Sir,

Two unusual cases of visual loss following severe non-surgical blood loss

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Non-arteritic anterior ischaemic optic neuropathy (NAION) is associated with systemic disorders such as hypertension, diabetes mellitus, chronic renal failure, atherosclerosis and vasculitis.¹ Many cases of NAION occur spontaneously; however, acute hypotension and anaemia are significant causative factors in the perioperative period.² Loss of vision after recurrent blood loss following surgery has been documented.³

Ischaemic optic neuropathy is an uncommon complication of severe blood loss, usually occurring in older patients but under special circumstances also occurring in young adults.^{1,4} The following cases of non-surgical blood loss illustrate these two entities and highlight the role of additional factors in the pathogenesis of the optic nerve damage.

Case reports

Case 1 A 37-year-old woman was seen in the Eye casualty with a 3-day history of blurred vision and photopsia of the left eye. She had been admitted under the Gynaecologists 4 days earlier with per vaginal bleeding. Her haemoglobin had fallen from 13.4 g/dl on admission, to 7.7 g/dl (normal range 11.5–16.5). On admission she was haemodynamically stable with a compensatory tachycardia of 120 bpm and blood pressure of 122/84. Blood loss was sufficient to warrant a transfusion of 4 units of blood. Hysteroscopy showed her to have a fibroid uterus. She was otherwise fit and well. She was taking regular Propranolol for the symptomatic relief of anxiety.

Visual acuities were 6/5 and N5 in the right eye and 6/6 N5 in the left. Colour vision using Ishihara plates was 16/17 in the right eye and 13/17 in the left. There was a small left relative afferent pupillary defect. Examination of the posterior pole revealed oedema of the left optic disc and dilated surface capillaries (Figure 1, top), with a small crowded right optic disc (Figure 2). Extraocular movements were entirely normal with no associated pain. Humphrey visual field analysis revealed extensive left sided visual field losses. A diagnosis of NAION secondary to hypovolaemia was made.

The patient was reviewed at 7 months and the visual acuities remained unchanged. However, visual field examination revealed gross constriction of the left visual field. Fundal examination revealed pallor of the left optic disc (Figure 1, bottom).

Figure 1 (Top) Fundus photograph on presentation showing oedema of the left optic disc with dilated surface capillaries. (Bottom) Fundus photograph taken 4 months after presentation showing pallor of the left optic disc.

Figure 2 Fundus photograph of the small crowded right optic disc, with no physiological cup.

Case 2 A 60-year-old man presented with epistaxis. He had no previous history of epistaxis or trauma. He was on no regular medication and was otherwise well. He was a heavy smoker of 40/day for the last 30 years. He had a strong family history of ischaemic heart disease. His blood pressure was 103/52 and pulse was 125 bpm. He was given one litre of normal saline and admitted. The ENT team eventually controlled the bleeding after 3 days, by nasal packing, cautery and balloon tamponade. His haemoglobin on admission was 17.7 g/dl and on discharge was 12.2 g/dl. He did not receive a blood transfusion during this time.

He re-attended casualty 1 week later complaining of a sudden deterioration of vision of the left eye, which he described as blurred especially inferiorly. There had been no further episodes of epistaxis, but he had felt tired and dizzy since discharge. There were no symptoms or signs of giant cell arteritis. Visual acuities were 6/6 in the right eye and 6/60 in the left. He had a left relative afferent pupillary defect, and a dense inferior altitudinal visual field defect (Figure 3). Dilated fundoscopy revealed a healthy right optic disc and an oedematous left optic nerve head especially superiorly. Two cotton wool spots were seen within the temporal vascular arcade at the right posterior pole and one cotton wool spot within the left temporal arcade.

He was very pale and short of breath. Blood pressure was 100/50. Heart rate was 110 bpm. Haemoglobin was 7.7 g/dl. An erythrocyte sedimentation rate (ESR) was performed and found to be 127 mm/h. A diagnosis of NAION secondary to hypotension and anaemia was made. He was admitted and transfused seven units of blood in total. His haemoglobin at discharge was 14.0 g/dl. Two weeks later vision was unchanged. The left optic disc was less swollen and the ESR was 46 mm/h.

He was reviewed 4 months later. Visual acuity in the left eye had improved to 6/24, however the dense inferior altitudinal field defect had remained unchanged. The left optic disc oedema had now resolved, leaving a pale superior optic nerve head. Haemoglobin was 17.2 g/dl and his ESR was 2 mm/h.

Comment

NAION is the most common acute optic neuropathy in patients over 50 years of age,⁴ with infarction of the optic nerve head. Factors involved with the arteritic variety are well understood, but less is known about the more common non-arteritic variety. Severe blood loss in itself is generally not sufficient to cause ischaemic optic neuropathy, indeed some authorities believe that it never follows a single haemorrhage in a

person in normal health.³ Co-existing vascular disease increases the risk in older patients, such as in Case 2.

