

## Purtscher Retinopathy and Fat Embolism Syndrome

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### ABSTRACT

Purtscher retinopathy and fat embolism syndrome are chorioretinopathies occurred after indirect ocular trauma. These diseases look like similar conditions but there are important differences which are help to differential diagnosis. In general, trauma is responsible from these diseases; but crush trauma play role in Purtscher retinopathy, while bone fracture play role in fat embolism syndrome. Retinal whitening is dominant funduscopic finding in these diseases; but whitening is wide and centrally in Purtscher retinopathy, while it is smaller and located more peripherally in fat embolism syndrome. These clinical findings associated with Purtscher retinopathy and fat embolism syndrome, resolve spontaneously in weeks or months but permanent visual loss can be seen in survivors. In this manuscript, we aimed to give general information about emergence and clinical course and differentiation of Purtscher retinopathy and fat embolism syndrome.

**Key words:** Purtscher retinopathy; Fat embolism syndrome

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### INTRODUCTION

Indirect trauma can cause chorioretinopathies. These diseases are rare but sight-threatening. Indirect ocular trauma-related chorioretinopathies are Terson syndrome, Purtscher retinopathy, fat embolism syndrome, shaken baby syndrome, whiplash maculopathy, and Valsalva retinopathy. Ophthalmologist must be aware of ocular negative effects of several trauma types including contusion, compression, crush, or acceleration/deceleration. Purtscher syndrome and fat embolism syndrome are similar conditions in terms of pathophysiological and clinical.

In this manuscript, we aimed to give general information about emergence and clinical course of Purtscher retinopathy and fat embolism syndrome; to emphasize the differences of these diseases.

### PURTSCHER RETINOPATHY

#### Etiology

Otmar Purtscher described Purtscher retinopathy as retinal angiopathy associated with head trauma, in a man falling down on the head from tree, since 1910. It is understood that Purtscher retinopathy can occurs after trauma in other parts of body, or several systemic diseases<sup>[1,2]</sup>. In todays, diagnosis of Purtscher retinopathy based on trauma history and funduscopy. Some cases are presented firstly to ophthalmologist due to visual loss. These cases masqueraded by visual loss, should be directed to internal medicine specialist to investigate the susceptible systemic condition. Acute pancreatitis, systemic lupus erythematosus (SLE), thrombotic thrombocytopenic purpura (TTP), HELLP, chronic renal failure, scleroderma, dermatomyositis, or amnion fluid embolism can be responsible for this clinic situation. In general, if responsible condition of this disease is other than crush trauma, when disease is called as Purtscher-like retinopathy<sup>[3]</sup>. Acute pancreatitis is the most

common reason of Purtscher-like retinopathy, but it can be seen following radical prostatectomy or anorexia nervosa<sup>[4,5]</sup>.

### Clinic and Pathophysiology

The most common presentation of Purtscher retinopathy is painless visual loss and presence of reactive afferent pupillary defect (RAPD). In Purtscher retinopathy, retinal findings do not occur in first 24-48 hours in general, but vary bilateral retinal abnormalities including optic disk edema, optic disk swelling, retinal whitening around optic disk, retinal hemorrhages, and pseudo-cherry red spot are occurred in a few days after trauma<sup>[6]</sup>. Purtscher retinopathy is a bilateral asymmetric disease. In superficial layers of inner retina, between arteriole and venule, clear bordered white islands are named as Purtscher fleckens and it indicates distal arteriolar occlusion. Purtscher fleckens provide the differentiation from central retinal artery occlusion because cleared borders between vessels are not seen in central retinal artery occlusion. In fundus fluorescent angiography, capillary drop-out and leakage is shown in area of soft exudates, and hypofluorescent is shown due to fluorescent blockage in area of retinal hemorrhage. Retinal capillary non-perfusion in fovea, causes visual loss<sup>[1]</sup>.

Soft exudates are constructed to retinal whitening in funduscopy. Retinal capillary destruction and arterial occlusion-related inner retinal atrophy are responsible from these soft exudates<sup>[7]</sup>. Trauma-related embolism cause endothelial damage and complement associated granulocyte aggregation<sup>[8,9]</sup>. These pathophysiological processes trigger the formation of embolism in retinal vessels<sup>[10]</sup>. Clinical and experimental studies shown that in Purtscher retinopathy small peripapillary retinal arterioles plugged by intravascular microparticles<sup>[11,12]</sup>. These intravascular microparticles are occurred associated with susceptible systemic condition.

