

# Unilateral multiple serous retinal detachments in polycythemia Vera patient

## Abstract

**Purpose:** To report a case with new signs that could be caused by polycythemia vera itself or just a coincidence and managing it with systemic steroids.

**Methods:** This was interventional case report, of a 59-year-old patient with polycythemia vera, presented with acute unilateral vision loss. Ophthalmological investigation revealed multiple serous retinal detachments with mild anterior chamber reaction these signs look similar to ocular signs of Vogt-Koyanagi-Harada disease, but in one eye and without systematic involvement of VKH. Polycythemia vera is a haematological disease that causes an increase number of red blood cells. The patient red blood cells were normal and he was compliance with Polycythemia vera treatment. All infectious tests were negative as well as autoimmune disease markers. Unilateral probable VKH disease was our diagnosis, patient was managed.

**Results:** Polycythemia vera is a haematological disease that have a few ocular symptoms. These signs include multiple serous retinal detachments with uveitis that respond well to corticosteroids.

**Conclusion:** Systematic corticosteroids which show a remarkable improvement in ocular signs and symptoms of Polycythemia vera that mimic VKH.

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**Abbreviations:** RE, right eye; LE, left eye; PV, polycythemia vera; VKH, vogt-koyanagi-harada; OCT, optical Coherence tomography; ESR, erythrocyte sedimentation rate; ANA, antinuclear antibody; RA, rheumatoid arthritis

## Introduction

Serous retinal detachments have a long list of different aetiologies. Inflammatory causes include (VKH) Vogt-Koyanagi-Harada, sympathetic ophthalmia and other choroidal inflammatory disease. Vascular causes include malignant hypertension, Coats' disease, age-related macular degeneration. Beside choroidal tumors like melanoma and haemangioma and structural anomalies like optic disc pit and coloboma.<sup>1</sup> Polycythemia vera (PV) is a chronic myeloproliferative disorder in which malignant stem cell proliferate in the bone marrow. The main characteristic is uncontrolled red blood cells production, in addition to white blood cells and platelets.<sup>2</sup> Ocular manifestations of PV are ranged from vessel occlusion to hemorrhage. Although the causal of ocular sign is not clear, hyperviscosity was the main element in ocular disease.<sup>3</sup> In this case we are presenting a presumed rare manifestation of PV which was treated by corticosteroids successfully.

## Case presentation

59-year-old patient presented to the ophthalmology clinic at Al Mouwasat University Hospital, complaining of sudden vision loss in the right eye from one day. The patient complained from central constant scotoma with a red painful eye. Best-corrected visual acuity (BCVA) was 20/50 in the right eye (RE) and 20/20 in the left eye (LE) on tumbling E Snellen chart. Intraocular pressure was 12 mmHg and

14 mmHg in the right and left eyes, respectively. Pupils were round, reactive and symmetrical, without relative afferent pupillary defect. Slit-lamp examination of the right eye revealed anterior chamber inflammation with 1+cells and ciliary injection and a quiet left eye. Fundus evaluation of the right eye had several focal posterior pole serous retinal detachments involving the macula with no vitreous cells while in the left eye was within normal limits.

Optical coherence tomography (OCT) confirmed serous retinal detachment at the macular area with a retinal thickness of 1178 micrometer (Figures 1&2). There were serious retinal detachments in temporal superior to optic nerve with a retinal thickness of 362 , nasal superior with 325 , and temporal with 256 (Figure 3). A fundus fluorescein angiography (FA) revealed angiographic features typical of Vogt-Koyanagi-Harada disease in the RE: multifocal blocking of background fluorescence from the choroid, serous detachments with late pooling of dye, optic disc hyperfluorescence, focal areas of delayed choroidal perfusion and multifocal areas of leakages with late pooling and leakage of dye in the subretinal spaces (Figure 4). Ophthalmic ultrasound revealed choroidal thickness and several foci of exudative retinal detachments (Figure 5).

