

# Pathogenesis and Neuroprotective Treatment in Purtscher's Retinopathy

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**Abstract:** Purtscher's retinopathy is characterized by sudden visual loss in severely traumatized patients and is associated with multiple areas of superficial retinal whitening located primarily in the posterior pole. Visual outcome in Purtscher's retinopathy is variable, and there is no well-defined treatment. We report on a patient with immediate blurred vision in the right eye after a traffic accident. Ophthalmoscopy revealed multiple whitish patches scattered over the macular and peripapillary areas in the right eye. Fluorescein angiography showed multifocal retinal arteriolar occlusion in the early phase and staining of the involved retinal vessels and optic nerve head in the late phase. Indocyanine green angiography (ICG) showed rarefaction of choroidal vessels in the peripapillary area of the right eye at early phase. The late phase ICG study revealed multifocal hypofluorescent patches in the macular and peripapillary areas. Megadose steroid therapy was given with good visual response in the first 2 weeks, and the patient's vision had recovered completely when followed-up 10 months later. Jpn J Ophthalmol 1998;42:318–322 © 1998 Japanese Ophthalmological Society

Key Words: Indocyanine green angiography, megadose steroid, Purtscher's retinopathy.

## Introduction

Purtscher's retinopathy, first described by Otmar Purtscher, is characterized by sudden visual loss in severely traumatized patients and is associated with multiple areas of superficial retinal whitening located primarily in the posterior pole.<sup>1</sup> Fluorescein angiography in Purtscher's retinopathy reveals focal areas of retinal arteriolar occlusion, patches of capillary nonperfusion, and leakage of the involved retinal vessels in late phase.<sup>2,3</sup> Similar fundus findings have since been reported to be associated with compressive chest injuries,<sup>2</sup> fat embolism<sup>4</sup> from multiple fractures, retrobulbar anesthesia,<sup>5</sup> acute pancreatitis,<sup>6</sup> childbirth,<sup>7</sup> and connective tissue disease.<sup>3</sup> Visual outcome in Purtscher's retinopathy is variable. Normal vision may not be recovered in patients with severe initial injury, when optic atrophy and macular infarction may persist in the late phase, 7-9 To date, there is no well-defined treatment for Purtscher's retinopathy.<sup>8</sup>

We report herein on a patient with Purtscher's retinopathy. Fluorescein angiography and indocyanine green angiography (ICG) were performed following trauma. Megadose steroid therapy was given with good visual response in the first 2 weeks, and there was complete visual recovery at the 10-month check-up.

# **Case Report**

A 17-year-old girl was admitted with the chief complaint of blurred vision in the right eye following a traffic accident 2 days prior. Blurred vision of the right eye and neck pain were noted immediately when she was brought to the emergency room of our hospital. A computed tomography (CT) scan of the neck revealed a fracture of bilateral pedicles of the second cervical spine. Chest and skull x-rays were normal. The neurosurgical department decided to give her conservative treatment for cervical spine fracture, and then she was transferred to our department for her eye condition. On admission, visual acuity was finger counting in the right eye; 6/6 in the

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left eye. She could read 15 of the Ishihara color plates with the left eye, but none with the right eye. The ocular alignment was orthophoric. Ocular movement was normal in both eyes. The anterior segment was normal except for relative afferent pupillary defect in the right eye. Fundus examination showed multiple patches of milky white retinal lesions scattered over the macular and peripapillary areas of the right eye (Figure 1). The optic nerve head was mildly swollen in the right eye. A small whitish patch was noted along the inferiotemporal arcade in the left eye. Fluorescein angiography of the right eye showed multifocal retinal arteriolar occlusion and capillary nonperfusion in the early phase (Figure 2). There was also a small hypofluorescent patch in the left eye, which became hyperfluorescent in the late phase. Moderate leakage of the involved retinal vessels and the optic nerve head were noted in the right eye at the late phase (Figure 3). Visual field examination revealed a large central scotoma in the right eye. Electroretinography results were normal in both eyes. Indocyanine green angiography was performed with a scanning diode laser ophthalmoscope (Rodenstock Instruments, Germany). After an intravenous injection of 25 mg ICG dye, transit of the dye through the choroid was detected by a 830 nm wavelength detector in the scanning laser ophthalmoscope. Videographic images were recorded on VHS tapes. At 1 minute, rarefaction of the choroidal vessels was present in the peripapillary area of the right eye (Figure 4). There was also a small patch of rarefied choroidal vessels along the



**Figure 2.** Retinal arteriolar occlusion and capillary nonperfusion were primary findings in early phase of fluorescein angiography.

inferiotemporal arcade of the left eye. Multifocal hypofluorescent patches were present in macular and peripapillary areas of the right eye at 8 minutes in the ICG angiographic study (Figure 5).

After diagnosis of Purtscher's retinopathy, megadose steroid therapy was given with methylprednisolone 250 mg every 6 hours intravenously for 3 days. Then the steroid amount was tapered gradually during the next 3 weeks. Two weeks later her vision had improved to 6/12 in the right eye. Most of the milky



**Figure 1.** Multiple patches of milky white retinal lesions were located primarily in macula and peripapillary areas in right eye.



**Figure 3.** Moderate leakage of fluorescein dye was noted along involved retinal vessels and optic disc in late phase of fluorescein angiography.



**Figure 4.** At 1'28" of ICG angiography, peripapillary area of right eye revealed distinct pattern of rarefaction of choroidal vessels.

white lesions resolved 1 month later. She was followed-up 10 months later. Vision of the right eye was 6/6. Some residual granularity could still be discerned in the central macular area of the retinal pigmented epithelium, and the optic nerve head appeared slightly pale (Figure 6).

