Malaysian Journal of Ophthalmology

© Malaysian Journal of Ophthalmology 2020; 4:289-294
Case report/Case series

Leukostatic retinopathy: a sight-threatening complication of chronic myeloid leukaemia with severe hyperleukocytosis

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Abstract

Retinopathy secondary to chronic myeloid leukemia (CML) commonly manifests as venous dilation and tortuosity, retinal hemorrhages, microaneurysms, and cotton-wool spots which are similar to features of non-proliferative diabetic retinopathy or hypertensive retinopathy. However, massive vitreous hemorrhage is rarely encountered, especially among those treated with chemotherapy. We report a case of a young CML patient in accelerated phase, presenting with bilateral painless sudden visual loss. Fundus examination showed bilateral dense vitreous hemorrhage. Laboratory results showed thrombocytopenia with a very low platelet count. Magnetic resonance imaging (MRI) of the brain and orbit showed subacute intraparenchymal hemorrhages and bilateral intraocular hemorrhages. We performed pars plana vitrectomy (PPV) and endolaser on the left eye, which had more extensive vitreous hemorrhage. At one-week follow-up, the patient unfortunately developed a retinal detachment. The patient underwent a second PPV with endolaser and insertion of silicone oil. Despite the prompt surgical intervention, the patient developed an ischemic retina resulting in poor visual prognosis. One month later, we performed PPV and endolaser on the right eye. Postoperatively, her vision improved significantly from hand movement to pinhole vision 6/45. Dense vitreous hemorrhage is a rare complication of childhood leukemia. General physicians should refer leukemic patients for ophthalmic evaluation. Awareness of potentially blinding complications of CML and prompt referral upon

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diagnosis is warranted for early detection and treatment. Reduced awareness of this potentially blinding complication may result in poor visual outcome.

Keywords: chronic myeloid leukemia (CML), pars plana vitrectomy (PPV), vitreous haemorrhage

Retinopati leukostatik: komplikasi ancam penglihatan disebabkan oleh leukemia myeloid kronik bererta hiperukositosis yang teruk

Abstrak

Retinopati sekunder untuk leukemia myeloid kronik (CML) biasanya ditunjukkan sebagai pengembangan salur vena dan tortuositi, pendarahan retina, mikroaneurisma, dan bintik-bintik kapas yang serupa dengan ciri-ciri retinopati kencing manis yang tidak proliferatif atau retinopati hipertensi. Walau bagaimanapun, pendarahan vitreous secara besar-besaran jarang ditemui, terutama di kalangan mereka yang dirawat dengan kemoterapi. Kami melaporkan kes seorang pesakit CML yang muda dalam fasa aselerasi, dengan menyaksikan kehilangan visual secara tiba-tiba tanpa rasa sakit. Peperiksaan Fundus menunjukkan pendarahan berlaku dalm kedua-dua mata. Hasil makmal menunjukkan trombositopenia dengan kiraan platelet yang sangat rendah. Pencitraan resonans magnetik (MRI) otak dan orbit menunjukkan pendarahan intraparenchymal subacute dan pendarahan intraocular bilateral. Kami melakukan vitreectomy pars plana (PPV) dan endolaser pada mata kiri, yang mempunyai pendarahan vitreus yang lebih banyak. Pada susulan satu minggu, pesakit itu malangnya mengalami lekang retina. Pesakit menjalani PPV kedua dengan endolaser dan memasukkan minyak silikon. Walaupun pembedahan dilakukan segera, pesakit mengalami pula retina iskemia yang mengakibatkan prognosis visual sangat rendah. Satu bulan kemudian, kami melakukan PPV dan endolaser di sebelah kanan mata pula. Selepas pembedahan, penglihatannya meningkat dengan ketara dari pergerakan tangan ke penglihatan pinhole 6/45. Pendarahan vitreous adalah komplikasi yang jarang berlaku pada psakit leukemia kanak-kanak.

Pakar perubatan am harus merujuk pesakit leukemia untuk penilaian oftalmik. Kesedaran tentang komplikasi CML yang berpotensi untuk meyebabkan kebutaan, dan rujukan segera adalah diperlukan setelah diagnosis untuk pengesahan awal dan rawatan. Kurangnya kesedaran tentang komplikasi yang berpotensi membutakan sebegini boleh menyebabkan hasil penglihatan yang rendah.

