A rare cause of acute severe hyponatraemia secondary to the syndrome of inappropriate anti-diuretic hormone (SIADH) secretion

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Introduction

Hyponatremia is the commonest electrolyte abnormality presenting to Medical Admissions and when acute, severe and symptomatic, is associated with high mortality. The syndrome of inappropriate antidiuretic hormone secretion (SIADH) is the most common underlying disorder in hospitalised patients with euvolaemic hyponatraemia and is a complication of many clinical conditions and drug therapies.

We discuss the cases of two patients presenting with acute severe hyponatraemia secondary to SIADH, admitted to the same hospital within a 2-week period.

The number of possible causes of SIADH is extensive and maybe difficult to determine despite thorough investigation. We present the chronological events including clinical history and biochemical results in these patients which highlight the importance of considering rarer causes for unexplained hyponatraemia in those with additional symptoms suggestive of underlying pathology

Background to cases

Patient 1 – 54 year old female

No significant past medical history, no regular medication

Presented 18th May:

- Severe abdominal pain
- Hypertension
- Serum sodium 132mmol/L

Deteriorated 21st May:

- Serum sodium 123mmol/L
- Seizure and 2222 call – transferred to ITU

Patient 2 – 27 year old female

No significant past medical history, no regular medication

Presented 2nd June:

- Increasing confusion, lethargy and abdominal pain
- Serum sodium 108mmol/L
- Seizure in ED – intubated and transferred to ITU

Analysis

	PATIENT 1	PATIENT 2
Serum sodium (mmol/L)	123	108
Creatinine (umol/L) (eGFR)	76	62
Liver function tests	Normal	Normal
Serum glucose mmol/L	Normal	6.4
Lipids	NA	↑ total cholesterol (7)
TSH (U/mL) +/- T3/4	0.67, fT4 21 pmol/l	1.9
9am cortisol (nmol/L)	480	872
Serum osmolarity (mOsm/kg)	255	251
Urine osmolarity (mOsm/kg)	614	582
Urine sodium (mEq/L)	195	194

	PATIENT 1	PATIENT 2
CXR	Normal	Normal
CT/MRI head	CT Head- normal	CT Head- normal
Further imaging/investigation	CT Abdo-pelvis- Normal	CT CAP- Bulky right adnexa and small cyst in the left adnexa

	PATIENT 1	PATIENT 2
Urinary porphobilinogen	1136umol/l (<10.7)	286.2 umol/l (<10.7)
Porphyrin Fractionation	91%	83%

Diagnosis: Acute intermittent porphyria (AIP)

Discussion

Acute Intermittent Porphyria (AIP) is an autosomal dominant condition that results from a partial deficiency in activity of porphobilinogen deaminase, the third enzyme in the pathway of haem synthesis. The deficiency leads to increased blood levels of porphobilinogen and decreased haem synthesis. Clinical manifestations classically include recurrent unexplained abdominal pain, peripheral neuropathy and neuropsychiatric symptoms. Hyponatraemia occurs in approximately 20% of cases of symptomatic AIP and is usually due to SIADH.

Both patients discussed were treated for acute severe hyponatraemia secondary to SIADH, with multiple boluses of 2.7% hypertonic saline and strict fluid restriction. Extensive imaging in each case failed to identify an underlying cause. The combination of unexplained SIADH, hypertension and abdominal pain raised the possibility of porphyria as a unifying diagnosis. It was no until they were commenced on haem arginate that immediate normalisation of sodium levels occurred.

The cases highlight that whilst a rare cause of hyponatraemia, in unexplained SIADH in young patients with additional symptomatology, a high index of suspicion is needed to accurately diagnose AIP. Early diagnosis, prompt treatment and avoidance of precipitating factors is essential for managing this sometimes fatal condition.

References

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BES2017 EP-042

Clinical biochemistry

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Poster presented at: