Retinopathy in severe aplastic anaemia

Harathy Selvan , Shorya Vardhan Azad, Priyanka Ramesh, Kusumitha Bhakthaganes

Ophthalmology, Dr Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi, India

Correspondence to
Dr Harathy Selvan;
sharathy2012@gmail.com

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DESCRIPTION

A 19-year-old girl presented to the emergency services with complaints of haematuria, gum bleeding and sudden visual loss. General examination revealed significant pallor and multiple petechiae all over the body. Her heart rate was 98 beats/min, blood pressure 102/58 mm Hg, respiratory rate 18 breaths/min and SpO₂ 99%. The best corrected visual acuity was counting fingers at 2 m in the right eye and at 1 m in left eye. The anterior segment of both eyes (BE) was within normal limits. On dilated fundus examination, BE showed extensive preretinal, intraretinal and subretinal haemorrhages, numerous Roth spots, hard exudates and masses of organising haemorrhages (figure 1). Macular optical coherence tomography showed BE subinternal limiting membrane bleeds, intraretinal oedema and neurosensory detachments (figure 2).

On investigating, her haemoglobin (Hb) was 28 g/L, haematocrit 8.2%, platelet count 1000/mm³ and leucocyte count 2.2×10^9 /L. She was referred to a haematologist for detailed evaluation and advised to refrain from eye rubbing, Valsalva manoeuvres and vigorous activities. Bone marrow examination revealed pancytopenia with a hypocellular marrow. A diagnosis of aplastic anaemia with anaemic retinopathy was made and treated with packed red blood cells and platelet rich plasma.

Anaemic retinopathy is fairly common, especially when the Hb levels are <80 g/L.¹ In the presence of anaemia and/or thrombocytopenia, the prevalence of retinopathy was found to be 28.3%, which further rose to 38% when both the conditions coexisted.² Nevertheless, in view of its subtle nature not affecting the visual acuity, it frequently remains undetected or under diagnosed. Ironically, anaemia and thrombocytopenia in the setting of aplastic anaemia can be potentially blinding. Retinopathy was observed in 69% of aplastic anaemia patients who showed an

Figure 2 Macular optical coherence tomography images. (A) Right eye and (B) left eye, showing subinternal limiting membrane bleeds, hyper-reflective inner retinal layers, intraretinal cystic spaces and neurosensory detachments (white *). Dense haemorrhage (white arrow) blocking the visibility of underlying structures in the left eye.

Learning points

- ▶ Anaemic retinopathy is fairly common, especially when the haemoglobin levels are <8 g%.
- ▶ Anaemia and thrombocytopenia in the setting of aplastic anaemia can be potentially blinding.
- ▶ Blood turbulence, endothelial injury and bleeding diathesis have been shown to contribute to the occurrence of haemorrhages in aplastic anaemia.

Hb <80 g/L and platelet counts of $<50 \times 10^9$ /L.³ They may present with retinal and vitreous haemorrhages, cotton wool spots, central retinal venous occlusion-like, optic disc oedema, macular oedema, peripheral retinal vasculopathy, orbital and lid hematomas.⁴⁻⁶ Blood turbulence, endothelial injury and bleeding diathesis have been shown to contribute to the occurrence of these haemorrhages.⁴ Retinopathy presenting as severe as in this case with sudden loss of vision is a rarity.

Management of anaemic retinopathy involves reassurance and careful observation, as the condition is bound to improve on treatment of the underlying aetiology and correction of anaemia.⁷ Small haemorrhages usually respond to blood transfusions while large haemorrhages threatening vision may require a hyaloidotomy or pars plana vitrectomy.⁴ Such severe presentations though rare, are important to be known, for prompt management to avoid permanent visual loss.

Contributors HS and SVA evaluated and managed the patient. PR and KB collected the data. All four authors were involved in the preparation and critical review of the manuscript.

Figure 1 Montage fundus images. (A) Right eye and (B) left eye, showing extensive preretinal, intraretinal and subretinal haemorrhages, numerous Roth spots (white arrows), neurosensory detachments (black *) and masses of organising haemorrhages (black arrows).

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ORCID iD

Harathy Selvan <http://orcid.org/0000-0003-4403-8767>

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