Clinical Presentation

• ID:
  – 2 day-old newborn delivered by C-section at 41 weeks of gestation due to failure to progress

• CC:
  – Multiple purple nodules on head, trunk and extremities, but otherwise physical exam normal.
  – Mother is a healthy 20 year-old primigravid, O negative, VDRL non-reactive, GBS negative, rubella non-immune, HIV non-reactive, hepatitis negative, gonorrhea and chlamydia negative
  – Pregnancy uneventful
Blueberry Muffin Baby
Differential Diagnosis

• Causes of Extramedullary Hematopoeisis
  – Infectious
    • ToRCH
  – Hematological
    • Hemolytic anemias, hereditary spherocytosis, intracranial bleeding, ABO incompatibility, twin-twin transfusion, hemangiomatosis
  – Malignancy/Proliferative
    • Congenital leukemia cutis, congenital rhabdomyosarcoma, Langerhans cell histiocytosis and myelodysplasia

Physical Examination
5mm to 1.5cm bluish, compressible, yet slightly firm, non-blanching, vascular papules
Histopathology
Lower power: Dense mid to deep reticular dermal infiltrate

Medium power: large pale cells with eosinophilic cytoplasm dissecting through collagen
High power: Cells containing abundant amphophilic cytoplasm with large and convoluted/reniform nuclei.

CD1a
**Histologic Diagnosis**

- Langerhans Cell Histocytosis
  - Congenital Self-healing Reticulohistiocytosis (Hashimoto-Pritzer)
  - Acute Disseminated LCH (Letterer-Siwe)
  - Chronic Multifocal LCH (Hand-Schüller-Christian)
  - Chronic Focal LCH (Eosinophilic Granuloma)

**LCH in the Neonatal Period**

- Congenital Self-Healing Reticulohistiocytosis
  - More likely present at birth
  - Nodular lesions more common
  - Cutaneous only
- Acute Disseminated LCH
  - Before age 2 years
  - Papulosquamous eruption of scalp, flexural areas, and trunk.
  - Pulmonary, hepatic and splenic involvement may occur.
Lab and Imaging Results

- Serologies for toxoplasma, rubella, CMV negative
- CBC and WBC differential normal with mild thrombocytopenia
- Hct, LFT, BMP normal
- Whole body scintigraphy, full body CT and US was negative

Diagnosis at Presentation

Congenital Self-Healing Reticulohistiocytosis (Hashimoto-Pritzker)
Histiocyte Society Guidelines on LCH Baseline and Reactivation

- Physical examination
- CBC and coagulation studies
- Liver function tests
- Skeletal radiographic survey
- Chest radiograph
- Abdominal ultrasound
- Urine osmolality
- If indicated by symptoms:
  - Bone marrow evaluation

Management: Congenital Self-Healing Reticulohistiocytosis

- Self-limited: topical and systemic therapies usually not required
- Regular Derm and Heme-Onc follow-up recommended to monitor for relapse or progression
  - Several reports of cutaneous-only LCH at presentation with development of multi-system involvement.
  - Increased risk for second malignancies
Follow-up: Our Patient

- At 2-month follow-up, all but 2 lesions on the face had resolved.
- At 4-month follow-up, all cutaneous lesions had resolved.
- No systemic findings

Congenital Self-Healing Reticulohistiocytosis

The End

References

- The Histiocyte Society. www.histio.org