

# Role of Multimodal Imaging in Purtscher Retinopathy

Dr. S. Saxena, Dr. A. Saxena, Dr. S. Basu

<sup>1</sup>Assistant Professor, Department of Ophthalmology, United Institute of Medical Sciences, Allahabad

<sup>2</sup>Assistant Professor, Department of Plastic Surgery, United Institute of Medical Sciences, Allahabad

<sup>3</sup>Consultant Ophthalmologist, Department of Ophthalmology, The Eye Foundation Hospital, Kochi

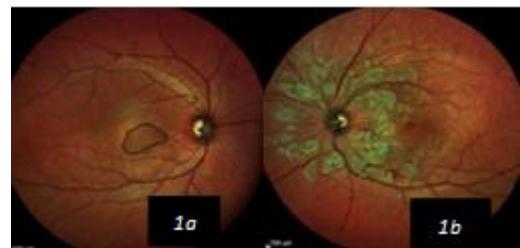
**Abstract:** *Purtscher retinopathy is a rare condition that occurs in patients with a history of trauma distant from the eyes.* <sup>1</sup> *Typical manifestations, including cotton wool spots, retinal hemorrhages and optic disc swelling, are confined to the posterior pole and often appear bilaterally. The pathogenesis underlying Purtscher retinopathy is not clear, but might be related to the embolic occlusion of precapillary retinal arterioles. Diagnosis is usually made on clinical grounds and supported by fluorescein angiography. For the majority of cases, visual function could recover to various extents without treatment.* <sup>2</sup> *Multimodal imaging can be a useful tool in identifying lesions which are very subtle on clinical examination in a visually symptomatic patient.*

**Keywords:** Hyper-reflective Foci, Purtscher, autofluorescence, steroids

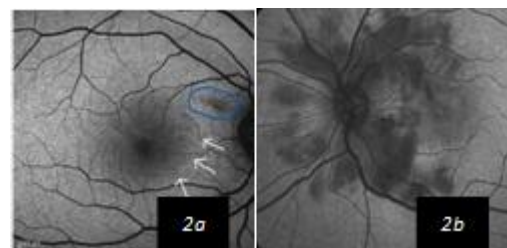
## 1. Case Report

We report a case of a 32-year-old man with no relevant previous ophthalmologic, systemic or familial history. The patient had a fall from bike six days back with traumatic head injury, yet no direct ocular trauma or acute lesions on brain computed tomography. He suffered contusion on his forehead. He came with the complaints of sudden painless diminution of vision in Left eye for three days. Best corrected visual acuity in the Right eye was 6/6p, N6, and Counting Finger 2 meters in his left eye. Patient complained a small scotoma in the right eye. Anterior segment examination was normal in both eyes. Funduscopic examination in the right eye revealed a subtle dark wedge-shaped area paracentral in location which was very much apparent on Colored fundus photography image (Fig.1a). Left eye fundus examination revealed polygonal areas of retinal whitening at peripapillary area and around macula with a clear demarcating area of non-affected retina on each side of the retinal arterioles. This was consistent with pathognomonic Purtscher flecken. There were areas of retinal whitening with ill-defined margins superficially over the vessels corresponding to cotton-wool spots. Few flame-shaped hemorrhages were also present with macular edema. (Fig.1b). Fundus Autofluorescence (FAF) imaging of right eye was suggestive for very subtle hyperautofluorescence close to fovea and hypoautofluorescence close to superior arcade and in the left eye there was hypoautofluorescence corresponding to Purtscher's flecken and blood and areas of hyperautofluorescence in paramacular areas. (Fig.2 a, b)

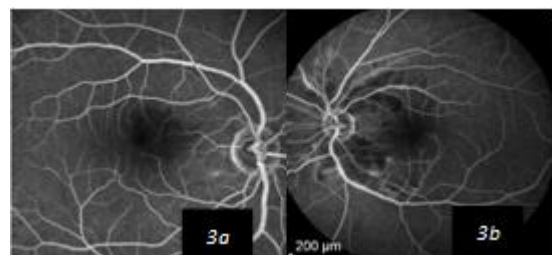
Fundus Fluorescein Angiography (FFA) of Right eye depicted enlargement of FAZ and small peripapillary capillary leak (Fig 3 a). Left eye, FFA depicted blocked choroidal fluorescence (either due to retinal whitening or blood), occluded retinal arterioles, area of capillary non perfusion, late leakage from the retinal vessels in area of ischemia and irregularly enlarged foveal avascular zone suggestive of macular ischemia (Fig.3b)



