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Unilateral vision loss in a patient with essential thrombocythemia

Essential thrombocythemia is a chronic myeloproliferative neoplasm that develops when megakaryocytes in the bone marrow produce an excess number of platelets.[1] This chronic hematologic disorder is associated with a number of thrombotic and embolic complications, and prior cases of vision loss have been documented.[1–6] We describe a rather unique case of essential thrombocythemia presenting with unilateral vision loss in a patient.

A 73-year-old male was referred to the ophthalmology clinic for a 1-week history of decreased vision in the left eye. The vision loss was described as a "green film" that obstructed his entire vision, with sparing of his superonasal visual field. Ocular history was remarkable for bilateral cataract surgery performed 30 years prior. His medical history included hypertension, dyslipidemia, coronary artery disease treated with a coronary artery bypass graft, right carotid endarterectomy, and a left parietal stroke 3 years earlier. His medications included atenolol, ramipril, alendronate, ranitidine, rosuvastatin, folic acid, and Aspirin. The patient denied any headaches, scalp tenderness, jaw claudication, eye pain, or weight loss.

Examination revealed a visual acuity of 20/60 OD improving to 20/40 with pinhole correction and counting fingers OS. There was a left afferent pupillary defect (relative afferent pupillary defect). Colour testing with Ishihara plates revealed 11/15 in the right eye and 0/15 in the left eye. Extraocular movements were full bilaterally. Slit-lamp examination demonstrated well-centred posterior chamber intraocular lenses with no evidence of posterior capsule opacification. Intraocular pressure was 24 and 25 mm Hg in the right and left eyes, respectively. Dilated fundus examination showed pale optic nerve swelling with associated rare disc hemorrhage. Compared with the right eye, there was also arteriovenous engorgement (Fig. 1). Optical coherence tomography also revealed disruption of the inner retina and outer nuclear layers, loss of foveal contour suggesting mild retinal edema, and left optic nerve head edema. Arteriovenous phase fluorescein angiography revealed decreased mid-peripheral choroidal perfusion and mild disc leakage in the left eye (**Fig. 1**). Goldmann visual field demonstrated a dense left central scotoma.

Laboratory testing at time of presentation demonstrated a normal C-reactive protein level, erythrocyte sedimentation rate, and B12 level. However, a complete blood count revealed an elevated white blood cell count (20.6×10^9/L) and significantly elevated platelets (102×10^{10}/L). Previous bloodwork demonstrated that the patient's platelets had been elevated for more than 1 year (63.9×10^{10}/L) prior to his presentation to the ophthalmology department. Subsequent computed tomography angiography revealed a chronic left parietal infarct in keeping with his previous cerebrovascular accident and severe (>95%) stenosis of the left common carotid artery bifurcation. A bone marrow biopsy was performed to rule out thrombocytosis but was inconclusive. However, subsequent genetic testing for a *JAK-2* mutation was positive, thereby confirming a diagnosis of essential thrombocytosis.

Essential thrombocythemia is a chronic myeloproliferative neoplasm characterized by excess platelet production by megakaryocytes in the bone marrow and can result in thrombohemorrhagic complications and possible progression to myelofibrosis or acute leukemia.[1] In this rare hematologic disorder, spontaneous thrombotic events are common, and systemic consequences include cerebrovascular accident, transient ischemic attack, deep vein thrombosis, pulmonary embolism, coronary artery ischemia, and hepatic, portal, splenic, and mesenteric vein thrombosus.[1,2] The World Health Organization criteria for essential thrombocythemia includes a platelet count of $\geq 450 \times 10^9$/L and a *JAK-2* mutation.[7] *JAK-2* is a nonreceptor tyrosine kinase that serves a vital role in transducing signals from various class I cytokine receptors; mutations in this gene disrupt normal functions that are for myelopoiesis.[8] Notably, the *JAK-2* mutation can be identified in more than 50% of patients who present with essential thrombocythemia.[8]

Previous reports in the literature have discussed the presentation of essential thrombocythemia in association with ocular manifestations, most commonly presenting with central retinal vein occlusion, branch retinal vein occlusion, or

