Three Cases of Missed Purtscher’s Retinopathy

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Abstract

Purpose: Visual loss associated with multiple traumas in the context of road traffic accident, especially when the maxillofacial and orbital areas are not directly injured, is an uncommon occurrence. The possibility of Purtscher’s retinopathy, i.e. retinal manifestation of mechanical trauma elsewhere in the body, as the cause of visual disturbance in these situations can often be overlooked.

Patients & Methods: We present three cases of Purtscher’s retinopathy where the diagnosis was initially missed. All three patients presented to Accident and Emergency department with visual loss in the left eyes, having sustained compressive chest and/or abdominal injuries in road traffic accidents. None had direct trauma to the orbital region. Serial color fundus photographs, fluorescein angiogram, Goldman visual fields, and electro diagnostic tests were performed on each patient, with a mean follow-up of 8.3 months (Range 6 to 10 months). The correct diagnosis of Purtscher’s retinopathy was made retrospectively in all three cases. Two patients had persistent central scotoma despite complete resolution of the retinal signs.

Conclusion: Purtscher’s retinopathy should be considered as a differential diagnosis in all cases of unexplained visual loss associated with multiple traumas. The retinal manifestations of Purtscher’s retinopathy can disappear in a short time interval. A retrospective diagnosis may be difficult in the absence of any fundal abnormalities if the diagnosis is initially missed on presentation.

Key words: Purtscher’s Retinopathy, Trauma, Central Scotoma, Cotton Wool Spots

Introduction

Patients suffering minor traumas in road traffic accidents frequently present to the accident and emergency department. However, it is unusual that these patients should complain of visual loss, especially when the maxillofacial and orbital areas are not directly injured.¹ The cause of the visual loss in these situations can often be baffling. The differentials that frequently arise are head injury associated damage to the visual projection, or traumatic optic neuropathy secondary to skull fracture involving the optic canal. Whilst these are perfectly valid ‘direct’ mechanisms of visual loss, the possibility of Purtscher’s retinopathy often escape consideration in the heat of the situation. Especially that the first ophthalmologist who attends to these cases tends to be a trainee.

Purtscher’s retinopathy is classically recognized as retinal manifestation of mechanical trauma occurring elsewhere in the body, and presents as an acute visual loss. The fundal appearances of white cotton wool patches, edema and hemorrhage concentrated around the optic disc are diagnostic in the right context. However, these retinal signs are transient.
If the diagnosis is initially missed, the correct diagnosis can only be inferred retrospectively, and only after exclusion of the above-mentioned direct mechanisms of visual loss by costly and unnecessary neuroimaging. We report three cases where the diagnosis of Purtscher’s retinopathy was initially ‘missed’.

Case Report

**Patient 1**

A 32-year-old driver crashed into a tree at 80 mph during an amateur car race. He was wearing a seat belt and helmet at the time. He presented to the Accident & Emergency department complaining of tenderness over the right side of his chest and the loss of central vision in his left eye. On examination, there was a seat belt graze over his thorax and bruising over the right third rib in the mid-axillary line. A chest x-ray confirmed a corresponding fractured rib. Otherwise there was no other significant injury. His Snellen visual acuities were recorded as right 6/6 and left counting finger. There was no evidence of any direct injury to the eye and orbit. Undilated direct ophthalmoscopy by the ophthalmic casualty officer showed the optic discs to be normal. The patient was referred to the eye clinic to for his unexplained visual loss.

When he was reviewed in the eye clinic the next day, the left visual acuity remained at counting finger. There was no relative afferent pupillary defect and the left fundal appearance seemingly remained unchanged (Figure 1). Goldman perimetry demonstrated a left central scotoma. A diagnosis of Purtscher’s retinopathy was made. Fluorescein angiography demonstrated areas of capillary non-perfusion at the macula (Figure 2). Six weeks after the accident, the fundal changes had largely resolved (Figure 3). His left vision remained poor at counting finger, and the scotoma persisted. Electrophysiological tests were carried out to rule out the possibility of a traumatic optic neuropathy. These tests, including full field Pattern Reversal Visual Evoked Potential, Flash Visual Evoked Potential and Electroretinogram all were normal.

Six months after the accident, his vision was recorded as right 6/6 and left 2/60.
His left vision improved to $6/6$ five months after the accident. Interestingly, a new cotton wool spot appeared along the superior temporal arcade in the left fundus, five months later, whilst the older lesion along the inferior temporal arcade resolved (Figure 8). His final Snellen visual acuity was $6/6$ right and left eye when he was discharged nine months following the accident.

**Patient 2**

A 37-year-old male was brought to the Accident and Emergency Department by the police. He was found wandering aimlessly near the scene of a road traffic accident. It later transpired that he was the driver of a vehicle, which crashed head-on into a bridge, killing the front seat passenger. He complained of loss of vision in the left eye and pain in his chest and jaw. Examination in casualty revealed a catalogue of injuries including a small right side pneumothorax, undisplaced fracture of the mandible and avulsion fracture of the fifth metatarsal base on the right.

He was not examined by an ophthalmologist until three days after the accident where his visual acuities were recorded as right $6/6$ and left $6/18$. There was no relative afferent pupillary defect. Amsler revealed a left central scotoma. Dilated fundal examination showed a few scattered nerve fibre layer hemorrhages, white cotton wool spots and edema at the macula in the left eye. Commotio retina was diagnosed by the resident on call.

Two weeks later, his left visual acuity deteriorated to $6/36$. There was a single cotton wool spot along the inferior temporal arcade in the left eye (Figure 5). Goldman perimetry showed a left central scotoma (Figure 6). A Fluorescein angiogram did not show any areas of non-perfusion (Figure 7). Electro physiology tests including Visual Evoked Potentials and Electroretinogram again did not demonstrate any significant optic neuropathy. The diagnosis was revised to Purtscher’s retinopathy.

