Abstract: Background. Langerhans-cell-histiocytosis is a proliferative disease of Langerhans-cells of uncertain etiology. Its low incidence and a variable clinical presentation, imitating other diseases often lead to delay of diagnosis and treatment: Case report. We report on an admission 6 weeks old boy. He had had contact to a varicella incubated child and three weeks later he suffered from a varicella-like rash. The polymorphic rash consisted of papulae, vesicles, pustules and petechiae. In addition hepatomegaly, splenomegaly, dyspnoe, fever, thrombopenia and anaemia were present. Skin biopsy revealed infiltrates of proliferating Langerhans-cells expressing CD1a and S100. Typical Birbeck-granules were shown by electron microscopy. Thus diagnosis of disseminated Langerhans-cell-histiocytosis was established. Discussion. Langerhans-cell-histiocytosis should be considered in severely affected newborns and infants demonstrating polymorphic rash. Persistent unidentified skin lesions require a skin biopsy.

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