The risk factors for NAION can be divided into two groups: ocular and systemic. The systemic factors that NAION is clearly related to include ischaemic heart disease, hypercholesterolaemia, hypertension, and diabetes mellitus.¹

There is increasing evidence that NAION is associated with nocturnal hypotension, or a failure of blood pressure to recover normally from its normal nocturnal dip which in turn may be exacerbated by over-zealous treatment of systemic hypertension.^{5,6} Thus, nocturnal hypotension may be the final insult in a multifactorial situation.⁵ On ambulatory measurements of diurnal blood pressure, patients with NAION consistently had a lower mean blood pressure than did control subjects and a lag in the usual rise in blood pressure in the morning to meet increasing daytime demands for perfusion.⁶ Chronic hypoperfusion of small end-arterial vessels that supply the optic nerve head may predispose to NAION, and may be caused by relative hypotension owing to overtreated hypertension or to abnormal vascular autoregulation. Blood pressure should thus be monitored carefully when treating hypertensive patients who are at risk for NAION, to avoid hypotension, especially on awakening in the morning.⁶ A better understanding of variations between blood pressure and intraocular pressure could provide more information about circulation to the optic nerve head itself. Moreover, the role of autoregulation at the optic nerve head (or lack of) as well as the role of vascular endothelium-mediated factors could be important aetiological factors.

The ocular risk factors for NAION include refractive state, intraocular pressure, and cup-disc ratio of the fellow eye. One study⁷ found that most fellow eyes (73%) had a cup-disc ratio not more than 0.1. Therefore, the crowded effect of the nerve head may induce a circulatory compromised disc, the 'disc-at-risk'.⁷ It may play an important role in NAION. It is thus interesting that Case 1 possessed small, anatomically predisposed optic nerves.⁸

Case 1 was unusual in that she was young and had no systemic vascular risk factors for NAION. In addition, she had only sustained one episode of bleeding, whereas generally it is felt that repeated episodes are a prerequisite to NAION.³ Younger patients usually have an illness in the background that puts them at an increased risk for the development of NAION. Moreover, acute systemic hypotension alone without anaemia is unlikely to cause optic nerve infarction.⁹ Pre-existing anaemia may be an additional factor when there has been previous chronic blood

Figure 3 Humphrey visual field analysis showing a dense left inferior altitudinal visual field defect.

loss, such as in cases following upper gastro-intestinal haemorrhage.² It is unlikely that Case 1 had an underlying chronic anaemia since the initial haemoglobin measured on admission in casualty was normal, before haemodilution effects could take place.

Prolonged hypotension, either intentional to reduce bleeding from the operation site or as a result of the blood loss in an anaesthetised patient, contributes in cases occurring after surgery.² Inability to compensate for blood loss, due to her systemic beta-blockade (propranolol), is likely to have been the important additional factor in Case 1. We propose that this blockade may well have compromised the ability to mount an adequate compensatory cardiovascular response. In particular, it is likely to have

compromised vasoconstriction and reflex tachycardia and thus helped to critically compromise optic nerve head perfusion.

It is also tempting to suggest that there is a congenital anomaly of the optic disc that makes it more susceptible to ischaemia than in normal individuals.⁸ Indeed Case 1 had 'discs-at-risk', the discs being small and crowded with a CDR of less than 0.1.

Case 2 highlights several important diagnostic and management issues. In the first instance it is often poorly appreciated that a significant epistaxis can cause infarction of the optic nerve head with potentially devastating irreversible visual consequences. This complication may well be avoidable if the visual risk is appreciated, leading to the prompt correction of

anaemia and hypotension. It is also important to assess an individual's additional risk factors for NAION following blood loss. The patient in Case 2 had significant atherosclerosis risk factors, in that he was a longstanding heavy smoker with a very strong family history of atherosclerotic complications. Thus, he was at greater risk of suffering critical end organ ischaemia following sudden blood loss. Furthermore he was not transfused during his initial admission despite a significant fall in the haemoglobin level and was also discharged before adequate time had been given for haemodilution effects to fully take place.

The distinction between the arteritic and non-arteritic forms of ischaemic optic neuropathy is often aided by measuring the ESR. In this case the ESR was significantly elevated and could thus pose a diagnostic difficulty. However our patient had neither any symptomatology nor specific clinical findings consistent with a diagnosis of giant cell arteritis. The markedly elevated ESR in Case 2 may have reflected the degree of haemodilution rather than indicating underlying giant cell arteritis.¹⁰ Common causes of an ESR over 100 mm/h include: multiple myeloma, carcinomatosis, giant cell arteritis, leukaemia, lymphoma and overwhelming sepsis. However, an important cause of raised ESR that is often overlooked is anaemia. In Case 2 the ESR progressively fell over a period of a week from 127 mm/h to 46 mm/h. Four months later the ESR was 2 mm/h. A high index of suspicion should be maintained in patients with elevated sedimentation rates and ischaemic optic neuropathies. If any clinical doubts exist, steroid treatment should be initiated and a temporal artery biopsy performed. In relation to Case 2, steroid treatment on the basis of a significantly raised ESR would have been inappropriate. The cotton wool spots detected on fundoscopy in Case 2 represented retinal nerve fibre layer infarcts secondary to anaemia.

