Sequential nonarteritic anterior ischemic optic neuropathy in patient on chronic hemodialysis

Ari Shemesh¹ · Edward Margolin¹

Received: 26 September 2018 / Accepted: 14 November 2018 / Published online: 21 November 2018
© Japanese Society of Nephrology 2018

Abstract

Nonarteritic anterior ischemic optic neuropathy (NAION) is a well acknowledged rare complication of chronic hemodialysis in patients with end-stage renal disease (ESRD). We present a unique case of a patient on chronic hemodialysis who presented with an NAION in the right eye, then 3 months later developed an NAION in the left eye followed in 1 month by second NAION episode in the left eye. Every episode of NAION was accompanied by intradialytic hypotension (drop in systolic blood pressure of over 20 mmHg) where visual loss was noticed at the end of the dialysis session. Clinicians should be aware of association between NAION and hemodialysis and that patients with dialysis-induced hypotension are at particularly increased risk of having NAION. Patients who had dialysis-associated NAION should be closely monitored to prevent occurrence of intra-dialytic hypotension as they are at the highest risk of sequential NAION and thus severe visual loss.

Keywords Nonarteritic anterior ischemic optic neuropathy · Dialysis-induced hypotension · Blindness after dialysis

Background

Nonarteritic anterior ischemic optic neuropathy (NAION) is the most common non-glaucomatous optic neuropathy in patients older than 50 years [1]. The precise etiology of NAION is unknown but physiologically feasible hypothesis is that it stems from inadequate perfusion of short posterior ciliary arteries which supply the blood to the optic nerve head. These small arteries are susceptible to injury by atherosclerosis, and epidemiological studies shows the disease is more prevalent in patients with cardiovascular risk factors [2]. In addition, anatomical factors like a congenitally small and crowded optic nerve head with a small cup-to-disc ratio (“disk at risk”) may be a contributing factor to the compromised blood flow through these arteries [3]. Since the short posterior ciliary arteries supply optic nerve head in two vascular semi-circles, optic nerve head ischemia results in segmental optic nerve swelling, correlated with an altitudinal defect on visual field exam [4]. Typically, patient with NAION presents with acute, painless monocular loss

of vision that is often first discerned on awakening. Vision may deteriorate over several hours or days. On examination there is a relative afferent pupillary defect (RAPD), and fundoscopy shows optic disc swelling which is often segmental with peripapillary hemorrhages.

Subsequent involvement of the fellow eye occurs in 14.7% of patients over 5 years from first incident [5], however, recurrences in the same eye are very uncommon (~6%) [6].

We present a unique case of patient receiving chronic hemodialysis who presented with NAION in the right eye, then 3 months later developed sequential NAION in the left eye and one month after that developed a second NAION in the left eye. Every episode of NAION was accompanied by intra-dialytic hypotension (drop in systolic blood pressure ≥ 20 mmHg).

Case presentation

35-year-old man who was on daily peritoneal dialysis for the past 1.5 years and three times a week hemodialysis for the past 6 months, with past medical history of hypertension, end-stage renal disease (ESRD) secondary to segmental glomerulosclerosis because of chronic anabolic steroid use presented with an acute onset of painless visual loss in the right eye that was noticed toward the end of his

✉ Edward Margolin
edmargin@gmail.com

¹ University of Toronto, Department of Ophthalmology and Vision Sciences, 801 Eglinton Ave West Suite 301, Toronto, ON M5N 1E3, Canada

Fig. 1 **a** Humphrey visual fields at initial presentation. The field is black in the right eye and is unreliable with non-specific depressed spots in the left eye. **b** Peripapillary ocular coherence tomography

at initial presentation demonstrating severely thickened retinal nerve fiber layer around the optic nerve in the right eye

hemodialysis session during which systolic blood pressure dropped 50 mmHg from baseline. On examination best-corrected visual acuity (BCVA) was light perception in the right eye and 6/6 in the left eye. There was a brisk right RAPD. Dilated fundusoscopic examination (DFE) demonstrated a diffusely swollen optic disc on the right with peripapillary hemorrhages and normal optic disc on the left with small cup-to-disc ratio. Visual field testing with Humphrey 24–2 algorithm (Carl Zeiss Meditec, Dublin, CA) demonstrated generalized depression on the right and was normal on the left (Fig. 1a). Peripapillary ocular coherence tomography (OCT) (Carl Zeiss Meditec, Dublin, CA) demonstrated severely swollen peripapillary retinal nerve fiber layer in the right eye confirming severe optic nerve head edema (Fig. 1b). Hemoglobin was 11.6 gram/d, and CRP was normal. MRI of the brain and orbits with gadolinium was interpreted as normal. Multiple measures again intra-dialytic hypotension were instituted: sodium levels were ramped from 150 to 140 mEq/L, dialysate temperature was reduced to 36°, and midodrine was administered when intra-dialytic hypotension was observed. Injections of epoetin alpha and intravenous iron infusions and blood transfusions on as needed basis were performed and patient was instructed not to eat prior to dialysis. Duration of dialysis was minimized and frequency was increased to five times weekly.

