Purtscher-Like Retinopathy with Cardioembolic Stroke: Case Report and Literature Review

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Abstract

Purtscher-like retinopathy (PUR) is a rare condition characterized by sudden vision loss with associated retinal white patches thought to be due to precapillary arteriolar occlusion. We present a case of PUR associated with a cardioembolic stroke in a patient following temporary cessation of anticoagulant therapy for a surgical procedure. Our patient presented with multiple risk factors for PUR and classic signs and symptoms including multiple peripapillary white retinal lesions near arterioles and sudden unilateral decrease in visual acuity. Optical coherence tomography showed inner retinal hyperreflectivity and thinning consistent with inner retinal ischemia, and fluorescein angiography showed delayed retinal filling. Her complement C5 factor was elevated on laboratory testing. Brain magnetic resonance imaging showed acute/subacute left occipital lobe ischemia thought to be from a cardioembolic stroke. Shortly prior to visual symptoms, our patient’s apixaban was held due to surgical drainage of a gluteal abscess. This case highlights the rare occurrence of PUR associated with cardioembolic stroke and the importance of cerebral imaging in a patient presenting with PUR of uncertain etiology.
Introduction

Purtscher retinopathy was first described in 1910 and is a condition that is associated with severe trauma, often cranial or thoracic [1]. Purtscher-like retinopathy (PUR) is a rare condition characterized by unilateral or bilateral sudden vision loss with associated peripapillary retinal white patches and hemorrhage in the absence of trauma. Although there is limited understanding on the pathogenesis of PUR, the diverse nontraumatic etiologies suggest that this condition is related to microembolization of precapillary arterioles [2].

PUR has been associated with various systemic diseases, including acute pancreatitis, myocardial infarction, and collagen vascular disorders [2, 3]. We describe here a rare case of a 75-year-old female with multiple risk factors for PUR who developed PUR in association with a cardioembolic stroke to the left occipital region. We review the literature surrounding this rare condition and discuss this ophthalmologic consideration when withdrawing antithrombotic agents in a pre-surgical setting.

Case Presentation

A 75-year-old woman with a past medical history of myelodysplastic syndrome, thyroid dysfunction, hypercholesteremia, hypertension, mitral valve prolapse, atrial fibrillation on chronic oral anticoagulation, previous traumatic large bone fractures, and recent gluteal abscess was referred to retina clinic by optometry for vision loss in her left eye. On presentation to retina clinic, the patient endorsed a 3-week history of painless vision loss in the left eye described as blurry, purple, lacy area covering the left eye visual field that she woke up with one morning. This had not changed since onset. She denied eye redness, pain, flashes or floaters, or symptoms suspicious for giant cell arteritis (jaw pain, scalp tenderness, weight loss, etc.). Her history was significant for multiple traumatic fractures (pelvis, femur, hip, arm) 3 years prior for which she underwent repair and required blood transfusions at that time. Additionally, 10 days prior to initial presentation she was diagnosed with a gluteal abscess that was drained in the hospital after pausing her anticoagulation therapy.

Her best corrected visual acuity was 20/20 in the right eye and 20/200 in the left eye. There was no afferent pupillary defect, and her intraocular pressure, confrontation visual fields, and extraocular movements were normal. Anterior segment was significant for cataracts in both eyes. The optic nerve head was noted to be without hemorrhage or edema in both eyes. The left eye fundus was noted to have fine peripapillary cotton wool spots, and Purtscher flecken primarily centered around the optic nerve and extending along the inferior and superior arcades and into the macula (shown in Fig. 1). There was no retinal arterial plaque or embolus. Fluorescein angiography of the left eye showed delayed choroidal filling, choroidal hypofluorescence due to overlying retinal whitening blockage, delayed arterial entry at 18 s, and delayed laminar flow at 27–29 s (shown in Fig. 2). Optical coherence tomography (OCT) of the left eye showed inner retinal hyperreflectivity and revealed a paracentral acute middle maculopathy (PAMM) sign with subsequent retinal thinning (shown in Fig. 3).

She underwent further investigation with imaging and laboratory workup including complete blood count, complete metabolic panel, hemoglobin A1c, liver function tests, thyroid-stimulating hormone, total protein, amylase, lipase, rheumatoid factor, antinuclear antibody, cytoplasmic antineutrophil cytoplasmic antibody, perinuclear antineutrophil cytoplasmic antibody, anti-double stranded DNA antibody, complement C5, rapid plasma reagin, HIV, and tuberculosis testing. The values were within normal limits/negative apart from a low white blood cell count (3.8 Thou/cmm) and hemoglobin (10.2 g/dL), elevated protein (8.1 gm/dL), mildly elevated antinuclear antibody (1.55), and markedly elevated complement C5 (62 U/mL).
Fig. 1. Color fundus photography of the left eye at the primary visit (a) shows Purtscher flecken (white arrows) superiorly and cotton wool spots (blue arrow) distributed throughout the fundus, findings which are better delineated on the red-free filter imaging (b).

Fig. 2. Fluorescein angiography of the left eye shows delayed choroidal filling and arterial entry at 18 s (a), delayed laminar flow at 29 s (b), and representative late frames at 1 m (c) and 3 m 35 s (d).
Based on the presence of Purtscher flecken, cotton wool spots, and elevated complement C5, a diagnosis of PUR with retinal ischemia was made.

