Langerhans Histiocytosis with Mandibular Mass and Cutaneous Lesions

Ana Paula Rocha[1], Tiago Milheiro Silva[1], Rui Alves[2], Maria João Brito[1]

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Ten-month-old boy was admitted with a two-month history of millimetric reddish-violet papulovesicular lesions observed on the face, trunk and limbs (Fig. 1). A painful mandibular mass with inflammatory signs and high fever were also present. He completed two antibiotic cycles with no improvement. Spontaneous drainage of mandibular purulent discharge to the oral cavity occurred, with severe pain, edema, and rubor as well as a persistent high grade fever. Blood tests showed hemoglobin 9.8 g/dL, leucocytes 19 x 10^9 cells/L (56% neutrophils and 30% lymphocytes), sedimentation rate 26 mm/h, and C reactive protein 2 mg/dL. Facial computed tomography showed an expansive lesion on the masseter muscle and hemimandible, with bone marrow invasion and cortical disruption (Fig. 2). He was treated with penicillin and clindamycin; surgical debridement with a bone biopsy was performed and revealed lymphohistiocytic infiltrate with histiocytic cells, multinucleated giant cells, lymphocytes and sparse neutrophils, with S100+, CD1a+, CD68+, and CD163+ neoplastic cells. Skin biopsy of the lesions also showed similar histological findings with S100+ and CD1a+ neoplastic cells.

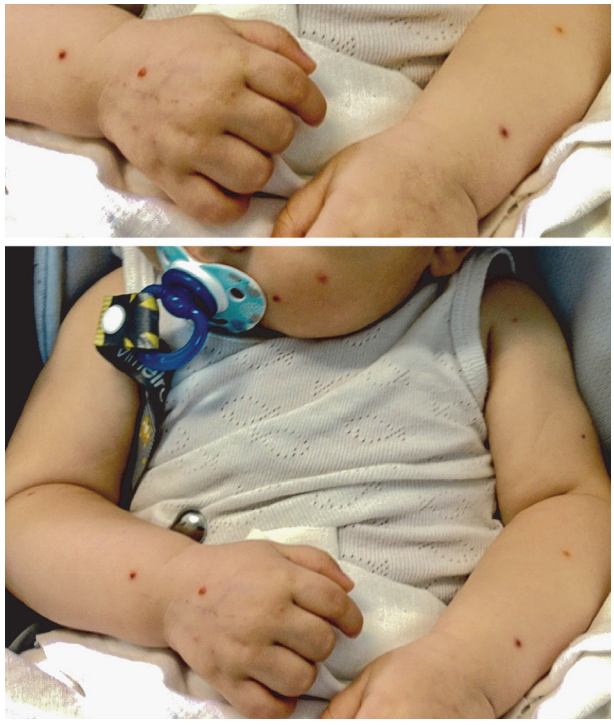

Figure 1. Reddish-violet papulovesicular lesions with a 2-month evolution.