### Discussion

In 1910, Otmar Purtscher described the visual loss in a severely traumatized patient that was associated with multiple superficial retinal white patches and retinal hemorrhages surrounding a normal-appearing optic disc.<sup>1</sup> In later reports, optic nerve head involvement was found to be a frequent association. In addition to the retinal arteriolar occlusion and capillary nonperfusion, staining of the optic nerve head has become an important finding of fluorescein angiography in patients with Purtscher's retinopathy.



**Figure 5.** At 8'32" of ICG angiography, multifoci of hypofluorescent patches (arrowheads) were scattered in macula and peripapillary areas in right eye.



**Figure 6.** Ten months later, only some retinal pigment epithelial granularity could be found in the posterior pole. There was a central depigmented scar at macula. Optic nerve head appeared slightly pale.

The severity of the optic nerve injury may influence the visual outcome. Optic atrophy may result in poor vision even when all retinal lesions resolve eventually. There are many proposed pathogenic mechanisms for retinal angiopathy, such as fat embolism, air embolism, granulocytic embolism, venous reflux, and angiospastic response.<sup>10</sup> Irrespective of the various pathogenic mechanisms, controversy persists as to whether the optic disc damage results from the nerve fiber loss of retinal angiopathy or from the ischemic injury of choroidal vascular compromise.<sup>11</sup> Choroidal perfusion has been noted to be normal in fluorescein angiography.<sup>2</sup> However, at long-term follow-up the alterations have not been limited to inner retinal layers. Peripapillary and central pigment epithelial granularity have been noted in some reports. <sup>6,11</sup> Furthermore, one case of histologic examination of the eyes performed 3 weeks after the onset of pancreatitis-associated retinopathy documented occlusion of both retinal and choroidal vessels.<sup>12</sup> Fat emboli lodged in vessels of the retina and choroid have also been demonstrated histologically in patients with Purtscher's retinopathy following multiple fractures.<sup>13</sup> Gomez-Ulla and colleagues<sup>14</sup> have recently shown the existence of a paramacular hypofluorescent patch of choroidal vascular abnormality by ICG angiography. In our patient, rarefaction of choroidal vessels in peripapillary and macular areas at early phase and multifocal hypofluorescent patches at late phase were demonstrated by an ICG angiographic study. Because of the blocking effect of the overlying edematous retina, we could not examine the choroidal vasculature more carefully. Nevertheless, from the aforementioned clinicopathologic evidence, we suggest that some of the clinical features of Purtscher's retinopathy, such as pigment epithelial granularity and optic atrophy, may ensue from the ischemic injuries of choroidal vasculopathy.

Different mechanisms probably lead to the retinal manifestation of Purtscher's retinopathy. Similar fundus findings have since been reported to be associated with compressive chest injuries,<sup>2</sup> fat embolism<sup>4</sup> from multiple fractures, retrobulbar anesthesia,<sup>5</sup> acute pancreatitis,<sup>6</sup> childbirth,<sup>7</sup> and connective tissue disease.<sup>3</sup> In patients with neck compression, the venous hydrostatic pressure may lead to active constriction and subsequent damage of the retina, either by direct endothelial damage or by retinal autoregulation.<sup>15</sup> The asymmetric involvement in our patient makes the hydrostatic pressure mechanism unlikely, albeit the head position at the time of trauma is an important factor in consideration of laterality. Burton<sup>11</sup> described four cases of unilateral Purtscher's retinopathy and suggested that arterial embolism seems to be the most plausible explanation. The risk of fat embolism in cervical spine fracture is rather low. There was no evidence of systemic fat embolism in our patient either. An alternative mechanism for arterial embolism has been suggested by the finding of complement-activated leukocyte aggregation with plasma samples from patients with pancreatitis-associated Purtscher's retinopathy.<sup>16,17</sup> Three previously reported causes for Purtscher's retinopathy-trauma, acute pancreatitis, connective tissue disease-are potentially able to activate the complement cascade.<sup>16,17,18</sup> An activated complement system can cause leukocytes to aggregate in vitro.<sup>19</sup> By injection of leukocyte aggregate, retinal microembolism with a picture similar to Purtscher's retinopathy has been produced in pigs.<sup>20</sup> Furthermore, activated leukocyte aggregate has been demonstrated to cause damage to the cultured endothelial cell through production of oxygen-free radical.<sup>21</sup> Thus, leukocyte aggregate provides a plausible pathogenic mechanism for Purtscher's retinopathy in patients with trauma, acute pancreatitis, and connective tissue disease.

Visual outcome in Purtscher's retinopathy is variable. Normal vision may not be recovered in patients with severe initial injury, in whom optic atrophy and macular infraction may persist in a late phase.<sup>7,8,9,10</sup> There is no well-defined treatment for Purtscher's retinopathy. Papaverine HCL, a peripheral vasodilator, has been used in a case of Purtscher's retinopathy based on the theoretical rationale of dilating ret-

inal arteriole to increase oxygen supply.<sup>5</sup> However, the therapeutic value of papaverine on ocular circulation is uncertain.<sup>22</sup> Atabay and coworkers<sup>8</sup> described late visual recovery in a patient who received megadose steroid treatment 3 weeks after the trauma. It has been proposed that the use of megadose steroid may stabilize the damaged cellular membrane in neuronal tissue, and thereby enable a degree of recovery from tissue insult.<sup>23</sup> Meanwhile, megadose steroid treatment can block the formation of complement-activated leukocyte aggregation and inhibit the production of oxygen-free radical.<sup>24,25</sup> In our patient, the visual recovery was successful under megadose steroid treatment. Nevertheless, the efficacy of this treatment in traumatic angiopathy still needs to be verified through systematic evaluation, with the outcome determinants such as initial extent and initial severity controlled.

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