

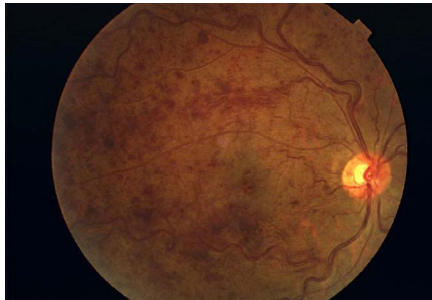
Intermittently blotchy

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Clinical context

A 40-year-old man with a history of fluctuating vision in his right eye for 1 year reported several blurring episodes lasting 1 hour, which resolved completely spontaneously between episodes. Visual acuity was 6/18, 6/12, N8 with no relative afferent pupillary defect. Intraocular pressure was normal.



Question 1

What are the signs shown in the photograph?

Question 2

What is your working diagnosis?

Question 3

Given that he has no other medical illness with normal systemic screen, and fundus fluorescein angiography (FFA) shows leakage of the peripheral veins and optic disc, what diagnosis would you most likely consider?

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Answer 1

Tortuous dilated superotemporal and inferotemporal vein, dot and blot, and flame-shaped haemorrhages in all four quadrants, hyperaemic optic disc, and dull macula.

Answer 2

Right central retinal vein occlusion (non-ischaemic) with macular oedema.

Answer 3

The feature of fluctuating vision, which returns to normal between episodes is the key history suggestive of papillophlebitis right (PP) in this patient. Another important observation is the lack of concordance between the good visual acuity and severity of retinal findings. Other features which point towards this diagnosis include the young age of the patient, unilateral presentation with disc hyperaemia and retinal venous engorgement, negative systemic screening, and FFA findings of optic disc and vein leakage.¹⁻³ PP is a rare form of partial central retinal vein occlusion (CRVO) affecting previously healthy women. It was first described by Lonn and Hoyt in 1966.² PP could be a form of inflammatory CRVO with more favourable outcomes. PP is postulated to be caused by inflammation of the optic disc, leading to central retinal vein compression and venous stasis. Although corticosteroids are the mainstay of treatment for PP, there is a lack of clear evidence to guide therapy. Some authors suggest combining it with anticoagulant therapy.^{3,4} Treatment is needed to prevent progression to frank CRVO with its complications. In this patient, oral ciclosporin A 5 mg/kg/day or 300 mg per day was prescribed together with 40 mg oral prednisolone od for 1 week and then tailed at 5 mg/week. He gave a history of previous prednisolone therapy that required levels of 30 mg daily to prevent relapse. His visual acuity improved to 6/12, 6/9, N5 after 1 week of therapy.

References

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