

# Journal of Molecular Biomarkers & Diagnosis

## **Case Report**

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# Case Report: An Unusual Case of Purtscher's Retinopathy

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#### Abstract

Purtscher retinopathy is a hemorrhagic and vasoocclusive vasculopathy and was first described as a syndrome of sudden blindness associated with severe head trauma. We present a case report of an unusual case of this condition where this is unilateral and without direct head trauma. We also discuss the current literature about this rare condition.

## Keywords: Purtscher retinopathy; Unilateral

# Introduction

Purtscher retinopathy is a haemorrhagic and vasoocclusive vasculopathy and was first described as a syndrome of sudden blindness associated with severe head trauma. These patients had multiple white retinal patches and retinal hemorrhages that were associated with severe vision loss [1].

Since its original description, Purtscher retinopathy has been associated with traumatic injury, primarily blunt thoracic trauma and head trauma, and numerous nontraumatic diseases.

#### Case History

A 49-year-old woman presented with sudden onset painless central scotoma in her left eye. She described a central circular area of dark vision 30 minutes after a fall from a horse. She was wearing a riding helmet and hit the ground head-first. No direct ocular or chest trauma was reported. She had no significant past ophthalmic or medical history.

Examination revealed unaided visual acuity of 6/6 Right eye 3/60 Left eye (no improvement with pinhole), bilateral lid ecchymosis, normal anterior segment and normal intraocular pressures. Dilated fundoscopy of the left eye showed multiple cotton wool spots and other white lesions separated from retinal vessels by clear zones characteristic of Purtscher's flecken. The changes were located around the disc and macular area while peripheral retina was being spared (Figure 1) Fundoscopy of the right eye was not remarkable.

On subsequent follow up 2 weeks later, her vision improved to 3/9 unaided, 6/12 pinhole. The retinal changes were partially resolved (Figure 2).



Figure 1: The changes around the disc and macular area while peripheral retina was being spared.



Figure 2: Partially resolved retinal changes with vision improved to 3/9 unaided, 6/12 pinhole.

# Discussion

Despite being described as early as 1910 by Purtscher, there has been paucity of data on the epidemiology of this condition. This is partially owed to the rarity of the disease, under-recognition and thus underreporting. A British Ophthalmological Surveillance Unit (BOSU) study found an incidence of 0.24 cases per million per year [1]. Purtscher's retinopathy can ensue after chest or head trauma in the absence of direct ocular trauma. On the other hand, Purtscher's like retinopathy is a term used to describe similar presentation in the context of other systemic disorders, including acute pancreatitis [2], SLE [3], HELLP syndrome and thrombotic thrombocytopenic purpura [4]. Most cases are bilateral, however unilateral involvement can occur.

Fluorescein angiography suggests that ischaemia due to microvascular occlusion is the cause. This could be an embolic phenomenon [5] or shear stress damage [6].

Patients present with rapid deterioration of vision with variable

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Received December 21, 2017; Accepted December 28, 2017; Published December 30, 2017

Citation: Jain S, Babiker S, Keenan TD (2018) Case Report: An Unusual Case of Purtscher's Retinopathy. J Mol Biomark Diagn 9: 374. doi: 10.4172/2155-9929.1000374

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degrees of visual acuity. The pathognomonic sign is the presence of flecken, which are inner retinal layer infarcts. In contrast to cotton wool spots which are nerve fiber layer infarcts with fluffy edges and overlap with the retinal vessels, these have demarcated margins and clear zones separating the lesions from the retinal vessels. Retinal haemorrhages have also been reported. Fundus fluorescein angiography reveals areas of hypo perfusion, delayed vascular filling combined with late leakage. Visual fields typically show central scotoma. Electrophysiology data are very limited, but reports show increased latency and decreased amplitude on visually evoked potential and reduced a and b wave on electroretinography.

Treatment with corticosteroids has been suggested, but no significant difference with or without treatment was found in a large study [7].

# **Conclusion and Learning Points**

It is of paramount importance to raise the awareness towards the different clinical presentations of this poorly understood disease. Ongoing reporting is indeed invaluable to consolidate the current body of evidence. Further research is needed to find a treatment for this devastating condition.

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