DOI: 10.7860/JCDR/2024/73623.19941

Case Report

Ophthalmology Section

Purtscher-like Retinopathy following Acute Pancreatitis: A Case Report

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ABSTRACT

Purtscher Retinopathy (PR) is also known as traumatic retinal angiopathy or lymphorrhagia retinae or retinal teletraumatism. It presents as occlusive retinal microvasculopathy distinguished by the presence of multiple retinal white lesions surrounding the optic nerve head and fovea, accompanied by perivascular clearing, often coinciding with intraretinal haemorrhages. Correlated conditions comprise of acute pancreatitis, crush injuries, severe head trauma, chest compression, long bone fractures, orthopaedic surgical procedures, Haemolysis, Elevated Liver Enzymes, and Low Platelets (HELLP) syndrome and battered baby syndrome. PR is a rare microvasculopathy, which is occlusive in nature and causes sudden and painless diminution of vision following serious trauma but might also be associated with non-traumatic aetiologies as well. The term Purtscher-like retinopathy is used when it occurs due to non-traumatic causes. Here, the authors present a case of a 25-year-old female, admitted to the hospital for severe abdominal pain, persistent nausea and multiple episodes of vomiting since two days. Her laboratory findings and radiological investigations revealed findings suggestive of acute pancreatitis. On day 2 of admission, she reported a bilateral diminution of vision. The ophthalmic evaluation revealed visual impairment and characteristic retinal findings of Purtscher flecken, cotton wool spots and few intraretinal haemorrhages mainly in the posterior pole. This case highlights the association between Purtscher-like retinopathy and acute pancreatitis.

Keywords: Cotton wool spots, Haemorrhages, Purtscher flecken, Purtscher retinopathy, Retinal microvasculopathy

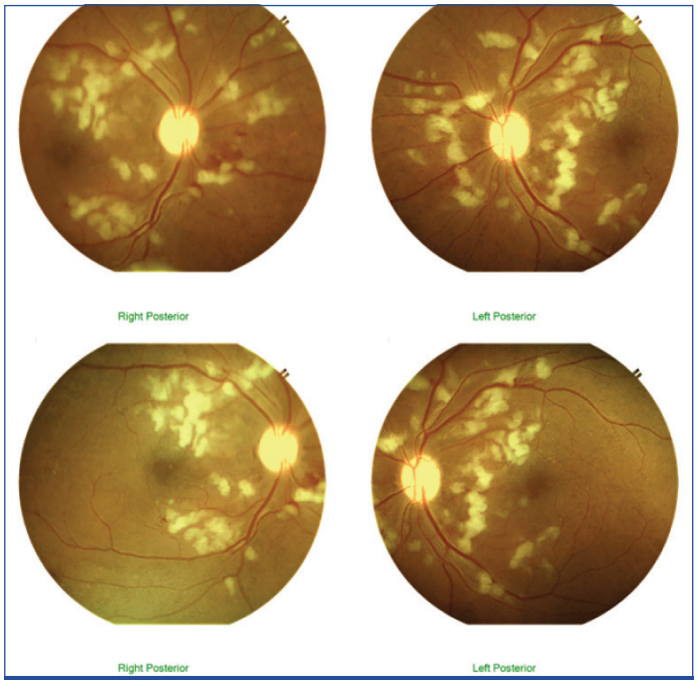
CASE REPORT

A 25-year-old female was referred to the ophthalmology outpatient department for diminution of vision in both eyes since one day. The diminution of vision was severe in nature and started suddenly without any pain, redness, floaters or flashes of light.

Two-day-ago, she had presented to the emergency department with complaints of severe epigastric pain not aggravated or relieved with any factors, nausea and two episodes of vomiting which was non-projectile in nature containing food particles. She was treated symptomatically with intravenous pantoprazole 40 mg and intravenous ondansetron 4 mg stat for the same. Patient denied history of similar episodes in the past. Patient also denied history of personal bad habits.

Blood investigations and radiological imaging were done. Laboratory investigations revealed significantly raised levels of lactate dehydrogenase 567 units/L, serum amylase 572 units/L and serum lipase 567 units/L. Her abdominal ultrasound findings suggested mild pancreatitis with moderate ascites. Hence, she was diagnosed as a case of acute pancreatitis and managed conservatively by fluid resuscitation (intravenous ringer lactate), intravenous antibiotic (piperacillin and tazobactam 4.5 gm three times a day), intravenous antacid (pantoprazole 40 mg once a day).

