Color Doppler Imaging of Retrobulbar Hemodynamics in a Patient with Glaucoma Duet Associated with Dural Carotid-cavernous Sinus Fistula

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Although elevated intraocular pressure (IOP) resulting from increased episcleral venous pressure is common in patients with dural carotid-cavernous sinus fistulas (DCFs), neovascular glaucoma (NVG) is rare. A 42-year-old woman with ocular congestive symptoms, including high IOP in her right eye, was diagnosed with right DCF with carotid angiography. Although the symptoms resolved after gamma knife radiosurgery was performed to treat the DCF, central retinal vein occlusion (CRVO) occurred 5 weeks later. Color Doppler ultrasonography indicated the absence of blood flow in the superior ophthalmic vein. This was unlike the retrograde pulsatile blood flow observed with Doppler ultrasonography before the gamma knife radiosurgery. The subsequent development of NVG was treated with a combination of peripheral retinal cryotherapy and diode laser transscleral cyclophotocoagulation. This is the first reported case of CRVO and NVG as complications of DCF after an uneventful gamma knife radiosurgery in a relatively young woman free of systemic cardiovascular or hematologic disorders. Color Doppler ultrasonography delineates orbital hemodynamic changes and helps provide a guide to treat complications associated with DCF.


KEY WORDS: • color Doppler ultrasonography • dural carotid-cavernous sinus fistulas • gamma knife radiosurgery • neovascular glaucoma

INTRODUCTION

Dural carotid-cavernous sinus fistulas (DCFs) are arteriovenous communications in low flow that usually run a benign and uncomplicated course [1]. Neovascular glaucoma (NVG) as a complication of DCF is rare, with only two cases reported in English-language literature [2]. One case occurred after complications while attempting an embolization of the superior ophthalmic vein. The second case...
occurred spontaneously in association with central retinal vein occlusion (CRVO). CRVO, as a complication of DCF, has been reported to occur spontaneously after carotid-jugular compression or after a transarterial or venous embolization [3–5]. We report a patient with spontaneous DCF who initially suffered from high intraocular pressure (IOP) due to elevated episcleral venous pressure resulting from retrograde arterial flow in the superior ophthalmic vein, as revealed by color Doppler ultrasonography. Gamma knife radiosurgery relieved the ocular congestion symptoms through the normalization of IOP, but CRVO, accompanied by an absent blood flow in the superior ophthalmic vein, occurred 5 weeks later and subsequently developed into NVG.

**CASE REPORT**

A 42-year-old woman reported the sudden onset of pain, protrusion, and movement limitation of her right eye. Except for having a red right eye for 2 years, her medical history was noncontributory. Best-corrected visual acuity was 20/20 in both eyes (OU). IOP was 30 mmHg in the right eye (OD) and 12 mmHg in the left eye (OS). There was proptosis, abduction limitation, and dilatation of the conjunctival and episcleral vessels in the right eye. The anterior segment and fundus were unremarkable other than a hyperemic disc. Gonioscopy revealed open angles OU. Color Doppler ultrasonography showed retrograde pulsatile flow in the right superior ophthalmic vein (Fig. 1A). Carotid angiography found DCF of the right internal carotid artery (Fig. 1B). Gamma knife radiosurgery was used to treat the fistula and was uneventful. Three days later, the medication-free IOP was 14 mmHg and the proptosis and vascular congestion returned to normal.

Five weeks after the gamma knife radiosurgery, the patient’s vision suddenly dropped to counting fingers at 10 cm OD. IOP was 13 mmHg OD and 12 mmHg OS. Ophthalmoscopy revealed a diffuse, flame-shaped, retinal hemorrhage and tortuous retinal veins, indicating CRVO OD. A hematologic evaluation showed nothing out of the ordinary. Doppler ultrasonography 1 week earlier failed to detect blood flow in the right superior ophthalmic vein, indicating thrombosis impeding venous drainage (Fig. 2A). Panretinal photoacoagulation was aborted due to dense hemorrhage, and rubeosis iridis developed 4 weeks later. Throughout the following month, IOP fluctuated between 35 and 51 mmHg with three glaucoma medications, including oral acetazolamide.

The patient underwent panretinal cryotherapy combined with diode transscleral cyclophotocoagulation. IOP decreased to 19 mmHg on the first postoperative day and the rubeosis iridis regressed 2 weeks later. Eight months after gamma knife radiosurgery, color Doppler ultrasonography revealed orthograde nonpulsatile flow in the right superior ophthalmic vein (Fig. 2B). At the 2-year follow-up, the patient’s vision was 20/600 and the medication-free IOP was 12 mmHg OD.

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**Fig. 1.** Dural carotid-cavernous sinus fistulas. (A) Color Doppler ultrasonography shows retrograde pulsatile blood flow in the right superior ophthalmic vein. (B) A right internal carotid angiogram shows filling of the cavernous sinus (arrowhead) arising from the meningohypophyseal trunk (arrow) in the early arterial phase.
**DISCUSSION**

Gamma knife radiosurgery is noninvasive and effective in treating DCF [6]. The focused high-dose radiation may induce a wide range of vascular effects in the target area, including tissue edema, thrombi, necrosis, and reparative endothelial cell growth and fibrosis. These changes will result in the occlusion of small, abnormal vessels, while larger arteries and veins are more radioresistant and will be less affected [6]. In our situation, the target area was in the posterior and lateral part of the cavernous sinus, away from its connection with the superior ophthalmic vein. Nevertheless, cavernous sinus thrombosis may extend into the superior ophthalmic vein, leading to CRVO.

There may be several factors leading to CRVO in DCF, including a predisposing high IOP, high-pressure shunting flow, or occlusion of the venous route [2–5,7]. In this case, color Doppler ultrasonography performed 4 weeks after gamma knife radiosurgery, when IOP was within normal limits, detected no blood flow in the superior ophthalmic vein, indicating vascular occlusion possibly due to thrombosis. It is likely that flow stasis associated with diminished retrograde shunting flow, as supported by decreased proptosis and normalized IOP 3 days after radiosurgery, facilitates thrombus formation. Although Sergott et al reported that these patients may be followed for anticipated improvement, our evidence challenges the suggestion that spontaneous resolution of the syndrome can be expected if it is caused by venous thrombosis [8].

This case highlights the variable clinical course of DCF. Orbital hemodynamics may change unpredictably. Since color Doppler ultrasonography is a noninvasive procedure, it may be repeated when necessary to follow orbital hemodynamic changes [9]. If the clinical symptoms worsen from increased arterial shunting, embolization therapy of the shunts may be beneficial [10]. Although some researchers suggest that patients should be monitored for anticipated improvement if ophthalmic vein thrombosis causes clinical deterioration, others are concerned that it may be an initial symptom of CRVO [7]. Further research is necessary to determine whether anticoagulant or fibrinolytic therapy will help prevent severe vision loss from CRVO once venous stasis retinopathy develops in association with thrombosis formation in the superior ophthalmic vein.

**Fig. 2.** Color Doppler ultrasonography after gamma knife radiosurgery. (A) No blood flow in the superior ophthalmic vein 4 weeks after radiosurgery. (B) Orthograde continuous blood flow in the same vein 8 months after radiosurgery.

**REFERENCES**


