

Bilateral Anterior Ischaemic Optic Neuropathy in a Child on Continuous Peritoneal Dialysis

Case report and literature review

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الإعتلال العصبي البصري الأمامي غير الشرياني بسبب نقص التروية الدموية لدى طفلة تجري الغسيل البريتوني المستمر

تقرير حالة و مراجعة للأدبيات

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ABSTRACT: Non-arteritic anterior ischaemic optic neuropathy (NAION) is a serious complication of continuous peritoneal dialysis (CPD) which can lead to poor vision and blindness. We report a five-year-old girl who had undergone a bilateral nephrectomy at the age of one year and was on home CPD. She was referred to the Paediatric Ophthalmology Unit of Sultan Qaboos University Hospital, Muscat, Oman, in 2013 with acute bilateral vision loss, preceded by a three-day history of poor oral intake. At presentation, the patient had severe systemic hypotension. An ophthalmological examination revealed severe bilateral visual impairment and NAION. She was treated with intravenous methylprednisolone and normal saline boluses. At a five-month follow-up, the visual acuity of the right eye had improved but vision in the left eye remained the same. Acute bilateral blindness due to NAION while on CPD is a rare condition in childhood. Paediatricians should be aware of this complication in order to ensure prompt management.

Keywords: Anterior Ischemic Optic Neuropathy; Blindness; Hypotension; Pediatrics; Continuous Ambulatory Peritoneal Dialysis; Case Report; Oman.

المخلص: يعد الاعتلال العصبي البصري الأمامي غير الشرياني بسبب نقص التروية الدموية من المضاعفات الخطيرة للغسيل البريتوني المستمر حيث أنه قد يؤدي إلى ضعف البصر والعمى. نعرض هنا حالة طفلة تبلغ من العمر خمس سنوات استؤصلت كليتها عندما كانت في السنة الأولى من العمر وبقيت على الغسيل البريتوني المستمر بالمنزل. وقد تم تحويل الطفلة في عام 2013 إلى وحدة طب العيون للأطفال بمستشفى جامعة السلطان قابوس بمسقط، عمان بعد إصابتها بانخفاض حاد ومفاجئ في البصر. وقد كانت الطفلة ولمدة ثلاثة أيام قبل فقدانها البصر تعاني من انخفاض معدل تناول الشراب والطعام عن طريق الفم. وعند فحص الطفلة بالمستشفى كانت تعاني من هبوط حاد في ضغط الدم. وأظهر فحص العينين عن ضعف حاد في البصر وإعتلال العصب البصري الأمامي غير الشرياني بسبب نقص التروية الدموية. عولجت الطفلة باستخدام دواء الميثيل بريدنيزولون الوريدي وجرعات كبيرة من المحلول الملحي النظامي. وبعد خمسة أشهر من المتابعة لوحظ تحسن في حدة البصر في العين اليمنى بينما بقيت العين اليسرى على حالها. وتجدر الإشارة إلى أن الانخفاض الحاد في البصر بسبب الاعتلال العصبي البصري الأمامي غير الشرياني بسبب نقص التروية الدموية لمرضى الغسيل البريتوني المستمر تعد حالة نادرة في مرحلة الطفولة. وينبغي أن يكون أطباء الأطفال على بينة من هذا التشخيص في مثل هؤلاء المرضى من أجل ضمان تدخل سريع وفعال.

الكلمات المفتاحية: الاعتلال العصبي البصري غير الشرياني بسبب نقص التروية الدموية؛ العمى؛ انخفاض ضغط الدم؛ الأطفال؛ الغسيل البريتوني المستمر؛ المتنقل؛ عرض حالة طبية؛ عمان.

ANTERIOR ISCHAEMIC OPTIC NEUROPATHY (AION) is characterised by sudden vision loss as a result of an infarction of the optic nerve head due to an impairment of blood flow.¹ The majority of cases are non-arteritic (90%); non-arteritic AION (NAION) is associated with numerous risk factors, including diabetes mellitus, atherosclerosis and arterial hypotension, all of which compromise the blood supply of the optic nerve head.¹ The main features of NAION include unilateral vision

loss usually occurring upon waking, ocular pain and headaches.^{2,3}

Bilateral NAION is rare but can occur among patients on dialysis, individuals who experience blood loss and those on medications such as phosphodiesterase type 5 inhibitors and interferon- α .^{4,5} Patients with end-stage renal disease are often managed via continuous peritoneal dialysis (CPD). Episodes of systemic hypotension occur frequently among patients on CPD, thus predisposing these