On ophthalmic evaluation, her best-corrected vision in the Right Eye (RE) was 6/18 and 6/36 in the Left Eye (LE). The anterior segment examination of both eyes was normal with normal pupillary reflexes. Dilated posterior segment examination of the RE revealed clear vitreous; optic disc had well-defined margins with a pink and healthy neuro-retinal rim, with a vertical cup-to-disc ratio of 0.3; dull foveal reflex; multiple, fluffy, near confluent polygonal areas of intra-retinal whitening suggestive of cotton wool spots extending upto three disc diameters along with flame-shaped intraretinal haemorrhages present superotemporal and inferotemporal to the optic disc, obscuring the underneath retinal vasculature along with retinal oedema. Dilated posterior segment examination of LE revealed the same findings as present in the RE mentioned above [Table/Fig-1].

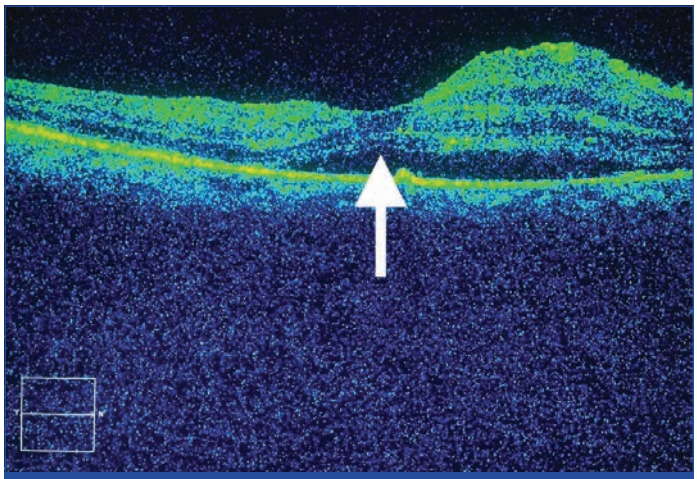


[Table/Fig-1]: Colour fundus photograph of the Right Eye (RE): visible swelling of the retina with multiple cotton wool spots and few intra-retinal haemorrhages and Colour fundus photograph of the Left Eye (LE): visible swelling of the retina with cotton wool spots and few intra-retinal haemorrhages.

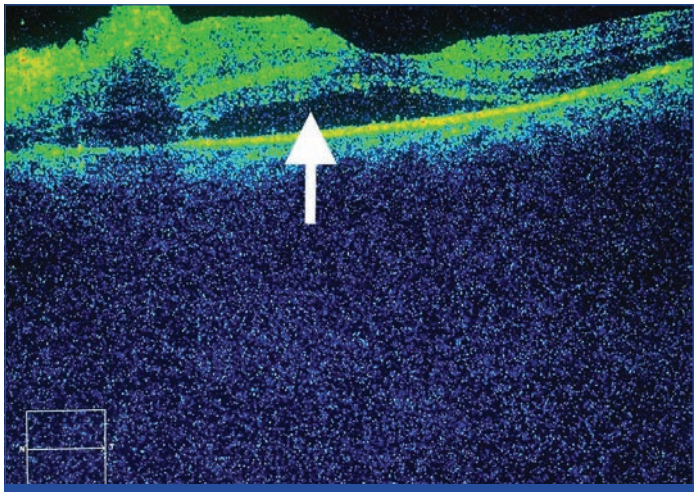
Fundus imaging of both eyes was done with a 3nethra flora mydriatic retinal camera. Spectral-domain Optical Coherence Tomography (OCT) done with Carl Zeiss CIRRUS MODEL 50) of both eyes showed a subfoveal collection of subretinal fluid and macular oedema on the nasal side LE >RE [Table/Fig-2,3]. The Central Retinal Thickness (CRT) in both the eyes was 331 µm.

