The AAP California Chapter 2 sent out a call for abstracts on scholarly projects by pediatric residents and medicine-pediatric residents from CHLA, Harbor-UCLA, Kaiser LA, Loma Linda, UCLA and USC; and medical students from the Southern California counties of Kern, Los Angeles, Riverside, San Bernardino, San Luis Obispo, Santa Barbara and Ventura. The accepted abstracts were showcased at the 2022 Advances in Pediatrics Virtual Symposium. We are thrilled to share with you the authors and their abstract each month.

Clinical Case Report: SIADH as a presentation of Neuromyelitis Optica

Divya Patel, MD; Joyce Koh, MD; David Lee, MD; John Luttrell, MD (CHLA)

A 23-year-old female with history of anxiety presents with 1 month of epigastric pain, emesis, weight loss, and intractable hiccups. Electrolytes on admission were reassuring, and she was admitted for feeding intolerance and nutritional optimization. On hospital day 11, she developed syndrome of inappropriate antidiuretic hormone secretion (SIADH) with sodium of 125, and endorsed symptoms of headache, dizziness, and paresthesias in bilateral distal extremities. Sodium was corrected with hypertonic saline, fluid restriction, and sodium supplementation. MRI brain showed abnormal hyperintensity in the mamillary bodies, concerning for vitamin B12 deficiency, and she was started on high dose vitamin supplementation. Symptoms progressed despite vitamin repletion and physical exam was notable for horizontal nystagmus, dysmetria, diminished strength and sensation, and hyporeflexia in bilateral extremities. SIADH persisted and she developed signs of bulbar weakness, including dysarthria, dysphagia, drooling, and somnolence. Repeat brain MRI showed extension of white matter hyperintensity in the brainstem, now concerning for demyelinating process including central pontine myelinolysis from rapid overcorrection of hyponatremia, despite appropriate sodium management. Over the next 4 days she developed new findings of disconjugate gaze, left facial allodynia, urinary incontinence, and right sided extremity weakness. Given signs of spinal involvement, MRI brain and spine was completed. Imaging demonstrated further progression of white matter enhancement in the brainstem extending to the proximal cervical spinal cord. Per neuroradiology team, these imaging findings were suggestive of a primary demyelinating disease. Diagnostic testing with lumbar puncture was ultimately pursued. CSF was notable for mild pleocytosis of 20 WBC, normal opening pressure, and negative infectious studies. Decision was made to start patient on high dose steroid therapy given high suspicion for demyelinating disease while awaiting remainder of CSF studies. CSF studies returned positive for oligoclonal bands in CSF (negative in serum) and positive Aquaporin 4 (AQP4) antibodies, findings consistent with a diagnosis of neuromyelitis optica. She underwent seven rounds of plasma exchange and showed improvement in symptoms. She was initiated on rituximab and transitioned to inpatient rehabilitation.

<u>Discussion:</u>

1. This patient's constellation of evolving neurologic exam findings correlated with her suspected CNS lesions. She showed symptoms of area postrema syndrome (hiccups, nausea,

vomiting), acute myelitis (limb weakness, bladder incontinence), and cranial nerve palsy (nystagmus, diplopia, and disconjugate gaze), all of which are features consistent with NMO.

2. Based on purely radiologic findings, an alternative diagnosis may have been made. It was important to follow the dynamic physical exam changes in the setting of trialed interventions, and to continue maintaining a broad differential when there were inconsistencies between the imaging reports and clinical course.

3. AQP4 is a water channel protein found in the hypothalamus, the region of the brain which regulates antidiuretic hormone release. AQP4 antibodies present in NMO may cause local inflammation in this area, leading to SIADH, as seen in this patient.

Poster Presenter:
Dr. Divya Patel
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Clinical Case Report: SIADH as a presentation of Neuromyelitis Optica

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Scroll to page 3 to view the poster.

The Hiccups and Downs of a Shifting Differential

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Case Presentation

- 23-year-old female with history of anxiety, panic attacks, and gastritis.
- Presents with 1 month of worsening epigastric pain, emesis, weight loss, and intractable hiccups.
- Admitted for feeding intolerance.

Physical Exam

Day 0	Soft abdomen with mild generalized tenderness. Neurologic exam normal.
Day 11	Develops headache, dizziness, facial allodynia, muscle cramps, and distal paresthesias. Exam with horizontal nystagmus, dysmetria, diminished strength, sensation, and symmetric hyporeflexia.
Day 21	Progressive somnolence and signs of bulbar weakness—dysarthria dysphagia, and drooling.
Day 25	New urinary incontinence and right-sided weakness.

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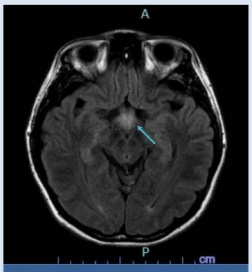
Hospital Course

On day 11 sodium drops to 125, labs consistent with **SIADH**.

Brain MRI with hyperintensity in the mamillary bodies suggestive of vitamin B1 deficiency.



MRI brain, FLAIR sequence

Vitamin B1 & **B12** are repleted. Sodium is gradually corrected. Despite interventions symptoms progress.

Brain and Spine MRI now show extension of hyperintense signal to the proximal cervical spinal cord, suggestive of inflammatory vs osmotic demyelination.

Homocysteine and **methylmalonic acid** levels return normal. **CSF** has mild pleocytosis (20 WBC), normal opening pressure, and negative infectious studies.

Day 35: CSF returns positive for oligoclonal bands and **Aquaporin 4 (AQP4) antibodies.**

Diagnosis

Neuromyelitis optica spectrum disorder (NMOSD)

- Antibody-mediated inflammatory disease of the CNS
- Characterized by **AQP4 antibodies**
- Affects optic nerves and spinal cord

Features of NMOSD in this patient

- **Area postrema syndrome** (hiccups, nausea, emesis)
- **Acute myelitis** (limb weakness, urinary incontinence)
- **Acute brainstem syndrome** (oculomotor dysfunction, cranial nerve palsies)
- **SIADH**

Take Home Points

- Maintain a broad differential when there are inconsistencies between the diagnostics and clinical course.
- Based on imaging alone, an alternative diagnosis may have been made.
- It was important to follow the evolving exam findings in the setting of trialed interventions.