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Visual disturbance as initial presentation of hairy cell leukemia

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PURPOSE. *Hairy cell leukemia (HCL) is a rare disorder that occasionally has visual symptoms after diagnosis. The authors present a case of HCL in which bilateral visual symptoms led to the initial diagnosis.*

METHODS. *Observational case report.*

RESULTS. *Bilateral decreased vision to 20/30 in the right eye and 20/40 in the left due to intraretinal and preretinal hemorrhages with no other systemic signs or symptoms prompted a hematologic evaluation in which HCL was found to be the causative disorder in a previously healthy 41-year-old man.*

CONCLUSIONS. *The authors present a rare case in which bilateral visual complaints led to the diagnosis of HCL. Thus, though uncommon, HCL should be considered in the differential diagnosis of otherwise unexplained retinal hemorrhages. (Eur J Ophthalmol 2009; 19: 318-20)*

KEY WORDS. *Hairy cell leukemia, Preretinal hemorrhage, Retinal hemorrhage*

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INTRODUCTION

Hairy cell leukemia (HCL) is a rare non-Hodgkin B-cell lymphoproliferative disorder with an incidence of approximately 600 cases per year in the United States. Patients usually present in one of four ways: 1) splenomegaly causing abdominal fullness which may be painful; 2) systemic complaints of fatigue, weight loss, and weakness; 3) bruising and bleeding which can be severe and recurrent; 4) incidental finding of enlarged spleen or abnormal blood counts on routine testing. It is four times more common in men than women and the median age at presentation is 52 years. Diagnosis can be confirmed by immunophenotyping from bone marrow biopsy (1).

There have been a handful of cases that document ocular involvement following HCL diagnosis and treatment. Two German reports by the same primary authors report retinopathy and monocular visual disturbance due to intraretinal hemorrhage as the initial symptom of hairy cell leukemia (2, 3). Findings in these reports included monocular intraretinal bleeding and edema (left eye only) and bilateral, soft exudates, microaneurysms, and capillary occlusions. Other reports have later documented ocular involvement including panuveitis and retinal vasculitis coincident with hairy cell leukemia either related to disease pathology or secondary to treatment (4-6). In the Czech literature, there are two reports, by the same authors, of leukemic infiltrates of the cornea in a single female patient with HCL (7, 8). However, ocular involvement is extremely rare and to our knowledge, we present the first case of bilateral blurry vision, preretinal hemorrhages, and macular thickening as the first presenting sign and symptom of HCL.

METHODS AND RESULTS

A 41-year-old man reported blurry vision and was evaluated by ophthalmology. Visual acuity was 20/30 in the right eye and 20/40 in the left. The remainder of the anterior segment examination was unremarkable. Posterior segment demonstrated bilateral preretinal central macular hemorrhages and blot retinal hemorrhages (Fig. 1). On OCT, there was notable bilateral thickening at the site of hemorrhage. Fluorescein angiography also

