Spectrum of clinical and subclinical choroidal abnormalities in patients with histiocytosis

Jasmine Francis, MD, FACS
New York, NY

Julia Canestraro, OD, Rampal Raajit, MD, David Abramson, M.D., Eli L Diamond, MD

Purpose:
To evaluate choroidal findings in patients with histiocytic neoplasms (NH) including abnormalities by ophthalmoscopy and optical coherence tomography characteristics (OCT) (subfoveal choroidal thickness (SFCT) and vascular architecture); and to determine if abnormalities change with histiocytosis-directed (kinase inhibitor) therapy.

Methods:
This retrospective review included patients with histiocytosis who completed an ophthalmic assessment and multimodal imaging at Memorial Sloan Kettering Cancer Center from June 2014 through March 2020. Clinically evident choroidal infiltrates by ophthalmoscopy were recorded. SFCT and was measured using enhanced depth imaging spectral domain optical coherence tomography (EDI SD-OCT) from the outer portion of Bruch’s membrane to the choroidal scleral interface. Choroidal vascular architecture was qualitatively examined. The main outcome measure was SFCT compared to matched controls, secondary outcome was change in SFCT on histiocytosis-directed (kinase inhibitor) therapy.

Results:
Ninety-four eyes of 47 patients (25 males, 22 females) with histiocytic neoplasms (Erdheim-Chester 28, Rosai-Dorfman 8, Xanthogranuloma 4, Mixed histiocytosis 7) were examined. 13.8% of eyes had clinically evident choroidal infiltration. In the entire cohort, the mean SFCT was 337.6 +/- 90.4mm compared with 248.0 +/- 81.5mm in the control group (p<0.0001). Brain or osseous sites of disease did not correlate with SFCT. OCT findings in the histiocytosis group included: 59.6% of eyes exhibited enlarged Haller’s layer with distortion of choroidal inner layers; and 39.4% of eyes had an outer choroidal infiltrate. In 68 eyes with follow-up imaging, the choroidal thickness significantly decreased by 31µm (p-value < 0.001) on histiocytosis-directed (kinase inhibitor) therapy.

Conclusions:
Patients with histiocytosis may have clinically evident infiltration of their choroid. Furthermore, patients with histiocytosis have increased SFCT compared with age and gender-matched controls that decreases with histiocytosis treatment. Choroidal abnormalities may be an underappreciated and measurable manifestation of histiocytosis and a marker of response to systemic treatment.