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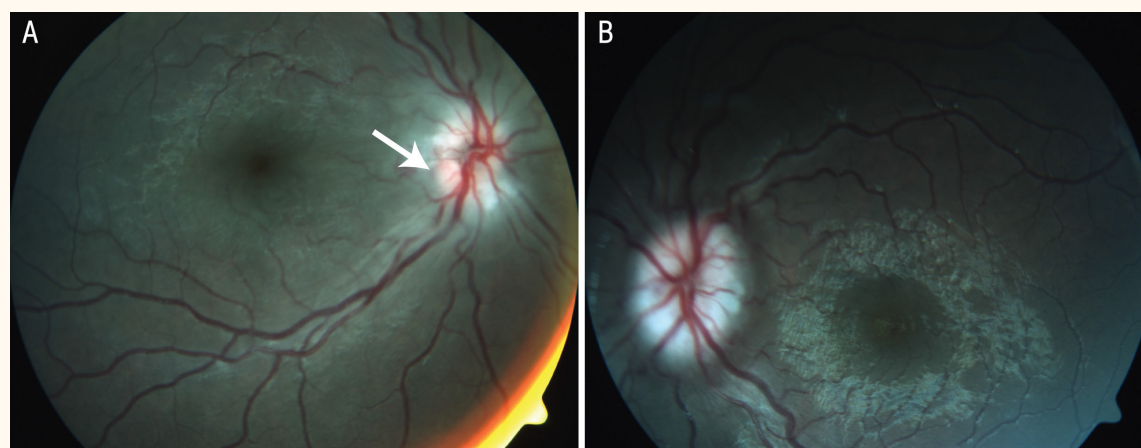


Figure 1: Funduscopy images of a five-year-old child on continuous peritoneal dialysis with acute bilateral vision loss. Bilateral pallid disc oedema with blurred margins was observed in both the (A) right and (B) left eyes. The right eye had an obscured peripapillary nerve fibre layer and tortuous vessels with flame-shaped haemorrhage close to the disc margin (arrow). The disc pallor was more severe in the left eye.

individuals to NAION.⁶ This report describes a five-year-old child who developed acute bilateral blindness due to NAION while on CPD and presents a review of the literature of this serious ophthalmic complication among children undergoing dialysis.

Case Report

A five-year-old girl was referred from the Paediatric Nephrology Unit to the Paediatric Ophthalmology Unit of the Sultan Qaboos University Hospital, Muscat, Oman, in 2013 due to acute bilateral vision loss, noted upon waking from sleep. The mother of the patient reported a history of poor oral intake, dizziness and blurred vision for three days prior to the acute deterioration of vision. There was no history of vomiting or diarrhoea. The patient had undergone a bilateral nephrectomy at the age of one year due to congenital nephrotic syndrome which had been diagnosed when she was 20 days old. Following the procedure, the patient had been on home CPD using Dianeal PD4 Glucose 2.27% (Baxter Healthcare Ltd., Thetford, Norfolk, UK) at six cycles per day for a total duration of 12 hours with a fill volume of 450 mL. Since then, she had suffered periodic episodes of systemic hypotension for which she had been admitted to hospital several times. Six months prior to the current presentation, the patient had been admitted to hospital for the management of posterior reversible encephalopathy syndrome.

At referral, the patient showed no signs of dehydration, although her blood pressure (BP) was very low for her age and height (50/25 mmHg). She received intravenous fluids until her BP returned to normal. A neurological examination was unremarkable apart from the visual impairment. In terms of visual

acuity, an ophthalmological examination revealed light perception in the right eye and counting fingers close to face in her left eye. Visual field testing using the confrontation method was not possible for the right eye due to her low visual acuity; however, the left eye showed superior and nasal field constriction. The pupils were dilated bilaterally, fixed with no direct light reaction in the right eye and sluggishly reacting to light in the left eye. A funduscopy showed bilateral pale optic disc oedema with blurred margins. There was an obscured peripapillary nerve fibre layer and tortuous vessels with flame-shaped haemorrhage close to the disc margin in the right eye, while the disc pallor was more severe in the left eye [Figure 1]. Blood investigations showed haemoglobin levels of 10.4 g/dL and urea levels of 21.4 $\mu\text{mol/L}$, which were considered appropriate as the patient was on CPD; as such, anaemia and uraemia were excluded as causes of the NAION. A magnetic resonance imaging (MRI) scan of the brain showed features consistent with posterior reversible encephalopathy syndrome. No other lesions were observed, thus ruling out infiltrative optic neuropathy and intracranial causes of NAION [Figure 2]. There was no evidence of optic nerve drusen on optical coherence tomography.

The patient was treated with intravenous methylprednisolone at 20 mg/kg/day for five days, followed by oral prednisolone at 2 mg/kg/day for one month as a tapering dose. To prevent anaemia, erythropoietin was also prescribed at an optimised dose. Normal saline boluses were administered to control the hypotension. Dialysis was continued at six cycles per day; however, the duration of each cycle was reduced from one hour to half an hour for two days. An ophthalmological review prior to discharge showed no significant changes. Funduscopy images revealed