Three months later the patient experienced an acute onset of painless visual loss in the left eye which was again noticed at the end of hemodialysis. There was a drop of 48 mmHg in systolic blood pressure during that dialysis session. At this time visual acuity was still light perception on the right and deteriorated to 20/30 on the left. DFE demonstrated a pale optic nerve head on the right and a segmentally swollen optic nerve on the left with peripapillary hemorrhages (Fig. 2). Visual fields (Humphrey 24–2 algorithm) demonstrated a black field on the right and now multiple scattered depressed spots on the left (Fig. 3a). Peripapillary OCT confirmed presence of very elevated retinal nerve fiber layer

around the left optic nerve with average thickness of 295 microns (Fig. 3b).

A systematic approach to minimize blood pressure fluctuations during hemodialysis was undertaken in consultation with the nephrology team. Since most known measures were already utilized, duration of dialysis sessions was shortened and frequency was increased to six times weekly. Despite this, one month later patient re-presented with new episode of significant deterioration of vision in the left eye which again occurred at the end of hemodialysis session. There was a drop of 38 mmHg in systolic blood pressure during that session. Visual acuity measured only light perception in each eye. DFE demonstrated a pale optic disc on the right and segmental optic nerve head edema superiorly with the pale inferior portion of the optic nerve on the left. Visual fields now demonstrated completely black visual field in the left eye (Fig. 4a). Peripapillary OCT demonstrated even more severely swollen peripapillary retinal nerve fiber layer than 1 month ago with the average thickness of 405 microns confirming the second episode of NAION in the left eye (Fig. 4b).

Three months later visual acuity remained light perception in both eyes and DFE demonstrated pale optic nerve heads in both eyes.

Discussion

NAION is well acknowledged rare ophthalmic complication of chronic hemodialysis in patients with end-stage renal disease (ESRD) [7–12]. Predisposing factors in this patient population are the presence of anemia, hypotensive effect of hemodialysis, and past history of NAION [13]. Intradialytic hypotension, it is commonly regarded as a decrease in systolic blood pressure ≥ 20 mmHg during hemodialysis or a decrease in mean arterial pressure by 10 mmHg, accompanied by clinical events that necessitate intervention by a nurse [7, 14]. The mechanism involved in the development

Fig. 2 Fundus photos demonstrating pale optic nerve on the right and segmentally swollen optic nerve on the left with peripapillary hemorrhages

Fig. 3 **a** Visual fields (Humphrey 24-2 algorithm) demonstrating black visual field on the right and scattered depressed spots on the left. **b** Peripapillary OCT demonstrating very elevated peripapillary retinal nerve fiber layer around the left optic nerve

Fig. 4 **a** Visual field in the left eye now demonstrating complete absence of responses to any stimuli. **b** Peripapillary OCT demonstrating worsening of the peripapillary retinal nerve fiber layer edema around the left optic nerve. No signal was obtained from the right optic nerve

of NAION in patients undergoing hemodialysis remains unclear since intradialytic hypotension is common and complicates 5–30% of all dialysis treatments but only very few patients who experience intradialytic hypotension develop

NAION [15]. The likely explanation is that systemic hypotension during hemodialysis causes hypoperfusion of short posterior ciliary arteries supplying the optic nerve head which then results in the ischemia of the optic nerve head in susceptible patients.

Clinicians involved in the circle of care of patients undergoing hemodialysis should be aware of this serious and potentially blinding complication and be alert to the importance of minimizing intradialytic hypotension especially in patients who have already experienced one episode of NAION. All efforts should be directed toward correction of systemic hypotension, anemia, and underlying vasculopathic risk factors in these patients to minimize their chance of developing sequential NAION [16].

The patient described in our report has become legally blind in both eyes as a result of sequential NAION which was followed by a recurrence episode of NAION in one eye. Recurrent NAION in the same eye is an extremely rare event occurring in only 3–6% of patients [3]. The fact that our patient had three separate episodes of NAION highlights that some patients likely have individual predisposition to this condition and that patients who had one episode of dialysis-associated NAION are at increased risk of having sequential NAION. Thus all patients who had one episode of dialysis-associated NAION should have all possible measures taken to minimize intra-dialytic hypotension which is likely the most important risk factor for second episode of NAION.

