# Central retinal arterial occlusion in a patient with pyoderma gangrenosum

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A 74-year-old male presented to us with a history of vision loss for 36 hours in the right eye (RE). The RE had a visual acuity of hand movements. The fundus revealed a pale retina, cattle tracking in the retinal vessels, and a cherry-red spot at the macula. The patient was a known case of pyoderma gangrenosum (PG) and had received intravenous methylprednisolone and cyclophosphamide at the onset of visual symptoms. An emergency anterior chamber paracentesis was performed following unsuccessful attempts of ocular massage. The patient improved to 6/9 in the RE 4 months after paracentesis. The patient had an aggressive course of PG, for which he needed a combination of oral steroid, immunomodulator therapy and biologicals. An association between central retinal arterial occlusion and PG has not been reported before, according to the best of authors' knowledge.

**Key words:** Anti-phospholipid antibody, central retinal arterial occlusion, cherry-red spot, ischemic tolerance of retina, paracentesis

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Central retinal arterial occlusion (CRAO) is an ophthalmic emergency and early treatment is crucial. Arterial occlusions have been reported in association with atherosclerosis, hypercoagulable states, and various inflammatory disorders including systemic lupus erythematosus, polyarteritis nodosa, Behçet's disease, Wegener's granulomatosis, Crohn disease, and Churg–Strauss syndrome. We report a patient with pyoderma gangrenosum (PG) who developed CRAO and underwent anterior chamber (AC) paracentesis after 36 hours of symptom onset.

## Case Report

A 74-year-old male presented to us with a history of sudden onset vision loss for 36 hours in the right eye (RE). The best-corrected visual acuity (BCVA) was hand movements with accurate projection of rays in the RE and 6/6p in the left eye (LE). The RE showed relative afferent pupillary defect. Intraocular pressure was 14 mmHg in the RE and 12 mmHg in the LE. Both eyes had posterior-chamber intraocular lens. There were no cells in the AC of either eye. RE had a pale retina, cattle tracking in the retinal vessels, and a cherry-red spot at the macula [Fig. 1a]. LE showed few small drusen. Spectral-domain optical coherence tomography (SDOCT, RTvue, Optovue Inc., California, USA) revealed an increased reflectivity of the inner retina [Fig. 2a]. The patient was a known case of PG for 1 year. He was given intravenous methylprednisolone (IVMP, 125 mg) and cyclophosphamide 500 mg at the onset of the visual decline by the rheumatologist, considering it a manifestation of inflammatory phenomenon in PG. The patient had a very aggressive/uncontrolled course of systemic disease and required multiple hospital admissions due to the same. Two weeks before the onset of visual symptoms, he had received IVMP 500 mg and cyclophosphamide 500 mg for PG. A clinical diagnosis of bilateral pseudophakia and CRAO in the RE was made. An emergency AC paracentesis was performed following unsuccessful attempts of ocular massage. On the 1st day after paracentesis, the BCVA of RE was 6/36. The superotemporal cattle-track appearance had disappeared [Fig. 1b]. The fundus fluorescein angiogram did not show any obvious cilioretinal artery. In the RE, the inferotemporal vessels did not fill up even at 8 min [Fig. 1c-e]. Echocardiography did not detect any clots or vegetation. The carotid Doppler demonstrated eccentric calcific plaque in the right internal carotid artery with <30% stenosis. Electrocardiogram was unremarkable. Hemoglobin was 11.8 gm/dl and total leukocyte count was 10,700/mm$^3$. Erythrocyte sedimentation rate (ESR) was 33 mm in the 1st h and C-reactive protein (CRP) was 21.9 mg/dl. There was no clinical evidence of giant cell arteritis. Blood pressure was 130/86 mmHg and the fasting blood sugar was 101 mg/dl. The ESR and CRP of the patient remained high in follow-up at 1 month (82 mm in the 1st h, 113.2 mmHg), 2nd month (101 mm in the 1st h, 63.3 mg/dl), and at 7th month (63 mm in the 1st h, 29 mg/dl). One month after the onset of CRAO, the skin lesions increased and the appetite of the patient reduced with increased inflammatory markers (ESR and CRP). In the course of disease, the patient showed good response to steroids, partial-to-good response to antitumor necrosis factor agents, poor response to mycophenolate mofetil, and combination of cyclophosphamide and steroid was considered to be less useful. Good response to cyclosporine was noted but it had

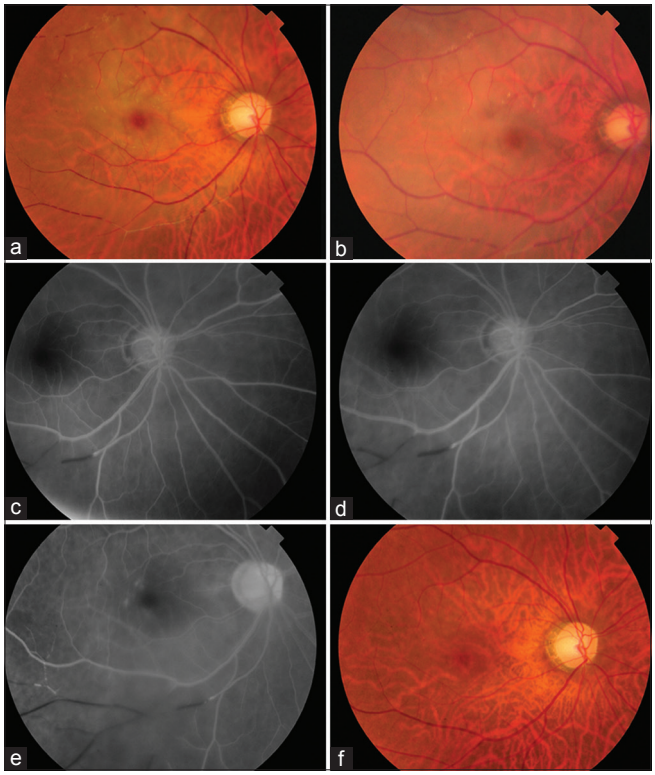


**Figure 1:** (a) At presentation, there was whitened retina, cherry-red spot at macula, and cattle tracking of blood columns. (b) One day after paracentesis, some apparent clearing of retinal pallor nasal to the fovea was noted. Superotemporal cattle-tack appearance had disappeared. (c-e) The fluorescein angiogram at 51 s, 61 s, and 8 min 21 s showed no filling of the inferotemporal retinal vessels. (f) At 4th month, mild optic disc pallor was noted.