Kata kunci: leukemia myeloid kronik, pendarahan vitreous, vitreectomy pars plana

Introduction

Leukostatic retinopathy secondary to severe hyperleukocytosis is an uncommon complication of chronic myeloid leukemia (CML). It is a rare complication and only reported twice worldwide.[1,2] In this case report, we describe a case of CML with bilateral visual loss caused by retinal ischaemia from severe systemic leukostasis.

Case report

A 13-year-old Malay girl was diagnosed with chronic myeloid leukaemia (CML) by bone marrow aspiration and BCR-ABL1 fusion gene test. She presented with bilateral acute onset of visual loss when she was admitted into the paediatric ward for intravenous chemotherapy. At presentation, vision in both eyes was hand movement. Anterior segment examination was unremarkable and intraocular pressure (IOP) was 14 mmHg in each eye. There was no view of the optic disc and

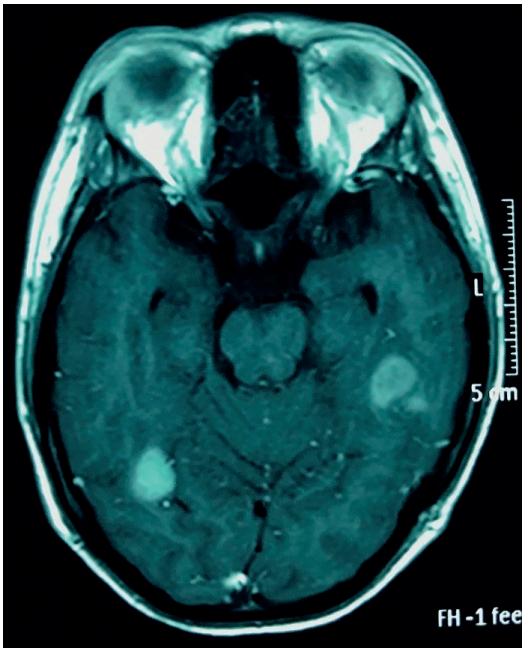

Fig. 1. Brain and orbit (axial) MRI. Intraparenchymal hemorrhages seen in the right posterior pons (pontomedullary junction), left temporal, bilateral occipital, and left thalamus (T1-weighted).

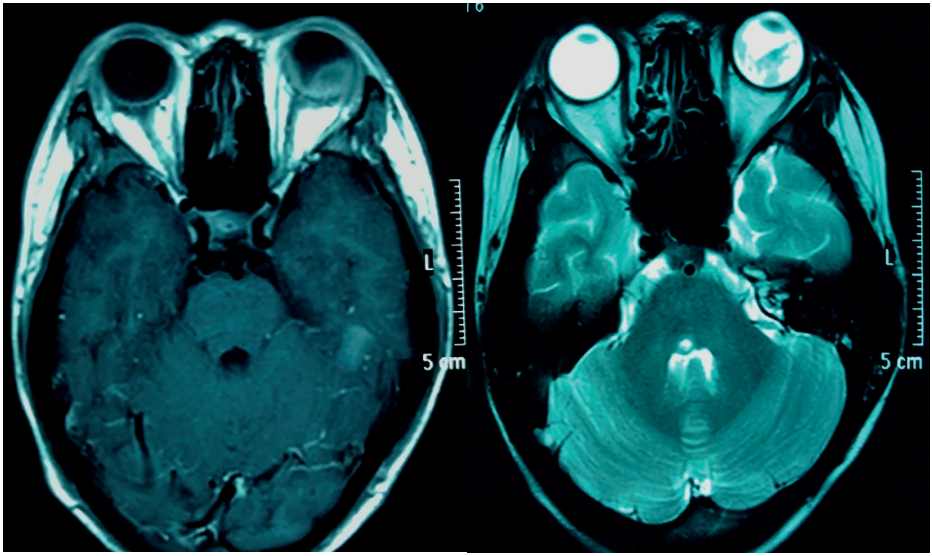

Fig. 2. Brain and orbit (axial) MRI. Heterogenous subretinal lesions seen at the posterior segment of the left eye, which are hyperintense in T1-weighted MRI image and hypointense in T2-weighted MRI image, suggestive of left eye subretinal haemorrhage.

