History
A 2 year old male presenting with papular rash on scalp of face, back and extremities, chronic otitis media, difficulty breathing, jaundice and failure to thrive.

Diagnosis
Langerhans Cell Histiocytosis (LCH) of the Thyroid Gland

Additional Clinical
Laboratory:
Tbil 10.2 mg/dL
AST 184 IU/L
ALT 112 IU/L
GGT 525 IU/L
Hepatitis, CMV, EBV (-)
Abdominal US:
Hepatomegaly and periportal edema.
Pathology:
Skin biopsy demonstrated monotonous cells staining positive for S100 and CD1a consistent with LCH.

Discussion
LCH is a disorder of immune dysfunction related to clonal proliferation of Langerhans cells. The typical patient is a young male caucasian although all ages, genders and races can be affected. Prognosis is related to the extent of systemic involvement. LCH can involve any organ. LCH of the thyroid is almost always associated with multisystem disease but is occasionally a unifocal process. Histiocytic infiltration results in thyromegaly and disorganized parenchyma or masses on imaging.

Findings
CT of the chest at initial presentation demonstrates an enlarged heterogeneous thymus, reticulonodular pulmonary infiltrates with scattered air cysts and a hypodense thyroid gland related to LCH. CT 2 months after initiation of therapy shows some restoration of normal thyroid architecture. Ultrasound demonstrates interstitial hypoechogenicity which is more focally prominent centrally.

Reference
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