The patient's past medical history remarkable for vitiligo 39 years ago. Abdominal tuberculosis presented as ulcerative colon which was diagnosed by biopsies 39 years ago and treated very well. In 2013, 4 years ago the patient had a hypersplenism. As a reason he had a splenectomy, which later led to diagnosis of polycythaemia vera with positive JAC2 J617F mutation by bone marrow biopsy. The disease was treated with Hydroxyurea 500mg one to 4 tablets a day for 4 years, adjusted monthly by haematologist. There was no history of

tinnitus, headache, neck stiffness, alopecia, trauma or previous ocular surgery. The purified protein derivative (PPD) test was negative. Serological autoimmune diseases tests: ESR, ANA, RA was negative too. MRI of brain, thoracic and abdomen was normal and neurological exam was within normal limits. Complete blood count (CBC) was within normal limits by monthly supervision of the haematologist and tight control with Hydroxyurea, WBC 7,700 cells/mcL, Hemoglobin 13.5grams/dL, platelet 256,000/mcL. We prescribed Corticosteroids (prednisolone 80 mg/day) and tapered on 5 months, after two weeks the patient's symptoms improved significantly, focal retinal detachments started to fade and the visual acuity improved. We followed up the patient for one year by tapering corticosteroid slowly. The eye exam after one year revealed visual acuity of 20/20 bilaterally, fundus in RE showed some choroidal atrophy (Figures 6&7) and the symptoms didn't reoccur after stopping the corticosteroids.

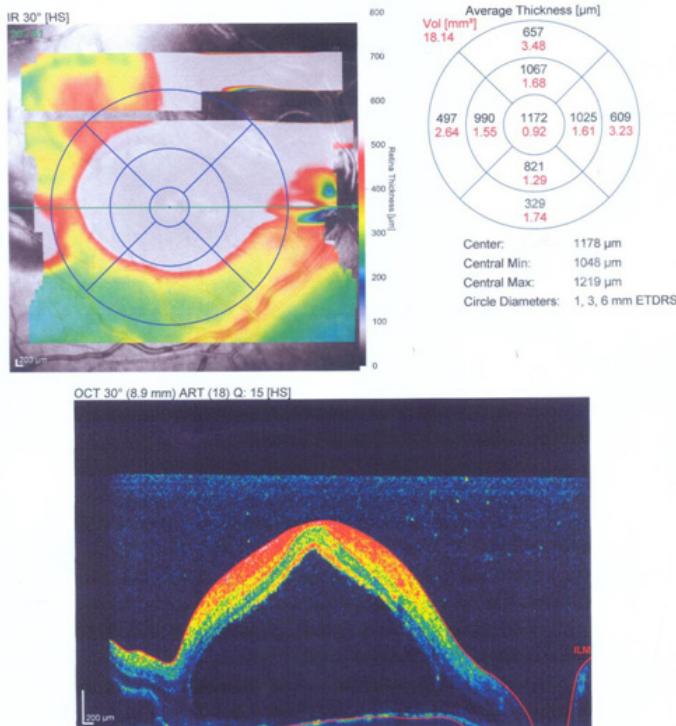


Figure 1 Optical coherence tomography (OCT) image of the right eye showing serous retinal detachment at the macular area.

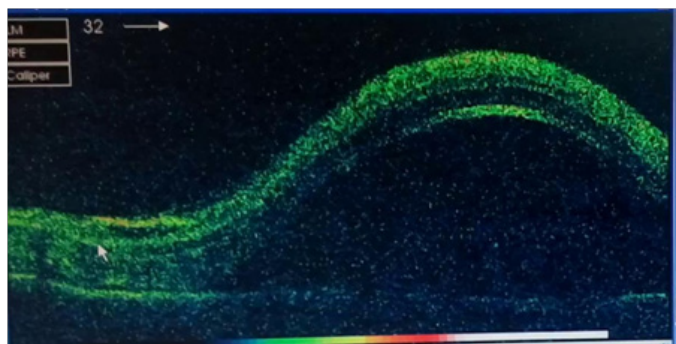


Figure 2 Optical coherence tomography (OCT) image of the right eye showing serous retinal detachment at the macular area.

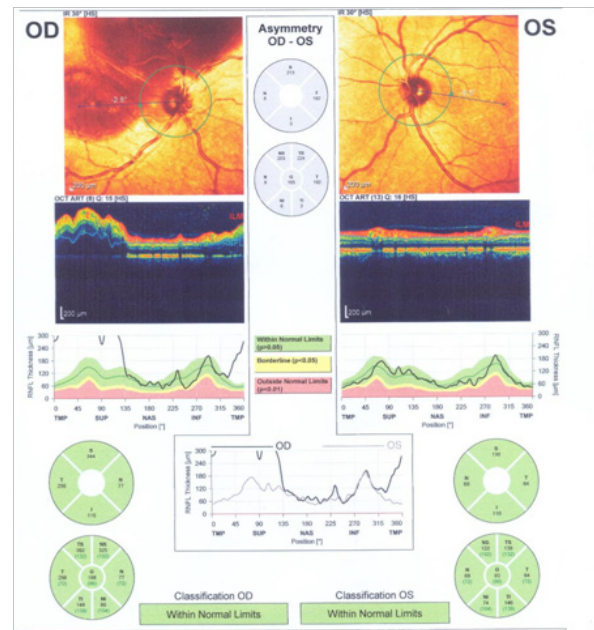


Figure 3 OCT of the optic disc showing serous retinal detachments in temporal and superior area of the optic nerve.

