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Case Report

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ACUTE BILATERAL BASAL GANGLIA LESION: LENTIFORM FORK SIGN IN END STAGE KIDNEY DISEASE WITH METABOLIC ACIDOSIS

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ABSTRACT

Neurological complications are not uncommon in end stage kidney disease population. Bilateral basal ganglia lesions have been frequently reported in patients with uraemic encephalopathy or metabolic acidosis. Lentiform Fork sign is a distinctive, as described in MRI picture that is not only seen in patient with uraemic encephalopathy, but also in other condition that presented with metabolic acidosis.

Keywords: bilateral basal ganglia lesion. Metabolic acidosis.

INTRODUCTION

Bilateral basal ganglia lesions have been described in end stage kidney patients, especially those who have diabetes or metabolic acidosis (1). The vulnerability of basal ganglia to metabolic acidosis and ureamic encephalopathy is well described in several case reports and literature reviews.

CASE REPORT

A 46-year-old Malay woman who had been receiving haemodialysis three times per week for past thirteen years was found lethargy, generalised weakness, slow response, and slurred speech, and respiratory distress. Additionally, she had missed her one haemodialysis session before presented to emergency department. On neurologic evaluation, the patient scored E3V4M6 on the Glasgow Coma Scores (GCS). The patient's medical history was notable only for end stage kidney disease with unknown primary disease and hypertension.

The patient's laboratory investigation results were shown in Table 1 with predominantly metabolic acidosis with lactic acidosis. Computed tomography (CT) brain on arrival showed hypodensities at both basal ganglia with no focus focal haemorrhage. The patient was treated with sustained low efficiency dialysis (SLED) and supportive care that led to improvement in blood chemistry. She was able to wean off mechanical ventilation on day 2 of admission. However, the patient's GCS scored E2V2M5on Day-4 and reintubated for airway protection. Repeated CT brain (Figure A) showed worsening bilateral basal ganglia hypodensities with effacement of the adjacent cerebral sulci. Magnetic resonance imaging (MRI) of the brain was done showed hyperintensity regions bilaterally in the basal ganglia surrounded by more hyperintense rim, suggestive of lentiform fork sign (Figure B).

We presumed that the new deterioration of her GCS was due to metabolic acidosis in origin given their radiological correlation, and continuous renal replacement therapy was commenced for her cerebral protection and intermittent intravenous mannitol was given for her cerebral edema.

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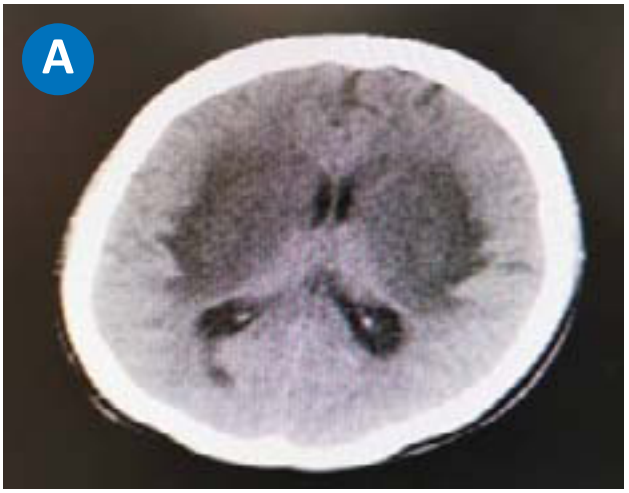
Table 1 showed the patient's investigation results:

Test		***Reference range***
Haemoglobulin	10.2	*12 - 15 g/dL*
Total white cell	9.07	*4 – 10 x 109/L*
platelet	299	*150 - 410 x 109 /L*
prothrombin	16.2	*11.6 – 14.1 sec*
aPTT	33.9	*31.4 – 43.3 sec*
Sodium	135	*135 – 150 mmol/L*
Potassium	4.7	*3.5 – 5.0 mmol/l*
Urea	18.9	*1.7 – 8.3 mmol/l*
Albumin	41	*35 – 50 g/L*
Random blood glucose	5.2	
Calcium	2.47	*2.10 – 2.60 mmol/l*
Phosphate	2.23	*0.80 – 1.45 mmol/l*
CRP	0.96	*< 0.80 mg/dL*
pH	6.976	*7.35 – 7.45*
PaO2	295	*75 – 100 mmHg*
PaCO2	31.6	*38 – 42 mmHg*
HCO3	7.9	*22 – 26 mmol/l*
Base excess	-20.7	*-3.0 – 3.0 mmol/l*
Lactate	15	*0.5 – 1 mmol/l*



