Isolated Bilateral Sub-Internal Limiting Membrane Haemorrhages at The Macula as A Presentation of Dimorphic Anaemia with Pancytopenia

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Abstract

A 33-year-old male presented with diminution of vision in both his eyes succeeding an episode of bleeding per-rectum due to haemorrhoids and ensuing dimorphic anaemia with pancytopenia for which he had received multiple blood transfusions. Clinical examination revealed bilateral isolated haemorrhages at the fovea which was localised to sub-internal limiting membrane plane on optical coherence tomography. This case-report emphasizes the varied clinical spectrum of anaemic retinopathy and the need of an acute fall in haemoglobin levels for anaemic retinopathy to occur. Investigating for vitamin B12 deficiency and pancytopenia is also warranted in the setting of anaemic retinopathy.

Key-words: Anaemic retinopathy, Dimorphic anaemia, Megaloblastic anaemia, Pancytopenia, Bilateral macular haemorrhages, Sub-internal limiting membrane haemorrhage.

KEY MESSAGES

- Isolated macular sub-internal limiting membrane hemorrhage is one of the rarer presenting sign of anaemic retinopathy.
- Acute reduction of hemoglobin levels usually precedes clinical manifestation of anaemic retinopathy.
- In a patient with anaemic retinopathy , it is important to screen for vitamin B12 deficiency and co-existent pancytopenia.

BACKGROUND AND INTRODUCTION

Anaemia occurs due to the reduction of the levels red blood cells (RBCs) or haemoglobin. Ineffective erythropoiesis, increased blood loss and haemolysis are the key mechanisms leading to anaemia. Anaemic retinopathy encompasses a wide spectrum of clinical signs which includes retinal haemorrhages at multiple anatomical levels, Roth spots, tortuous vessels, cotton-wool spots, hard exudates, and optic neuropathy.[1]

We report a case of dimorphic anaemia with severe pancytopenia which presented with bilateral isolated macular haemorrhages as the only ocular presentation of anaemic retinopathy which is very rare and to best of our knowledge has not been reported earlier.

CASE HISTORY

A 33-year-old male patient presented with acute onset painless non-progressive diminution of vision in both his eyes (BE) for five days. Past history includes history of haemorrhoids associated with repeated blood transfusions including recent transfusion a month back.

Patient complained of vision loss after the acute bleeding episode. Systemic symptoms include the complaints of dyspnoea and weakness for the past few months.

General examination revealed the presence of significant pallor. Ocular examination revealed best corrected visual acuity (BCVA) 20/20, N6 in both eyes (BE). Anterior segment examination showed the presence of conjunctival pallor in BE while rest of the findings including pupillary examination were within normal

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limits. Fundus examination revealed the presence dark-red well-defined hemorrhages at the fovea in BE. There was no evidence of vascular changes or hemorrhages elsewhere in both fundi (Figure 1a-d).

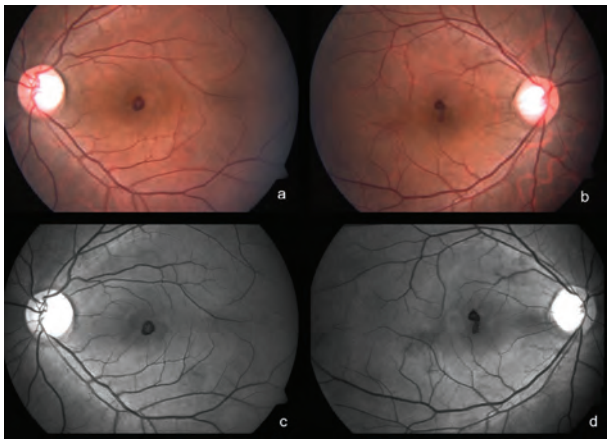

Figure 1: Fundus photography in anaemic retinopathy Colour fundus photographs (CFP - a, b) and red-free images (c, d) depicting macular haemorrhages in both eyes. Note the absence of other clinical signs of anaemic retinopathy

Optical coherence tomography (OCT) revealed the presence of hyper-reflective lesions (figure 2a,b – white arrows) at the fovea in the sub-internal limiting membrane (ILM) plane in BE . These foci exhibited back-shadowing suggestive of hemorrhage and the integrity of the retina layers were maintained (Figure 2 a, b). Laboratory investigations during the acute episode, one month prior to presentation, revealed Hb-3.3gm% along with pancytopenia, reduced serum vitamin B12 and ferritin levels.

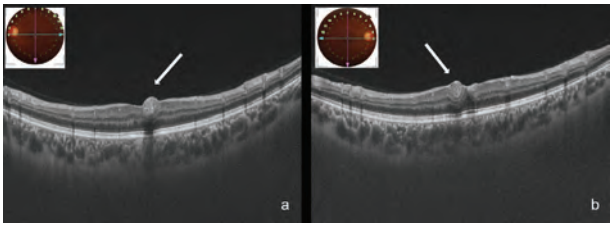

Figure 2: Optical coherence tomography (OCT) in anaemic retinopathy. Optical coherence tomography (OCT)vertical scan- demonstrating the presence1e of hyper-reflective lesions (white arrows in a,b) in the sub-internal limiting membrane(ILM) plane representing macular hemorrhages. Note the variegated reflectivity within the lesions , the well-preserved retinal integrity and back shadowing associated with the lesions in both eyes.