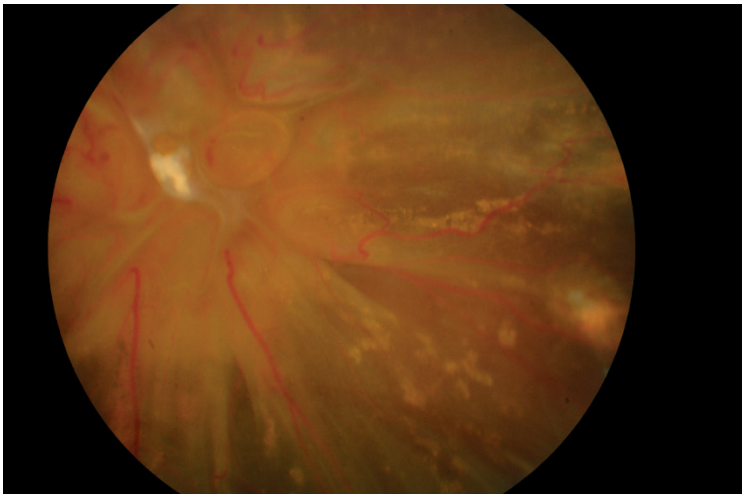

Fig. 3. Postoperative left-eye fundus photo. Retinal detachment with the retinal blood vessels appearing tortuous, dilated, and straightened due to contraction of the underlying proliferative vitreoretinopathy membrane.

macula on fundus examination. Ultrasound B scan was unable to be performed in the paediatric ward. However, magnetic resonance imaging (MRI) of the brain and orbit showed subacute intra-parenchymal white matter haemorrhages and left eye subretinal haemorrhages (Figs. 1 and 2). Laboratory results showed severe hyperleukocytosis, with total white blood cells of 678 × 10^9/L (normal range 4-10 × 10^9/L) and neutrophilia of 551 × 10^9/L (normal range: 2-7 × 10^9/L). The patient received ten cycles of intravenous chemotherapy with oral imatinib 400 mg OD and achieved haematological remission. Two months after imatinib treatment, there was persistent bilateral vitreous haemorrhage with hand movement vision. We performed *pars plana* vitrectomy (PPV) and endolaser on the left eye (worse eye). Intraoperative findings included vitreous haemorrhage, subhyaloid haemorrhage, intraretinal haemorrhage, and perivascular retinal infiltrates. Postoperatively, vision remained at hand movements. At one-week follow-up, the patient developed a retinal detachment in the operated eye (Fig. 3). The patient underwent a second PPV with endolaser and insertion of silicone oil. One month later, we performed PPV and endolaser on the right eye (better eye). Intraoperative findings were similar to the left eye. Postoperatively, her vision improved significantly from hand movement to pinhole vision of 6/45 in the right eye.

Discussion

Leukostasis is one of the fatal complications of CML, more often in the blastic phase. It is typically characterized by partial or total occlusion of systemic microcirculation by aggregation of leukemic cells and thrombi leading to respiratory or neurological symptoms. Leukostatic retinopathy is a rare ocular manifestation of CML and this term has only been recognized in recent years.[1,2] It has been reported that this ocular complication is related to local circulatory stasis, which leads to retinal ischemia and blindness.[1,2]

In the case of our patient, full blood count revealed she had hyperleukocytosis on presentation. She demonstrated ocular manifestations of retinal ischemia such as vitreous haemorrhage, subhyaloid haemorrhage, and significant tortuous veins with diffuse intraretinal haemorrhages. All of these ocular signs suggested that she might have severe retinal microvascular stasis, which led to retinal ischemia. Eventually, she ended up with poor vision after PPV. We hypothesise the mechanism of retinopathy in this case was likely caused by retinal ischemia secondary to leukostasis in CML. During follow-up in the eye clinic, optical coherence tomography of the left-eye macula was similar to chronic central retinal artery occlusion, manifested by diffuse thinning and disorganization of the inner retinal layer, which is consistent with retinal ischemia. One of the shortfalls was inability to perform fundus fluoroscein angiography in our patient due to her unfavourable systemic condition.

Performing PPV in a leukemic child is always challenging due to the high risk of retinal tears and incomplete posterior vitreous detachment. Apart from retinal tears and detachment, other rare surgical complications include epiretinal membrane formation, as well as cataract and macula hole formation. However, a recent report suggests that early vitrectomy facilitates rapid and optimal visual recovery, provided that the general condition of the child is good enough to undergo surgery.[3]

In conclusion, leukostatic retinopathy is a sight-threatening condition. General physicians should refer leukemic patients for ophthalmic evaluation early. Awareness of potentially blinding complications of CML and prompt referral upon diagnosis is warranted for early detection and treatment. Detection of early stage disease would enable vision preserving treatment to be commenced. Reduced awareness of this potentially blinding complication may result in a poor visual outcome.

Acknowledgements

We would like to thank the Director General of Health Malaysia for his permission to publish article.

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