Purtscher-like retinopathy: A rare ocular finding in nephrotic syndrome

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Abstract:
In this report, we present a case of Purtscher-like retinopathy in a 12-year-old male child with nephrotic syndrome. He was a known case of steroid-dependent nephrotic syndrome, frequent relapser with spontaneous bacterial peritonitis, who presented with complaints of difficulty in vision in both eyes. The fundus examination showed multiple cotton-wool spots and Purtscher fleckens centered around the optic nerve head with superficial retinal hemorrhages in both eyes. Fluorescein angiography confirmed the diagnosis of Purtscher-like retinopathy.

Keywords:
Nephrotic syndrome, Purtscher’s retinopathy, Purtscher-like retinopathy

Introduction
Purtscher’s retinopathy is a rare condition, classically described in patients with a history of trauma.[1] Purtscher-like retinopathy includes the nontraumatic causes of this retinal disease. It is described in a variety of conditions including acute pancreatitis, fat embolism syndrome, chronic renal failure, connective tissue disorders, and childbirth, among others.[2‑4] To our knowledge, only two cases of Purtscher-like retinopathy associated with nephrotic syndrome have been described worldwide.[5,6] Herein, we report a rare case of bilateral Purtscher-like retinopathy in a child with nephrotic syndrome, for the first time in India.

Case Report
A 12-year-old male child was admitted in the Pediatric Intensive Care Unit with complaints of fever, rapid breathing, and increasing swelling of face and body [Figure 1]. He had a 7-year history of nephrotic syndrome with frequent relapses. On examination, he was found to have pallor and generalized pitting edema. Physical examination revealed tachycardia, subcostal retraction, distension of abdomen, and a blood pressure of 140/90 mmHg. Ultrasonography of abdomen confirmed the diagnosis of ascites and in addition showed bilateral pleural effusion and hepatomegaly with bilateral renal parenchymal disease Grade 1. Laboratory studies revealed anemia (Hb 10%), nephrotic range proteinuria, normal blood counts, blood urea of 42 mg/dl, and creatinine of 0.9 mg/dl. Urine culture showed Escherichia coli >10⁵ CFU/ml. Diagnosis of steroid-dependent nephrotic syndrome, frequent relapser with spontaneous bacterial peritonitis with urinary tract infection, was made. He was treated with intravenous ceftriaxone (100 mg/kg/day), intravenous crystalline penicillin G (2 lakh units/kg/day), 20% infusion of albumin (1 g/kg), oral prednisolone (2 mg/kg/day), and oral furosemide (1 mg/kg/day) tablet enalapril (0.5 mg/kg) and tablet levamisole (1.5 mg/kg/day).

On the 4th day of admission, the patient presented to emergency eye services with complaints of difficulty in vision in both eyes. On ophthalmic examination, his visual acuity (VA) was 20/30 in both eyes. Slit-lamp examination was unremarkable, and pupillary reactions...
were normal. The patient had right eye congenital ptosis. Fundus examination showed multiple polygonal, discrete areas of retinal whitening between arterioles and venules (Purtscher-flecken) along with several cotton-wool spots at the posterior pole, in the peripapillary area. Cotton-wool spots could be differentiated from Purtscher-flecken as they are smaller, with ill-defined edges, and are located superficially over vessels. Few superficial retinal hemorrhages were also seen in both eyes [Figures 2 and 3]. There was no evidence of optic disc edema. The peripheral fundus was normal. Fluorescein angiography revealed masking of the underlying choroidal fluorescence in the early phase followed by peripapillary leakage in the areas of retinal lesions in late phase [Figure 4]. All these features were consistent with the diagnosis of Purtscher-like retinopathy. Except for the treatment of the underlying nephrotic syndrome, no additional treatment for retinopathy was given.

After 2 months, the patient’s VA remained 20/30. Fundus examination showed complete resolution of retinal lesions along with pallor of disc, attenuation of retinal arterioles, obvious retinal nerve fiber layer defects, and pigmentary changes in the nasal retina [Figures 5-7].

Discussion

Nephrotic syndrome is a common renal disorder in childhood. The clinical and biochemical features of nephrotic syndrome result from heavy proteinuria >40 mg/m²/h, resulting in hypoalbuminemia (<2.5 g/dl), hypercholesterolemia, and edema. Ocular manifestations of nephrotic syndrome are mostly related to prolonged steroid usage. Posterior subcapsular cataract, increased intraocular pressure, ptosis, keratitis, etc., are commonly associated with corticosteroid usage. Although there is an increased risk of systemic arterial and venous thromboembolism in nephrotic syndrome, retinal vascular involvement has not been reported.

The pathogenesis of Purtscher retinopathy remains unknown. Purtscher proposed that the white retinal changes may be secondary to lymphatic extravasations from retinal vessels in the course of increasing intracranial pressure. Currently, microembolization of the precapillary arterioles is considered the most likely cause of Purtscher flecken. Complement activation is a feature
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Retinal hemorrhages, cotton-wool spots, typically restricted to posterior pole, bilateral involvement, and occlusion and leakage of the retinal vessels on fluorescein angiography are common characteristics seen which can lead to variable degree of vision loss.

There are rare reports of Purtscher-like retinopathy in nephrotic syndrome. Viola et al. reported severe Purtscher-like retinopathy in septicemic disseminated intravascular coagulation (DIC) in a child with nephrotic syndrome.[6] They speculate complement activation and the increase of cytokines and inflammatory mediators in case of sepsis, with subsequent worsening of the coagulation, may have induced DIC causing Purtscher-like retinopathy. In another report, it has been described in mild chronic renal failure associated with nephrotic syndrome.[5] Authors suggested severe hypoprothrombinemia, hypoalbuminemia, and hyperfibrinogenemia caused by severe nephrotic syndrome in association with an upper respiratory tract infection causing complement activation as possible mechanisms for pathogenesis. Both cases reported bilateral severe retinal involvement with profound diminution of vision, while our case was milder in nature regarding the degree of vision loss. The cause of retinopathy remains unclear. Complement activation due to spontaneous bacterial peritonitis and urinary tract infection can be considered a possible causative event.

At present, no definite guidelines exist as to the treatment of this condition. There are isolated reports of successful treatment using intravenous high-dose steroids.[9] Steroid treatment may be considered in patients with severe ophthalmic symptoms. Most patients typically do get better without any treatment, with consequential recovery of vision of at least 2 and 4 Snellen lines in 50% and 23%, respectively.[8] In a systematic review performed by Miguel et al., there was no statistically significant difference between VA improvement for patients treated with corticosteroids compared with observation.[10] They concluded that observation and treatment of the underlying etiology is the most reasonable therapeutic option without risk of adverse drug effects.

Conclusion

Our case illustrates a rare but clinically significant ocular involvement in nephrotic syndrome in the form of Purtscher-like retinopathy. Hence, all patients of nephrotic syndrome presenting with visual complaints must be evaluated thoroughly for Purtscher-like retinopathy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/
have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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