

Case Report

Spontaneous hyphema and pupillary block in a patient with a left ventricular assist device

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Summary

The left ventricular assist device (LVAD) has been a standard of care for the management of patients with advanced heart failure since the 1990s. An increased risk of spontaneous bleeding related to the device has been noted, ranging from minor epistaxis to major thoracic and mediastinal hemorrhages. To our knowledge, intraocular hemorrhage has not been previously reported. We report a 72-year-old patient with an LVAD who subsequently developed a spontaneous intraocular hemorrhage that manifested as hyphema, pupillary block, and acute intraocular pressure elevation.

Case Report

A 72-year-old man with a history of ischemic cardiomyopathy with prior 5-vessel coronary artery bypass and a continuous-flow left ventricular assist device (LVAD) placed 6 months prior awoke with 10/10 pain and loss of vision in the right eye. Recent medical history included an ischemic stroke suffered 1 week after the LVAD procedure and 2 episodes of gastrointestinal (GI) bleeding with arteriovenous malformations (AVMs) found on endoscopy. Ocular history included uneventful bilateral cataract extraction and intraocular lens (IOL) implantation approximately 10 years prior and non-exudative, age-related macular degeneration in both eyes. There was no recent trauma. His medicines included 2 mg warfarin daily with a goal international normalized ratio (INR) of 1.8–2.5, which had been reduced to 1.8–2.2 after the GI bleeds.

On initial evaluation, visual acuity in the right eye was light perception; intraocular pressure (IOP) in the right eye by Tono-Pen (Reichert Technologies, Depew, NY) was 87 mm Hg. The anterior chamber was deep with a 1.5 mm crescent-shaped layering hyphema. The iris was flat, without neovascularization, and the angle was open, with a deep chamber by Van Herick slit-lamp examination. The pupil was unreactive, and the entire pupillary aperture was filled with a blood clot obstructing the view to the posterior segment (Figure 1). B-mode ultra-

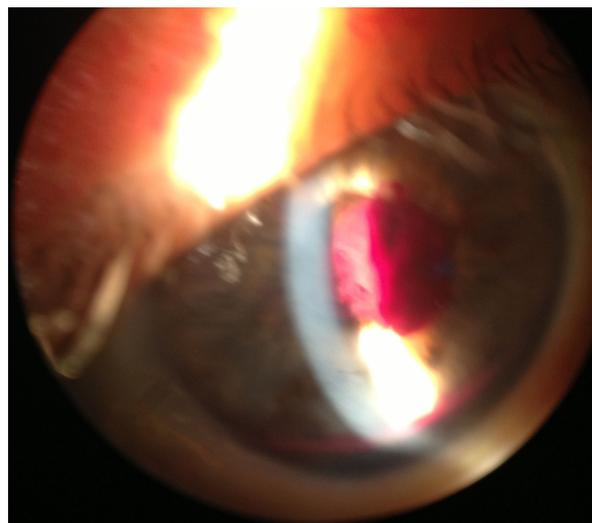


Figure 1. Bedside photography with a handheld portable slit-lamp showing a 1.5 mm crescent-shaped layering hyphema in the anterior chamber with a blood clot filling the pupillary aperture and obstructing the view to the posterior segment.

sonography revealed a vitreous opacity consistent with blood in the anterior vitreous. The left eye was unremarkable, with an open angle. The IOP dropped to 46 mm Hg 1 hour after administration of oral acetazolamide 500 mg and topical timolol 0.5%, 1 drop times 2

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doses every 5 minutes. Two hours later, IOP was 17 mm Hg; the patient's pain abated with additional oral acetazolamide and topical brimonidine 0.2%, timolol 0.5%, latanoprost 0.005%, prednisolone, and atropine solutions. INR on admission was found to be elevated at 1.7, slightly below the goal therapeutic range for the LVAD. Prothrombin time was 14.9. In collaboration with cardiology, warfarin was stopped because of the ocular bleed. Computed tomography imaging was negative for intracranial hemorrhage. The patient was admitted to the coronary care unit and monitored closely for signs and symptoms of thrombosis.

Twelve hours later, the patient's 10/10 eye pain returned. The IOP measured 58 mm Hg, with a shallow anterior chamber and 360 degrees of anterior iris bowing. Repeat B-scan was unchanged. An acute angle closure secondary to pupillary block from the blood clot was suspected. Anterior chamber paracentesis and laser iridotomy were considered but deferred due to very shallow anterior chamber, high risk of rebleeding, and potential hemodynamic compromise during transport to the laser facility. Additional acetazolamide and intravenous mannitol were administered with cardiology approval and close monitoring of fluid, electrolyte, and respiratory status before and after administration. Six hours later, IOP had returned to 26 mm Hg, with resolution of pain and deepening of the anterior chamber (Figure 2).

In the subsequent days, the IOP remained in the 20–30 mm Hg range as the patient received acetazolamide 250 mg every 6 hours and topical timolol 0.5% twice daily, brimonidine 0.2% 3 times daily, dorzolamide 2% 3 times daily, and latanoprost 0.005% at night. The inferior layered hyphema decreased to 10%, with gradual deepening of the anterior chamber and resolution of the pupillary blood clot. Gonioscopy was unremarkable, without neovascularization of the angle. Three months after discharge, IOP was 7 mm Hg on topical timolol, with a deep anterior chamber and complete resolution of the hyphema. Visual acuity had improved to his baseline of 20/200, limited by a geographic macular scar that had not been previously visible.