**Figure 1:** Fundus Photo 1a) Right eye, wedge shaped paracentral area marked with black. 1b) Left eye, Purtscher retinopathy with fleckens



**Figure 2:** FAF imaging a) Right eye, paracentral hyperautofluorescence marked with white arrows and area of hypo autofluorescence marked with blue circle. 2b) Left eye, hypoautofluorescence corresponding to areas of Purtscher flecken and blood along with areas of hyperautofluorescence in paramacular areas



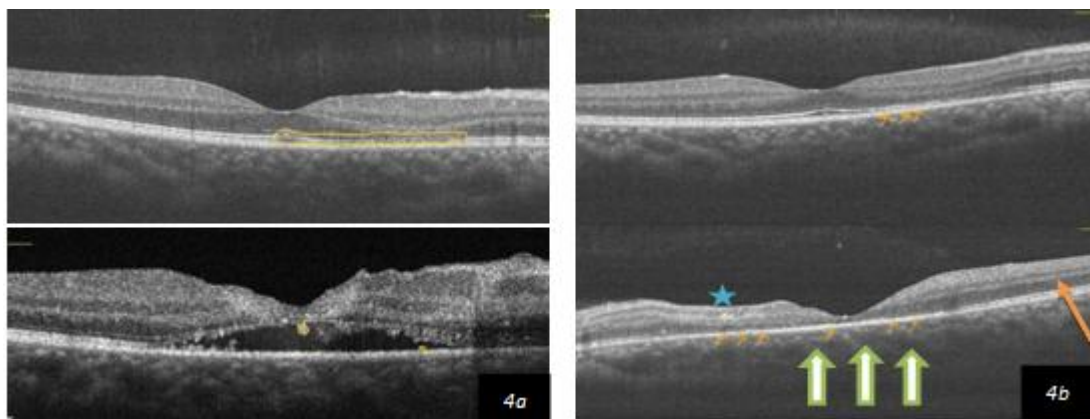
**Figure 3:** FFA 3a) Right eye, depicted enlargement of FAZ and small peripapillary capillary leak. 3b) Left eye, FFA depicted blocked choroidal fluorescence (either due to retinal whitening or blood), occluded retinal arterioles, area of capillary non perfusion, late leakage from the retinal vessels in area of ischemia and irregularly enlarged foveal avascular zone.

vessels in area of ischemia and irregularly enlarged foveal avascular zone suggestive of macular ischemia

In the acute phase, Wide field swept source Optical coherence tomography (OCT) of Right eye showed focal disruption of the ellipsoid zone nasal to fovea (Fig.4a). Left eye, OCT suggestive of hyperreflectivity in the inner and middle retinal layers suggestive of paracentral acute middle maculopathy. Also, there is presence of disorganization of retinal inner layers (DRIL) with multiple hyperreflective retinal foci (HRF) mainly in outer retinal layers with neurosensory detachment (Fig.4b).

Based on history and clinical findings, the patient was diagnosed as a case of Bilateral Purtscher's Retinopathy. He was managed with oral Prednisolone 40 mg daily for one week followed by gradual tapering. After one week, his

BCVA improved to 6/60 in L/E and 6/6 in R/E with mild resolution in scotoma. One week later, it showed further improvement to 6/36 in L/E. Fundus examination revealed resolving hemorrhages and cotton wool spots in L/E. At follow-up visit of 2months, BCVA was 6/6 in right eye and 6/36 in L/E. On OCT of the right eye, there was focal RPE atrophy (Fig.4c), in left eye, there was outer retinal atrophy and photoreceptor loss (Fig.4d). On Fundus photography and FAF of the right eye there was reduction in size of the wedge shaped defect and very minimal areas of hypoautofluorescence at nasal parafoveal area (Fig 5a, c) and in Left eye, there was a decrease in number and size of cotton wool spots and hemorrhages and FAF showed linear areas of Hyperautofluorescence and areas of Hypoautofluorescence suggestive of involvement of RPE (Fig 5b, d).



**Figure 4:** OCT a) Right eye, focal disruption of the ellipsoid zone nasal to fovea (yellow area marked). b) Left eye, hyperreflectivity in the inner and middle retinal layers suggestive of paracentral acute middle maculopathy (orange arrow), presence of disorganization of retinal inner layers (DRIL; blue star) with multiple hyperreflective retinal foci (HRF) mainly in outer retinal layers (yellow dots) with neurosensory detachment (white arrows). c) right eye focal RPE atrophy (yellow arrows). d) left eye, outer retinal atrophy and photoreceptor loss (yellow arrows).



