ACQUIRED HYPOTHYROIDISM IN A TODDLER

An Unusual Presentation of LCH

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CASE PRESENTATION

A previously healthy, developmentally appropriate 23-month-old male was admitted to hospital for evaluation of persistent rash, periorbital edema, and an enlarging neck mass. His scalp rash had developed over three months and was unresponsive to multiple treatment modalities. His neck mass rapidly progressed over one month. He had bilateral periorbital edema with purulent discharge. He was also experiencing intermittent ear drainage and bleeding, gum hypertrophy and mucosal ulcers. Past medical history was unremarkable. He had a normal newborn metabolic screen and he consumed an iodine replete diet.

On physical exam, he appeared generally unwell. A strikingly large lower neck mass was present and obscured the typical suprasternal anatomical landmarks. The mass was soft with no palpable nodules.

Initial work-up revealed an elevated TSH (19.8 mU/L) and Free T4 of 8.4 pmol/L. Repeat studies 5 days later showed further increase of TSH (28.63 mU/L) and Free T4 (6.8 pmol/L). Anti-thyroid peroxidase antibodies and ant-thyroglubulin antibodies were negative. Pediatric Endocrinology was consulted to initiate treatment for hypothyroidism. Further investigations were required to determine the underlying etiology prior to initiating therapy .

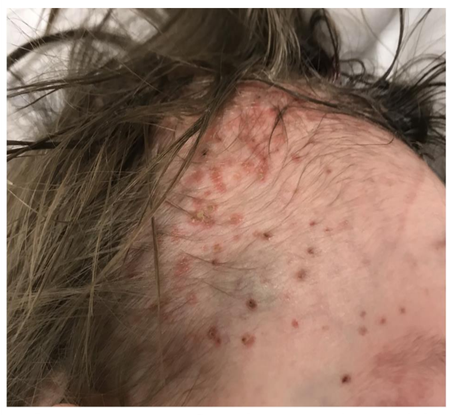

Figure 1. Scalp rash with petechial, pustular and papular morphologies

Figure 2. Periorbital edema and bruising

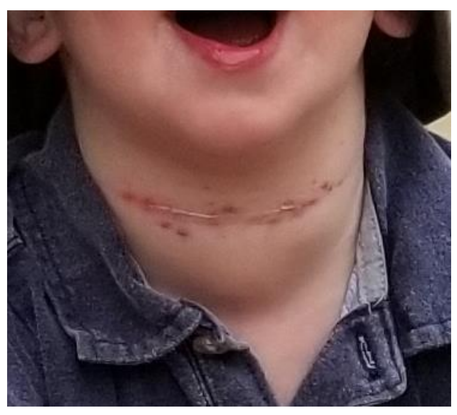

Figure 3. Prominent neck mass with overlying linear rash

IMAGING AND BIOPSY RESULTS

Initial Imaging:

A neck and thyroid ultrasound demonstrated a diffusely enlarged and hypervascular gland indicating increased metabolic activity which was atypical in a biochemically hypothyroid patient.

Biopsies:

An initial skin biopsy was non-diagnostic.

Fine needle aspiration of the neck mass (performed by Dr. Safwat Girgis) yielded the diagnosis of **Langerhans cells histiocytosis (LCH)**. No normal thyroid elements were identified. (Figure 4).

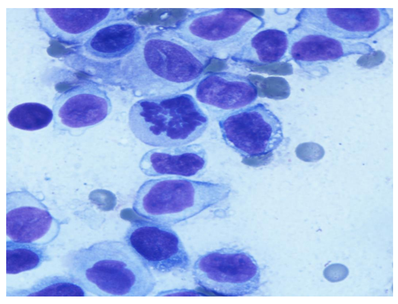

Figure 4. Pathology results of the neck mass FNA showing atypical histiocytes

CLINICAL COURSE

The patient's acquired hypothyroidism was due to LCH infiltrating the thyroid and compressing residual glandular tissue. The patient required PICU admission and was urgently treated with corticosteroids to reduce airway compression.

Management of his multi-system LCH included prednisone and weekly vinblastine. His neck mass diminished significantly. TSH normalized on levothyroxine replacement.

A repeat PET scan after 12 weeks of therapy demonstrated new metabolic activity in the pituitary stalk with mild thickening, however; he has no symptoms to suggest diabetes insipidus or pituitary insufficiency.