Discussion

While spontaneous hyphema is well documented in the setting of anticoagulation and anterior segment abnormalities such as juvenile xanthogranuloma and iris vascular lesions,^{1–5} hyphema or other ocular hemorrhage has not been previously reported in patients with LVADs. However, nonocular bleeding complications with both the traditional pulsatile LVAD and the newer

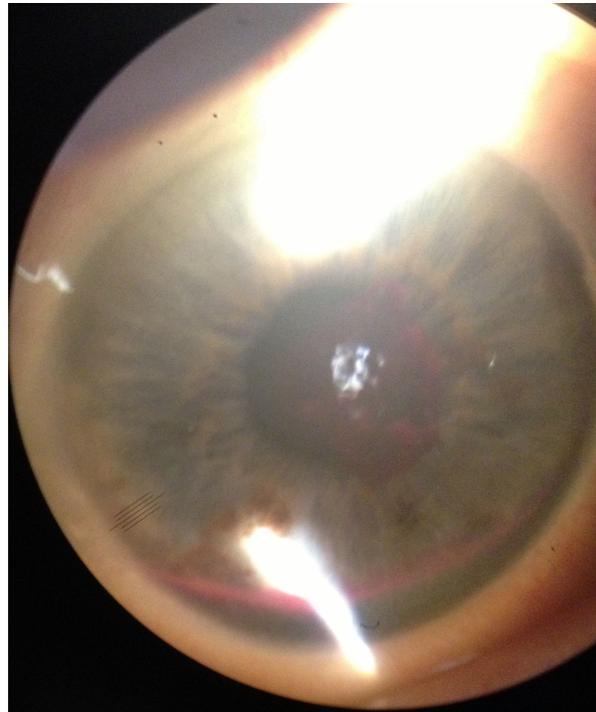


Figure 2. Three hours after the administration of intravenous mannitol, the shallowing of the anterior chamber improved, and the clot at the pupillary aperture began to dissolve.

continuous-flow LVAD have been reported.^{6,7} Angiodysplasia and AVMs are common sources of GI bleeding, which had previously occurred in our patient as well.

The LVAD can create an acquired von Willebrand's disease by compromising high molecular weight von Willebrand's factor (vWF) multimers, with proteolysis from shear stress, increased ADAMST-13 (a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13) activity, and impaired vWF-ristocetin platelet aggregation.^{6,8} The continuous-flow LVAD is also associated with fibrinolytic system activation and decreased platelet counts,⁶ although both platelet count and platelet function assay were normal in our patient, and vWF factor levels were unavailable.

Spontaneous hyphema related to pseudophakic uveitis-glaucoma-hyphema syndrome, even with a posterior chamber IOL, has been reported in the setting of abnormal iris vascularity, zonular laxity, and anteriorly rotated ciliary processes in plateau iris configuration.^{9,10} However, subsequent dilated examination revealed a well-centered in-the-bag IOL without any evidence of haptic chafing. The patient was without neovascular changes

suggestive of ocular ischemic syndrome and had no history of diabetes. The ocular bleed in this patient likely resulted from the change in systemic immunologic and thrombostatic function from direct contact of the blood circulation with the LVAD biomaterial.⁷ Though the mildly elevated INR may have contributed to the intraocular bleed, at least one study showed that INR was not predictive of bleeding events after LVAD implantation.¹¹ Another study indicated that older age (>65), lower pre-operative hematocrit (≤ 31), ischemic cardiomyopathy, and female sex were risk factors for bleeding with the LVAD.¹² Although spontaneous hyphema has been reported in the setting of elevated INR alone, in these cases the INR elevation was more significant, with INR >5 and/or prothrombin time >18.^{5,13,14} Iris vascular anomalies, such as localized vascularized iridolenticular adhesions in exfoliation glaucoma, are anatomic predisposing factors in the setting of warfarin use.¹⁴ Cases of significant suprachoroidal bleeding and angle closure associated with anticoagulation and exudative or end-stage macular degeneration have been reported,^{15–17} but the patient's B-scan and history of nonexudative AMD argue against this etiology.

In conclusion, sudden ocular pain in the LVAD patient should alert the clinician to the possibility of spontaneous hyphema, even without history of antecedent trauma, significant INR elevation, or iris vascular abnormalities. LVAD patients should be screened for gastrointestinal and hematologic risk factors for bleeding, and routine eye examination is essential to their follow-up care.

Literature Search

The authors searched the US National Library of Medicine at the National Institutes of Health and the PubMed OLDMEDLINE subset (Cumulated Index Medicus and the Current List of Medical Literature) for the period 1980–2014 using the following terms alone and in combination: *acute, spontaneous, ocular, intraocular, ophthalmic, periorbital, periocular, hemorrhage, bleed, bleeding, hyphema, pupillary block, intraocular pressure, glaucoma, angle closure, LVAD, left ventricular assist device, HeartMate, continuous flow, pulsatile.*

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