**Figure 5:** Fundus Photo a) right eye, reduction in defect size, b) left eye, decrease in number and size of cotton wool spots and hemorrhages Figure 5 FAF c) right eye minimal areas of hypo autofluorescence at nasal parafoveal areas. d) left eye, Linear areas of hyperautofluorescence and few areas of hypoautofluorescence suggestive of involvement of RPE.

## 2. Discussion

Purtscher's retinopathy was first described in 1910 by Otmar Purtscher, in a patient with severe head trauma.<sup>3</sup> Purtscher retinopathy is a vision-threatening condition most commonly observed in young or middle-aged males as a consequence of trauma, head or chest injuries, thoracic compression, and bone fractures.<sup>3</sup> The term "Purtscher-like retinopathy" is used to describe the retinopathy seen in conditions other than trauma, acute pancreatitis, systemic lupus erythematosus, lymphoproliferative disorders, bone marrow

transplantation, orthopaedic surgeries, intravitreal injection, fat embolism syndrome, childbirth, connective tissue disorders, and renal failure.<sup>4</sup> The diagnosis of PR is based mainly on the clinical image and evaluation of the fundus of the eye. The main symptom by the patients is sudden, painless decrease in vision, following immediately after trauma or within 24-48 hrs, which may affect one or both eyes ranging from minimal impairment to hand motions visual acuity.<sup>5</sup>

Pathophysiology not elucidated completely, suggested mechanism is the occlusion of retinal arterioles by either

aggregated granulocytes and/or platelets, following complement activation, or to fat micro emboli or fibrin clots.<sup>4, 6</sup>

Fundus of the patients shows Purtscher flecken, areas of retinal whitening in the superficial aspect of the inner retina, between the arterioles and venules clearly demarcated by the area within 50 µm from the normal retina due to occlusion of precapillary arterioles, cotton-wool spots, retinal haemorrhage, macular oedema and optic disc swelling.<sup>5</sup>In our case, fundus photography of the right eye was showed wedge shape defect corresponding to patient's scotoma. Now a days fundus autofluorescence is an important imaging tool. Only 1 report of a Purtscher-like retinopathy case (following pancreatitis) was found in the literature, which described the results of FAF examination.<sup>7-14</sup>In our case the clinical findings in the right eye were subtle however autofluorescence was clearly showing areas of hyperautofluorescence and areas of hypoautofluorescence suggestive of retinal pigment layer dysfunction.

SD-OCT provides supplementary findings for diagnosis, Purtscher flecken appear as hyperreflective inner retinal layer, cotton wool spots appear as hyperreflective areas in inner layers of retina.<sup>3</sup> SD-OCT also provides information on extent of macular edema, any neurosensory detachment of retina and thickness of retina which can be used as a prognostic factor. In our case Swept Source-OCT was used, which showed better visualization of the layers of retina and other important findings like hyper reflective foci (HRF), PAMM, DRIL in the left eye which actually helped in prognosticating the case.

HF, which are discrete, focal, hyper reflective lesions, are observed in some retinal diseases, such as AMD, RVO, central serous choroidretinopathy, Stargardt disease, retinitis pigmentosa as well as Diabetic Retinopathy.<sup>8-10</sup> Pathophysiology of HF, is still not fully understood. It is suggested that inflammation activates microglia cells and induce them to swell as well as spread to all retinal layers to become HRF.<sup>11</sup> In DR, HF was thought to be small intraretinal proteins or lipid deposits which form after inner blood-retinal barrier breakdown.<sup>12</sup> HF can act as a biomarker of poor final visual outcome. To our knowledge HRF is not reported in cases of Purtscher's retinopathy till now, and further study needs to be done.

Fluorescein angiography can show capillary nonperfusion, retinal and/or disc edema, perivascular staining, and late leakage from injured retinal vessels providing further information about the severity of the disease.<sup>3, 5, 6</sup>In our case, fluorescein angiography, left eye depicted blocked choroidal fluorescence, occluded retinal arterioles, area of capillary non perfusion and macular ischemia.

In the majority of the cases, the acute lesions resolve spontaneously within 1–3 months after the onset and may be replaced by mottling of the RPE, temporal disc pallor, or attenuation or sheathing of the retinal vessels.<sup>5</sup>There are no consensual guidelines of the therapeutic approach of this condition.<sup>6</sup>The majority of researchers recommend the administration of steroids during treatment. There are reports concerning improvement of visual acuity after the use of

steroid therapy. However, there are no randomised studies confirming the effectiveness of such a management.<sup>13</sup> In our case, after giving oral Prednisolone 40 mg, BCVA in the Left eye improved from 6/60 in L/E to 6/36 and also there was reduction in visually significant scotoma in the Right eye. The prognosis in PR depends mainly on the duration and the extent of severity of the changes observed during the early period of the disease.