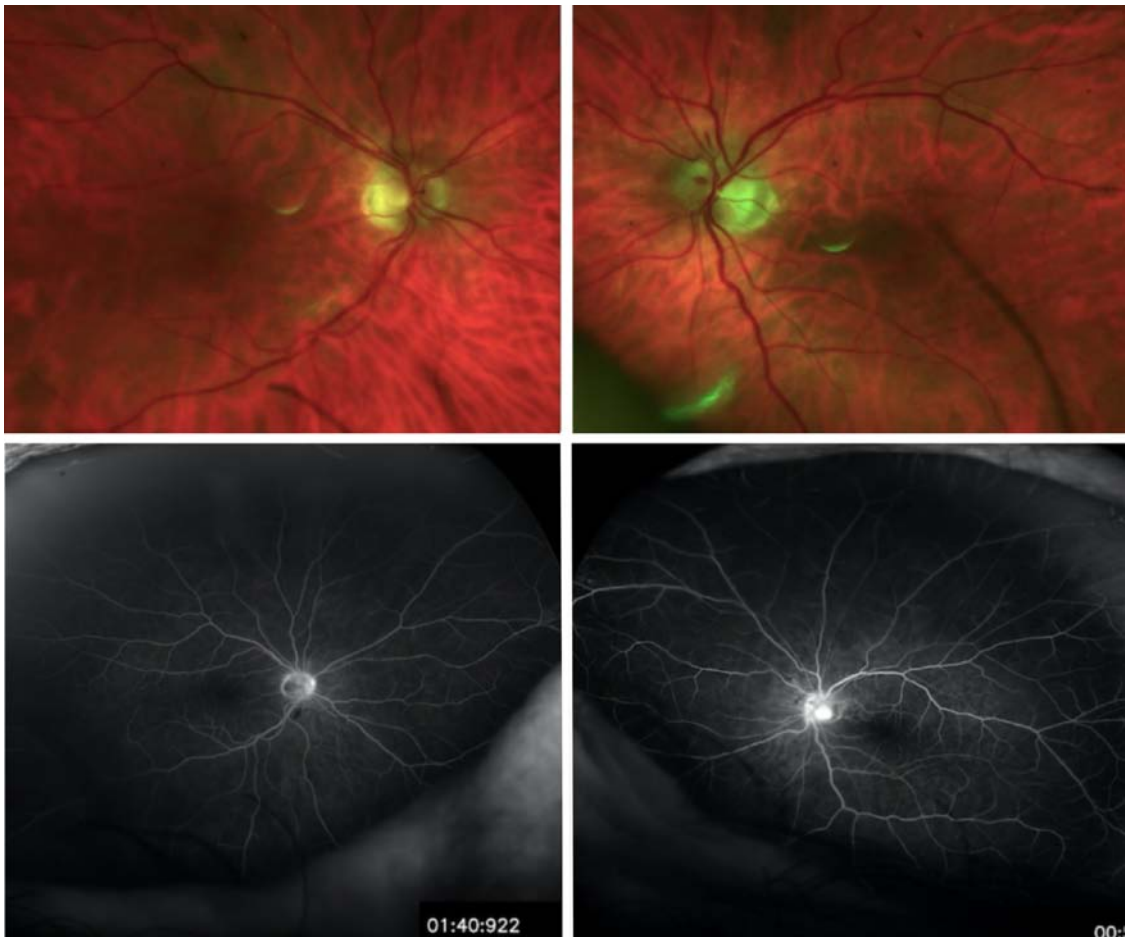

Fig. 1—Fundus images: (A) right eye; (B) left eye. Intravenous fluorescein angiography: (C) right eye; (D) left eye.

central retinal artery occlusion.[3–6,9,10] The differential diagnosis for this presentation includes occult giant cell arteritis, toxic or nutritional optic neuropathy, and ocular ischemic syndrome.

This case highlights an unusual presentation because the patient came to our clinic shortly after the onset of symptoms with severe vision loss and minimal retinal signs. Initial examination did not demonstrate any significant hemorrhage, vascular tortuosity, or a cherry red spot. The only significant retinal findings were subtle clinically. Given that there was occlusion of the left internal carotid artery, ocular ischemic syndrome is included in the differential diagnosis; however, many features do not support this diagnosis. For instance, the patient denied orbital pain, and clinically, conjunctival injection, neovascularization of iris or angle, midperipheral retinal hemorrhages, or microaneurysms were absent. The findings in this case suggest a central ophthalmic artery or central artery occlusion, both with reperfusion on the fluorescein angiogram. Optical coherence tomography showed disc swelling with subtle macular edema, and intravenous fluorescein angiography showed decreased choroidal signal in the periphery—both of which may be more consistent with a central ophthalmic artery event.

To date, investigation of the literature of ophthalmic artery occlusion secondary to essential thrombocytosis is limited, and we believe that this is one of the first reports of such a phenomenon. Unfortunately, the patient did not regain vision in the left eye. It should be noted that his platelet levels were previously elevated and continued to increase for 16 months until vision loss occurred. As such, this case also highlights the importance of comprehensive testing in preventing disease morbidity and preserving patient quality of life. Although the presentation to the ophthalmology clinic was acute, the patient's vision may have been slowly deteriorating. Of note, the patient's platelets were documented to be elevated more than 1 year ago. If this abnormality were adequately investigated and addressed, perhaps vision loss could have been reduced or prevented. It took more than a year for the patient to eventually present with noticeable vision loss and have subsequent work-up of his condition. Given the initially large differential diagnosis, this was a challenging case that ultimately led to an uncommon finding of vision loss in a setting of essential

thrombocythemia.. Thrombocythemia should be evaluated with a comprehensive systemic examination and appropriate laboratory work-up aids in order to prevent morbidity and mortality.

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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.