Spectrum of clinical and subclinical choroidal abnormalities in patients with histiocytosis

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No Financial Interests or Relationships
Off label Use Drugs
14% of non-LCH histiocytosis patients have clinically evident infiltration of the choroid.

However, the vast majority of these patients (80%), have statistically significant increase in SFCT compared to age- and gender-matched controls.

With systemic treatment, SFCT decreases.

Choroid morphology:
- outer choroidal infiltrative densities (40%)
- enlarged Haller’s layer w/ inner ch. distortion (60%)

There is no association with CNS nor osseous disease.
Histiocytic Disorders: Introduction

- Clonal, hematopoietic disorders
- Accumulations of activated histiocytes in affected tissues

- Langerhans Cell Histiocytosis
- Non-Langerhans Cell Histiocytosis
  - Erheim Chester Disease
  - Rosai Dorfman disease
  - Xanthogranuloma
# Histiocytic Disorders: multisystem disease

## Langerhans Cell
- **CNS (6%)**
  - Hypothalamic-pituitary axis
- **Lung (10%)**
- **Spleen (13%)**
- **Liver (16%)**
- **Lymph nodes (19%)**
- **Skin (39%)**
- **Bone (77%)**

## Non-LCH (ECD)
- **CNS (40%)**
- **Lung (46%)**
- **Orbit (22%)**
- **Maxillary sinus (59%)**
- **Heart (57%)**
- **Skin (27%)**
- **Bone (95%)**
Histiocytic Disorders: Ocular manifestations

- **Orbit**
- **Eyelid**
- **Other...**
Histiocytic Disorders: Published Choroid


Barak et al, Retin Cases Brief Rep, 2012
Histiocytic Disorders: Published Choroid
Study Question

In histiocytosis:
1. Does the OCT of the choroid have characteristic findings?
2. Does this change with treatment?

Histiocytosis Service
Memorial Sloan Kettering

https://www.mskcc.org/cancer-care/types/histiocytosis

Eli Diamond MD
Patients included:

- Histiocytosis Patients 98
  - LCH 34
    - Ophtho 14
  - Non-LCH 64
    - Ophtho 47

94 eyes

- This was compared to an age and gender matched control set of patients
13 of 94 eyes (14%) had **clinically evident** choroidal infiltration (some more evident than others). In all cases, the findings are post-equatorial. In the 9 patients, 4 had bilateral findings.
Mean subfoveal choroidal thickness (SFCT):

Normal controls = 250µm
Non-LCH patients = 337µm.

The difference, $p = 0.00008$

- Non-LCH patients had thicker SFCT, as measured by OCT
Examples of choroidal thickness

NORMAL: mean 250μm

HISTIO: mean 337μm
(80% of eyes had SFCT > 250μm)
Other occult choroidal findings in non-LCH eyes

40% of eyes have an outer choroidal density (infiltrate?)

60% of eyes have an enlarged Haller’s layer (the large choroidal vessels) with distortion of the inner choroidal layers
Results

- 34 non-LCH patients (with imaging at baseline AND follow up): significant decrease in SFCT on systemic treatment

The mean change was a decrease of 31μm (p-value = 0.000005)

The length of time on treatment (and f/u exam) did not appear to influence the amount of SFCT
Results

Is CNS/osseous disease a marker for disease burden and would that correlate with choroidal thickness?

- No significant difference in SFCT between patients with or without CNS disease (p-value = 0.35)

- No significant difference in SFCT between patients with or without osseous disease (p-value = 0.35)
Why is the choroid thicker in histio?

• We know the choroid is a site for accumulation of activated histiocytes (in the clinically evident cases). In the subclinical eyes, could the thicker choroid represent disease involvement?

• For instance, lymphoma and myeloproliferative diseases can infiltrate the choroid

• And/or could the choroid be a site where these abnormal activated histiocytes are being generated?

• We know the choroid can be a site of extramedullary hematopoiesis (both in neonates and adults- although our knowledge is limited to enucleation specimens). Therefore, presumably, the choroid could be a source of these cells
Conclusion/Summary

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