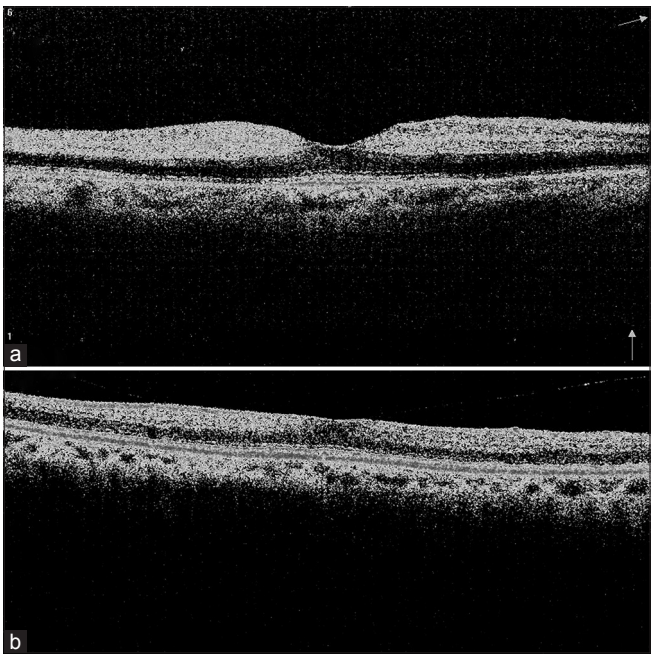


**Figure 2:** (a) The optical coherence tomography at presentation showed hyperreflectivity of inner retinal layers. (b) At the 4th month, the optical coherence tomography showed loss of foveal depression and retinal thinning.

to be stopped due to acute kidney injury. He subsequently received etanercept with some response.

The BCVA in the RE improved to 6/18 at 1 month and 6/9 at 4 months. At 4 months, mild pallor of optic disc and mild pigmentary changes at the fovea were noted [Fig. 1f]. The SDOCT revealed loss of foveal depression and retinal thinning in the RE [Fig. 2b]. At the last follow-up at 10th month, the patient was receiving oral prednisolone, methotrexate, and IV infliximab for PG.

## Discussion

PG is a rare inflammatory disease of skin belonging to a spectrum of neutrophilic dermatoses which also include Behçet's disease. Leukocytoclastic vasculitis is seen in about 40%[1] cases with PG. According to the best of authors' knowledge, there is no prior report of an association of CRAO with PG. PG has been reported to be associated with inferior vena caval thrombosis in antiphospholipid syndrome,[2] portal vein thrombosis in chronic myelomonocytic leukemia,[3] and aortoiliac embolism[4] in a patient with acne conglobata and myocardial infarction. A case of superficial venous thrombosis in pregnancy has been reported which ultimately resulted in PG lesions due to Koebner phenomena.[5] Chronic inflammatory diseases may predispose to a hypercoagulable state due to upregulation of procoagulant factors and reduced anticoagulant mechanisms.[2] Furthermore, inflammation of vessels may cause CRAO, as in giant cell arteritis. In our patient, there was no evidence of active retinal arterial inflammation, leukemia, venous thrombosis, or acne conglobata, and the pathogenesis appeared to be related to arterial thrombosis. A limitation of this report is that antiphospholipid antibody or anticardiolipin antibody was not tested.

We, however, agree that a single case report does not necessarily prove a cause-effect relation of the diseases and may even be coincidental. It has been shown by Hayreh and Zimmerman that "massive irreversible retinal damage occurs in CRAO lasting about 240 min."[6] However, spontaneous reperfusion of both the central retinal artery and lateral posterior ciliary artery can happen as late as 4 days after documented ischemia with a final BCVA of 20/30.[7] This is because, unlike the experimental models, CRAO in human may not be complete. Furthermore, there may be some ischemic tolerance of the retina mediated by the upregulation of neuroprotective agents and antiapoptotic factors due to previous brief episodes of retinal ischemia.[8] Our patient presented >24 h after the onset of sudden visual loss. We decided not to give up and to give a trial of AC paracentesis. Good final visual acuity (≥6/9) could be achieved in this case, which can be noted in only 12.5% of all eyes with CRAO.[7] However, a large series evaluating 74 patients concluded that paracentesis did not provide additional visual gain.[9] In our case, anatomical and visual recovery on the 1st day after paracentesis may point toward its beneficial role. However, the visual recovery, in this case, could also have been related to the control of inflammatory process with steroids and various immunomodulator therapy/biologicals or the natural history of CRAO[10] rather than AC paracentesis. High ESR and CRP at presentation may denote an underlying activity of PG which may have predisposed to CRAO.

## Conclusion

In conclusion, we report a rare case of PG who developed CRAO and achieved good final BCVA after paracentesis at 36 hour of onset of symptoms and management with systemic steroids and immunomodulator/biological therapy.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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