CASE REPORT

Bilateral posterior ischaemic optic neuropathy after severe diabetic ketoacidosis, cardiopulmonary resuscitation and respiratory failure

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SUMMARY

A 44-year-old male European with type I diabetes mellitus fell into diabetic ketoacidosis. In the emergency room, he developed an episode of asystole and respiratory failure requiring one cycle of cardiopulmonary resuscitation and extracorporeal membrane oxygenation (ECMO). Waking up 7 days later, he presented a bilateral complete loss of vision. Ophthalmological examination including fundoscopy on days 1 and 10, after extubation, showed bilateral large round pupils non-reactive to light and a normal fundus. Neuroimaging studies, including MRI and MRA of the brain, were all within normal limits. A lumbar puncture and comprehensive serological testing excluded an infectious or rheumatic cause. An empirical high-dose intravenous steroid treatment administered for 5 days had no effect on his vision. His eye examination at 1.5 months follow-up showed a normal fundus except for progressive bilateral optic nerve disc pallor, which pointed towards the diagnosis of a posterior ischaemic optic neuropathy.

BACKGROUND

Ischaemic optic neuropathy is an acute ischaemia of the optic nerve, in either its posterior or anterior portion. Posterior ischaemic optic neuropathy (PION) is less common than anterior ischaemic optic neuropathy.¹ Differentiation between these two types of ischaemia is based on different sources of blood supply to these two parts of the nerve. The anterior part, which is defined as the optic nerve head, is supplied by the posterior ciliary artery. The posterior part, which is defined as everything but the optic nerve head, is supplied from several sources and has no blood supply by a defined artery.^{2,3}

PION usually manifests as a sudden-onset visual loss with possible bilateral involvement. According to a retrospective chart review from 2001, there are three main types of PION: perioperative, arteritic and non-arteritic. Arteritic PION usually occurred in patients with a giant cell (temporal) arteritis, while non-arteritic PION was associated with a systemic vascular disease.⁴

CASE PRESENTATION

The patient was found in diabetic ketoacidosis (DKA) at home after he had tried to maintain his blood glucose level without long-term insulin. The patient was known to have long-standing diabetes mellitus type I, and alcohol and benzodiazepine

abuse. At admission, his body temperature was 33.1°C; blood pressure was 97/37 mm Hg with a pulse rate of 83/min; blood pH was 6.9; blood glucose level was 68 mmol/l, and he had a positive toxicology screening for benzodiazepine use. His eyes were described as equal, round and bilaterally reacting to light. In the emergency room, he suffered from an episode of asystole requiring one cycle (2 min) of cardiopulmonary resuscitation, but no adrenaline at that moment. He was then intubated, hydrated with intravenous fluids, and treated with an insulin drip. He also received catecholamines, including dobutamine and noradrenaline, for 1 day. He received venovenous ECMO because of an evolving respiratory failure owing to an aspiration pneumonia. His DKA and respiratory failure resolved over the next few days. After 7 days in the intensive care unit, upon waking, he had a bilateral complete loss of vision.

INVESTIGATIONS

Ophthalmological examination including fundoscopy on days 1 and 10 after extubation showed bilateral large round pupils that were non-reactive to light and a normal fundus. Neuroimaging studies, including MRI and MRA of the brain, were all within normal limits, including a normal presentation of the optic nerve. A lumbar puncture and comprehensive serological testing (antinuclear antibody, antineutrophil cytoplasmic antibodies, anti-cardiolipin antibodies, syphilis and Lyme disease in the liquor) ruled out any infectious or rheumatic cause, and showed only a slight blood-brain barrier disturbance. The patient had no clinical symptoms consistent with temporal arteritis, and sonography of the temporal arteries was normal.

TREATMENT

An empirical high-dose intravenous steroid treatment for 5 days had no effect on his vision. We started with a long-term treatment with 100 mg acetylsalicylic acid/day.

