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Case Report

Purtscher-like retinopathy with renal impairment: a case report and review of the literature

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Short Title: Purtscher-like retinopathy with renal impairment

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Abstract

Introduction

Purtscher-like retinopathy (PLR) is a rare retinal vasculopathy characterized by acute vision loss. It is typically associated with systemic diseases such as renal impairment. The combined incidence of Purtscher retinopathy (PuR) and PLR is estimated at 0.24 cases per million annually. The hallmark of PLR is sudden-onset visual acuity reduction accompanied by retinal findings, including Purtscher-flecken, hemorrhages, and cotton-wool spots.

Case Presentation

We report a 46-year-old male with a history of chronic hypertension, dyslipidemia, cryoglobulinemia, and multiple viral infections, presenting with sudden bilateral vision loss. Fundoscopy revealed retinal swelling, hemorrhages, and exudation. Laboratory findings indicated impaired renal function (eGFR by CKD-EPI Cr 19 mL/min/1.73 m^2), cryoglobulinemia, and signs of chronic kidney disease. A renal biopsy confirmed membranoproliferative glomerulonephritis with immune complex deposition. The patient was treated with corticosteroids, therapeutic apheresis, and supportive care. Visual acuity partially improved in one eye during hospitalization.

Conclusion

PLR is a rare condition often linked to systemic diseases such as renal failure. Its pathophysiology involves retinal microvascular damage, potentially mediated by complement activation. Diagnosis is based on characteristic fundoscopic findings and associated systemic conditions. Corticosteroids remain the most commonly used treatment, although evidence for their efficacy is limited. This case highlights the rare overlap between PLR and renal impairment, emphasizing the importance of early recognition and multidisciplinary management. Further research is needed to elucidate the pathophysiology and optimize treatment protocols for PLR.

Introduction

Purtscher-like retinopathy (PLR) is a rare retinal vasculopathy characterized by sudden vision loss. It is associated with non-traumatic systemic diseases, including renal failure, the latter being rare. The combined incidence rate of Purtscher retinopathy (PuR) and PLR is estimated to be approximately 0,24 cases per million individuals annually [1,2]. The characteristic presentation is a sudden decrease of vision acuity within 48 hours of the onset of an associated condition with fundoscopic findings such as Purtscher-flecken, flame-shaped and dot-and-blot retinal hemorrhages, and cotton-wool spots [2,3].

We present a rare case of nephritic syndrome and neuroretinitis in a 46-year-old male patient with chronic hypertension, dyslipidemia, cryoglobulinemia, and multiple viral infections.
These conditions interacted complexly, underscoring the need for comprehensive diagnostics.

The overlap of PLR and renal impairment is extremely rare, despite a possible share of mechanisms like microvascular damage or complement activation. By presenting this case, we contribute valuable insights to the limited literature, highlighting the complex interplay of systemic conditions and emphasizing the importance of further research to understand this uncommon association better.

Case Report/Case Presentation

A 46-year-old male patient was hospitalized in the ophthalmology department due to bilateral vision deterioration following episodes of untreated high blood pressure (up to 200/140 mmHg) over the past 3–4 months. The patient complained of sudden foggy vision. His medical history was unremarkable with no pre-existing eye diseases.

Initial examination

Initial physical examination revealed severe hypertension (200/140 mmHg). On ophthalmoscopic examination his best-corrected visual acuity was 0.06 (approximately equivalent to 20/330 in the Snellen chart) in the right eye and 0.01 (approximately equivalent to 20/2000 in the Snellen chart) in the left. Anterior segment examination was unremarkable and intraocular pressures were within normal range. Fundoscopy revealed unclear boundaries of the retinal nerve fiber layer in both eyes, massive swelling, exudation in the center, and pigmented foci with surrounding hypopigmentation and scattered hemorrhages.

