

## CASE REPORT

# Uraemic optic neuropathy – a rare presentation of uraemia

Adaobi Uzoamaka Solarin, Adetola M Adesanya, Moturayo Adekunle, Alaba Oladimeji, Olusegun Henry Gbelee, Barakat Adeola Animashaun

Department of Paediatrics, Lagos State University Teaching Hospital, Ikeja, Nigeria.

### ABSTRACT

Uraemic optic neuropathy (UON) is an acute but reversible loss of vision in patients with end-stage renal failure. Hypoperfusion of the posterior ciliary arteries, the major blood supply to the optic nerve head, results in ischaemic injury to the nerve. Anaemia, hypertension and elevated nitrogen urea level have been identified as factors contributing to optic nerve neuropathy. Toxic uraemic metabolites affect nerve conduction and their removal with dialysis results in improved vision. Optic neuropathy is a rare complication of uraemia in children. This is the first case of optic neuropathy related to uraemia in a child with end-stage renal disease in Nigeria.

**Keywords:** Uraemia; reversible blindness; haemodialysis.

### CASE REPORT

An 11-year-old girl diagnosed with human immunodeficiency virus type 1 at 4 years of age presented to the paediatric emergency unit of Lagos State University Teaching Hospital, in Ikeja, with bilateral leg swelling and facial swelling of two weeks' duration. There was a history of acute onset of cough, difficult breathing and reduced urinary output. She had been on antiretroviral drugs (lamivudine, zidovudine and nevirapine) with very poor drug compliance. Both parents had died 4 years previously of HIV infection and the primary care-giver is the paternal aunt.

On admission, she was in severe respiratory distress, markedly pale and afebrile (36.7°C). She had facial puffiness and bilateral pitting pedal oedema up to the shin. She had a respiratory rate of 54 breaths per minute, and oxygen saturation was 77% on room air and 83% on intranasal oxygen. There were widespread crackles in both lung fields. On cardiovascular system evaluation she manifested small-volume peripheral pulses, a pulse rate of 148 beats per minute, raised jugular venous pressure, and cardiomegaly with a displaced apex beat. Blood

pressure was 130/92 mmHg (both systolic and diastolic pressure were greater than the 99th centile). Gross ascites was evident and she had a tender hepatomegaly of 6 cm below the right costal margin in the midclavicular line. Urine output was 1.1 mL/kg/h. Dipsticks testing of the urine revealed heavy proteinuria.

Laboratory findings included an elevated serum urea concentration of 36 mmol/L and creatinine of 975 µmol/L; the packed cell volume was 16%. The fluid overload, with acute pulmonary oedema, was thought to be due to the combination of renal failure and cardiac failure. HIV-associated nephropathy was considered the most likely cause of the renal disease in view of the poor compliance with antiretroviral drugs and the heavy proteinuria.

Intravenous diuretics were administered and urgent haemodialysis initiated. A transfusion of packed red cells was given during the dialysis procedure and her blood pressure was controlled with oral amlodipine. Owing to severe financial constraints, there was a delay in delivering additional sessions of dialysis despite indications for its need.

Received 15 November 2017; accepted 05 March 2018; published 21 March 2018.

Correspondence: Adaobi Uzoamaka Solarin, [asolar234@gmail.com](mailto:asolar234@gmail.com).

© The Author(s) 2018. Published under a [Creative Commons Attribution 4.0 International License](https://creativecommons.org/licenses/by/4.0/).

Five weeks after admission, she complained of sudden loss of vision in the right eye. There was no associated fever, headaches, convulsions or abnormalities on neurological examination. Blood pressure was 100/60 mmHg. Serum urea and creatinine were 74.8 mmol/L and 2210 µmol/L, respectively. A review by an ophthalmologist revealed no perception of light in the right eye, an absent pupillary reflex, a clear cornea with normal anterior chamber depth, and a pink disc with blurred margins. Regarding the left eye, she could count fingers, had a clear cornea with normal anterior chamber depth, and a pink disc with distinct margins. An assessment of HIV retinopathy was made. Within 24 hours of the right eye being affected, however, loss of vision developed also in the left eye.

