

Leukostasis syndrome in CLL

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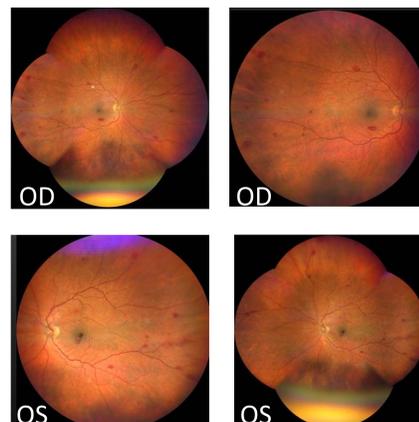
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Introduction

- Chronic lymphocytic leukemia (CLL) is a neoplasm of mature B lymphocytes and often has a benign clinical course.
- Elevated WBC alone is not an indication for treatment. (Often can be $> 100 \times 10^9$)
- We present a case of leukostasis syndrome (symptomatic hyperleukocytosis), a hematological emergency which occurs rarely in CLL.

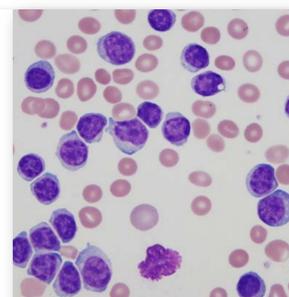
Case Presentation

A 52-year-old man with a history of hypertension and CLL presented to ophthalmology clinic with acute blurred left eye vision. Fundoscopic exam was notable for scattered white centered intraretinal hemorrhages in both eyes, consistent with leukemic retinopathy.



Fundoscopy exam photos at presentation demonstrating thick central intraretinal hemorrhage and scattered peripheral retinal hemorrhages

- An urgent outpatient hematology consultation was arranged, and the patient was referred to the hospital when routine laboratory studies revealed a WBC of $518.9 \times 10^9 / L$ with 97% lymphocytes.
- Hemoglobin was 5.4 g/dl and platelet count was $56 \times 10^9 / L$.
- On physical examination, the patient appeared well, lungs were clear to auscultation, abdomen was soft with profound splenomegaly.
- CT abdomen/pelvis revealed diffuse lymphadenopathy, hepatosplenomegaly, a segment of mildly dilated small bowel and mild diffuse pulmonary opacities.

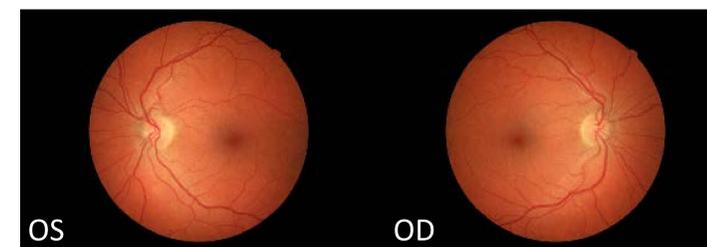


Peripheral smear with many lymphocytes and smudge cells

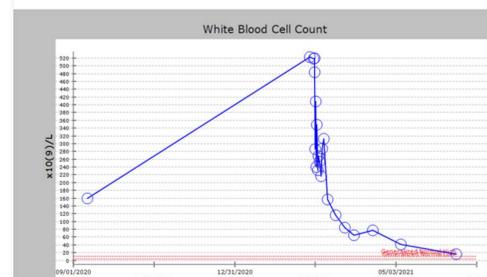


Thickened loop of small bowel

- The patient's WBC rapidly decreased to $231.8 \times 10^9 / L$ after three rounds of leukapheresis. Additionally, chlorambucil and acalabrutinib were given to treat CLL.
- Allopurinol and intravenous fluids were given to prevent the development of tumor lysis syndrome (TLS), a common complication of leukostasis syndrome.
- The patient was discharged with close hematology follow up, and his vision had improved from 20/150 to 20/40 at his 6-month ophthalmology follow-up visit.



Fundoscopy exam photos at 6 months show significant improvement in retinal hemorrhages corresponding to improvement in visual acuity.



The patient's WBC had normalized at the three-month hematology follow-up visit.

Discussion

- Leukostasis is a clinical diagnosis that often manifests as neurological or pulmonary symptoms
- Leukemic leukocytes can cause injury to vascular walls, resulting in thrombosis and occlusion of both large and small vessels. Additionally, leukemic cells can consume oxygen at a high rate which exacerbates microvascular ischemia.
- Urgent hematology consultation is indicated for leukostasis syndrome.
- Leukapheresis is a procedure that mechanically separates and removes WBCs, resulting in a rapid, but temporary reduction in leukocrit.
- Leukapheresis requires specialized equipment and blood bank consultation.
- There is no clear evidence that leukapheresis improves long-term mortality.
- The definitive treatment of leukostasis is targeted therapy for the underlying hematological malignancy.

References

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