Otolaryngologic Presentation of Multifocal Non-Langerhans Cell Histiocytosis
Jason Toy BS¹, Michael Larson MD¹, Melissa Mark MD², Maria Aguiar MD³, Thomas Gallagher DO⁴
¹Department of Otolaryngology, Eastern Virginia Medical School, ²Division of Hematology and Oncology, Children’s Hospital of The King’s Daughters, ³Division of Pathology, Children’s Hospital of The King’s Daughters, ⁴Department of Otolaryngology, Children’s Hospital of The King’s Daughters

Introduction
• Non-Langerhans Cell Histiocytosis (non-LCH) is a classification of proliferation of histiocytes that cannot be characterized as Langerhans cells (CD1a+/Langerin+/S100+).¹
• Non-LCH encompasses a spectrum of diagnoses that are differentiated by the locations effected by the disease process:²
  - Predominant cutaneous involvement
  - Skin involvement with a major systemic component (present in this case)
  - Predominant extracutaneous involvement
• The presentations each histiocytosis can vary widely, even when comparing diagnoses within one of these specific subgroups.
• The primary histiocyte cells in each specific type of histiocytosis are identified using immunohistochemical markers. Although pathogenesis of these conditions is not fully understood.

Case Presentation
• A 6-year-old previously healthy female presented with a 2-year history of bilateral proptosis, progressively worsening on the left with acute redness and tearing.
• No associated pain, itchiness, or visual impairment. Denied fevers, chills, night sweats, weight or appetite changes.
• Physical exam positive for bilateral proptosis L>R, inability to close left eye, and a full abdomen. Eyes were non-tender
• Laboratory results significant for Total Bilirubin 1.5, AST 59, and LDH 1094.

Case History
• Emergency department CT of the orbits to assess for optic glioma, showed bilaterally enlarged optic nerves, bilateral proptosis L>R, and a left maxillary sinus mucous retention cyst.
• MRI of the orbits, face, and neck w/wo contrast shows infiltrative soft tissue mass in neck, extending to bilateral maxillary sinuses and orbits L>R with displacement of optic nerves (Figure 1).

Discussion
• Non-LCH diseases are defined by their immunohistochemical signature and typically follow a documented pattern of involvement. This case is unique as its features are shared between two histiocytosis processes.
  - Head, neck, and orbital masses that are S100+, CD1a-, and Langerin-, occurring in children is characteristic of extradural Rosai-Dorfman disease.²
  - Bilateral renal and significant non-head and neck involvement is more suggestive of Erdheim-Chester disease. However, this patient is missing symmetric long bone lesions that are often seen and is significantly younger than normal presentation.³
• Erdheim-Chester disease and Rosai-Dorfman disease share some common features and have been seen concomitantly in a small number of cases, however, this is typically seen in older adult patients.⁴ Highlighting the unique presentation of this patient’s case.
• Treatment for non-LCH varies based on the site(s) involved and degree of involvement, ranging from simple resection to systemic chemotherapy when disease is disseminated and high-risk organs are involved, as seen in the present case.

Diagnosis and Management
• Given the pathology and radiologic findings a diagnosis of a non-LCH was made, however the exact type is difficult to ascertain.
• Regardless of specific diagnosis, the multifocal nature of the disease process and ophthalmic involvement dictates need for systemic chemotherapy, Clodarabine dosed 25mg/m².
• Current regimen is six 5-day cycles 28 days.
• Only significant new symptoms throughout course has been an isolated episode of palpitations, Echo, EKG, and cardiac MRI all within normal limits.
• The patient is on PJP prophylaxis with Bactrim and receives Neulasta at the completion of each treatment cycle secondary.

References

Contact
Jason Toy BS
825 Fairfax Ave Suite 310
Norfolk, VA 23507
Toy@EVMED.edu

Figure 1: Coronal and axial MRI views demonstrating soft tissue mass involving the face, bilateral maxillary sinuses, and orbits.

Figure 2: Left maxillary soft tissue mass, biopsied via endoscopy.

Figure 3: Sagittal MRI showing L spine and sacral spinal canal involvement.