Purtscher retinopathy effects posterior pole especially. This condition is explained with special anatomical features of retinal vessels. Except 2 capillary layers, retinal vessels course into nerve fiber layer. These capillary layers present in ganglion cell layer and inner nuclear layer and these capillary layers connected with another. In macula, there is 3<sup>rd</sup> capillary layer and it present between other 2 capillary layers. 4<sup>th</sup> capillary layer present in the most superficial layer of retina and it move away from optic disk to nasally for 2 disk diameters, to direction of temporal vascular arcs for 4 disk diameters, and to temporally for 1 disk diameter. There is fewer anastomosis between peripapillary vessels in these regions. Because of these specific anatomic features, peripapillary retina is more sensitive for embolic occlusion<sup>[13]</sup>.

### Treatment and Course

In past, it is thought that high dose steroid is provided neuronal membrane stabilization, and is blocked complement activation-related granulocyte aggregation. But effectiveness of steroid in middle or long term cannot be shown<sup>[10,14]</sup>. In both 2 groups using and not using high dose steroid, visual acuities were found similar in 1<sup>st</sup>, 2<sup>nd</sup>, and 6<sup>th</sup> months<sup>[15]</sup>. Although hyperbaric oxygen and vasodilator drugs are considered as useful even if limited, the most effective treatment strategy is treating the susceptible systemic condition when the formation of new embolism and additional retinal damage can be blocked<sup>[16,17]</sup>. Retinal findings are resolved in 1-3 months spontaneously and the visual functions recovery in the majority of the patients, but infarct in macula and optic disk is irreversible. Macular pigmentation, sheathing of vessels, optic atrophy, and neovascular glaucoma can be seen as a legacy of Purtscher retinopathy.

## FAT EMBOLISM SYNDROME

Fat embolism syndrome is described as systemic and ocular findings following long bone fracture. It is occurred 5 percent of all fractures and retinal findings occur 60 percent of all patients with fat embolism syndrome<sup>[2]</sup>. In general, fat embolism syndrome occurs a few days later fracture of long bone, but it has been reported following tarsal bone fracture. Fat embolism syndrome is mortal for 20 percent of patients<sup>[18]</sup>.

Diagnosis of fat embolism syndrome is quietly complicated and there are many diagnostic criteria and scoring systems. Systemic findings of this diseases include petechial rash, alteration in central nervous system condition (such as paralysis, delirium, coma), respiratory insufficiency, fever, tachycardia, anemia, and elevation of erythrocyte sedimentation rate<sup>[11]</sup>.

After the bone fracture, coagulation and hypovolemia is the main problems. Complement activation, and saponification of fat storage due to elevation of plasma lipase, are associated to the pathophysiological process. Releasing of pulmonary fat droplets in late stage and recurrent embolism formation cause fat embolism syndrome. Insufficient fixation of fracture contributes recurrent embolism formation that is sourced from hydrolyzed fatty acids. So, all organs be exposed to mechanical and biochemical damage of these fat droplets<sup>[19]</sup>.

Bilateral intraretinal hemorrhages and soft exudates are most common funduscopic findings in fat embolism syndrome. This disease looks alike Purtscher retinopathy in terms of clinical findings, but in fat embolism syndrome retinal lesions are smaller, fewer, located in more periphery, and they are seen earlier<sup>[2]</sup>. Because the average diameter of fat emboli is 5-10  $\mu\text{m}$ , while average diameter of retinal arteriole is 45  $\mu\text{m}$ , so fat emboli goes more away. In Purtscher retinopathy retinal arterioles are affected primarily, while fat embolism syndrome is disease of vessels with smaller diameter. Visual symptoms are weaker and the most of patients are asymptomatic in fat embolism syndrome<sup>[20]</sup>. These findings resolve in a few weeks and ophthalmologists are seen this clinical condition rarely. Persistent paracentral scotoma is the most common clinic presentation that occurs after the acute stage<sup>[2]</sup>.

## CONCLUSION

Purtscher retinopathy and fat embolism syndrome are similar systemic conditions and they are diagnosed with history and detailed ocular and systemic examination. These diseases differentiate when considering type of trauma (crush or fracture); presence of systemic diseases or symptoms; shape, size and localization of retinal whitening.

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