A 13-month-old boy with recurrent upper respiratory infections, persistent fevers, and chronic rash presented to our facility. The patient had been treated with several months of antibiotics and topical creams with no improvement. In the emergency room, he had a prominent rash (panel A), jaundice, and tachypnea. Complete blood count showed white blood cells $3.5 \times 10^3/\mu L$, absolute neutrophils $0.35 \times 10^3/\mu L$, hemoglobin $8.4 \, \text{g/dL}$, and platelets $12 \times 10^3/\mu L$. Total bilirubin was $8.5 \, \text{mg/dL}$. Systemic Langerhans cell histiocytosis (LCH) was suspected. Skin biopsy showed histiocytic infiltration (panel B) positive for CD1A (panel C). Bone marrow biopsy revealed presumed histiocytic infiltration (panel Dii) positive for CD163 and PGM1 (CD68), but negative for CD1A (panel Di).

Multisystemic LCH is a rapidly progressing illness in which Langerhans cells proliferate in various tissues. The hallmark is CD1A$^+$ as seen in the skin but not in the marrow. LCH with macrophage activation in the bone marrow has been rarely described and can exist in different levels of severity. The marrow infiltrate might represent a proliferation of cells secondary to cytokine production previously reported with LCH in younger children and associated with poor prognosis. The rash of LCH is frequently misdiagnosed. Clinicians should be aware that atypical infiltrates might be activated macrophages. The skin and the marrow infiltrate have responded to chemotherapy.
Langerhans cell histiocytosis with atypical histiocytic marrow infiltration

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