Laboratory findings

Blood tests showed normocytic normochromic anemia, elevated erythrocyte sedimentation rate, impaired kidney function (with eGFR by CKD-EPI Cr 19 mL/min/1.73 m^2), dyslipidemia, hypoalbuminemia as well as mildly increased lipase and lactate dehydrogenase. Urine analysis showed leukocyturia, microhematuria, and proteinuria of 1 g/L. Infection screening revealed seropositivity for CMV (IgM, IgG), Herpes Simplex (IgM, IgG), EBV, and Varicella zoster (IgG). Tests for viral hepatitis, HIV, syphilis, toxoplasmosis, bartonellosis, and Lyme disease were all negative. These findings reflect latent infections without clinical relevance to the eye lesions, and the retinopathy is unrelated to infectious agents, including CMV or B. burgdorferi. Autoimmune screening markers were negative, although cryoglobulinemia, normal complement C4, borderline C3c levels, highly positive kappa, and lambda-free light chains (normal ratio) were found. Given the presence of acute kidney injury of unclear etiology, 7 days after initial presentation the patient has been transferred to the Nephrology Department for further evaluation. On transfer, visual acuity of the right eye improved to 0,06, but the left eye remained unchanged. Further investigation was performed and both chest X-ray and head CT were normal, besides involutional brain changes. Brain MRI suggested left optic neuritis and severe non-specific foci in the cerebellar hemispheres and white matter. Ultrasound examination of the heart suggested LV dilatation and slightly increased filling pressure. Extracranial cerebral circulation ultrasound exam revealed low-grade stenosis of the right MCA and atherosclerotic arteriopathy.

Pathological findings

Due to the patient's severe renal findings, a renal biopsy was performed, and histopathology indicated focal non-active membranoproliferative and sclerosing glomerulonephritis with isolated immune complexes/cryoglobulins (shown in Fig. 1). Electron microscopy revealed podocyte foot process fusion, cryoglobulin-like immune complexes, and focal arterial endothelial proliferation with a possible component of thrombotic microangiopathy. Additionally, cholesterol crystals in the tubules and foam cells in the stroma were observed.

Diagnosis

The diagnosis of Purtscher-like retinopathy (PLR) was based on the patient's sudden bilateral vision loss, characteristic fundoscopic findings (retinal nerve fiber layer swelling, exudation, and hemorrhages), and the absence of trauma. The systemic involvement of chronic kidney disease with cryoglobulinemia, supported by renal biopsy findings of membranoproliferative glomerulonephritis and immune complex deposition, provided a plausible etiology. Infectious and autoimmune causes were excluded through targeted testing, further supporting the diagnosis of PLR secondary to systemic vascular damage from renal disease.

Treatment

The patient was treated for hypertension, dyslipidemia, anemia, and renal dysfunction. A Methylprednisolone pulse therapy and 5 therapeutic apheresis sessions for cryoglobulinemia were performed. However, kidney function did not improve and remained the same as at the time of arrival, with creatinine levels now at approximately 400 µmol/L and an eGFR by CKD-EPI <15 ml/min/1.73m^2. To this day the patient is being monitored by a multidisciplinary team.

The visual acuity of the right eye was 0.15 11 days after admission, while the left eye remained unchanged with severe visual impairment. This shows a notable improvement in one eye, which could indicate a positive response to treatment or resolution of some contributing factors to the initial visual impairment.

After discharge, the patient undergoes monthly follow-up appointments with a nephrologist and an ophthalmologist. Genetic testing was performed, analyzing genes associated with hereditary cardiovascular diseases, but no pathogenic or likely pathogenic variants explaining the patient's clinical presentation were identified. Currently, no genetic diagnosis has been established.

Discussion

Methodology

The literature was reviewed using PubMed and Google Scholar databases to identify articles about Purtscher-like retinopathy with renal involvement. The search was performed using the keywords "Purtscher-like," "retinopathy," "renal failure," "nephritic," "nephrotic," and "renal involvement." To ensure relevance, studies published within the last 10 years, available as full-text articles, and written in English were filtered to include studies published within the last 10 years. CARE reporting guidelines have been followed.

Aim

The literature around Purtscher-like retinopathy is scarce, mostly consisting of case reports. Our literature review aims to analyze reported cases of Purtscher-like retinopathy that coincided with acute or chronic renal impairment from the past 10 years.

Discussion

Purtscher-like retinopathy has many etiologies, unlike its counterpart, Purtscher's retinopathy, which is defined by a trauma event in the patient's medical history within the past 24-48 hours [3].

