Purtscher-like retinopathy secondary to dermatomyositis

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CASE REPORT

A 18-year-old Caucasian male with no previous past medical history presented with two weeks of progressively worsening blurry vision, proximal muscle weakness, myalgia, ataxia, rash on knuckles and back, dark colored urine, and dysphagia. The patient denied any ocular history, medication use, or illicit drug use. He was now bed bound and unable to move his arms or legs independently. The patient recently completed high school and denied any recent travel, change in diet, sick contacts, or family history of rheumatologic disorders.

On admission, the patient had creatinine kinase of 29,006 U/L, elevated liver enzymes, thrombocytopenia, and elevated CRP with subsequent diagnosis of rhabdomyolysis. On physical examination, he exhibited a shawl sign and Gottron’s papules. The patient also had a quadriceps muscle biopsy performed which showed non-specific myopathic changes, but results were confounded since the patient was initiated on steroid therapy prior to biopsy. The patient was diagnosed with dermatomyositis with the above mentioned clinical findings.

On ocular examination, vision measured 20/250 in the right eye and 20/300 in the left eye at distance. There was no acute pathology on anterior segment examination. Dilated fundus examination showed numerous cotton-wool spots, retinal hemorrhages, and Purtscher-flecken confined to the posterior pole (Figure 1). These findings prompted the diagnosis of Purtscher-like retinopathy. The patient was discharged to an outpatient rehab facility on oral prednisone and treated with infusions of rituximab monitored by Rheumatology. At two week follow up, the patient’s vision was 20/40 and 20/300 at distance and white retinal lesions had decreased in number and size. The patient was receiving daily physical therapy which allowed him to transfer independently and ambulate longer distances with a walker.

DISCUSSION

Purtscher retinopathy, first described by Otmar Purtscher in 1910 in a patient with cranial trauma, clinically presents as a chorioretinopathy with retinal findings including cotton-wool spots, retinal hemorrhages, optic disc edema, and areas of inner retinal whitening (Purtscher-flecken) [1, 2]. Miguel et al., published updated diagnostic criteria for Purtscher retinopathy which requires 3 out of 5 of the following: Purtscher-flecken, retinal hemorrhages in low to moderate number, cotton-wool spots confined to posterior pole, probable explanatory etiology (e.g. trauma, chest compressions, etc.), and complementary investigation compatible with diagnosis. By definition, Purtscher-like retinopathy occurs without a history of trauma and etiologies include acute pancreatitis, chronic renal failure, connective tissue disorders such as lupus, scleroderma, dermatomyositis, and preeclampsia [1]. There are no evidence-based guidelines on treatment, and therapy is focused on treating the underlying etiology. High-dose, intravenous steroids remain a commonly initial treatment regimen, but the efficacy of steroids has not been established in prospective trials and remains controversial [1, 3].
Retinopathy associated with dermatomyositis is an uncommon entity first described by Bruce in 1938 [4]. Retinopathy associated with dermatomyositis is rare and has only been described in a few case reports in the literature. Yan et al. described an 18-year-old Chinese female diagnosed with Purtscher-like retinopathy associated with dermatomyositis that presented with decreased vision and lower extremity weakness that was subsequently treated with systemic corticosteroid [5]. Bader-Meunier et al. reported a 7-year-old with juvenile dermatomyositis and thrombotic thromobocytopenic purpura treated with emergent plasmaphoresis, which resulted in the improvement of the retinopathy [6]. Barreiro-Gonzalez et al. described an 11 year-old female with dermatomyositis diagnosed with Purtscher-like retinopathy after cotton wool spots, flame hemorrhages, and venous tortuosity were found on dilated fundus examination [7]. In patients with dermatomyositis, the retinopathy usually resolves without vision loss, but permanent vision loss can occur from severe macular hemorrhage and optic atrophy [8].

CONCLUSION

Purtscher-like retinopathy is an uncommon retinal pathology that is associated with multiple systemic diseases. While systemic steroids are commonly used in initial treatment, therapy should be focused on treating the underlying cause of the retinopathy. The association with dermatomyositis is rare and has only been described in few case reports in the literature.

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Keywords: Dermatomyositis, Purtscher-like retinopathy, Retina

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REFERENCES


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Logan Russ Vincent – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.
Data Availability
All relevant data are within the paper and its Supporting Information files.

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