4-month-old with Failure to Thrive and a Rash

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Rashes in young children are incredibly common, and while the underlying diagnoses are often straightforward, unusual cases or those that fail to respond to conventional treatment should prompt close scrutiny. We report a case of a 4-month-old female with a persistent scalp rash and a history of a milk protein allergy who was admitted for failure to thrive and suspected pneumonia.

On exam, the patient had 10 discrete, yellow, crusted papules on her scalp, several of which were hemorrhagic. There was macerated skin and several ulcerative lesions located in the neck folds and submental skin. The anterior chest had seven skin-colored, umbilicated papules. There were no lesions in the diaper area or on the extremities.

Biopsy confirmed a diagnosis of Langerhans Cell Histiocytosis (LCH). LCH is a rare, inflammatory neoplastic disorder of myeloid dendritic cells that results in infiltration of various organs of the body. Bone is the organ affected most frequently (80%), and the skull is the most commonly involved bone in all ages.[1] Other systemic manifestations include fever, lymphadenopathy, hepatomegaly, splenomegaly, or pulmonary lesions.[1] The oncogenic mutation *BRAF* V600E is found in approximately 64% of LCH cases.[2] Importantly, the cells responsible for LCH arise from the myeloid progenitor cells of the bone marrow rather than from the Langerhans cells of the skin.

Cutaneous disease is the most common manifestation of LCH in patients less than 2-years-old, however, LCH is rarely limited to the skin. Systemic involvement occurs in 87-93% of patients with cutaneous findings, and children less than 3-years-old tend to have acute disseminated multisystem disease.[1] Involvement of the liver, bone marrow, or spleen signals a worse prognosis, with 5-year survival rates less than 77%.[1] Older children and adults tend to have disease involving a single organ with survival rates nearing 100%.[1]

In young children, LCH classically presents with scaly, red-brown, seborrheic dermatitis-like papules or as an eczematous, erythematous, scaly eruption on the scalp and flexural folds. Erosions in

the flexural folds are common. LCH has a wide variety of potential cutaneous morphologies, though, and petechiae, purpura, or hemorrhagic crusts should raise suspicion. Approximately 4 to 5 children per 1 million are affected, but despite its rarity, LCH should be considered in pediatric patients presenting with atypical rashes or those recalcitrant to treatment.[3] A high clinical suspicion for systemic disease is essential in children less than 3-years-old to prevent a delay in diagnosis and expedite appropriate management.

References:

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Figure 1. Scattered skin-colored to erythematous papules, several with hemorrhagic crust on the forehead and left frontal scalp.

Figure 2. Closer image of the central frontal scalp with yellow, scaly, crusted papules with hemorrhage.

Figure 3. Red, macerated skin of the right neck with ulceration deep in the neck fold.