Peripheral smear demonstrated pancytopenia, dimorphic anaemia with few hypersegmented nuclei and toxic granules. Bone marrow biopsy was suggestive of erythroid hyperplasia with dyserythropoetic changes and features suggestive of megaloblastic anaemia. Patient received five units of blood transfusion along with iron, folic acid and methylcobalamine supplements and is currently under follow-up with a haematologist and a general surgeon for his haemorrhoids and is scheduled for surgery. Considering his current Hb levels (9.8gm%) and the nature of haemorrhages secondary to acute anaemia, the patient was counselled regarding his condition and the need for anaemia control. Observation and review after one month were advised.

DISCUSSION

Anaemia , with (38%) or without thrombocytopenia (28%), is implicated as the cause of a wide plethora of clinical findings grouped together as anaemic retinopathy .As the severity of anaemia increases and more acute the onset, the risk of retinopathy also increases, especially when the hemoglobin (Hb) level is below 6 gm/dL.[1]

The major pathological changes are as follows:

1. Anaemia causes retinal hypoxia, which causes infarction of the nerve fiber layer leading to cotton wool spots.
2. Hypoxia also leads to vascular dilatation, increased transmural pressure owing to hypoproteinaemia and microtraumas to the vessel walls, which cause retinal oedema and haemorrhages.
3. Thrombocytopenia, when associated with anaemia, leads to defective coagulation and haemorrhages.[2,3] The severity of retinopathy is directly related to the severity of associated thrombocytopenia in these cases as evidenced by rapid resolution of retinopathic changes with normalisation of platelet count with initiation of treatment even when the red cell population has not normalised completely.[4]

Vitamin B12 deficiency can lead to pancytopenia, when severe, due to defect in the DNA synthesis. Megaloblastic anaemia usually presents with flame shaped haemorrhages, cotton-wool spots, Roth spots and hard exudates. In a study conducted by Satish et al, they reported flame shaped haemorrhages (37.50%) as the most common form followed by deep haemorrhages (31.25%), Roth spots (18.75%) and sub-hyaloid haemorrhage (8.33%) in patients (n=48) with iron deficiency anaemia and megaloblastic anaemia. Fundal pallor was the second most common retinal manifestation seen in 15 out of 48 patients (31.25%) with anaemia.[5] The occurrence of iron deficiency anaemia along with megaloblastic anaemia

(dimorphic) with thrombocytopenia in our patient was further worsened by repeated blood loss due to haemorrhoids.

Bilaterally symmetrical macular sub-ILM haemorrhages are less frequently encountered in dimorphic anaemia with thrombocytopenia. Similar clinical presentation was reported in a case of megaloblastic anaemia without thrombocytopenia.[6] Bilateral macular haemorrhages are quite uncommon with a few reports associated with cases of head trauma, Valsalva manoeuvre, diabetic retinopathy, retinal branch vein occlusion, ruptured macro-aneurism of the retina, post refractive surgery, drug induced anaemia and leukemia.[7]

Management of anaemic retinopathy is mainly aimed at reassuring the patient and careful observation for worsening/improvement as the condition resolves spontaneously with the correction of anaemia and treatment of implicated aetiology. Small haemorrhages usually respond well to blood transfusions while larger haemorrhages threatening vision may require a Nd:YAG hyaloidotomy or pars plana vitrectomy since it can lead to permanent visual impairment due to pigmentary macular changes and formation of epimacular membranes.[8]

CONCLUSION

Anaemic retinopathy may be the presenting symptom of anaemia and hence needs thorough systemic evaluation which includes a hemogram and peripheral smear. The presence of bleeding at various levels of the retina coupled with a relevant systemic history helps us arrive at an early diagnosis. The occurrence of anaemic retinopathy indicates the acute nature of the implicated aetiology of anaemia. Bilateral macular sub-ILM haemorrhages are one of the rarer clinical manifestations of anaemic retinopathy and need a higher-index of suspicion to diagnose and manage efficiently.

LEARNING POINTS

1. Anaemic retinopathy is fairly common, especially in developing countries like India.
2. Anaemia and thrombocytopaenia in the setting of dimorphic anaemia can be potentially blinding.
3. As the severity of anemia increases, the risk of retinopathy increases, especially when platelet count is low.
4. Bilateral macular sub-ILM haemorrhages are one of the rarer clinical manifestations of anaemic retinopathy and need a higher-index of suspicion to diagnose and manage efficiently.

CONFLICTS OF INTEREST

There are no conflicts of interest.

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None

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