Haemodialysis was started, and was associated with the return of vision in the right eye, and then in the left eye a few hours after dialysis. The post-dialysis urea was 19.3 mmol/L and the creatinine 1382 µmol/L.

Chronic dialysis could not be sustained because of inadequate financial resources and the patient died eight weeks after her initial presentation. Her vision remained intact until this time.

## DISCUSSION

Uraemic optic neuropathy (UON) is an extremely rare manifestation of renal failure with few reported cases [1]. It presents an emergency and early recognition of this complication with immediate institution of treatment can potentially reverse the visual impairment and prevent long-term sequelae [1]. Its pathophysiology is poorly understood but it has been suggested that it may be caused by uraemic toxins affecting optic nerve conduction [2].

The anaemia, hypertension and elevated urea, which were seen in the index case, are risk factors for UON [3,4]. Characteristic findings in optic neuropathy are reductions in the pupillary response to light and oedema of the optic disk [5]. In the index case, the absent pupillary response, the absence of other neurological deficits, the sudden loss of vision in the setting of worsening of uraemia and the prompt improvement with dialysis support the diagnosis of uraemic optic neuropathy. Optic neuritis from bacterial infection was unlikely as there were no symptoms or signs of acute infection.

Anterior ischaemic optic neuropathy may have a presentation similar to UON [6] and can occur in haemodialysis patients [7], where hypotension and hypoperfusion cause ischaemic injury to the optic nerve [8]. In the index case, pulmonary oedema and heart failure with severe anaemia were the initial presentation, raising the possibility of

ischaemic optic neuropathy. However, heart failure and anaemia had been addressed with diuretics and a blood transfusion before the onset of sudden visual loss. Systemic hypertension also interferes with the autoregulation of arterial perfusion to the optic disc and optic nerve [9]. The index case had hypertension but at the time of the blindness the blood pressure had improved on treatment.

## CONCLUSIONS

Uraemic optic neuropathy is a rare complication that is reversible with prompt treatment. Sudden loss of vision in patients with advanced renal failure should raise suspicion about this preventable cause of blindness.

## Ethics approval

Permission for publication was granted by the patient's guardian and the hospital authorities.

## REFERENCES

1. Lee KG, Vaithilingam I. Bilateral optic neuropathy – A rare uraemic manifestation of end-stage renal disease. *NDT Plus*. 2011; 4:455.
2. Saini JS, Jain IS, Dhar S, Mohan K. Uremic optic neuropathy. *J Clin Neuroophthalmol*. 1989; 9(2):131-133.
3. Haider S, Astbury NJ, Hamilton DV. Optic neuropathy in uraemic patients on dialysis. *Eye*. 1993; 7:148-151.
4. Doluoglu OG, Saricaoglu MS, Oztekin CV, Karakurt A, Akdemir AO, Koc E. Cortical blindness due to bilateral occipital infarcts in a renal failure patient with prostate cancer: a rare complication of hemodialysis. *Case Rep Med*. 2013. Article ID 539761.
5. Cohen AB, Pless M. Optic neuropathy with disc edema. *Continuum*. 2009; 15(4):22-46.
6. Korzets Z, Zeltzer E, Rathans M, Manor R, Bernheim J. Uraemic optic neuropathy. *Am J Nephrol*. 1998;18:240-242.
7. Leon LR, Marinaro C. Previous ischaemic optic neuropathy in haemodialysis. *Nefrologia* 2009; 29:614-616.
8. Smith JL, Goldhammer Y. Hypertensive optic neuropathy. *Trans Sect Otolaryngol Am Acad Ophthalmol Otolaryngol*. 1975; 79(3):OP520-523.
9. Pillunat LE, Anderson DR, Knighton RW, Joos KM, Feuer W. Autoregulation of human optic nerve head circulation in response to increased intraocular pressure. *Exp Eye Res*. 1997; 64:737-744.