Purtscher’s-Like Retinopathy Following Acute Pancreatitis

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Abstract

Background Purtscher’s retinopathy is a rare condition that results from trauma to the head, neck or thorax. Purtscher’s-like retinopathy is similar in appearance to Purtscher’s retinopathy but occurs in association to various systemic conditions, including acute pancreatitis. Both are a result of occlusion of precapillary arterioles by small and intermediate sized emboli and present with retinal whitening and intraretinal hemorrhages throughout the posterior pole. Other findings may include optic nerve edema, macular edema, macular ischemia and serous macular detachment. Patients are often symptomatic for reduced vision and visual field loss.

Case Report A 23-year-old African American male presented with reduced vision in both eyes following hospitalization for the treatment of acute pancreatitis secondary to alcohol abuse. Fundus examination revealed extensive peripapillary retinal whitening in both eyes with a few intraretinal hemorhages. Optical coherence tomography of the macula revealed a serous detachment in both eyes. Improvement in visual acuity and partial resolution of the macular subretinal fluid was observed upon follow up.

Conclusion Purtscher’s and Purtscher’s-like retinopathy are thought to be self-limiting conditions that resolve without ocular treatment. Following resolution of the acute retinal findings, patients can be left with visual acuity and/or visual field loss. While there is currently no accepted treatment for the retinopathy itself, it is important to treat the associated systemic condition.

Introduction The first case of Purtscher’s retinopathy was described in 1910 by Otmar Purtscher in a patient who had incurred severe head trauma. While classic Purtscher’s retinopathy is thought to occur after trauma to the head, neck or thorax, without direct ocular injury, similar retinopathy can also occur in an array of non-traumatic conditions, in which case the term “Purtscher’s-like retinopathy” is used. Some of the more common systemic conditions that can lead to Purtscher’s-like retinopathy are acute pancreatitis, autoimmune disease, fat embolism syndrome, pancreatic adenocarcinoma, renal failure and amniotic fluid embolization while other rare causes are thrombotic thrombocytopenia purpura, hemolytic uremic syndrome and cryoglobulinemia.

Case History A 26-year-old African American male presented to the eye clinic with sudden onset blurry vision left eye (OS) worse than right eye (OD). The patient had been hospitalized eight days ago for the treatment of acute pancreatitis secondary to alcohol abuse. Following his first night at the hospital, he woke up with blurry vision. On our examination he was found to have uncorrected visual acuity of 20/30 OD and 20/70 OS with no improvement on pinhole. All other entrance tests and anterior segment findings were within normal limits. Dilated fundus exam revealed extensive peripapillary polygonal plaque-like white lesions (Purtscher’s flecken), cotton wool spots and a few flame shaped intraretinal hemorhages in both eyes (OU). Optical coherence tomography (OCT) of the macula showed an area of serous macular detachment OU (Figure 2a,b). Given the case history and fundus appearance, the patient was diagnosed with Purtscher’s-like retinopathy following acute pancreatitis. Same day laboratory results for serum lipase, a diagnostic measure for pancreatitis, were severely elevated: 1721 U/L (Ref 11-82 U/L), 20 times the upper limits of normal. Therefore, he was admitted for further treatment. At 10 day follow-up, the patient’s vision was 20/30 OD and 20/60 OS. The OCT scan of the macula revealed resolution of the serous macular detachment OU and improvement in the detachment OS. OCT angiography showed ischemia nasal to the macula OU.
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The patient was to return in one week for fluorescein angiography, but has since been lost to follow up.

**Discussion**

**Epidemiology**

Purtscher's and Purtscher's-like retinopathies are rare, with an estimated incidence of 0.24 per million per year. The actual incidence is suspected to be higher due to possible underreporting.

**Purtscher's-Like Retinopathy and Acute Pancreatitis**

A link between Purtscher's-like retinopathy and acute pancreatitis was first suggested in 1975. Acute pancreatitis results from an inability of the pancreas to release the enzymes and hormones it produces due to an obstruction to outflow. Accumulation of pancreatic substances leads to activation of digestive enzymes and autodigestion of the pancreas with subsequent inflammation and activation of the complement system. After gallstones, alcohol abuse is the second most common cause of acute pancreatitis and the most common cause of chronic pancreatitis. Symptoms of acute pancreatitis include nausea, intractable vomiting and epigastric pain. High serum and urine amylase and lipase levels in addition to epigastric tenderness are typical of acute pancreatitis. The diagnosis can be confirmed with abdominal ultrasound or computed tomography scan. Treatment of acute pancreatitis includes intravenous fluids, pain management and nutritional support. Patients are also instructed to abstain from alcohol use as more frequent consumption after the initial episode is related to higher risk of developing chronic pancreatitis and later pancreatic cancer.