Based on her clinical history, fundus evaluation and ophthalmic investigations and lack of other systemic conditions which can give rise to similar clinical picture, she was diagnosed as a case of purtscher-like retinopathy following acute pancreatitis. Patient was

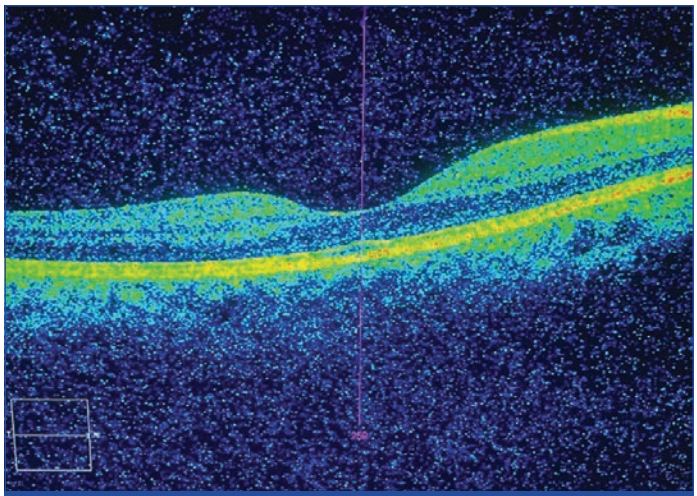
started on 1% nepafenac eyedrops three times a day in both eyes, oral prednisolone 1 mg/kg/day. She was advised a weekly follow-up for initial two weeks followed by a monthly review with slow tapering of oral steroids. In the 3rd month, her best corrected visual acuity in RE was 6/9 partial and LE was 6/12. An SD-OCT of both eyes was performed again which revealed a normal foveal contour with complete resolution of macular oedema [Table/Fig-4,5]. The CRT had reduced to 217 µm in RE and 216 µm in LE.



[Table/Fig-2]: OCT of the Right Eye (RE) macula shows subretinal fluid and macular oedema on the nasal side.



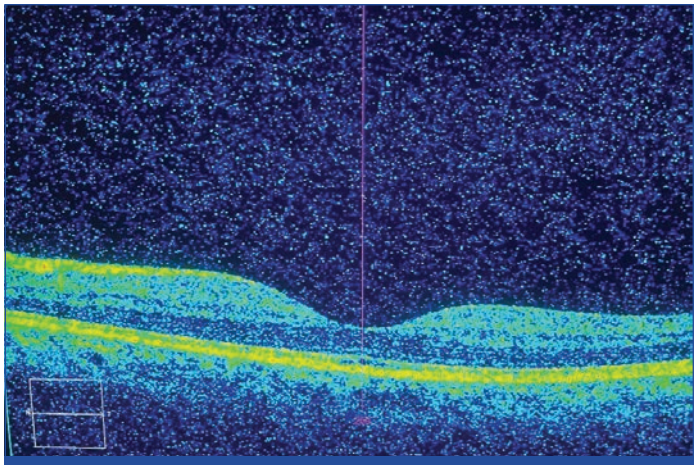
[Table/Fig-3]: OCT of the Left Eye (LE) macula shows inner retinal layer hyper-reflectivity, thickening in the inner-retina, folds in the internal limiting membrane on the nasal side with presence of subretinal fluid.



[Table/Fig-4]: OCT of the Right Eye (RE) macula shows normal foveal contour and resolution of oedema at three months follow-up.

DISCUSSION

The PR was originally described by Otmar Purtscher, an Austrian ophthalmologist in 1910 where he linked it to head trauma [1].



[Table/Fig-5]: OCT of the Left Eye (LE) macula shows normal foveal contour and resolution of oedema at three months follow-up.

Diagnosing PR relies on assessing the fundus along with relevant medical history. Patients commonly report sudden, painless vision deterioration, which can impact one or both eyes. While symptoms affecting both eyes occur in 60% of cases, instances of unilateral retinopathy are also documented. Characteristic changes in the fundus include Purtscher flecken, cotton wool spots due to microvascular infarct of the retinal nerve fiber layer, macular oedema, optic disc swelling, and retinal haemorrhages. The distinctive purtscher flecken are regions of retinal whitening sharply delineated within 50 µm of the normal retina which occur due to occlusion of precapillary arterioles [2].

