Asymptomatic Leukemic Optic Nerve Infiltration as Presentation of Acute Lymphoblastic Leukemia Relapse

Florin Grigorian¹, Ang Li², Adriana P Grigorian¹

¹University Hospitals Eye Institute, Cleveland, OH, USA, ²Case Western Reserve Medical School, Cleveland, OH, USA

Introduction:
We present a 4 year old boy with history of CNS acute lymphoblastic leukemia (ALL) in remission, diagnosed with leukemic optic neuropathy at a routine eye exam.

Methods:
Vision was equal in both eyes, measuring 20/30 by Allen, considered normal given the age limitations. Color vision (HRR) was normal and equal in both eyes and pupils were equally reactive to light and accommodation without relative afferent papillary defect (RAPD). Fundus examination was normal in the right eye and revealed a white mass covering the left optic disk, completely obscuring it and peripapillary vascular sheathing. The rest of the fundus was normal.

Results:
The MRI showed no enhancement of the optic nerves or brain tissue. There was a questionable diffusion restriction of the left optic nerve head. The peripheral blood smear showed 1% blasts (5% diagnostic for relapse) and the CSF revealed rare cells suspicious of blasts. Chemotherapy was started (Methotrexate and Vincristine). No ocular radiation was performed due to the asymptomatic nature of the condition. At 3 weeks follow up the vision exam was the same and the mass covering the left optic nerve was replaced by mild gliosis with good visualization of the optic nerve head. The fluorescein angiography did not demonstrate any defects. The CSF pathology later revealed blasts as the dominant cells and the bone marrow biopsy revealed hypercellularity confirming the relapse.

Conclusions:
Tumoral optic nerve infiltration may be the first sign of ALL relapse. In our case the vision, color vision and pupillary reflexes were normal and maintained throughout the evolution of the disease. Of note, the absence of enhancement on MRI does not exclude neoplastic infiltration. We recommend routine ophthalmological exam for all patients with history of ALL to exclude optic nerve involvement without systemic symptoms or signs.

References:

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