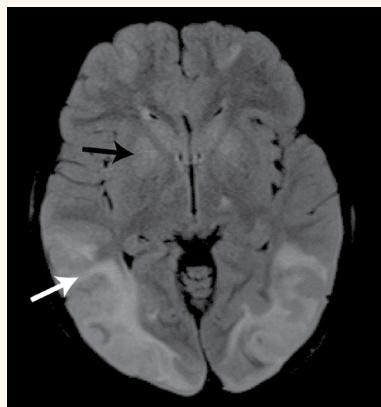


Figure 2: Magnetic resonance imaging of the brain of a five-year-old child on continuous peritoneal dialysis with acute bilateral vision loss. Hypotense T1 and hyperintense T2 signals were noted in the bilateral parietal, occipital cortical and subcortical areas, consistent with vasogenic oedema suggestive of posterior reversible encephalopathy syndrome (white arrow). Bifrontal and bilateral basal ganglionic hyperintensities were also present, likely due to calcification (black arrow). There was no evidence of cerebral venous thrombosis.

that the NAION was resolving with persistent bilateral disc pallor that was still more severe in the left eye compared to the right eye. Upon discharge, the patient's parents were advised of the importance of keeping their child hydrated and monitoring her BP in order to prevent further CPD complications. At a five-month follow-up, improvement was noted in the visual acuity of the right eye (0.6). However, vision in the left eye remained at counting fingers close to face.

Discussion

Apart from NAION, other causes of acute bilateral vision loss include giant cell arteritis, optic nerve *drusen*, uremic optic neuropathy, infiltrative optic neuropathy and intracranial space-occupying lesions leading to optic nerve compression.⁷⁻¹⁰ However, the age of the patient in the current case was not consistent with a diagnosis of giant cell arteritis, which typically presents in old age.¹¹ Moreover, NAION secondary to anaemia or uremic optic neuropathy were excluded following blood investigations while optic nerve *drusen*, infiltrative optic neuropathies and intracranial space-occupying lesions were ruled out via optical coherence tomography and MRI. Due to the presentation, NAION was considered to be the underlying aetiology for the visual impairment.

Although bilateral NAION is a rare disease among children, a few cases have been previously described in the literature; most children present with loss of light perception, visual fixation, ocular pursuit movements

and bilateral mydriasis unreactive to bright light.¹²⁻¹⁵ In general, NAION is associated with low BP and hypovolemia.¹²⁻¹⁶ It can occur following parainfectious or post-vaccine optic neuritis as well; however, visual loss in these cases is usually bilateral and more severe at onset.¹⁷ NAION has also been documented among children undergoing major surgical procedures.¹⁶ It is important to rule out other possible causes of NAION, such as uremic optic neuropathy, increased intracranial pressure and drug toxicity.⁸⁻¹⁰ Nanji *et al.* reported an association with optic nerve *drusen*, which are calcific deposits that form in the optic nerve head secondary to abnormalities in axonal metabolism and degeneration.⁷ All of these factors were ruled out in the current patient; however, she had had periodic episodes of hypotension preceding the vision loss. As such, the cause of NAION was attributed to hypotension-induced low perfusion and *ischaemia* of the optic nerve.

The pathophysiology of AION is due to the susceptibility of the optic nerve to *ischaemia* and the tendency of the retina to develop an ischaemic injury; any increase in intraocular pressure or decrease in perfusion may lead to *ischaemia* of the optic nerve and retina.¹⁸ Moreover, the condition has also been reported in association with local anatomical and systemic factors such as the presence of a small optic nerve head with little cupping, intraocular hypertension, systemic hypertension, hypovolemia and anaemia.¹³ Hypotension is considered among the main causes of decreased perfusion to the micro-circulation of the optic disc and retina.¹ Chronic hypotension has been reported in children with NAION.¹⁴ Hypotension has also been shown to occur during sleep; among 925 cases of NAION, 73% reported visual loss on waking from sleep.¹⁹ In the present case, the patient was found to be hypotensive on admission and reported the loss of vision upon waking from sleep. Many causes can lead to chronic hypotension among patients on CPD, including hypovolemia, the use of anti-hypertensive medications and removal of vasopressors during dialysis.²⁰

Poor visual outcomes following NAION have been reported among both children and adults on CPD.^{6,10,21} The treatment of AION involves the reversal of hypotension in order to maintain adequate perfusion to the optic nerve. The use of steroids is controversial, with some studies showing a positive outcome.^{3,22,23} The use of combined levodopa and carbidopa has been found to improve visual acuity in patients with AION.²⁴ In addition, significant improvement in vision has been reported with steroids and levodopa.¹² Steroids and prostaglandins have also been reported to be effective.²⁵ The current patient was managed

with intravenous saline in addition to intravenous methylprednisolone followed by oral prednisolone. Since the main aetiology of the condition in this case was hypotension, her parents were counselled to pay close attention to her hydration and BP levels after discharge in order to identify hypovolemia and prevent complications.

Conclusion

Children undergoing CPD are at risk of sudden blindness due to NAION, mainly as a result of chronic hypotension. Visual outcomes after treatment are generally poor. Therefore, prompt management of hypotension and hypovolemia in children on CPD is necessary in order to prevent the development of this serious complication.

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