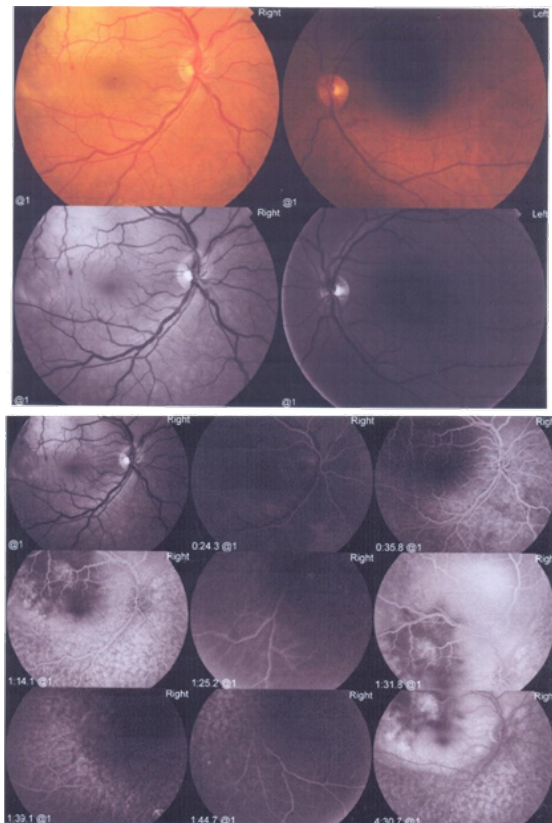
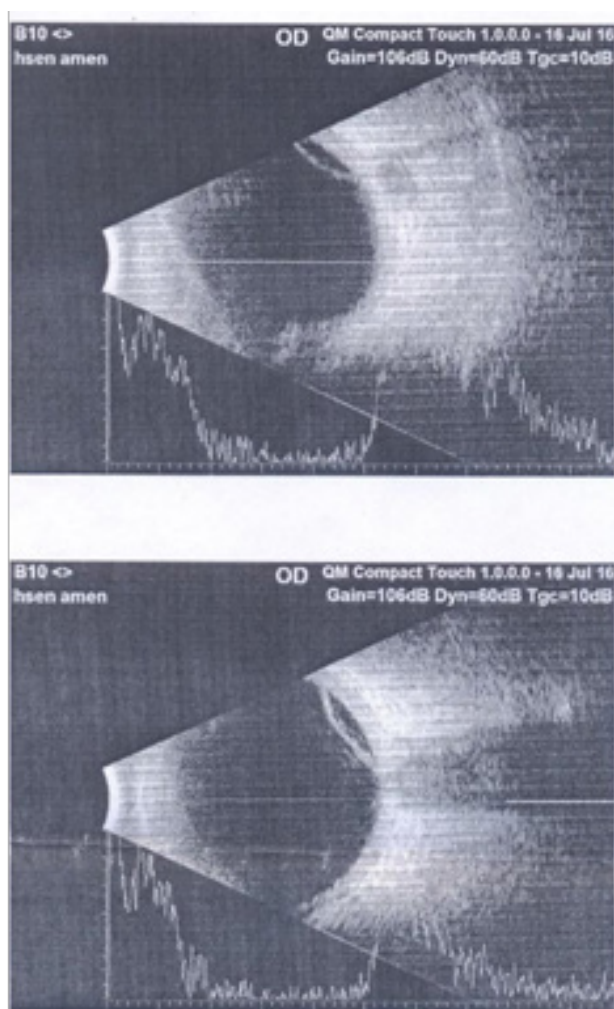