The SD-OCT imaging can serve as an additional diagnostic tool for PR. It allows for the visualisation of varying severity of macular oedema, which, when significantly increased, can result in the breakdown of retinal layers, detachment of the neurosensory retina and retinal pigment epithelium, ultimately leading to reduced visual acuity. Seung K and Hoon L documented a case of PR featuring diffuse serous macular detachment [3]. In the present case, SD-OCT revealed similar changes, including subfoveal neurosensory retinal detachment due to the accumulation of subretinal fluid.

An additional valuable assessment in PR involves utilising Fundus Fluorescein Angiography (FFA) which enables visualisation of the circulation within the retina and choroid. Observable changes encompass the obstruction of choroidal fluorescence, blocked arterioles, regions of non-perfusion, as well as delayed leakage from retinal vessels in ischaemic areas and swelling of the optic disc. However, FFA could not be performed due to the patient's systemic condition in this case; therefore, FFA findings were not compared with the previous study.

Currently, microembolisation of the retinal and choroidal vessels stands as the leading hypothesis for the onset of PR. Depending on the underlying cause of the retinopathy, the occlusive material may include air, lipids, clusters of leukocytes, platelets, fibrin, or immunoglobulins. Trauma and inflammatory conditions such as acute pancreatitis can trigger the activation of the complement system, the involvement of which in the pathogenesis of retinopathy has been validated through laboratory analyses.

The prognosis for retinopathy varies and can be unpredictable. In most patients, fundus changes typically resolve within two months, and visual acuity returns to its previous state [2]. However, severe cases may exhibit persistent alterations in the retinal pigment epithelium, along with macular and optic nerve disc atrophy or neovascularisation, resulting in irreversible vision loss. Functional impairments such as non-acute central vision and scotoma in the visual field persist in approximately 50% of cases [4].

As per findings by Holak HM et al., the prognosis of PR hinges primarily on the duration and severity of early-stage changes.

Rapid resolution of oedema is associated with a favourable visual acuity prognosis, regardless of other factors. The duration of acute retinal changes emerges as the pivotal factor influencing long-term prognosis [5].

A diagnostic criterion of PR has been suggested. Three out of the following five criteria should be present: Purtscher flecken; retinal haemorrhages, low to moderate number (1-10); cotton wool spots (typically restricted to the posterior pole); probable or plausible explanatory aetiology; complementary investigation compatible with the diagnosis [1]. In the present case, all criteria were met and we called it purtscher like retinopathy, because it occurred because of atraumatic reason.

A case report published by Sharma S et al., reported a 28-year-old male with alcohol consumption history, who was diagnosed with acute necrotising pancreatitis developed purtscher like retinopathy with serous macular detachment and was treated symptomatically. Resolution of fundus findings were noted on further follow-up [6].

A case report published by Almpani M et al., reported a 33-year-old female pre-diagnosed with anxiety and attention deficit disorder developed acute pancreatitis and was admitted for the same, complained visual impairment. On further examination, she was diagnosed with purtscher-like retinopathy. OCT findings were consistent with the same [7].

PR or purtscher's like retinopathy is exceptionally rare occurring at a rate of 0.24 cases per million, highlighting its unique nature [8]. Furthermore, similar fundus findings are also found in other conditions giving rise to differential diagnoses of grade 3 or 4 hypertensive retinopathy, partial central retinal arterial occlusion, lupus retinopathy, HIV retinopathy etc., [1].

CONCLUSION(S)

In summary, Purtscher's-like retinopathy is a rare condition observed in patients with acute pancreatitis. The clinical history and findings in this case suggested acute pancreatitis as the most likely cause, diagnosed on the basis of history, symptoms and laboratory investigations. OCT, being a quick and non-invasive procedure, can assess the extent and involvement of retinal layers and macula. In the present patient, topical nepafenac drops and systemic oral steroids showed promising improvement in visual acuity and the fundus picture. Hence, it becomes of utmost importance to look for any systemic associations in the patient to treat the condition effectively.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Jun 24, 2024
- Manual Googling: Jul 16, 2024
- iThenticate Software: Jul 19, 2024 (11%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Jun 18, 2024**
Date of Peer Review: **Jul 06, 2024**
Date of Acceptance: **Jul 23, 2024**
Date of Publishing: **Sep 01, 2024**