Langerhans Cell Histiocytosis

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Outline

• What is Langerhans cell histiocytosis?
• Radiology of skull and lung lesions
  – Patient MK
• Different types of common skull lesions
  – Patient JS
• Involvement of other bones
  – Patient TJ
• Where else do we see LCH?
What is LCH?

- *Histiocytosis*: A group of disorders characterized by the proliferation of histiocytes (macrophages)

- *Langerhans cell*: Dendritic cell found in the epidermis

Ardavin, 2003
What is LCH?

• *Langerhans cell histiocytosis*: Clonal proliferation of Langerhans cells in one or many organs: Bone, skin, lungs, brain, soft tissues, other organs

• Thought to be caused by abnormal response to infection
A disease of many names

- Histiocytosis X (Lichtenstein, 1953)
  - *Eosinophilic granuloma*: Localized bone disease
  - *Hand-Schüller-Christian disease*: Skull lesions, proptosis, and diabetes insipidus
  - *Letterer-Siwe disease*: Disseminated histiocytosis
- Langerhans cell histiocytosis (1973)
What is LCH?

- **Epidemiology**
  - Most common in children 1-4 years old
  - Incidence: 2-5 patients per million per year
- **Presentation**
  - Bone pain, rash, dyspnea
- **Diagnosis**
  - Need biopsy for diagnosis
  - EM: Birbeck granules

http://www.som.tulane.edu/classware/pathology/medical_pathology/New_for_98/Lung_Review/Lung-55.html
Treatment and prognosis

• Restricted (no visceral organs involved)
  – Treatment: Observation, local radiation, corticosteroid injection, excision
  – Prognosis is good

• Extensive (visceral organs involved)
  – Treatment: Chemotherapy, corticosteroids
  – Worse prognosis: 50% mortality with disseminated disease
Patient MK

• Two-year-old girl who presented with a rapidly-growing bump on her head
• Head CT showed parietal bone lesions

http://www.gwc.maricopa.edu/class/bio201/skull/latskul.htm
Patient MK: CT head w/ contrast

- Well-defined lytic lesions in the R and L parietal bones
- Bone is the most common site of LCH lesions; skull is involved in 30%
- Classic skull lesions: Round, osteolytic, with sharp borders
- Ddx: Mets, venous lakes
Patient MK: CT head w/ contrast

- What classic feature of LCH skull lesions is seen here?
Patient MK: CT head w/ contrast

- Classic feature of LCH skull lesions: On tangential views, a beveled edge appearance

Children’s Hospital Boston
LCH of skull: Plain skull film

- In this similar patient, we see well-defined lytic lesions, one with a beveled edge.
Patient MK

• MK was confirmed to have LCH by biopsy
• Patients with LCH need a skeletal survey:
  – CXR
  – AP films of all long bones
  – AP film of pelvis
  – Lateral spine films
• Bone lesions need CT evaluation to determine treatment
• Look for visceral involvement if the patient has symptoms
Patient MK

- MK’s skeletal survey showed a lesion in her scapula
- A chest CT to evaluate the scapular lesion also revealed lung involvement
Patient MK: CT chest w/ contrast

- MK’s chest CT shows multiple nodular opacities in the RML
- Nonspecific, but likely LCH in this patient
- This is evidence of extensive disease, so MK received chemotherapy
Pulmonary LCH

- Lung is involved in 23-50% of patients
- Work up pulmonary symptoms with CXR, then CT
Pulmonary LCH: CXR

- Classic CXR findings: Reticulonodular opacities most prominent in the upper and mid lung fields
- LCH spares the lower lobes

http://www.histio.org/society/LCH/Adult/vassallo1.shtml
Pulmonary LCH: CT chest w/ contrast

- Classic chest CT findings in extensive disease: Reticulonodular changes, large cysts, and destruction of lung parenchyma
- Cysts → bullae → pneumothorax
- Ddx: PCP, TB, LIP

Schmidt et al., 2004
LCH of skull

- Patient MK had skull and lung findings
- Let’s return to the skull, an important site of involvement for LCH
- We’ve seen LCH involving the parietal bones

http://www.gwc.maricopa.edu/class/bio201/skull/latskul.htm
LCH of skull

• There are three other common sites of LCH lesions in the skull, each associated with a classic presentation or finding

• **The first:** Mastoid involvement can spread to the bones of the middle ear → deafness

http://www.gwc.maricopa.edu/class/bio201/skull/latskul.htm
LCH of skull: 3D CT

- *The second:* Mandibular involvement causes “floating tooth” appearance

Kilborn et al., 2003
Patient JS

- *The third*: Orbital wall involvement, as illustrated by patient JS
- One-year-old boy who presented with a rash, which on biopsy showed LCH
- Skeletal survey showed L orbital involvement
Patient JS: CT head w/ contrast

- Soft tissue mass centered in the greater wing of the L sphenoid bone, involving the lateral orbital wall, with associated bone destruction

- Let’s review some anatomy...
Skull anatomy

- JS’s lesion involved the greater wing of the sphenoid on the L

Sphenoid bone:

http://www.csuchico.edu/anth/Module/skull.html
Skull anatomy

- The greater wing of the sphenoid forms part of the lateral orbital wall

http://www.oculoplastics.com/topics/tumors_orbit/orbital_tumors_anatomy.htm#volume
LCH of skull

• *The third common skull lesion*: Orbital wall involvement $\rightarrow$ proptosis (downward displacement of orbit)

• *Hand-Schüller-Christian disease*: Skull lesions, proptosis, and diabetes insipidus

• Summary of common skull lesions:
  – Mastoid involvement $\rightarrow$ deafness
  – Mandibular involvement $\rightarrow$ floating tooth
  – Orbital wall involvement $\rightarrow$ proptosis
LCH of bones

- What about the rest of the skeleton?
- Radiologically, these lesions look different than skull lesions
- LCH can occur in any bone except those of hands and feet
- Most common sites of involvement: Skull, ribs, pelvis, femur

Howarth et al., 1999
Patient TJ

- Six-year-old boy who presented with pain in his left arm after falling on it
Patient TJ: Plain film L humerus

- TJ has a lesion in the mid-diaphysis of the L humerus
Patient TJ: Plain film L humerus

- Classic early long bone lesions: Aggressive-looking, with endosteal scalloping, cortical thinning, and periosteal reaction

- Ddx: Ewing’s sarcoma, osteosarcoma, osteomyelitis, lymphoma, met

- Classic later lesions: Sclerotic margins, no periosteal reaction
Patient TJ: T1 MR L humerus

- Ill-defined, hypointense lesion involving the marrow and cortex of the humerus, with an associated soft tissue mass
- Typical T1 MR appearance for LCH outside the skull
- 30% have an associated soft tissue mass
Patient TJ: T2 MR L humerus

- Hyperintense lesion with laminated periosteal reaction
- Ddx: Ewing’s sarcoma, osteosarcoma, osteomyelitis, met, lymphoma
- TJ’s biopsy showed LCH
Where else do we see LCH?

• We have focused on bone and lung, the most common sites for radiologically-identifiable LCH lesions

• LCH can be seen radiologically in:
  – CNS, especially pituitary
  – Liver, spleen, and biliary system
  – Lymph nodes
  – Soft tissues
  – GI tract
Acknowledgements

• Gillian Lieberman, M.D.
• Pamela Lepkowski
• Larry Barbaras
• Christine Duncan, M.D.
• Matthew Jolley, M.D.
• Fabio Komlos, M.D.
• Alexander Guimaraes, M.D., Ph.D.
References