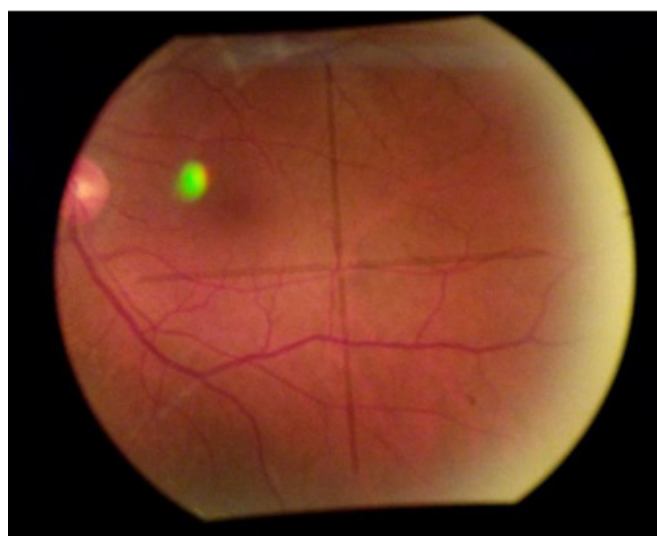
Figure 4 Multifocal blocking of background fluorescence from the choroid, serous detachments with late pooling of dye, focal areas of delayed choroid perfusion and multifocal pinpoint hyper fluorescent areas of leakages with late pooling and leakage of dye in the sub retinal foci throughout the posterior pole.



**Figure 5** Ophthalmic ultrasound showing choroid thickness and several foci of exudative retinal detachments.



**Figure 6** Right eye some choroid atrophy.



**Figure 7** left eye normal fundus.

## Discussion

Ocular presentations of Polycythemia vera are usually complications of hyperviscosity which cause transient ocular blindness.<sup>3</sup> There are several case-reports of ocular signs of PV as an initial sign of the disease or in the course of disease progression of those include central retinal artery occlusion<sup>4-6</sup> isolated cilioretinal artery occlusion,<sup>7</sup> multiple cotton wool spots with delayed arterial and venous circulation time,<sup>8</sup> peripheral retinal neovascularization,<sup>9</sup> bilateral papilledema<sup>10</sup> bilateral anterior ischemic optic neuropathy,<sup>11</sup> Subarachnoid hemorrhage and diplopia,<sup>12</sup> and preretinal and intraretinal hemorrhages in longstanding polycythemia vera as associated with anemia<sup>13</sup> Differential diagnosis of multiple serous retinal detachment is broad it is include Vogt-Koyanagi-Harada disease (VKH), sympathetic ophthalmia, posterior scleritis, choroidal inflammatory diseases, malignant hypertension, disseminated intravascular coagulopathy, age-related macular degeneration and Coats' disease, uveal effusion syndrome and posterior scleritis and giant cell arteritis.<sup>1</sup>

Our first impression was an ocular Vogt-Koyanagi-Harada disease (VKH) depending on clinical examination finding which was typical for VKH. The OCT finding was large areas of serous retinal detachments (Figure 1&3). While Fluorescein angiography (FA) photos show early hypofluorescence by blockage in the site of lesions, followed by a late hyperfluorescence. Currently the most important part in diagnosis of acute VKH is presence of exudative retinal detachment.<sup>14</sup> The skin involvements in VKH occur after ocular manifestations. Skin involvement usually include vitiligo, poliosis and alopecia. Our patient had vitiligo 39 years ago, so these part of VKH criteria is not present in our patient. On the other hand, vitiligo patient has an increased risk of comorbid autoimmune diseases.<sup>15</sup> The VKH is a bilateral disease, However, there were several reports of unilateral eye involvement of VKH with systemic symptoms that developed after several years.<sup>16</sup> Incomplete VKH disease was reported in a case of chronic myeloid leukemia in remission phase which made VKH more likely than eye involvement by leukemia itself.<sup>17</sup>

Serous macular detachment can be the first sign of some haematological diseases such as acute lymphoblastic leukemia.<sup>18–20</sup> Patient's PV was controlled very well and his complete blood count (CBC) was within normal limits at the presentation which is made it hard to draw a conclusion of a relationship between the PV and ocular signs. The previous TB doesn't have an association with this ocular finding as the PPD test was negative and the disease was treated successfully. The PV could have a role in serous retinal detachment which needs to be proven in large scale number of cases.

## Conclusion

Ocular sign of polycythemia vera are few but important to be aware of. Unilateral multiple serous retinal detachments that mimic unilateral VKH disease has been described in some case reports without giving it a separate entity and without linking it to suspect systematic disease. More observations should be done on both VKH disease and its causes and polycythemia vera and its ocular manifestations.

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None.

## Conflicts interest

The authors declare that they have no competing interests.

## Consent to publish

Written informed consent was taken from the patient to publish this case report and related photographic evidence.

## Ethics approval and consent to participate

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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