VISION LOSS 2 WEEKS AFTER UNEVENTFUL OPTIC NERVE SHEATH FENESTRATION SURGERY MAY BE ATTRIBUTED TO RETINAL REPERFUSION INJURY

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Purpose: The purpose of this study was to report a rare case of retinal reperfusion injury after optic nerve sheath fenestration surgery.

Patients and Methods: Single patient case report.

Results: A 58-year-old African American woman presented with severe visual loss in the right eye 2 weeks after optic nerve sheath fenestration surgery. Retinal examination at presentation showed severe arterial narrowing and the appearance of new intraretinal hemorrhages. Fluorescein angiography demonstrated delayed venous filling.

Conclusion: Severe visual loss after optic nerve sheath fenestration procedure is uncommon. This case may represent the first reported case of reperfusion injury in human eyes.


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Case Report

A 58-year-old African American woman was referred to the University of Louisville’s Retina Service for evaluation of an unexplained visual loss in the right eye that occurred 2 weeks after an uneventful optic nerve sheath fenestration (ONSF) procedure on the right.

The patient was known to have suffered from idiopathic intracranial hypertension over the last 10 years. Previous medical treatment by oral acetazolamide was discontinued due to patient’s intolerance. She therefore underwent lumboperitoneal shunt procedure; however, the shunt had to be removed due to a methicillin-resistant Staphylococcus aureus infection. The patient noticed that her vision is deteriorating progressively over the last 6 months before the surgery. Her opening intracranial pressure (ICP) as measured 2 weeks before the surgery confirmed the presence of high ICP. Her preoperative visual fields showed marked visual field constriction. Optic nerve sheath fenestration procedure on the right side was then performed due to the presence of progressive vision loss and failure of other means to control her ICP. The vision of the right eye was 20/30 at the time of surgery.

The patient’s medical history was positive for type 2 diabetes mellitus, systemic hypertension, kidney stones, chronic obstructive pulmonary disease, and osteoporosis. She was on multiple systemic medications including furosemide, Glimepiride, hydralazine, allopurinol, and hydrocodone.

Funduscopic preoperative examination showed bilateral advanced papilledema and severe arteriolar narrowing (Figure 1), secondary to systemic hypertension. Optic nerve sheath fenestration surgery performed by the oculoplastic service was uneventful, without any evidence of hemorrhage, direct vascular injury, or obvious optic nerve trauma.

At the immediate postoperative period, the patient had initial visual improvement. However, her vision started to decrease gradually over a period of 2 weeks. There was no catastrophic sudden visual loss; rather, the patient noticed gradual visual deterioration. Two weeks postoperatively, the vision in the right eye dropped to hands motion. A relative afferent pupillary defect was noted. Funduscopic examination of the right eye revealed that the swelling of the disk has indeed subsided, but a dull foveal reflex and scattered new microscropic intraretinal hemorrhages in the posterior pole along the superior temporal arcade were noted (Figure 2). The left eye examination remained unchanged. Optical coherence tomography of both eyes was within normal limits.

Fluorescein angiography revealed delayed venous filling mainly involving the superior temporal branch, multiple hypofluorescent spots corresponding to the retinal hemorrhages, and a few small

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hyperfluorescent spots suggestive of microaneurysms (Figure 3). No macular edema or macular ischemia was noted.

The differential diagnosis in this case would be of a combined central retinal artery occlusion and branch retinal vein occlusion secondary to ONSF procedure. Other possibilities to consider are ophthalmic artery ischemia, compressive postoperative hematoma, traumatic optic neuropathy, and retinal reperfusion injury. The patient underwent extensive medical and radiologic workup including magnetic resonance angiography of the brain and orbits and carotid artery sonography, which showed no evidence of edema or hematoma around the optic nerve and no evidence of carotid artery occlusion.

Discussion

Optic nerve sheath fenestration is a surgical technique that is indicated to save vision in patients with increased ICP that is not responding to other treatments. The basic mechanism by which ONSF works is by creating a window in the dural sheath of the optic nerve. This opening acts as a fistula through which cerebrospinal fluid leaks in the orbit and reduces the compression around the optic nerve. It is reserved usually for patients who show signs of progressive loss of vision secondary to optic nerve compression but with residual vision.

The procedure is highly successful in stabilizing or improving the vision. It has been estimated that 88%–94% of patients either stabilize or have visual improvement after the procedure. However, there is a considerable rate of reoperation, 10%–32%, due to failure of the initial procedure and progression of the disease. The procedure carries the risk of certain complications, which may be up to 45%. However, most of the complications are trivial and not visually significant. The most commonly reported complications include diplopia due to manipulation of extraocular muscles during the procedure, mainly the medial rectus, atonic pupil, and anisocoria that are believed to be due to interruption of the blood supply to the ciliary ganglion. Orbital cellulitis has also been reported. Unfortunately, the procedure may result in permanent and severe loss of vision in less than 2% of cases. The most commonly reported causes of this visual loss were central retinal artery occlusion, branch retinal artery occlusion, and traumatic optic neuropathy.

This patient had advanced long-standing papilledema secondary to increased ICP. Hypertension-
related papillopathy should be considered in the differential diagnosis but seemed unlikely because the retina lacked other manifestations of hypertensive retinopathy such as cotton wool spots, retinal hemorrhages, and macular edema. She presented with gradual-onset vision loss in the early postoperative period with the appearance of retinal hemorrhages.

The presence of extreme preoperative conditions leading to chronic low retinal oxygenation due to the patient’s chronic obstructive pulmonary disease, chronic hypoxia, and uncontrolled systemic hypertension, aggravated by chronic papilledema triggered us to consider of the possibility that this patient may have suffered from retinal ischemia–reperfusion injury. Although papilledema is not a usual cause of retinal ischemia per se, the presence of long-standing papilledema may have further compromised the retinal blood flow to the ischemic retina. When the papilledema was suddenly relieved by ONSF surgery, abrupt reperfusion to the ischemic retina may have caused reperfusion injury to the retina, leading to gradual clinical visual loss.

Reperfusion injury has been demonstrated in other tissues other than the eye including the neural tissues after reperfusion of ischemic strokes and the myocardium after reperfusion therapy of acute myocardial infarction. The postulated mechanism that underlies this type of injury includes severe oxidative stress and electrolyte and acid–base disturbances. Animal experiments showed the possibility of ischemia–reperfusion injury in the eye induced by oxygen-free radicals. It also showed the role of different antioxidant mechanisms in reducing the severity of oxidative injury. In our case, releasing the optic nerve compression and retinal artery compression ONSF procedure may have led to a massive and sudden increase of the blood supply to the ocular tissues, followed by alterations in the interior milieu and cell death. The reperfusion-induced hyperoxia results in the generation of oxygen-free radicals in high concentration, which results in further cellular damage. That can manifest clinically in the form of hemorrhagic retinal ischemia.

To our best knowledge, this entity has never been reported in human eyes and this case is the first reported case with documentation of the possibility of ischemia–reperfusion injury. Preexisting conditions of severe chronic retinal ischemia secondary to systemic diseases may contribute to clinically evident reperfusion injury after surgical intervention. Further research is required to study the pathophysiology and the underlying mechanism of this type of injury in the human retina.

**Key words:** reperfusion, papilledema, optic nerve, ischemia.

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**Fig. 3.** Red-free (A), fluorescein angiography arterial phase (B), mid-AV phase (C), and recirculation phase (D) of the right eye show delayed venous filling along the superotemporal branch (black arrows), hypo-fluorescent spots corresponding to retinal hemorrhages (white arrows) and a few microaneurysms (arrowheads).
References