Incidence and epidemiology

The combined incidence of PuR and PLR is estimated to be 0.24 cases per million population per year. It may be even higher because not all cases are symptomatic and therefore reported [1,3]. Serhan et al. state that sex distribution in PuR and PLR is even between males (50.89 %) and females (49.11%) [4], similarly Miguel et al. have found that 60% of the cases (PuR and PLR combined) are of male patients [2], but Agrawal and McKibbin state that the condition is more common among males, as ⅔ of their included cases were men [1]. In the case of PLR with renal impairment, there is no current data on sex distribution. From our review, we could conclude that the gender distribution is

closer to even, as 5 of the 9 included cases were female patients. Agrawal and McKibbin have estimated that PuR and PLR are most commonly seen in young or middle-aged people, as the mean age of their cases was 42 years [1], Serhan et al. estimated it to be lower - 34.62 [4] years, Miguel et al. have found that the mean age is similar to the latter, 34 years [2]. Among the 9 PLR cases with renal involvement in our review, the patient age range was between 12-74 years, the mean age being 38 and the median being 34. Therefore, the original conclusion of Agrawal and McKibbin that these diagnoses are more common in young people still stands. In our reported case, the patient's age was 49, which is notably higher than the majority of patients identified in the cases reviewed in the literature.

Etiology

The most common etiologies of PLR include acute pancreatitis, renal failure, and autoimmune diseases. Among these, systemic lupus erythematosus (SLE) accounts for 13.1% of combined PuR and PLR cases, though only 3% in some reports [2,4]. Acute pancreatitis is also a significant cause, contributing to 20% of PuR and PLR cases [1], 11.9% of combined PuR and PLR cases [4], 66.82% of PLR cases [5], and 19.1% of PuR and PLR combined cases [1,2,4,5]. Other conditions associated with PLR include malignancy, hemolytic uremic syndrome (HUS), COVID-19, the Valsalva maneuver, and thrombotic thrombocytopenic purpura [2]. However, renal disease is rarely stated as the causative factor for PLR and cases of PLR with renal involvement remain uncommon. We reviewed 9 cases from the past 10 years of patients with PLR that were primarily tied to acute or chronic renal pathology. The presenting pathologies in these cases were: acute kidney injury caused by recurrent urinary tract infections in the background of chronic kidney disease [6], secondary malignant hypertension caused by IgA nephropathy [7], scleroderma renal crisis with malignant hypertension [8], hypertension following end-stage renal failure [9], peritoneal dialysis-associated peritonitis and hypertension following end-stage renal failure [9], nephrotic syndrome [10,11], severe acute renal failure following hydronephrosis [12] and cryoglobulinemia [13]. In the case that we report, the patient had cryoglobulinemia with nephritic syndrome, which is a rare presentation in patients with PLR. Many patients with PLR, just like the patients in the reviewed cases, have multiorgan involvement at the time of presentation, and in most cases, it is complicated to pinpoint which disease is the culprit of PLR [4]. Our reported case is a rare example of PLR primarily associated with kidney disease.

Pathophysiology

The pathophysiology of PLR is complex and not yet fully understood, though several hypotheses have been proposed. One prominent theory involves embolic occlusion of the retinal pre-capillary arterioles and microvascular infarction of the retinal nerve fiber layer. Emboli may consist of leukocyte aggregates, fat droplets, fibrin, platelets, or other particles, such as cholesterol crystals, as seen in our patient [2,3].

Complement activation has also been implicated as a significant factor. It is suggested that complement-mediated vascular injury promotes the formation of leukocyte aggregates up to 50 µm in diameter, which are large enough to obstruct retinal arterioles (approximately 45 µm in diameter). Complement activation is observed in several conditions associated with PLR, including connective tissue disorders and renal failure, supporting its role in the disease mechanism [3].

However, embolic occlusions in retinal arteries may not persist long enough to induce retinal infarction, suggesting that additional factors likely contribute to the observed vascular damage [3]. These may include hyperviscosity, sudden increases in intracranial pressure, and vascular endothelial dysregulation [33].

Despite the insights provided by these hypotheses, the precise mechanisms underlying PLR remain unclear [2].

Clinical presentation

The diagnosis of PLR is primarily clinical, characterized by a sudden decrease in visual acuity of varying severity. The most common retinal findings in PLR include cotton-wool spots (reported in 58-92% of cases), retinal hemorrhages (50-83%), Purtscher flecken (50-53%), macular edema (13%),

swelling of the optic nerve (20%), pseudo-cherry red spots, and optic disk swelling [1,3,4]. Approximately 50- 60% of cases are bilateral [1,4]. Of our reviewed 9 cases with renal involvement, as well as in the reported case, all patients presented with a bilateral decrease in visual acuity except for one case, where the laterality of visual involvement was not mentioned [6–13].

