

CASE REPORT

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Lentiform fork sign: A distinctive neuroradiologic manifestation of uremic encephalopathy—A case report

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ABSTRACT

Introduction: The lentiform fork sign is a unique magnetic resonance imaging (MRI) appearance of the basal ganglia believed to be due to acute metabolic acidosis and seen in several conditions that result in metabolic acidosis such as uremic encephalopathy.

Case Report: We present the case of a 76-year-old hypertensive and diabetic male patient with end-stage renal failure on dialysis, whose brain MRI revealed bilateral symmetrical T2W and fluid-attenuated inversion recovery hyperintensities in the basal ganglia, surrounded by a hyperintense rim delineating the lentiform nuclei, giving rise to the lentiform fork sign.

Conclusion: We describe this under-reported unique sign, the lentiform fork sign, in an end-stage renal disease patient, whose brain MRI revealed this sign to highlight the significance of this sign, which has been widely reported to be associated with uremic encephalopathy.

Keywords: Basal ganglia hyperintensity, Lentiform fork sign, MRI, Uremic encephalopathy

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INTRODUCTION

Lentiform fork sign is a rare neuroradiological sign that has been hypothesized to be due to acute metabolic acidosis and seen in several conditions that result in metabolic acidosis, such as uremic encephalopathy, diabetes mellitus, methanol and ethylene glycol intoxications, and acidopathies such as propionic acidemia (PA) and pyruvate dehydrogenase deficiency [1, 2]. The lentiform fork sign is an imaging finding observed on brain MRI, showing involvement of the basal ganglia [3, 4]. It is believed to be due to its high metabolic demand, which predisposes it to hypoxic, toxic, and metabolic disorders [3, 4].

Uremic encephalopathy is an acquired toxic syndrome resulting from a metabolic disorder in patients with untreated or poorly treated acute or chronic kidney disease. Accumulation of endogenous uremic toxins in patient with chronic renal failure plays significant role in the pathogenesis of uremic encephalopathy [5]. Uremic encephalopathy commonly targets the basal ganglia, with a typical pattern of involvement of the lentiform nucleus, known as the lentiform fork sign, as they will have a hyperintense signal on T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequence of brain MRI [2], which are routine sequences performed in brain examinations. Although, the lentiform fork sign is neither pathognomonic nor diagnostic, but characteristic of uremic encephalopathy thereby limiting the myriad of differential diagnoses of basal ganglia hyperintensities. We describe this under-reported unique sign, the lentiform fork sign, in an end-stage renal disease patient, whose brain MRI revealed this sign to highlight the significance of this sign, which has been widely reported to be associated with uremic encephalopathy.

CASE REPORT

A 78-year-old hypertensive, diabetic, and end-stage renal disease, male patient was referred to the Department of Radiology for investigations on account of unexplained weakness and irritability. An initial unenhanced brain computed tomography (CT) scan revealed bilateral symmetrical hypodensities involving the lentiform nuclei (Figure 1).

Brain MRI done 5 hours later revealed bilateral symmetrical T2W and FLAIR hyperintensities in the basal ganglia surrounded by a hyperintense rim delineating the lentiform nuclei, giving rise to the lentiform fork sign (Figure 2A and B). Laboratory evaluation revealed elevated blood urea 27.2 mm/L (normal: 0–8.3 mm/L), serum creatinine 930 $\mu\text{mol/L}$ (normal: 62–106 $\mu\text{mol/L}$), and uric acid 447 $\mu\text{mol/L}$ (normal: 202.3–416.5 μmol).



Figure 1: Brain CT without contrast showing bilateral hypodense basal ganglia with a linear area of lower hypodensity outlining the lentiform nuclei.

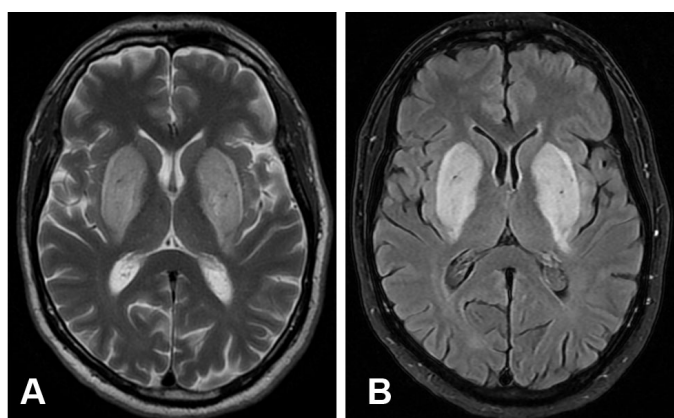


Figure 2: Brain MRI, axial sections. (A) T2-weighted sequence and (B) FLAIR showing the basal ganglia with hyperintense signal, symmetrical and bilateral, surrounded by a hyperintense rim delineating the lentiform nucleus, representing the lentiform fork sign.