The occurrence of the same pattern of optic nerve damage after severe blood loss suggests that watershed infarction is an important mechanism in the pathogenesis of NAION. The following hypothesis for three general types of visual loss after hypotension has been proposed.⁹ Profound hypotension without anaemia or arteriosclerosis causes watershed infarctions in parietal and occipital lobes, with the optic nerves tending to be spared. By contrast, brief hypotension in a patient with arteriosclerosis is likely to cause juxtalamina optic nerve infarction, indistinguishable from spontaneous anterior ischaemic optic neuropathy (Case 2). Whereas, hypotension in an anaemic patient without arteriosclerosis risk factors, favours infarction in the orbital optic nerve.

However, Case 1 had no pre-existing anaemia, nor

any arteriosclerosis risk factors. We propose that the use of beta-blockers and a small crowded optic disc-at-risk were the important additional factors to the hypotension.

Management of NAION involves reduction of risk factors to prevent development of the disease. The major goal in the treatment of NAION secondary to hypovolaemia is to reverse the systemic hypotension and to correct anaemia.² In Case 2 the visual acuity partially recovered, but prompt intervention has been reported to result in partial recovery of vision in hypotension-induced ischaemic optic neuropathy. The importance of lowering the intraocular pressure by using acetazolamide and topical timolol, in addition to the correction of hypotension and anaemia has been reported.¹¹

Awareness of potential visual morbidity associated with chronic anaemia or acute systemic hypotension secondary to blood loss, even in young patients, and the realisation of the importance of rapid treatment, may lessen the incidence and severity of NAION. Prompt intervention can result in partial recovery of vision. Spontaneous NAION has no immediate treatment other than the treatment of any associated atherosclerosis risk factors. There is however some evidence that Aspirin may have a role to play in reducing the frequency of second eye involvement in patients with spontaneous non-arteritic anterior ischaemic optic neuropathy.¹²

The reasons for unilateral rather than bilateral visual loss in the above cases are unknown. Possible explanations may be that one disc was more at risk than the other, or the unaffected disc may be damaged in a subclinical manner.

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Possible cat scratch disease causing neuroretinitis and CRVO in a child

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Bartonella are gram-negative rods.¹ Of the 14 known species, five are associated with human disease. *B. henselae* causes cat scratch disease and *B. quintana* causes trench fever. *B. henselae* infections often present with fever, malaise and lymphadenopathy. A history of contact with a cat is not always present. Eye involvement can cause Parinaud oculoglandular fever, neuroretinitis^{2,3} and an inflammatory mass of the optic nerve head.⁴

Case report

A 9-year-old male presented with a history of poor vision for 2 days, with no other specific medical or ocular history of note. There was no pain, lymphadenopathy, fever or trauma. Physical examination revealed a healthy male with normal blood pressure, urinalysis, without hepatosplenomegaly, lymphadenopathy or anaemia.

Ophthalmological examination revealed a visual acuity of 6/5 in the right eye and 3/60 in the left with a dense relative afferent pupillary defect. There was no anterior segment or vitreous inflammation. The left retina had widespread hemorrhages with tortuous vessels with an area of possible focal retinitis involving the papillo-macular bundle, and a early macular star (Figure 1). The right fundus was normal.

Investigations, including: FBC, U & E, clotting and thromophilia screen, LFT, autoantibody screen, serum cholesterol, HDL and triglycerides, ESR, serum homocysteine, MRI scan, echocardiogram and ocular ultrasound, were normal. Fundus fluorescein angiogram was not possible. Indirect immunofluorescence titres for anti-*B. henselae*, taken 3 weeks after initial presentation, were one in 64. There was a cross-reactivity with *B. quintana* which was reported as one in 128 IgG (IgM negative).

A tentative diagnosis was made of neuroretinitis secondary to *B. henselae* with a subsequent central retinal vein occlusion. A 2-week course of 250 mg Clarythromycin b.d. was given. Further questioning elicited a history of contact with a stray kitten while on holiday in Italy 5 weeks prior to presentation. The titers were taken 8 weeks after this exposure. Subsequent titers were unchanged. Three months later, the vision had improved to 6/6. The retinal appearances had returned to normal with the exception of mild increased tortuosity.

Figure 1 Left ocular fundus.