Diagnostics

Agrawal and McKibbin state that the fundus examination results combined with one of the possible causative diseases present are enough to make the diagnosis [3]. On the other hand Miguel et al. proposes that an approach with criteria is more appropriate. There are no established guidelines for diagnosis, however, the condition can be diagnosed when at least three of the following five criteria are met: 1) Purtscher-flecken, 2) intraretinal hemorrhages, 3) cotton wool spots, 4) plausible etiology, 5) compatible complementary examination [2].

Management and treatment

There are no evidence-based treatment guidelines for PuR or PLR at the moment. It is thought that most patients recover spontaneously regardless of treatment, and data shows no evidence of effective treatment in comparison to observation [1–4]. The most popular treatment option in literature is the intravenous administration of Corticosteroids [2,4,5]. In 1979 Hammerschmidt et al. conducted a study that demonstrated the ability of corticosteroids to inhibit the process of complement-induced granulocyte aggregation. By inhibiting this aggregation, corticosteroids reduce the likelihood of microvascular occlusion, minimizing tissue damage caused by reduced blood flow (ischemia) and inflammatory injury [14]. However, to this day, there is no quality data stating that treatment with corticosteroids is tied to better outcomes, and the reason could be that the pathophysiology of PLR is still not clear [2–5]. In 4 of the 9 cases we reviewed, patients were treated with corticosteroids, two other cases were already being treated with corticosteroids and received no additional corticosteroids, and in one case, treatment was not specified. In only 3 cases, patients had no or limited improvement in vision acuity, and all of them were treated with corticosteroids. In our case, the patient was treated with corticosteroids and had an improvement in vision acuity over 11 days, which could coincide with the resolution of the dominating disease or, on the other hand, demonstrate effective treatment with corticosteroids. Improvement of symptoms in reported cases where the patients have been treated with corticosteroids has been attributed to the ability of corticosteroids to stabilize damaged neuronal membranes and microvascular channels and to inhibit granulocyte aggregation related to complement activation. PLR in itself is thought to be more likely to be linked with fatal outcomes, as it is more frequently tied to multiorgan failure or advanced disease, which ultimately leads to the patient's death. Until we have better research to show the effect of various treatment options, observation remains the safest route to avoiding adverse drug reactions [2].

Prognosis

Miguel et al. state that there are prognostic factors such as male sex and causative pathology [2], Agrawal and McKibbin have found that fundoscopy findings such as optic disc swelling could also influence the outcome [3]. On the contrary, Serhan et al. saw no trends of this sort in their systemic review and stated that prognosis is not tied to the causative etiology and is more likely to be tied to the patient's personal history [4].

Conclusions

Purtscher-like retinopathy (PLR) remains a rare and underrecognized retinal vasculopathy, which is linked with a multitude of systemic conditions, including acute and chronic renal impairment. With our report, we present a unique case of PLR in a patient with chronic kidney disease. To this day, there are no standardized diagnostics and treatment protocols for PLR, nevertheless, it is of utmost importance to recognize pathognomonic fundoscopic findings as Purtscher-flecken of PLR and the signs of associated systemic diseases on time. If it is done correctly, it could aid in the choice of treatment and minimize the risks of everlasting vision impairment. Our literature review showcases the rarity of PLR with renal failure and the possible challenges in pinpointing the causal disease in patients with multisystemic involvement. This case report and literature review underscore the

importance of further investigating the pathophysiology, diagnostics, and efficacy of treatment options, in hopes of better understanding this disease and developing universal evidence-based treatment options for it.

Statements

Statement of Ethics

Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The original consent form is available as a separate document.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Melita Virpšaitė and Giedrė Žulpaitė contributed to the drafting and writing of the case report. Marius Miglinas critically reviewed the work for important intellectual content, provided final approval of the manuscript, and ensured the accuracy and integrity of the work. All authors meet the criteria for authorship and agree to be accountable for all aspects of the work.

Data Availability Statement

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

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Figure Legends

Fig. 1. Pathological findings of renal biopsy