The lentiform fork sign has been reported in several conditions associated with metabolic acidosis including uremic encephalopathy. The MRI, clinical history of end-stage renal disease, and laboratory investigations raised the possibility of uremic encephalopathy. The patient was treated along this line and he made a full recovery with the resolution of the MRI finding.

DISCUSSION

The lentiform fork sign is a distinctive neuroradiological sign which on MRI is characterized by bilateral symmetrical hyperintensities in the basal ganglia surrounded by a hyperintense rim delineating the lentiform nuclei [2]. The lentiform fork sign results from vasogenic edema of the lentiform nuclei [2]. The white matter surrounding the basal ganglia, along with the internal and external capsules, shows hyperintensity on T2W and FLAIR images, creating the typical fork appearance. On T2/FLAIR images, the linear hyperintensity of the external capsule constitutes the prong of the fork, while the hyperintensity of the internal capsule, internal and external laminae fused together to form the stem of the fork [6]. This sign has been depicted on the brain MRI of the patient under review.

The susceptibility of the basal ganglia to a range of toxins and metabolic disturbances attributable to their high metabolic demand has been documented [3, 4]. The pathophysiological bases of the lentiform fork sign have been linked to metabolic acidosis [2], which disrupts the blood–brain barrier and the unique imaging appearance is ascribed to a difference in metabolic vulnerability between neurons of the basal ganglia and the astrocytes of surrounding white matter [5].

Although the lentiform fork sign is not specific for uremic encephalopathy; however, it has been widely reported to be associated with uremic encephalopathy. Uremic encephalopathy is an acquired toxic syndrome that occurs in patients with untreated or inadequately treated acute or chronic renal disease. The patient under review is a diabetic, hypertensive, and end-stage renal disease patient on dialysis who suddenly developed unexplained behavior. His comorbidity and diabetes mellitus, according to some authors, may have contributed to making the basal ganglia to be more sensitive to uremic toxins and metabolic acidosis due to endothelial dysfunction [6, 7].

The lentiform fork sign has been described not only in uremic encephalopathy but also in cases of metabolic acidosis from other causes such as diabetes mellitus, methanol, and ethanol poisoning. Metabolic acidosis, commonly observed in these disorders, appears to be a major contributing factor to the pathogenesis of the lentiform fork sign [2]. In the absence of worsening comorbidities and life-threatening clinical presentation, the prognosis has been reported to be good [8]. These abnormalities of the basal ganglia are said to be reversible

with adequate treatment including dialysis. Grasso et al. [1], Kim et al. [5], Saini et al. [8], and Okada et al. [9] documented complete resolution of the imaging findings on follow-up MRI following treatment. Lentiform fork sign can be seen in patients with conditions that result in metabolic acidosis, hence, a good clinical history is indispensable to narrow down the gamut of differential diagnosis.

CONCLUSION

Lentiform fork sign is an MRI finding in the basal ganglia that has been hypothesized to be triggered by metabolic acidosis and has been widely reported in uremic encephalopathy due to chronic or acute renal failure. Radiologists and other clinicians should be familiar with this unique, hitherto under-reported MRI appearance to narrow down the specific etiology from the myriad of conditions grouped under the heading “basal ganglia hyperintensities,” to properly guide management.

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Author Contributions

Awajimijan Nathaniel Mbaba – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Khalid Mohamed Abdalla – Conception of the work, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Hamza Mustapha Ahmed – Design of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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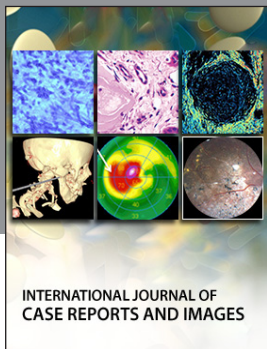
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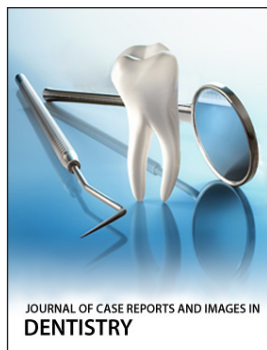
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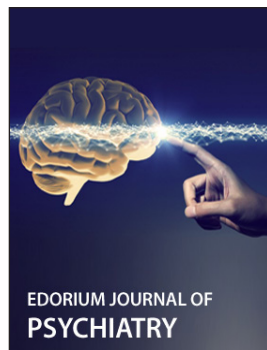
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