About 2% of patients with acute pancreatitis present with ocular complications. Retinopathy due to pancreatitis is thought to be a result of leukoembolization following activation of the complement system. These emboli are small to intermediate (~60-80μm) in size, allowing them to pass through larger branch arteries, but lodge in the precapillary arterioles. This leads to multiple small occlusions and infarctions, producing the characteristic appearance of Purtscher's-like retinopathy.

**Signs and Symptoms**

Patients with Purtscher's-like retinopathy often report an acute, painless decrease in vision. Visual field loss, such as central, paracentral or arcuate defects, may also be noted. Fundus examination reveals white lesions in the inner retina along with some intraretinal hemorrhages. The white lesions can be cotton wool spots and/or Purtscher's flecken. Cotton wool spots are flame shaped infarcts at the retinal nerve fiber layer and obscure the underlying retinal vessels. On the other hand, Purtscher's flecken do not obscure the retinal vessels; they are polygonal plaque-like lesions and are the result of precapillary arteriole occlusions in the inner nuclear layer. Additionally, the pathognomonic finding of Purtscher's flecken has a unique characteristic: a clear demarcation zone between the white
lesion and the adjacent retinal vessel (Figure 4). This area of normal retina represents the perivascular capillary-free or avascularized zone, which extends about 50 µm on either side of a vessel. Its presence further substantiates that the primary pathology in Purtscher’s-like retinopathy is the occlusion of the precapillary arterioles by small to intermediate-sized emboli.15 The retinal hemorrhages present are most frequently flame shaped, but dot/blot hemorrhages can also be found. The amount of hemorrhage is typically minimal. Other associated findings can include optic nerve edema, macular edema, macular ischemia and serous macular detachment.

The retinal findings typically present 24 to 48 hours after onset of the associated systemic condition and persist for about 1-3 months.15 They are usually bilateral, but can also be unilateral. Retinopathy is generally limited to the posterior pole due to the anatomic blood supply of the macula and peripapillary retina. Throughout most of the retina, this vascular network consists of two capillary layers with interrelated anastomoses. However, there are three capillary layers at the macula and four capillary layers around the optic nerve head, allowing for a thicker nerve fiber layer in the arcuate and papillomacular bundles.4, 15 This multi-layer capillary network extends two disk diameters nasal and four disk diameters temporal to the optic nerve. Unlike the majority of the retina, these capillaries have fewer arteriolar feeds and anastomoses and are thus more susceptible to embolic occlusion, as seen in Purtscher’s-like retinopathy.15

Differential Diagnoses
Differential diagnoses for Purtscher’s-like retinopathy may include central retinal artery occlusion, central retinal vein occlusion and commotio retinae.16, 17

A central retinal artery occlusion (CRAO) presents with unilateral retinal whitening in the posterior pole in the absence of intraretinal hemorrhages. The retinal whitening is confluent and is a result of occlusion of the central retinal artery by a large embolus.15 The occlusion commonly occurs at the narrowest portion of the central retinal artery, where it enters the sheath of the optic nerve head. Conversely, in Purtscher’s-like retinopathy, the areas of retinal whitening appear as distinct patches and there is no visible embolus.18

A central retinal vein occlusion (CRVO) leads to dilated, tortuous retinal veins with numerous intraretinal hemorrhages and cotton wool spots in one eye.19 It is a result of blockage of the central retinal vein at or behind the lamina cribrosa. The occlusion can be due to thrombosis or, less commonly, inflammation. Thrombosis of the vein develops as a result of abnormal blood flow caused by compression from the lamina cribrosa or an adjacent retinal artery.20 The vessel appearance and the absence of Purtscher’s flecken aids in differentiating CRVO from Purtscher’s-like retinopathy. Furthermore, a CRVO involves the entire retina and can lead to the development of anterior segment neovascularization.20

Commotio retinae is opacification of the retina produced by disruption of the photoreceptor outer segments following blunt trauma to the globe. It can occur anywhere in the retina, appearing within 24 hours, and resolves spontaneously in a few days with no sequelae. The neurosensory retina appears gray-white with or without intraretinal hemorrhages and retinal pigment epithelium mottling.21 In contrast to Purtscher’s-like retinopathy, the retinal whitening seen in commotio retinae is sheen-like and allows visualization of the underlying retina.22 In addition, there is no direct ocular trauma with Purtscher’s-like retinopathy.