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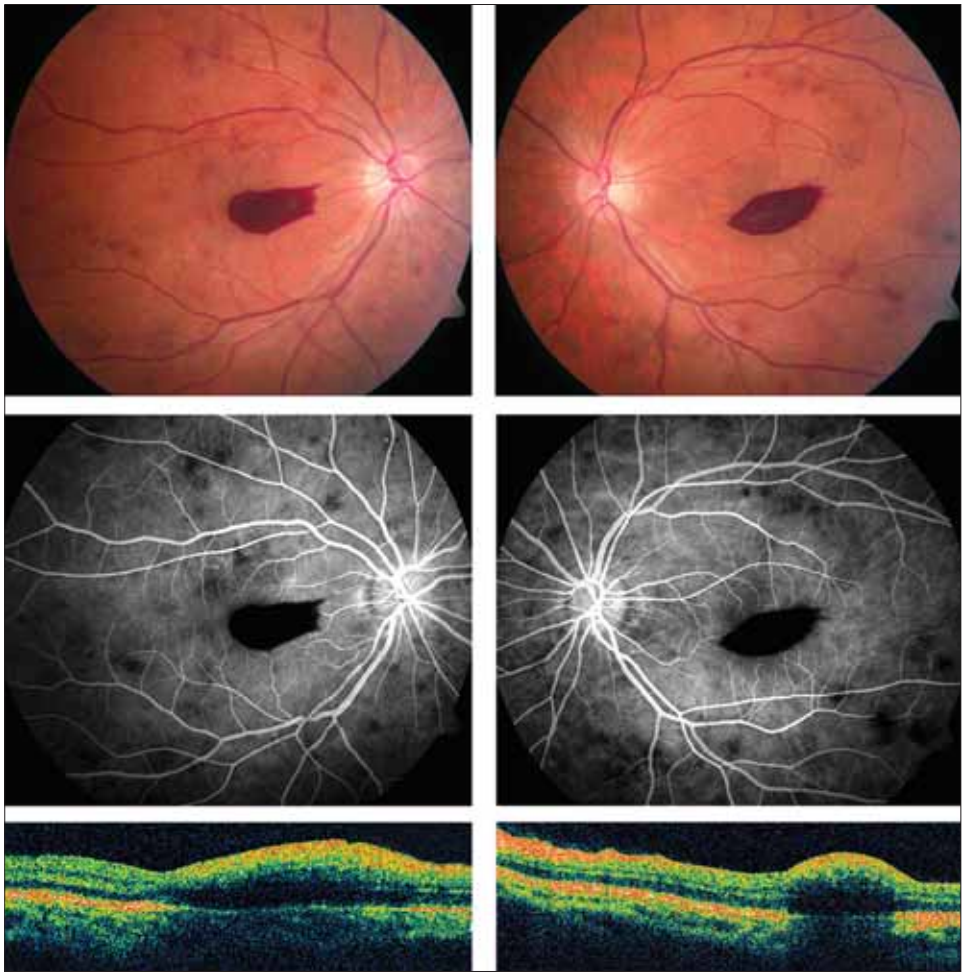

Fig. 1 - *Color fundus photographs* **(top)** *of the right and left eyes showing central preretinal macular hemorrhages and scattered dot hemorrhages with fluorescein angiogram* **(middle)** *and optical coherence tomography* **(bottom)** *showing the resulting blockage and central thickening, respectively.*

showed blockage at the site of hemorrhage.

He was referred for further workup and found to have severe anemia, thrombocytopenia, and severe splenomegaly; interestingly, he did not report fatigue or weight loss. Of note, his hematocrit, hemoglobin, and platelet levels were extremely, low measuring 9.3%, 3.1, and 40,000, respectively, while his white blood cell count was normal at 6900. Bone marrow biopsy confirmed the diagnosis of HCL with abundant characteristic “hairy” appearing mononuclear cells that stained positive for CD19, CD20, and ANXA1 and negative for CD5 and CD10.

DISCUSSION

A number of patients with HCL experience unusual bruising and bleeding. In the case we present, a patient presented with bilateral decreased vision due to preretinal hemorrhages, leading to the diagnosis of HCL in the absence of other vascular abnormalities and before other memorable symptoms. This is the first reported case of initial HCL presentation with visual complaints in the United States. Thus, it is worthwhile that HCL be considered in the differential diagnosis of otherwise unexplained retinal hemorrhages.

There are two German reports, by the same authors (Bertram et al), of HCL presenting as retinopathy (2, 3). In their case, retinal hemorrhages, capillary occlusions, and soft exudates were found on ophthalmoscopy and angiography in both eyes; however, the decreased vision was only in the left eye and attributed to intraretinal hemorrhage and edema. In the present case, there were also bilateral retinal hemorrhages and capillary occlusions, but there was also bilateral preretinal hemorrhage and associated macular edema and decreased vision, without any exudation in either eye. Whereas in both cases further examination and laboratory workup revealed splenomegaly, anemia, and thrombocytopenia, it is of note that the pa-

tient in this report had no complaints of fatigue or weight loss and had a normal white blood cell count. This underscores that although HCL is often associated with a leukopenia, the WBC may be normal, or, as in the German report, elevated.

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