### 3. Conclusion

Patient with decreased visual acuity following trauma requires ophthalmologic evaluation to rule out Purtscher's retinopathy. Despite the lack of support of standard treatment for this condition, it justifiable to give steroid therapy in patients with PR. Careful and continuous clinical observation of each patient still remains an important aspect of management. Clinical examination is often not sufficient for evaluating the patient's condition. Multimodal imaging then becomes important in making the diagnosis, in monitoring the progression of disease, and as a surrogate outcome measure of the efficacy of an intervention.

ETHICS: Consent was taken from the patient and his attendant and the full privacy was maintained throughout the treatment course.

### References

- [1] Miguel AI, Henriques F, Azevedo LF, et al. Systematic review of Purtscher's and Purtscher-like retinopathies. *Eye (Lond)* 2013; 27: 1–13. [PMC free article] [PubMed]
- [2] Agrawal A, McKibbin MA. Purtscher's and Purtscher-like retinopathies: a review. *Surv Ophthalmol* 2006; 51: 129–136. [PubMed]
- [3] Roskal-Wałek J, Gierada M, Mackiewicz J. Purtscher's retinopathy—case report and literature review. *Medical Studies/Studia Medyczne*. 2017 Jan 1; 33 (2): 127-32.
- [4] Campo SM, Gasparri V, Catarinelli G, Sepe M. Acute pancreatitis with Purtscher's retinopathy: case report and review of the literature. *Digestive and Liver Disease*. 2000 Nov 1; 32 (8): 729-32.
- [5] Schachat AP, Wilkinson CP, Hinton DR, Sadda SR, Wiedemann P, editors. *Ryan's Retina E-Book*. Elsevier Health Sciences; 2017 Apr 17.
- [6] Massa R, Vale C, Macedo M, Furtado MJ, Gomes M, Lume M, Meireles A. Purtscher-like retinopathy. *Case reports in ophthalmological medicine*. 2015; 2015.
- [7] Giani A, Deiro AP, Sabella P, Eandi CM. Spectral domain-optical coherence tomography and fundus autofluorescence findings in a case of Purtscher-like retinopathy. *Retinal Cases & Brief reports*. 2011; 5 (2): 167-70.
- [8] De Benedetto U, Sacconi R, Pierro L, Lattanzio R, Bandello F. Optical coherence tomographic hyperreflective foci in early stages of diabetic retinopathy. *Retina*. 2015; 35 (3): 449–53.
- [9] Bolz M, Schmidt-Erfurth U, Deak G, Mylonas G, Kriechbaum K, Scholda C. Optical coherence tomographic hyperreflective foci: a morphologic sign of lipid extravasation in diabetic macular edema. *Ophthalmology*. 2009; 116 (5): 914–20.

- [10] Chen KC, Jung JJ, Curcio CA, Balaratnasingam C, Gallego-Pinazo R, DolzMarco R, Freund KB, Yannuzzi LA. Intraretinal Hyperreflective foci in acquired Vitelliform lesions of the macula: clinical and histologic study. *Am J Ophthalmol.*2016; 164: 89–98.
- [11] Coscas G, De Benedetto U, Coscas F, Li Calzi CI, Vismara S, Roudot-Thoraval F, Bandello F, Souied E. Hyperreflective dots: a new spectral-domain optical coherence tomography entity for follow-up and prognosis in exudative age-related macular degeneration. *Ophthalmologica Journal international d'ophtalmologie International journal of ophthalmology Zeitschrift fur Augenheilkunde.*2013; 229 (1): 32–7.
- [12] Nishijima K, Murakami T, Hirashima T, Uji A, Akagi T, Horii T, Ueda-Arakawa N, Muraoka Y, Yoshimura N. Hyperreflective foci in outer retina predictive of photoreceptor damage and poor vision after vitrectomy for diabetic macular edema. *Retina.*2014; 34 (4): 732–40.
- [13] C. Atabay, T. Kansu, and G. Nurlu, “Late visual recovery after intravenous methylprednisolone treatment of Purtscher’s retinopathy, ” *Annals of Ophthalmology*, vol.25, no.9, pp.330– 333, 1993.
- [14] Guerra, R., Silva, I., Marback, E., Maia Jr., O. and Marback, R., 2014. Fundus autofluorescence in blunt ocular trauma. *Arquivos Brasileiros de Oftalmologia*, 77 (3).