While the above described conditions are some of the more common differentials, numerous conditions can present with cotton wool spots and/or other white lesions and intraretinal hemorrhages, hence a thorough case history and review of systemic conditions can help narrow down the list of differentials and arrive at the correct diagnosis.

Diagnosis and Imaging
Diagnosis of Purtscher’s-like retinopathy is often made based on clinical findings. For further assessment, different imaging modalities can be utilized to help confirm the diagnosis.

Fluorescein angiography may show non-perfusion of the smaller retinal arterioles or capillaries and then late leakage from the retinal vessels in areas of ischemia.15 Additionally, the cotton wool spots, Purtscher’s flecken and hemorrhages will exhibit hypofluorescence. While fluorescein angiography is the gold standard for evaluating retinal vasculature, OCT and OCT angiography can be used as less invasive alternatives.
In the acute stages of Purtscher’s-like retinopathy, OCT will show hyper-reflectivity of the inner retina corresponding to cotton wool spots and Purtscher’s flecken, indicating ischemia in each respective retinal layer. Following resolution of acute retinal findings, long-term thinning and atrophy may occur, particularly in those with ocular sequelae. OCT is also helpful in assessing the presence of subretinal fluid, especially at the macula.

OCT angiography often exhibits capillary dropout in both the superficial and deep vascular plexus of the retina. These areas of capillary dropout appear hyporeflective and correspond with the cotton wool spots and Purtscher’s flecken observed on clinical exam. The irregular capillary non-perfusion may be permanent even after clinically visible retinal changes have cleared, especially in those with sustained vision or visual field loss.

Furthermore, areas of choroidal hypofluorescence have been noted on indocyanine green angiography and support the idea that the choroidal circulation can also be affected. Infarction of the choriocapillaris compromises the outer retinal layers and can lead to outer retinal changes, such as the accumulation of subretinal fluid.

**Treatment and Prognosis**

Most cases of Purtscher’s and Purtscher’s-like retinopathy are left untreated and typically resolve on their own with the possibility of sequelae. Recovery of at least two Snellen lines is seen in about half of cases without treatment. Xia et al. conducted a literature review to determine the efficacy of available treatments. A total of 76 studies, involving 88 cases and 139 eyes, were included in the analysis to determine changes in visual acuity and potential side effects with the use of systemic or intravenous corticosteroids, hyperbaric oxygen therapy or traditional Chinese medicine. Steroids are believed to stabilize and speed the recovery of damaged neuronal membrane and microvascular channels, while hyperbaric oxygen is believed to increase tissue oxygenation during early ischemia. The study revealed improvement in vision at 1-3 months, 4-6 months and after 6 months in both groups who did and did not receive treatment, confirming that Purtscher’s and Purtscher’s-like retinopathy are indeed self-limiting. Of the studies that were included in the review, 43% of clinicians preferred to monitor patients without treatment, while 57% chose to treat them. Among those who chose to treat patients, the majority utilized corticosteroids. However, further research is needed to determine whether corticosteroids can change the natural course of Purtscher’s and Purtscher’s-like retinopathy, as most currently available studies are case reports with small sample sizes and high risk for bias. In addition, the underlying systemic disease varies from case to case, making it difficult to draw any conclusions, as the particular treatment for the systemic condition itself can influence improvement of ocular signs and symptoms.

Long term sequelae of Purtscher’s and Purtscher’s-like retinopathy may include optic atrophy, retinal pigment epithelial atrophy, retinal nerve fiber layer thinning, foveal thinning and retinal vessel attenuation or sheathing. Prognosis is poor in cases with macular infarction, longer duration of acute retinal changes, optic disc swelling, choroidal hypoperfusion, severe retinal capillary nonperfusion, outer retina involvement or a prior episode of Purtscher’s retinopathy in the same eye.

**Conclusion**

While Purtscher’s retinopathy is classically associated with severe trauma, a similar presentation can be seen in association with various other conditions. Patients often present with reduced vision and classic retinal findings. Following resolution of the acute retinal changes, patients can have residual visual acuity and visual field loss. There is currently no accepted treatment for Purtscher’s and Purtscher’s-like retinopathy. The best course of action is to treat the underlying condition and monitor for ocular resolution. Thus, it is extremely important to obtain a detailed history from the patient as well as employ a multidisciplinary team to provide comprehensive care.

**References**