**Figure 4.** Patient 1. Left color fundus photograph six weeks after accident. There was considerable resolution of the retinal signs.

**Figure 5.** Patient 2. Left color fundus photograph showing a single cotton wool spot along the inferior temporal arcade two weeks after the accident.

**Figure 6.** Patient 2. Left Goldman field two weeks after accident demonstrating a central scotoma.

**Figure 7.** Patient 2. Left fluorescein angiogram at two weeks failed to show any area of non-perfusion.
Figure 8. Patient 2. Left color fundus photograph five months after the accident. A new cotton wool spot has developed along the superior temporal arcade whilst the original one resolved.

Patient 3

A 38-year-old ‘banger’ racer was involved in a collision during a race where he was hit from behind and shunted at 40 mph into a fence. He came to the Accident and Emergency department on the same day complaining of pain in the upper part of the abdomen and loss of vision in the left eye. On examination, there were bruises and tenderness in the epigastric area, but no guarding was demonstrated. His visual acuity was right 6/6 and left 6/24. There was no relative afferent pupillary defect. There was also no evidence of any direct trauma to the orbits. Fundal examination showed a few dot and blot hemorrhages at the left macula, but was otherwise unremarkable. No definitive diagnosis for the visual loss was given at the time.

When he was re-examined two days later these fundal changes had disappeared. The visual acuity was recorded as 6/6 right and 6/18 left six weeks later. The left macula changes noted previously had completely resolved (Figure 9). Goldman field showed a left central scotoma (Figure 10). Fluorescein angiography was also unremarkable (Figure 11). A retrospective diagnosis of Purtscher’s retinopathy was presumed.

His left visual acuity remained at 6/36 ten months following the accident.

Retinopathy secondary to indirect mechanical trauma occurring elsewhere in the body had been recognized by Purtscher, an Austrian ophthalmologist, as early as 1910.2 He described this in a patient who had sustained head trauma accompanied by loss of vision, an entity he later referred to as ‘angiopathia retinae traumatica’.3 The appearance described by Purtscher was of multiple oval white spots and intraretinal hemorrhages surrounding the optic disc. Since then, similar fundal appearances has been described in a wide range of precipitating events such as trauma, including both compression injuries and bone fractures, pancreatitis, childbirth, and renal failure.4

The pathogenesis of Purtscher’s retinopathy is still uncertain. Purtscher theorized that it was due to an increase in the cerebrospinal fluid pressure that was transmitted from the subarachnoid space to the optic nerve, producing lymphatic stasis in the perivascular spaces of the retinal vessels.2,3 This is clearly not a valid explanation for all the other conditions mentioned above that result in the retinopathy.

The ophthalmoscopic picture suggests ischemia in the posterior pole manifesting as patches of edema and hemorrhages. One possible cause is embolization of the peripapillary terminal arterioles.

Figure 9. Patient 3. Left color fundus photograph six weeks after the accident. No retinal lesion was present.

Figure 10. Patient 3. Left Goldman field demonstrated a persistent central scotoma six weeks after the accident.
This can be due to fat embolization, such as in trauma involving fractures, and platelet/fibrin clumps in conditions where there is systemic complement activation, such as in acute pancreatitis. The other possible mechanism for producing the retinopathy is through increases in arterial hydrostatic and/or venous intravascular pressures.<sup>5,6</sup> Multiple injuries are frequently accompanied by crushing or compressive forces. The effect of sudden increase in pressure within the arterial part of the circulation can lead to nerve fibre layer hemorrhages and exudate and cotton wool spots (resembling hypertensive retinopathy), whereas mild retinal edema can be produced by hydrostatic insults on the venous systems.

We present three cases of Purtscher’s retinopathy where the diagnosis was initially overlooked. The mean follow-up period was 8.3 months (Range 6-10 months). All three cases sustained compressive injuries to the chest or abdomen. The first case also suffered a broken rib. None of the three cases had direct trauma involving the orbits or globes. They were therefore ‘susceptible candidates’ for the development of Purtscher’s retinopathy. The fact that it was not diagnosed at presentation in all three of our cases was probably because Purtscher’s retinopathy remains a rare occurrence in everyday ophthalmic practice despite the modern age of massed transportation and car travel.

The diagnosis of Purtscher’s retinopathy can easily be missed if examination of the patient is delayed; in mild form of the retinopathy such as in patient 3, the fundal signs had all but disappeared by the third day of the accident. On the other hand, delayed retinal manifestations can occur up to five months following the accident, as in patient 2, despite the resolution of the initial retinal changes and the patient’s symptoms.

It is interesting to note that the left eye was involved in all of the cases. This may be due to the fact that they all sustained similar compressive injuries to the thorax and abdomen, and the arrangements of the vascular anatomy is such that the left side of the vasculature in the head and neck is more vulnerable to sudden increases in hydrostatic pressure.

The prognosis is usually favorable in Purtscher’s retinopathy where substantial recovery of visual function can be expected. It is also recognized that some cases are left with permanent visual loss.<sup>7</sup> In our series of three patients, only one patient made a full recovery. It is likely that Purtscher’s retinopathy represents a spectrum of retinal insults of varying severity, and hence the variable prognosis.

**Conclusion**

Purtscher’s retinopathy should be considered as a differential diagnosis in cases where visual loss is associated with multiple traumas, especially where the maxillofacial and orbital region is not injured. All such cases should have a careful ophthalmic assessment including a dilated fundal examination. If a definitive diagnosis of Purtscher’s retinopathy can be made in the acute situation, unnecessary and costly investigations can potentially be avoided.
References