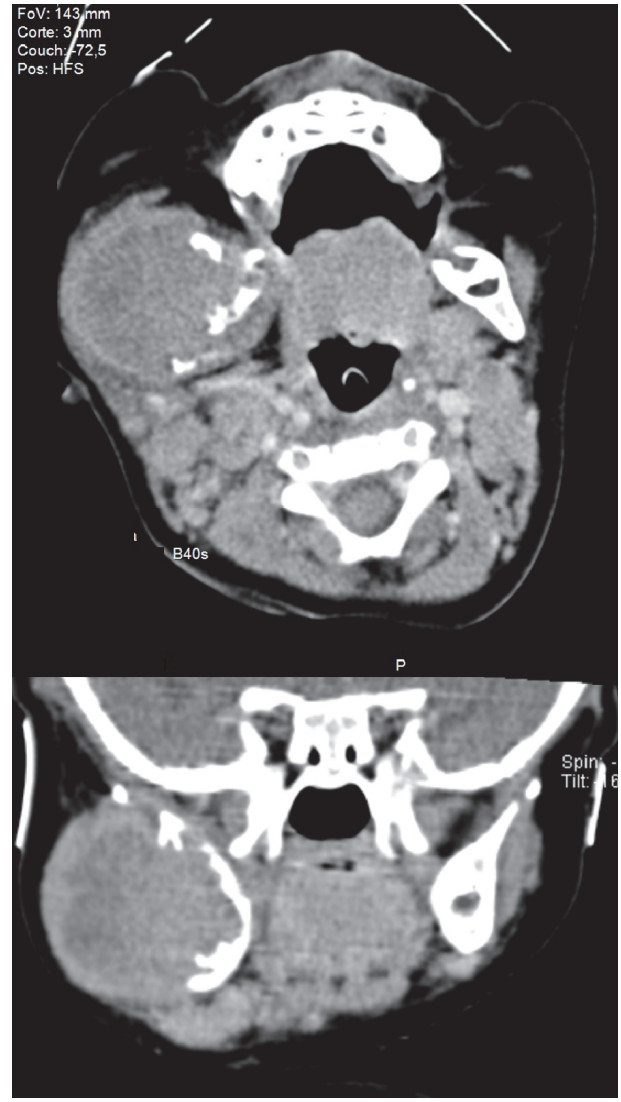

Figure 2. Facial computed tomography, soft-tissue window, coronal and sagittal images; soft-tissue expansive lesion with mandibular bone destruction.

1. Infectious Diseases Unit, Medical Pediatrics Department, Women's, Children's and Adolescent's Division, Dona Estefânia Hospital, Central Lisbon Hospital Center, Lisbon, Portugal
2. Surgery Unit, Women's, Children's and Adolescent's Division, Dona Estefânia Hospital, Central Lisbon Hospital Center, Lisbon, Portugal
Corresponding Author
Ana Paula Rocha
paulasacrocha@gmail.com
Infectious Diseases Unit, Dona Estefânia Hospital, Rua Jacinta Marto, 1169-045 Lisbon, Portugal
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Langerhans cell histiocytosis (LCH) was diagnosed based on clinical and anatomopathological criteria. Bone scintigraphy revealed foci of hypercaptation on the eighth rib, while chest radiograph and abdominal ultrasound were normal. The disease progressed while on LCH-IV 2010 protocol (new skin lesions, middle ear and skull base involvement), consequently second line therapy was started, and complete remission was achieved two months after its conclusion.

Langerhans cell histiocytosis is a clonal proliferation of cells phenotypically similar to Langerhans cells,[1-5] and may infiltrate several organs like ganglia, bone, skin, central nervous system and lung.[2,4,5] Cutaneous involvement is encountered in 40%-50% of cases, and occurs more frequently in younger children.[2,3,5] In this case, although skin lesions were the first sign of the disease, only the mandibular mass led to the diagnosis. Although rare, Langerhans cell histiocytosis should be excluded when a mandibular mass with no infectious cause occurs, especially when associated with chronic skin lesions.[1-3,5] Multisystem involvement has worse prognosis and must be excluded.[1,4]

Keywords: Infant; Histiocytosis, Langerhans-Cell/diagnosis; Histiocytosis, Langerhans-Cell/therapy; Mandibular Diseases; Skin Diseases

WHAT THIS REPORT ADDS

- Cutaneous involvement in Langerhans cell histiocytosis (present in 40% of cases) might be the first sign of the disease, especially in the young child, generally like an eczematous rash or ulcerated lesions, and it should always prompt the exclusion of other sites of involvement.
- Although rare, Langerhans cell histiocytosis should be excluded when a patient has a mandibular mass and chronic skin lesions.
- The presence of inflammatory signs may delay the diagnosis, raising suspicion of osteomyelitis; therefore, when there is no response to antibiotics, a bone biopsy is mandatory.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Consent for publication

Consent for publication was obtained.

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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