CLINICAL VIGNETTE

A Case of Pancreatitis Associated Purtscher's Retinopathy

Matthew Daly, BS and Jonathan Balakumar, MD

Case

A 26-year-old male presented with epigastric pain and visual loss. He has a history of significant alcohol abuse, intermittently drinking two bottles of whiskey daily for the prior three months. He was seen at an outside hospital and admitted for pancreatitis. During his hospitalization he awakened with blurry vision and left against medical advice attributing his visual loss to the medications he was receiving. The patient continued to have bilateral blurry vision and epigastric pain and presented to the optometry clinic where he was noted to have extensive cotton wool spots of both eyes and diagnosed with Purtscher's Retinopathy (PR). He was then transferred to the Emergency Department for further care.

In the Emergency Department, the patient was afebrile with a pulse 88/bpm, blood pressure 124/79 mm Hg, respiratory rate of 16 and oxygen saturation 100% on room air. He was well appearing, in no distress with unremarkable cardiac, pulmonary and abdominal exams. He had bilateral blurry vision and lipase of 1721 U/L. The patient was admitted for intravenous hydration, and supportive care with an unremarkable hospital stay with improvement in visual acuity.

Discussion

Purtscher's retinopathy (PR) also referred to as angiopathia retinae traumatica, is a traumatic angiopathy that was first noted in 1910 by Otmar Purtscher as white retinal patches and hemorrhages associated with visual loss following severe head trauma.[1] While trauma remains the primary cause leading to PR,[2] it has been reported in a variety of settings. It is typically seen in young or middle-aged men with traumatic injury or organ failure with connective tissue disease.[2] Our case of Purtscher's Retinopathy was associated with pancreatitis in a patient who presented with epigastric pain and visual abnormalities. While the relationship between PR and pancreatitis has been previously documented,[3-7] this relationship has not been well explored.

Signs pertaining to PR can be categorized as acute or chronic. Acute signs include cotton-wool spots, Purtscher flecken, and retinal hemorrhages. Cotton wool spots are white ischemic infarcts of the nerve fiber layer, whereas Purtscher flecken are separate, distinct regions of retinal whitening. Retinal hemorrhages may appear as dot and blot hemorrhages in the deep retinal layers or flame hemorrhages in the superficial nerve fiber layer.[8] Chronic signs consist of optic disc abnormalities consistent with papilledema, as well as, optic disc pallor and atrophy of retinal pigment epithelium.[2,8] PR symptoms fall into several general categories. First, the vision loss can be either bilateral or unilateral. Second, the vision loss may present as a loss of visual acuity, visual field, or combination of the two.

PR often presents similar to fat embolism syndrome,[8] in which patients are visually asymptomatic but are noted to have cotton wool spots and small blot hemorrhages. These patients typically present with other signs of systemic fat embolism. PR is generally confined to the peripapillary area and posterior pole. Additionally, patients are typically aware of visual problems immediately after onset.[8] The prognosis for vision loss varies. The vast majority of patients have spontaneous visual recovery without treatment, however several cases have reported improvement with high-dose, intravenous steroids as well as hyperbaric oxygen therapy.[2] Our patient with PR improved with supportive care alone.

There are several theories as to the pathogenesis of Purtscher's retinopathy. While the scientific community has yet to agree on a single cause, one widely supported theory states PR is caused by retinal arteriolar embolisms. This explains the sudden onset of visual changes as a result of multifocal lesions which lead to the obstruction of retinal vessels. These microcirculation effects have only been studied in animal models. Essentially, microparticles form clots enter retinal arteries and lead to artery branch occlusions, resulting in ischemic spots, retinal infarction, and retinal hemorrhage.[9] These clots, or emboli, can form from any number of media, including air, fat, and fibrin. It is also well understood that the body's own physiological immune response can lead to formation of leukocyte aggregate emboli that form following complement activation.[8] Other theories of development of PR include increased intracranial pressure causing extravasation of lymph from retinal vessels, as well as venous reflux and secondary arteriolar spasm causing acute ischemic damage.[8]

Our case potentially supports retinal arteriolar embolism theory of PR pathogenesis. Acute pancreatitis typically begins with acinar cell injury and can progress to multiorgan dysfunction. The steps leading to this final result involve an inflammatory response mediated by a cascade of acute inflammatory mediators, including complement proteins.[10] Complement activation leads to leukocyte aggregation and vessel occlusion.[4,8] Complement associated microvascular damage has been noted

to affect vision in a variety of diseases. For example, a wide variety of autoimmune diseases, including systemic lupus erythematous and multiple sclerosis, involve ocular manifestations as a result of microvascular damage caused in part by immune complex formation and complement activation.[11] Both classical and alternative complement pathways are activated in acute pancreatitis with varying magnitudes depending on the disease subtype.[12] Autoimmune pancreatitis, for example, primarily employs the classical pathway, leading to high levels of circulating immune complexes, whereas chronic pancreatitis frequently shows lower levels due to activation of a separate complement path.[12] Pancreatitis disease severity has not been shown to be associated with increased levels of complement activation.[13] Therefore, if the retinal arteriolar embolism theory of PR pathogenesis is the primary mechanism of development of PR in patients with pancreatitis, there may not be a direct relationship between disease severity and development of PR.

The primary symptoms of pancreatitis include severe epigastric pain and vomiting.[14] Often patients can recover with supportive care. This case provides an example of a rare complication of visual loss associated with pancreatitis. Although, our patient's vision recovered, PR has lead to permanent visual loss in others.

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