

Vortex Vein Varix Simulating Choroidal Melanoma

BY KAITLIN A. PATRICK, BS; AND CAROL L. SHIELDS, MD

There are numerous lesions that can simulate choroidal melanoma, labeled as pseudomelanoma.^{1,2} In a series of 1739 cases of pseudomelanoma, the most common simulators included choroidal nevus (49%), peripheral exudative hemorrhagic chorioretinopathy (8%), congenital hypertrophy of the retinal pigment epithelium (6%), and hemorrhagic detachment of the retina or retinal pigment epithelium (5%).² Choroidal vortex vein varix has been documented to simulate melanoma but represents less than 1% of pseudomelanomas.² Shields et al² found that 7 out of 1739 patients with pseudomelanomas had a vortex vein varix referred for misdiagnosis of melanoma.²

Vortex vein varix is a physiologic dilation of a choroidal draining vortex vein that usually occurs in specific fields of gaze and can appear as an elevated, dark choroidal mass simulating choroidal melanoma.^{3,4} In opposite fields of gaze, the dilated vein can flatten or collapse, confirming the diagnosis of a varix and differentiating the varix from a solid choroidal melanoma.²

CASE

A 67-year-old white woman presented with occasional photopsia for 5 months. The patient had been diagnosed with a suspicious choroidal nevus (rule out choroidal melanoma) in the left eye (OS) and was referred for management.

On examination, best-corrected visual acuity (BCVA) was 20/30 in the right eye (OD) and 20/20 OS. Intraocular pressures were normal. In the OD, there was a small red choroidal mass at the 5 o'clock equator that appeared flat like a vortex ampulla but filled with blood when the patient gazed inferonasally, classic for a vortex vein varix (Figure 1). The variceal lesion measured 2 mm in basal dimension and 1.4 mm in ultrasonographic thickness, but flattened in primary gaze. In the OS,

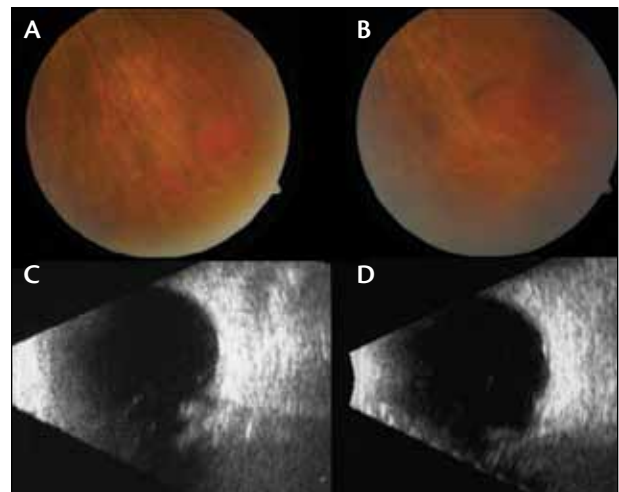


Figure 1. A 67-year-old woman with bilateral vortex vein varices. Findings in the right eye (OD) with changes in gaze. Fundus photograph OD in primary gaze (A). Fundus photograph of the OD looking inferonasally, showing a filled vortex vein varix measuring 2 mm by 2 mm in base (B). Ultrasonography of the OD in primary gaze (C). Ultrasonography of the OD looking inferonasally, showing a filled vortex vein varix measuring 1.4 mm in ultrasonographic thickness (D).

there was a prominent vortex ampulla in the 7-o'clock equatorial region that similarly filled to a varix when gaze was in that direction (Figure 2). The varix measured 5 mm in basal diameter and 2 mm in ultrasonographic thickness when filled, but collapsed to flat in primary gaze. Both varices filled and expanded when the patient looked in the quadrant of the vascular abnormality. Optical coherence tomography and indocyanine green angiography captured the variceal structures (Figure 3). The fluctuating expansion and collapse of the varices

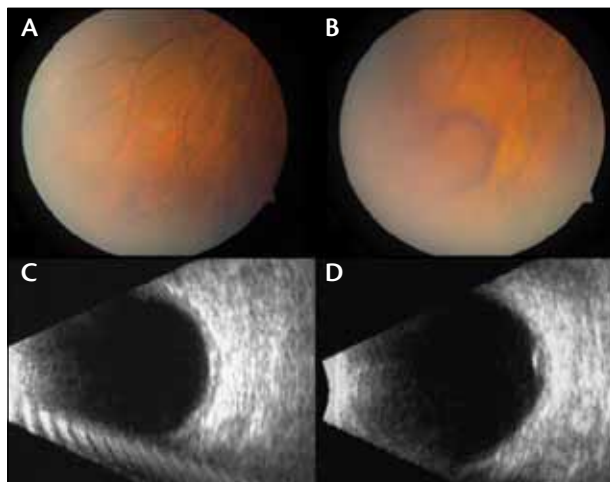


Figure 2. Findings in the left eye with directional changes in gaze. Fundus photograph OS in primary gaze (A). Fundus photograph of the OS looking inferonasally, showing a filled vortex vein varix measuring 5 mm by 4 mm in base (B). Ultrasonography of the OS in primary gaze (C). Ultrasonography of the OS looking inferonasally, showing a filled vortex vein varix measuring 2 mm in ultrasonographic thickness (D).

established the diagnosis of vortex vein varices in both eyes and not melanoma. Observation was advised. At nearly 3 years following initial presentation, the patient remains asymptomatic and the varices remain unchanged.