OUTCOME AND FOLLOW-UP

His eye examination at 1.5 months follow-up showed a persisting complete loss of vision with a normal fundus, except for progressive bilateral optic nerve disc pallor. Otherwise, he recovered completely.

DISCUSSION

A combination of several clinical findings is highly suggestive of PION. Patients typically present with

To cite: Wirth CD, Leitner C, Perrig M. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-008291

a sudden and painless loss of vision, usually of one eye, with no light perception and pupils non-reactive to light. They also typically show normal fundus and optic nerve on initial ophthalmoscopy, with optic nerve atrophy in subsequent weeks and months.⁵ All of these conditions were present in this patient except for the monolateral manifestation.

DKA as a severe complication of diabetes mellitus is rarely associated with an optic nerve atrophy: the first case we found was a 15-year-old diabetic patient who survived DKA, complicated by acute severe encephalopathy, hypopituitarism and atrophy of the optic nerve.⁶ The next case was an 82-year-old woman who developed bilateral optic neuropathy following a massive gastrointestinal haemorrhage, resulting in myocardial and cerebral infarction and complicated by DKA.⁷ Another case was a 50-year-old man who lost vision of his right eye after severe DKA and only retained partial vision in his left eye. Bilateral optic atrophy was diagnosed.⁸ Another case is a 58-year-old man with insulin-dependent diabetes mellitus and myocardial infarction, who was admitted with the diagnosis of acute pancreatitis complicated by sepsis and DKA. Bilateral ischaemic optic neuropathy was diagnosed.⁹ The most recent case found was a 35-year-old man with DKA who developed rhabdomyolysis, and acute renal and respiratory failure. Bilateral PION was diagnosed.¹⁰

The pathogenesis behind non-arteritic and perioperative PION is not completely clear. The central portion of the retrobulbar part of the optic nerve has a very poor vascularisation, which makes it more vulnerable to ischaemia during hypotension.¹¹ Ischaemic optic atrophy after surgery is a rare disease and was only significantly associated with age, male gender and surgery type.¹² Bilateral involvement was present in 70% of perioperative cases, and patients' loss of vision was worse than that in non-arteritic cases.⁴ Many authors discuss the effects of severe hypotension and anaemia as being the main factors for perioperative PION.^{13–14} Other pathogenic factors described for ischaemic optic neuropathy include use of vasopressors and prolonged cardiopulmonary bypass time in open-heart operations.¹⁵

In our case, DKA was accompanied with the need for resuscitation, which leads to severe hypotension. That might be the main cause for the PION, but the use of catecholamines and ECMO might also be important factors.

Patients with non-arteritic PION are older than patients with a perioperative PION. They usually have only one eye affected, and they present with evidence of a systemic vascular disease or vascular disease risk factors, including hypertension, diabetes mellitus, hypercholesterolaemia, cardiac and cerebrovascular disease.⁴ This patient had no other known risk factors for atherosclerosis than his type 1 diabetes, and no documented diabetic organ manifestation. In synopsis, our patient presented a more typical clinical picture of a patient with perioperative PION than non-arteritic PION. However, to prevent further vascular events, we initiated a prophylactic anticoagulation with low-dose acetylsalicylic acid.

The prognosis depends upon the subtype of PION. Perioperative PION is usually bilateral, severe and irreversible,¹⁶ and no effective treatment has been found.¹⁷ On the other

hand, non-arteritic and arteritic PION is improved by the use of systemic steroid therapy, and there have been cases of spontaneous improvement in vision.^{17–18} The vision of our patient did not improve after the high-dose steroid therapy.

Learning points

- This case adds to the growing list of non-surgical and non-arteritic posterior ischaemic optic neuropathy (PION). We found no other case in the literature that described a bilateral PION caused by diabetic ketoacidosis (DKA), cardiopulmonary resuscitation and extracorporeal membrane oxygenation.
- Early treatment of DKA, severe hypotension and respiratory failure should be the cornerstone to prevent PION in these patients.
- An arteritic cause for a sudden loss of vision should always be excluded.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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