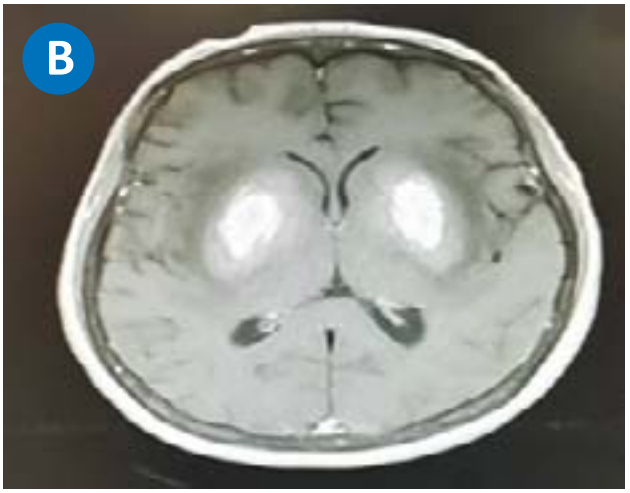


Figure A CT brain contrast revealed worsening bilateral basal ganglia hypodensities with involvement of the adjacent white matter and thalami. There was effacement of the adjacent cerebral sulci, frontal horns of both lateral ventricles and third ventricles. Figure B MRI of the brain (non-contrast) showed both globus pallidus demonstrated heterogenous hyper and hypointensity, involving of both putamina in lesser extends. Heterogenous avid enhancement of both globus pallidus and the peripheral enhancement in T2-weighted MRI

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2 weeks later, her GCS improved with E4VTM6 and repeated CT brain showed lesser degree of cerebral edema. However, her condition was complicated with ventilated associated pneumonia due to prolonged ventilation. The patient was succumbed on day-18 of hospitalization.

DISCUSSION

Bilateral basal ganglia lesions have been well described in end stage renal disease patients, commonly presented in patients with diabetes mellitus and metabolic acidosis. The clinical presentation is usually acute onset, which involved neurological disorders like drowsy or movement disorder. Lentiform fork sign has been described as a distinctive radiological finding in end stage kidney disease (ESKD) with uraemic encephalopathy or metabolic acidosis, which may suggest metabolic cause or metabolic acidosis can be an important aspect in the pathogenesis (3).

In literature review, 90% of patients at presentation had associated with metabolic acidosis (4). The most interesting character of this syndrome is the consistency of the neuroimaging finding with CT brain uniformity revealed bilateral hypodense basal ganglia lesion and MRI brain showed bilateral hypointense area in the basal ganglia on T1-weighted images and hyperintense area on T2-weighted images (1-5).

The exact pathogenesis of appearance of Lentiform fork sign in ESKD is still in mystery. Metabolic acidosis possible increases susceptibility basal ganglia to insults, and high frequency of the co-existing diabetes may further increase the risk. In Gyanendra review, several of the acidotic condition related to the Lentiform Fork sign, are well known associated with increased cerebral lactate production. As an ultimate common pathway, through the changes in vascular reactivity, metabolic acidosis may disrupt the brain blood barrier leading to vasogenic edema and later cytotoxic edema, proportionate the severity of acidosis on presentation. In this case report, the clinical course is acute onset but reversible with the resolution of metabolic acidosis and cerebral edema.

The main treatment for this presentation included correction of uremia and metabolic acidosis as supportive therapy, and prevention of worsening of cerebral edema has been the focus of management. Mode of renal replacement therapy depend on the severity of cerebral edema of the patient. The neurological symptoms usually improved gradually: 20% showed complete resolution, 30% showed no neurological improvement and the rest showed partial recovery. Neurological and clinical improvement do not correlate with severity of radiographically changes (4).

CONCLUSION

Lentiform fork sign is an important radiological finding to differentiate the important aetiology from other conventionally causes for basal ganglia lesions. Metabolic acidosis in ESKD patients with radiological finding of lentiform fork sign can help to exclude other long list causes of bilateral ganglia lesions.

Conflict of interest: There is no conflicts of interest to declare.

Consent to participate: The author declares the next of kin of the patient provided authorities for use of her medical records as research.

References

1. Kumar G, Goyal MK (2010) Lentiform fork sign: a unique MRI picture. Is metabolic acidosis responsible? Clin Neurol Neurosurg 112(9):805–812. doi:10.1016/j.clineuro.2010.06.006
2. Fabiani G, Teive HA, Munhoz RP (2013) Lentiform fork sign and fluctuating, reversible parkinsonism in a patient with uremic encephalopathy. Movement Disorder 28(8):1053. doi:10.1002/mds.25503
3. Manish R, Chetan M. Lentiform fork sign in a case of end-stage renal disease. Kidney International (2012) 82, 365; doi:10.1038/ki.2012.119
4. Li JY, Yong TY, Sebben R, Khoo E, Disney AP (2008) Bilateral basal ganglia lesions in patients with end-stage diabetic nephropathy. Nephrology (Carlton) 13(1):68–72. doi:10.1111/j.1440-1797. 2007.00838.x
5. Jaryal A, Thakur S. Lentiform fork sign: Uraemia alone or multifactorial causation? Haemodialysis International 2020: 24: E10-E12. DOI:10.1111/hdi.12810