DISCUSSION

The vortex vein varix is a benign condition that should be considered in the differential diagnosis of choroidal melanoma. The vortex vein varix has been found in patients as young as 23 years of age, but that are usually seen in older patients such as the patient discussed in this case.³⁻⁵ The dilation and bulging of the vortex vein ampulla are usually asymptomatic and are not often recognized clinically. When the dilation of the vortex vein ampulla is large enough that it simulates a choroidal melanoma, it is called a varix of the vortex vein ampulla.³

An ampulla is an aneurysmal dilation of a vein of various shapes and sizes.³ Venous tributaries in the choroid drain into the vortex vein ampulla, which leads to the vortex vein.³ The vortex vein is located within the scleral canal and exits the orbit through the ophthalmic vein.³ There is usually 1 vortex vein per quadrant of the eye, but Lim et al have shown that up to 70% of eyes can have more than 4 vortex veins.^{6,7} The additional vortex veins were found more commonly in the nasal quadrant than the temporal quadrant, which increases

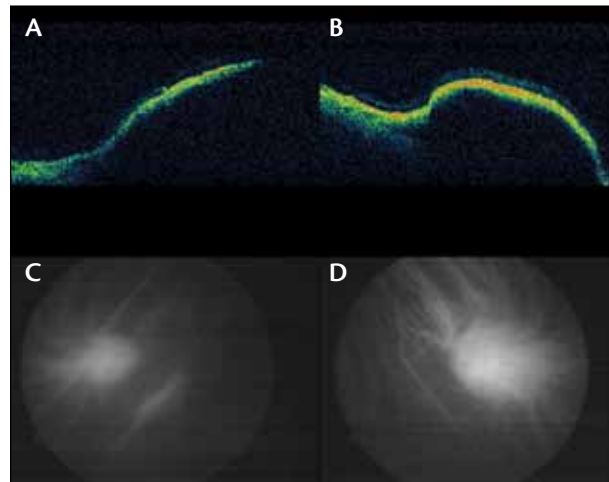


Figure 3. Optical coherence tomography (OCT) and indocyanine green angiogram of the vortex vein varix in the right eye (OD) and left eye (OS). OCT of the filled vortex vein varix of the OD (A). OCT of the filled vortex vein varix of the OS (B). Indocyanine green angiogram showing hyperfluorescence of the vortex vein varix in the OD (C). Indocyanine green angiogram showing hyperfluorescence of the vortex vein varix in the OS (D).

the chances of a dilated ampulla occurring in the nasal quadrant.⁶ Shields et al found that vortex vein varices are usually found at an equatorial location in the nasal fundus, which is consistent with the findings in our patient.³

The dynamic nature of the vortex vein varix is helpful in differentiating it from choroidal melanoma.³ The lesion becomes more prominent when the eye gazes in the direction of the vein and then becomes less prominent in primary gaze.⁶ Additionally, a light pressure applied to the globe can drain the blood out of the varix and make the varix become less prominent or disappear.^{3,6} The gaze-evoked enlargement and disappearance of the varix by ultrasonography is useful for diagnosis.⁶

Gunduz et al reported 4 cases of vortex vein varices that were referred with the diagnosis of small choroidal melanoma.³ These 4 patients had unilateral vortex vein varices, whereas our patient had bilateral vortex vein varices.³ In all cases, ultrasonography exhibited acoustic solidity of the lesion, and indocyanine green angiography showed gaze-evoked fluctuation of the hyperfluorescence of the ampulla in the venous filling frames in the lesion, demonstrating that the origin of the lesion was the vortex ampulla.³ Gunduz et al noted that color Doppler imaging, performed in 1 patient, showed a vascular lesion that filled when the eye was looking toward the lesion.³ When the eye returned to primary gaze the lesion resolved, showing no sign of residual vascularity.³

The etiology of ocular vortex vein varix has not yet been determined, but it is theorized to be from several causes, including most often kinking of the extrascleral portion of the vortex vein due to a change of gaze.^{3,4} Shields et al speculated that partial obstruction of the vortex vein by the superior or inferior oblique muscle could lead to varix formation.¹ Other contributing factors could be a gaze-evoked narrowing of the scleral emissary canal, or an increase in ocular venous pressure from prone position or from use of the Valsalva maneuver.³⁻⁵

Although the pathogenesis of vortex vein varix is unknown, the dynamic nature of a vortex vein varix, illustrated by the gaze and pressure-evoked changes in the lesion, can be diagnostic and confirmed by other ancillary testing methods. Using testing methods such as ultrasonography, fluorescein angiography, indocyanine green angiography, and color Doppler imaging can help to differentiate a vortex vein varix from other pseudomelanomas.³ The vortex vein varix is a benign condition that is often asymptomatic, yet it is important to be differentiated from choroidal melanoma to prevent unnecessary medical treatments and spare patients unwarranted anxiety. ■

Kaitlin Patrick, OMS II, is a candidate for a Doctor of Osteopathic Medicine, Class of 2015, and Co-President of Pulmonics, the Philadelphia College of Osteopathic Medicine.

Carol L. Shields, MD, is the Co-Director of the Ocular Oncology Service, Wills Eye Institute, Thomas Jefferson University. She is a Retina Today Editorial Board member. Dr. Shields may be reached at +1 215 928 3105; fax: +1 215 928 1140; or carol.shields@shieldsoncology.com.



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