Learning points

- Clinicians should be aware that patients with dialysis-induced hypotension are at an increased risk of having NAION which is a sight threatening disease with poor prognosis.
- Patients with ESRD on chronic hemodialysis who have already suffered an episode of NAION in one eye during dialysis session are at much increased risk of having a NAION in the fellow eye.
- In patients with ESRD on chronic hemodialysis who had NAION in one eye it is of utmost importance to minimize intra-dialytic hypotension, e.g., decrease in systolic blood pressure ≥ 20 mmHg or a decrease in mean arterial pressure by 10 mmHg, and not just preventing systemic hypotension (systolic blood pressure < 90 mmHg). Correction of anemia and co-existing vasculopathic risk factors should be undertaken as well.
- If NAION occurs in the second eye, there is a high risk of patient becoming legally blind.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Research involving human participants and/or animals Not applicable.

Informed consent Not applicable, no identifying information presented.

References

- Hattenhauer MG, Leavitt JA, Hodge DO, Grill R, Gray DT. Incidence of nonarteritic anterior ischemic optic neuropathy. *Am J Ophthalmol*. 1997;123(1):103–7.
- Jacobson DM, Vierkant RA, Belongia EA. Nonarteritic anterior ischemic optic neuropathy. A case–control study of potential risk factors. *Arch Ophthalmol*. 1997;115(11):1403–7.
- Burde RM. Optic disk risk factors for nonarteritic anterior ischemic optic neuropathy. *Am J Ophthalmol*. 1993;116(6):759–64.
- Olver JM, Spalton DJ, McCartney AC. Microvascular study of the retrolaminar optic nerve in man: the possible significance in anterior ischaemic optic neuropathy. *Eye (Lond)*. 1990;4(Pt 1):7–24.
- Newman NJ, Scherer R, Langenberg P, Kelman S, Feldon S, Kaufman D, et al. The fellow eye in NAION: report from the ischemic optic neuropathy decompression trial follow-up study. *Am J Ophthalmol*. 2002;134(3):317–28.
- Hayreh SS, Podhajsky PA, Zimmerman B. Ipsilateral recurrence of nonarteritic anterior ischemic optic neuropathy. *Am J Ophthalmol*. 2001;132(5):734–42.
- Servilla KS, Groggel GC. Anterior ischemic optic neuropathy as a complication of hemodialysis. *Am J Kidney Dis*. 1986;8(1):61–3.
- Hamed LM, Winward KE, Glaser JS, Schatz NJ. Optic neuropathy in uremia. *Am J Ophthalmol*. 1989;108(1):30–5.
- Haider S, Astbury NJ, Hamilton DV. Optic neuropathy in uraemic patients on dialysis. *Eye (Lond)*. 1993;7(Pt 1):148–51.
- Korzets A, Marashek I, Schwartz A, Rosenblatt I, Herman M, Ori Y. Ischemic optic neuropathy in dialyzed patients: a previously unrecognized manifestation of calcific uremic arteriolopathy. *Am J Kidney Dis*. 2004;44(6):e93–7.
- Nieto J, Zapata MA. Bilateral anterior ischemic optic neuropathy in patients on dialysis: a report of two cases. *Indian J Nephrol*. 2010;20(1):48–50.
- Bartlett S, Cai A, Cairns H. Non-arteritic ischaemic optic neuropathy after first return to haemodialysis. *BMJ Case Rep*. 2011. <https://doi.org/10.1136/bcr.04.2011.4072>.
- Basile C, Addabbo G, Montanaro A. Anterior ischemic optic neuropathy and dialysis: role of hypotension and anemia. *J Nephrol*. 2001;14(5):420–3.
- Kooman J, Basci A, Pizzarelli F, Canaud B, Haage P, Fouque D, et al. EBPG guideline on haemodynamic instability. *Nephrol Dial Transplant*. 2007;22(Suppl 2):ii22–44.
- Sands JJ, Usvyat LA, Sullivan T, Segal JH, Zabetakis P, Kotanko P, et al. Intradialytic hypotension: frequency, sources of variation and correlation with clinical outcome. *Hemodial Int*. 2014;18(2):415–22.
- Al Zubidi N, Zhang J, Spitze A, Yalamanchili S, Lee AG. Pallid disc edema and choroidal perfusion delay in posthemodialysis nonarteritic ischemic optic neuropathy. *Can J Ophthalmol*. 2013;48(5):e120–3.