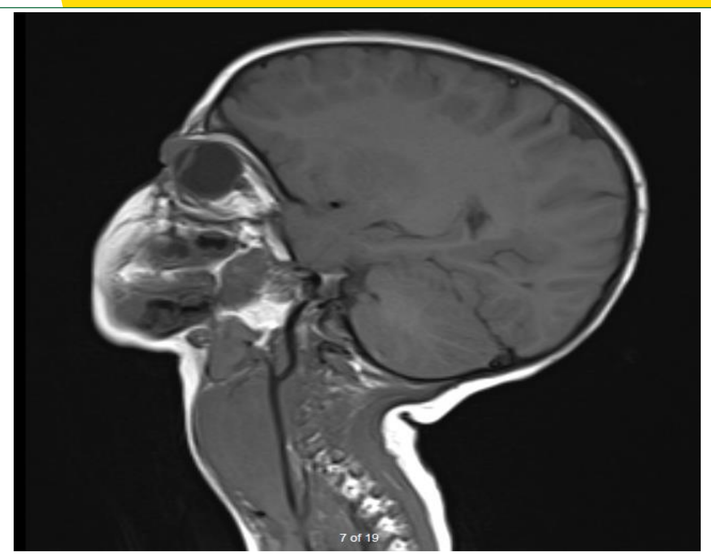

Figure 5. Sagittal MRI brain and neck demonstrating prominent neck mass

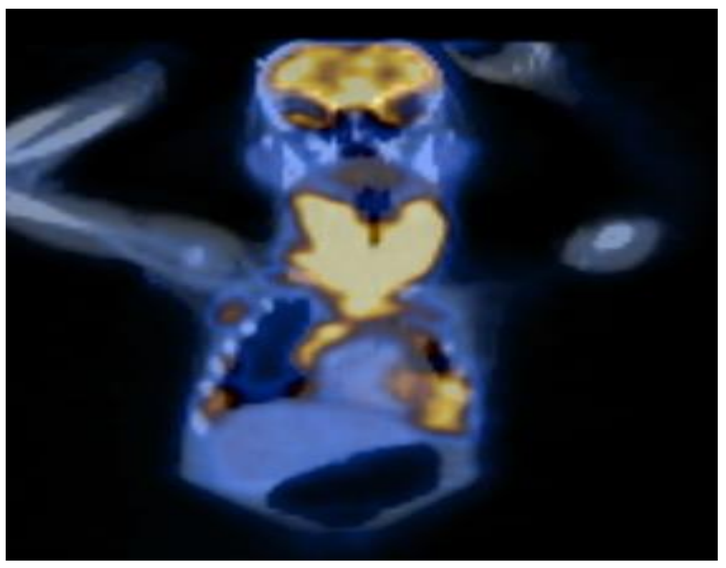

Figure 6. PET- CT demonstrating multiple foci of hypermetabolic abnormalities (scalp, left maxilla, anterior and posterior mediastinum, right axilla and bilaterally pleural spaces)

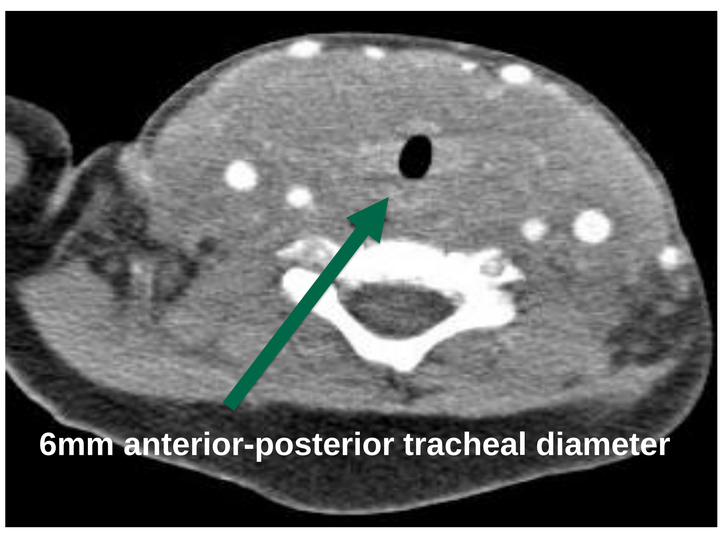

Figure 7. Axial CT neck shows a soft tissue abnormality within anterior neck, effacing and infiltrating the thyroid. There is significant mass effect on the subglottic trachea.

CLINICAL PEARLS

- Toddlers rarely present with acquired primary hypothyroidism
- Massive goiters are also rare and if present, infiltrative disorders should be considered as an etiology
- It is crucial to consider and urgently manage external compression of the trachea and neck vasculature when significant thyroid enlargement is present.

References

1. Makras et al. Endocrine manifestations in Langerhans cell histiocytosis. *Trends in Endocrinology and Metabolism.* 2007.
2. Allen et al. Langerhans-Cell Histiocytosis. *NEJM.* 2018

We extend our sincere gratitude to the patient and his family for allowing us to share his story and photographs.