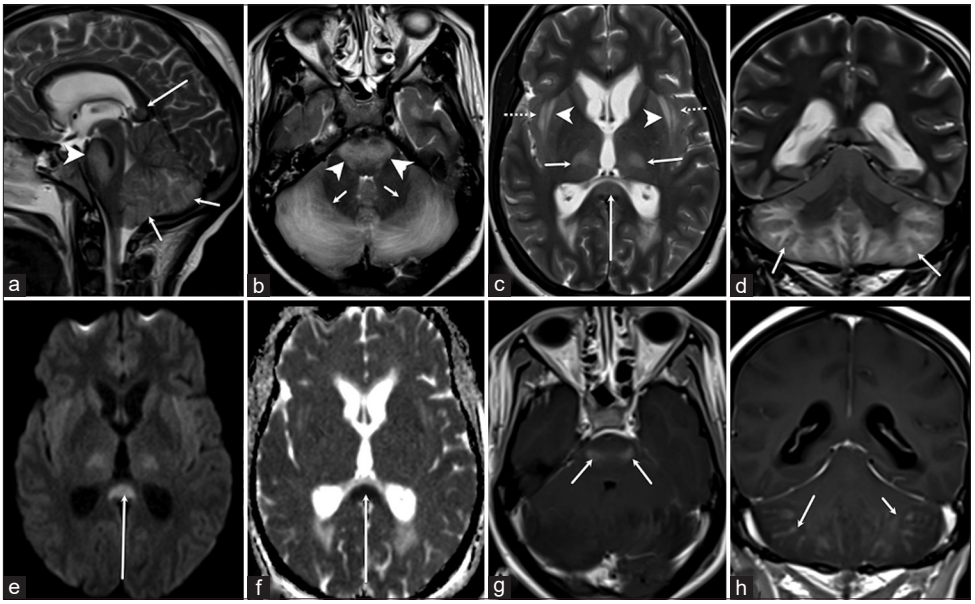

Figure 1: Sagittal (a), axial (b and c), and coronal (d) T2-WIs show hyperintensity in the center pontine tegmentum (arrowheads in a and b), inferior vermis and cerebellum (short arrow in a, b and d), central splenium of corpus callous (long arrow in a and c), bilateral symmetrical posterolateral thalami (short arrows in c), external capsules (arrowheads in c) and extreme capsules (broken arrow in c). The splenial lesion is bright on the diffusion-trace image (e) and dark on the Apparent Diffusion Coefficient (ADC) map (f), suggesting restricted diffusion; the rest of the lesions show facilitated diffusion. Following gadolinium administration, axial (g) and coronal (h) T1-WIs show enhancement in the central pons (arrows in g) and cerebellar white matter (arrows in h)

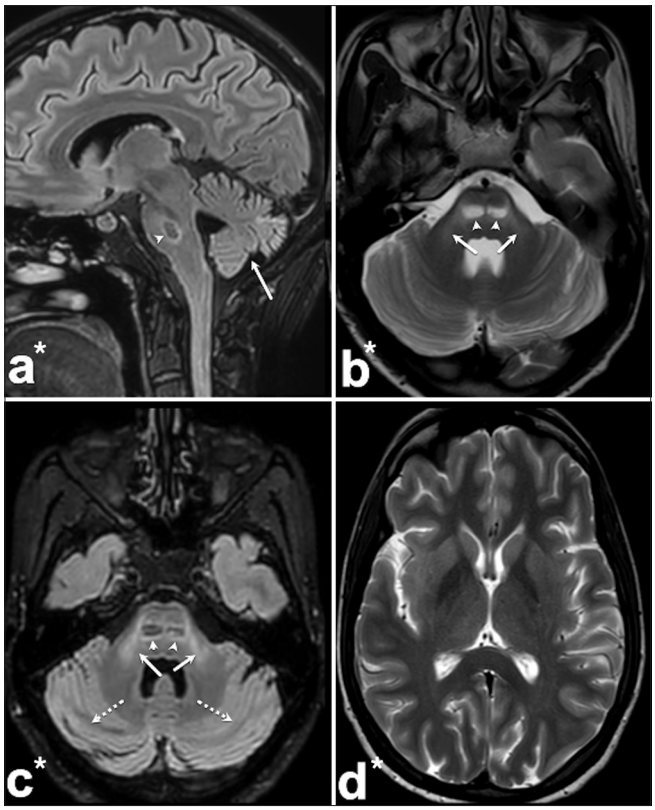

Figure 2: Sagittal FLAIR (a*) images show mild atrophy of the inferior vermis (arrow) and focal hypointensity with a hyperintense rim in central pons (arrowhead) reminiscent of "goggles." Axial T2-WI (b*) at the level of pons shows central pontine hyperintensity, which suppresses in axial FLAIR image (c*), suggesting cavitation. Axial T2-WI (d*) at basal ganglia show complete resolution of thalamic, splenium, and external/extreme capsule signal abnormalities

understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Divyani Garg, Ayush Agarwal, Ajay Garg[1]

Departments of Neurology and [1]Neuroimaging and Interventional Neuroradiology, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence: Dr. Divyani Garg, Room 60, CN Center, AIIMS, New Delhi - 110 029, India.
E-mail: divyanig@gmail.com

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