Brain magnetic resonance imaging revealed acute/subacute ischemia and infarct in the left occipital lobe. Carotid doppler showed 60–70% stenosis on both sides. Echocardiogram showed a normal ejection fraction. Computed tomography-angiography of the head/neck revealed mild vessel wall calcifications seen at the cavernous carotid arteries bilaterally without significant limitation to flow. There were moderate left-sided and mild right-sided carotid bifurcation calcifications seen with mild to moderate focal limitation to flow. It was concluded by her neurologist that she likely experienced a cardioembolic stroke to the left occipital region while off apixaban for her gluteal abscess drainage.

The patient returned to retina clinic 2 weeks later. Her left eye visual acuity was noted to be 3'/200. On exam, her optic nerve head still appeared normal, and the cotton wool spots previously described were improving. Two weeks following this, her vision was noted to be 2'/200 and the cotton wool spots had further improved. Her OCT showed retinal thinning with inner retinal hyperreflectivity in the left eye, consistent with prior ischemia (shown in Fig. 3).

Discussion

We describe a rare case of PUR in the setting of cardioembolic stroke. The most commonly described PUR etiologies are acute pancreatitis, renal failure, and autoimmune disease causing fat emboli, leuko-aggregation, or other mechanism of arteriolar occlusion [2]. Additional pathogenic theories include capillary endothelial damage, hyperviscosity, sudden increased intracranial pressure, or sudden expansion of retinal veins [2]. Purtscher flecken are polygonal intraretinal whitening with a juxta-vascular clear zone caused by arteriolar occlusion and are considered pathognomonic for PUR in addition to the cotton wool spots commonly described. There is often subtle retinal vascular leakage [2].

The pathophysiology for PUR remains indeterminate though is thought to be due to underlying systemic disease causing embolization from leukocyte aggregates, fat, fibrin, or platelets causing arteriolar occlusion and ischemia [4]. Activation of the complement cascade with the formation of C5a-induced leukocyte, platelet, and fibrin causing embolization has been proposed as a possible mechanism [4, 5]. Our patient’s laboratory workup was significant.
for elevated complement C5 and offers support for this hypothesis. Additional workup (including fibrinogen level, bleeding time, prothrombin time, partial thromboplastin time, international normalized ratio, or additional complement factors) was not performed by the primary team and represents a limitation of this study.

Literature review shows reports of PUR following ischemic events including myocardial infarction, ischemic stroke, carotid dissection, and post cerebro- and cardiovascular surgical interventions [3, 6–8]. Our case stands out as the patient did not have a recent diagnosis of stroke, and it was her ophthalmic diagnosis that prompted further workup that identified the cerebrovascular accident, thus emphasizing the important role of the ophthalmologist in properly diagnosing this condition and referring the patient for workup of associated comorbidities. Literature review reveals that the suspected mechanisms include emboli causing occlusion and/or increased coagulation activity leading to increased thromboembolic phenomena. Given the anatomic and physiologic similarity of cerebral and retinal vasculature, the association between retinal and cerebral vascular disease bears credence to this pathogenic theory of PUR [3].

In addition, the OCT sign seen in our patient consistent with PAMM due to the hyperreflectivity of the inner nuclear layer has been described in other PUR cases [9]. This supports the microvascular embolization theory in PUR pathophysiology and is ultimately associated with retinal thinning and persistently poor vision after the initial insult [10]. Similar OCT findings have been described after central retinal artery occlusions; however unlike central retinal artery occlusion, PUR is associated with elevated C5 and pathognomonic Purtscher flecken [2, 4, 5, 11]. All these findings offer support for our diagnosis of PUR in the case of this patient.

Shortly preceding her visual symptoms, our patient’s apixaban was stopped to allow for surgical treatment of her gluteal abscess. It is known that withdrawal of anticoagulation can lead to increased likelihood of ischemic stroke [12]. While temporary cessation of anticoagulation is often indicated for various clinical scenarios, caution and appropriate reinstitution is critical to minimizing the risk of adverse events. Our patient’s presentation in the retina clinic initiated an investigative workup following her PUR diagnosis and led to the discovery of her recent left occipital lobe infarction. Though rare, cerebrovascular infarction should be considered in patients presenting with PUR of uncertain etiology and should prompt appropriate cerebral and cardiac imaging.

While long bone fractures are a known cause of Purtscher retinopathy due to fat emboli, this phenomenon has classically been described in the acute setting. Our patient suffered significant traumatic long bone injury 3 years prior. To our knowledge, a delayed Purtscher retinopathy due to fat emboli has not been described in the literature and is unlikely to present remotely from initial injury.

In conclusion, we present a rare case of PUR following anticoagulation therapy pause resulting in a cardioembolic stroke. Stroke and PUR share pathogenic mechanisms and may be underreported. Microemboli resulting from anticoagulation cessation may lead to ischemic stroke in addition to PUR. A full workup including cerebral and cardiac imaging should be completed following PUR of uncertain etiology in order to rule out the potentially drastic consequences of cerebral ischemia.

**Statement of Ethics**

Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images. This retrospective review of patient data did not require ethical approval in accordance with local/national guidelines.
Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Kevin Elwood prepared the manuscript including writing the case presentation and reviewing the data. Andrew Dieu prepared the figures and legends and wrote parts of the manuscript. Clara Kuranz wrote parts of the manuscript. Mihai Mititelu wrote parts of the manuscript and figure legends and reviewed all aspects of the report, including the medical data.

Data Availability Statement

All data generated and analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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