CASE REPORT

Anterior chamber exudation in chronic myeloid leukaemia

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SUMMARY

Anterior chamber leukaemic hypopyon is a rare occurrence in chronic myeloid leukaemia. We discuss two cases marked by rapid exudation inside the anterior chamber, which were subsequently diagnosed as chronic myeloid leukaemia. The hypopyon in both the cases resolved on induction of chemotherapy.

BACKGROUND

Leukaemia is known to have varied ocular features, and may at times be associated with visual loss. Commonly, these features are observed in the posterior segment as a resultant of direct cellular invasion, immune suppression, non-perfusion, bleeding or toxicity of chemotherapy.¹ However, anterior chamber (AC) exudation with hypopyon is a less frequent manifestation of the disease, typically seen in relapsing cases of acute leukaemia, though it has been rarely described in chronic myeloid leukaemia (CML) as well.^{2–13} We discuss two such cases with posterior segment manifestations, complicated by a sudden occurrence of exudates in the AC appearing like a hypopyon, consequently diagnosed as CML.

CASE PRESENTATION**Case 1**

An 18-year-old young, healthy-looking man presented with sudden and painless loss of vision in the left eye since 2 weeks. There was no history of systemic illness. He was earlier diagnosed with Eales' retinal vasculitis elsewhere and had been worked up extensively for associations like tuberculosis, sarcoidosis and viral infections, all of which were found to be negative. There was no abnormality detected on physical examination. Best corrected visual acuity (BCVA) was 6/6 in the right eye (OD) and 6/60 in the left eye (OS). The anterior segment of either eye was unremarkable while intraocular pressures (IOPs) were within normal limits. The fundus examination of either eye showed clear media with a normal optic disc, 360° perivascular cuffing and exudation (vein > artery; OS > OD) (figure 1). Multiple pre-retinal bleeds with few pale centred haemorrhages suggestive of Roth spots and macular oedema were also noted (figure 1). Fundus fluorescein angiography revealed normal macular perfusion with blocked fluorescence corresponding to the haemorrhages along with paravascular leakage.

Considering the common differential diagnosis of Roth spots, a complete hemogram with peripheral blood smear, antinuclear antibody and

echocardiogram was ordered. To our surprise, the patient came back the next day with complete loss of vision in the left eye. On examination, total hyphaema along with streaks of white exudative material over it were noted (figure 2). The exudates appeared like a hypopyon (1.5 mm) superior to the blood level. On examination, OD was the same as before. The IOP was 20 mm Hg in OS. Sonography of OS revealed an anechoic vitreous cavity.

Case 2

A 28-year-old apparently healthy man presented to us with sudden painless loss of vision in both the eyes since 1 month. BCVA was hand motions close to face OU. The anterior segment was within normal limits in both eyes. On fundus examination, he was found to have vitreous haemorrhage in both eyes. Comprehensive systemic workup was performed for pulmonary tuberculosis, sarcoidosis, viral infections including HIV and hepatitis infections, diabetes mellitus, hypertension and homocysteinemia, all of which were negative. Keeping in mind his Indian subcontinent origin, he was presumed to have Eales' retinal vasculitis. The patient was planned for pars plana vitrectomy but, to our surprise, during the preoperative inpatient stay the patient developed a 2 mm mobile hypopyon in the AC of the right eye. The AC of the patient had been examined 3 hours prior to this occurrence and had been found to be normal.

INVESTIGATIONS

Both these patients had no signs of systemic disease and had been thoroughly evaluated for systemic associations of retinal vasculitis. Very high serum leucocyte counts and myeloblasts were detected in peripheral blood film. The second case was positive for Philadelphia chromosome.

Figure 1 (A and B) Fundus photographs depicting leukaemic vasculitis and macular oedema. Roth spot-like haemorrhages (encircled) can be seen in the left eye (OS).

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Figure 2 Clinical photograph of the left eye (OS) showing total hyphaema. White exudative material can be seen dispersed within the hyphaema, while a superior hypopyon can also be seen (white arrow).

TREATMENT

After haematology consultation, chemotherapy was administered for CML. AC of both the patients cleared gradually within 2 weeks. While the first case was observed, the second case underwent vitrectomy OD for vitreous haemorrhage and OS retina was laser photocoagulated multiple times. During the surgery, neovascular fibro vascular proliferation was noted nasal to the optic disc.

