Lentiform fork sign in a uremic patient with a high anion gap metabolic acidosis with seizures: a case report from North West of Ireland

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Abstract

Background: Lentiform fork sign is a neuroradiological abnormality which is encountered in the clinical practice associated with uremic encephalopathy, dialysis disequilibrium syndrome and metabolic acidosis.

Case presentation: We describe here a case of this neuro-radiological abnormality which was encountered in a patient with uraemia and high anion gap metabolic acidosis who presented with generalised convulsion and later had some tremor in her hands. In our patient, there were few predisposing factors which might have possibly resulted in this abnormality chronic kidney disease, diabetes mellitus, and metabolic acidosis.

Conclusion: The Lentiform fork sign is a rare occurrence which can be related to a long list of toxic and metabolic causes but in conjunction with metabolic acidosis in chronic kidney disease patients, it can narrow down this list of alternate diagnosis.

Keywords: Lentiform fork sign, Uremic encephalopathy, Seizures, Anion gap, Status epilepticus

Background

Basal ganglia are metabolically highly active deep grey matter structures. There are various toxic and metabolic insults (uraemia, hypoxia, drug overdose, infections) that result in abnormalities in basal ganglia on neuroimaging [1–3]. The clinical features or uremic encephalopathy include tremors, chorea-athetoid movements, convulsions and coma in patients with renal diseases.

Lentiform fork sign is a neuroradiological abnormality which can be associated with uremic encephalopathy, dialysis disequilibrium syndrome and metabolic acidosis [3–5]. The sign is characterised by symmetrical hyperintensities in bilateral basal ganglia surrounded by hyperintense rim which delineates Lentiform nucleus [2, 3].

The underlying cause for which is postulated to be secondary to vasogenic edema.

We describe here a case of this neuro-radiological abnormality which was encountered in our patient with Uraemia and high anion gap metabolic acidosis who presented with a generalised convulsion and later had some tremor in her hands.

Case presentation

A 74-year-old lady with known CKD stage 3b was brought by ambulance to the emergency department after a first prolonged generalised tonic clonic seizure that lasted 90 minutes. On arrival she had a low Glasgow Coma Score (GCS) of 3/15. She was intubated and transferred to the intensive care unit. She had a history of diarrhoea and vomiting over the previous 7 days. Her family also reported her being fatigued over the previous week.

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Her past medical history also included hypertension, type 2 diabetes mellitus, osteoporosis, chronic obstructive pulmonary disease, anxiety disorder and benign paroxysmal positional vertigo.

Her regular medications at home included Venlafaxine 75 mg once daily, Metformin 500 mg twice daily, Zolpidem 10 mg at night and Lercandipine 10 mg once daily.

Investigations

Laboratory investigations on admission (Table 1) showed an acute kidney injury, with associated high anion gap metabolic acidosis. Her acidosis was attributed to a combination of lactate accumulation and acute renal failure. Serum ketones were not elevated. Neuroimaging done by CT-brain and MRI-brain are shown in Fig. 1.

| Electrolytes                  | Result       | Reference ranges |
|-------------------------------|--------------|------------------|
| Sodium                        | 137 mmol/L   | 136–145 mmol/L   |
| Potassium                     | 4.6 mmol/L   | 3.5–5.1 mmol/L   |
| Chloride                      | 97 mmol/L    | 98–107 mmol/L    |
| Bicarbonate                   | 9.2 mmol/L   | 22.0–29.0 mmol/L |
| Blood glucose                 | 8.5 mmol/L   | 3.9–7.1 mmol/L   |
| Serum acetone                 | Negative     | –                |

Renal functions

- Blood urea: 39 mmol/L (2.8–8.1 mmol/L)
- Creatinine: 657 umol/L (45–84 umol/L)
- Glomerular filtration rate (GFR): 5 ml/min/1.73 sqm (> 90 ml/min/1.73 sqm)

Arterial blood gases

- pH: 7.04 (7.35–7.45)
- HCO3: 9.2 mmol/L (22.0–29.0 mmol/L)
- CO2: 4.5 kPa (4.27–6.40 kPa)
- O2: 10.3 kPa (11.07–14.40 kPa)
- O2 sats: 91.6%

- Lactate: 10.7 mmol/L (1.0–1.8 mmol/L)
- Anion gap: 30.8 mmol/L (12–15 mmol/L)

Full blood count

- White cell count: 12.4 10^9/L (4.0–11.0 10^9/L)
- Hemoglobin: 7.3 g/L (11.5–16.5 g/L)
- Platelets: 318,000 10^9/L (140–450 10^9/L)

Liver function tests

- ALT: 8 U/L (0–33 U/L)
- AST: 13 U/L (0–32 U/L)
- Alk. phosp: 143 IU/L (35–105 IU/L)
- GGT: 23 U/L (< 40 U/L)
- Total bilirubin: 8 U/L (0–15 umol/L)
- Total protein: 62 U/L (66–87 g/L)

Table 1 Showing blood investigations at the time of arrival

Discussion

There have been different toxic and metabolic derangements which are attributed to abnormalities in the basal ganglia [1]. The Lentiform fork sign is one such neuroimaging abnormality which is previously described related to chronic kidney disease.

In a case series of 11 patients with chronic kidney disease it was encountered commonly in patients who had metabolic acidosis at some point [2, 3]. Diabetes mellitus is reported to have some role in causation of this basal ganglia abnormality which our patient had however her blood glucose at the time of arrival was mildly deranged, the underlying mechanism has been postulated to be due to an underlying endothelial dysfunction in chronic diabetics [4–6].

In our patient, there were few predisposing factors which might have possibly resulted in this abnormality chronic kidney disease, diabetes mellitus and metabolic acidosis. The tremor that was observed could be due to the basal ganglia insult or just the direct effects of the renal derangements she presented with.

In our patient, the serum potassium was within the normal range despite several days of diarrhoea preceding admission. It is likely that her whole-body potassium stores were low, and her serum concentration reflected a degree of extracellular potassium shift in the face of metabolic acidosis, lack of insulin effect and possible alpha-adrenergic surge post seizure [7]. Her lactate level was presumed secondary to seizure and hypoxemia, but
it should be noted that she was taking metformin prior to admission.

The pathophysiological basis for the basal ganglia abnormality is linked to vasogenic and cytotoxic edema [2]. The neuroimaging modalities CT brain and MRI brain reveal this neuroimaging abnormality and with aggressive dialysis and renal optimization these changes can be reversible [8]. Our patient did not undergo any further imaging to look for an interval difference.

**Conclusion**
The Lentiform fork sign is a rare occurrence which can be related to a long list of toxic and metabolic causes but in conjunction with metabolic acidosis in chronic kidney disease patients it can narrow down this list of alternate diagnosis.

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Authors’ contributions
SM, LDK, JL and SS were involved in writing of manuscript diagnosing and treating the patient. Rest of the authors MF, IA, BC, MAM, SS, SK, MHA, JL and KM were involved in care and treatment of the patient. All authors have read and approved the manuscript.

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Consent for publication
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Competing interests
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