OUTCOME AND FOLLOW-UP

Six months after presentation, BCVA was 6/6 OD and 6/12 OS in the first case, and 6/18 OD and 6/6 OS in the second eye. The patients are taking haematology consultations regularly for need of chemotherapy and were systemically stable at last follow-up.

DISCUSSION

Across its spectrum of ocular manifestations, leukaemia may involve the optic nerve, retina, choroid, ciliary body, iris and anterior chamber, cornea, sclera or the conjunctiva.^{1 14} While retinal and uveal manifestations are common, those of avascular tissues are rather rare. In AC, perhaps hyphaema is a more frequent finding than hypopyon.¹ Most cases of 'leukemic hypopyon' have been reported in patients with an established diagnosis of acute leukaemia, with few being seen in CML as well. Its occurrence in CML has been described either as a presenting sign or in the very early course of the disease.^{15 16} This difference between acute and chronic leukaemia is likely to be due to the insidious nature of CML. In either case, ocular examination even in the absence of symptoms is necessary as ophthalmic signs may be indicative of a future downhill course.¹⁷

Two previously reported cases of CML with AC hypopyon described by Santoni *et al*¹⁶ and Lipton *et al*¹⁸ had been treated with chemotherapy recently and had presented with 'kerato-hypopyon' while in blastic transformation. Previously, AC exudation has been documented to be the presenting sign of CML in a case with myelocytes, band forms and promyelocytes determined on blood film examination.¹⁵ A single case of CML-related hypopyon in a child with anterior uveitis refractory to topical steroids has also been seen.¹⁹ In our cases, the patients presented with visual loss and during the course of management were found to have myeloblasts on peripheral blood film examination. It is generally believed that hypopyon of acute leukaemias may not always respond to induction therapy, the AC being an immune-privileged sanctum to

systemic chemotherapy penetration.²⁰ However, in our cases of CML, there was a gradual resolution of the hypopyon after the induction of chemotherapy.

Cytopathology of the hypopyon in one of the aforementioned cases had revealed predominance of lymphocytes and few plasma cells.¹⁵ In the other case with multiple ocular recurrences, it revealed immature lymphocytes with a high nuclear-cytoplasmic ratio and prominent nucleoli.¹⁸ AC exudation in CML is rare but should be taken seriously as it may be representative of blast crisis.^{16 18 20} The sudden occurrence of AC exudation is most likely due to direct invasion of the AC by tumour cells.²⁰ As seen in our first case, this may be accompanied by spontaneous hyphaema due to the accompanying bleeding tendency.

Usually, hypopyon is associated with infective (eg, endophthalmitis and keratitis) or inflammatory (eg, Behcet's disease-associated uveitis) conditions. These diseases are, however, associated with other pathognomic ocular signs. Significant whitish AC exudation in both our cases had developed over a very short course (2–3 hours in the second case). Thus, rapid development of AC hypopyon in the absence of other ocular features should alert the physician to the possibility of a haematological malignancy. Although 'ghost cells' may also accumulate in cases with vitreous haemorrhage, presenting a false appearance of a hypopyon, they usually occur in cases of chronic haemorrhage persisting over months as against the short history of our cases.²¹

Associations of retinal vasculitis and differential diagnoses of Eales' disease are well known.²² Bilateral disease in the Indian subcontinent is frequent, and a young healthy man presenting with bilateral vitreous haemorrhage or proliferative retinal vasculopathy is often assumed to be a case of Eales' disease after initial assessment, as we did in our second case. Even, in our first case, the patient had been initially diagnosed with Eales' disease elsewhere, but the presence of Roth spots (figure 1) alerted us to the possibility of leukaemic vasculitis.²³ Therefore, every such case should be diagnosed only after careful exclusion of associations and comprehensive tailored workup based on clinical findings like AC exudation and Roth spots. A simple investigation such as a haemogram with a peripheral smear must not be overlooked in such cases.

We regret that we do not have images of the second case.

Learning points

- ▶ An unexplained anterior chamber hypopyon is a crucial clinical pointer to the likelihood of leukaemia.
- ▶ Apart from acute leukaemias, it may also be seen in chronic myeloid leukaemia during the early course of disease.
- ▶ The visual prognosis may, however, be independent of its occurrence.

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