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LETTER

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A rare cause of haemorrhagic vesicular lesions in childhood: Langerhans cell histiocytosis

A 50-day-old female patient was referred to our clinic because of the complaints of a rash on the anterior aspect of body and extremities, loss of appetite, restlessness for more than 15 days, and axillary body temperature reaching 38.5°C for the last three days. Physical examination on admission revealed pathological findings such as 2/6 systolic murmur and hepatosplenomegaly (HSM) palpable 7 cm below the ribcage. In the dermatological examination, there were crusted vesicular lesions, some of which were haemorrhagic, in the anterior aspect of the body, genital area, and palmoplantar region and gingival ulcers on the oral mucosa (Figure 1). In the laboratory examination, white blood cell count was 7400/μL, haemoglobin level 7.7 g/dL, platelet level 128 000/μL, ALT level 52 U/L, AST level 69 U/L and C-reactive protein level 27 mg/L. There were no lymphopenia and neutropenia. No additional findings were found except HSM in the ultrasonographic examination of the abdomen. Serology examination of cytomegalovirus, herpes, varicella, toxoplasma and Ebstein-Barr virus regarding HSM, fever and cytopenia aetiology were found to be negative. There was no reproduction in the blood cultures. Respiratory viral panel PCR sent to detect whether there was an enteroviral disease was found to be negative. Punch biopsy from the lesions on the anterior of the body was performed, and histopathological examination of the lesional skin revealed infiltration consisting of large eosinophilic cytoplasm, irregular and oval-shaped histiocytic cells, some with double or lobulated nuclei, forming slots in the diffuse pattern in the upper dermis. CD1a was detected as surface antigen (+) and S100 protein (+) in immunohistochemical examination (Figure 2). The patient was diagnosed with Langerhans cell histiocytosis with the existing findings. There were no pathological findings in the peripheral smear and bone marrow biopsy performed for systemic involvement, and no bone involvement was found on radiological examination.

Langerhans cell histiocytosis (LCH) is a rare disease characterised by clonal neoplastic proliferation of Langerhans-type cells.[1,2] The true incidence of LCH is unknown. Although it is seen in all age groups, it is most commonly diagnosed in children between the ages of 1-3 years.[1,3,4] Approximately 70% of childhood cases have a single system involvement. The skeletal system is the most commonly involved. Cases with multiple system involvement are frequently under the age of 2 years.[2,5] Skin involvement and hepatosplenomegaly were detected in our patient.

The skin is the second most frequently involved organ with a frequency of 30%-60%. LCH cases presenting only with skin involvement are around 10%. The most common skin findings are brownish-purplish papules and an eczematous rash resembling candida infection. Also, pustular, purpuric, petechial, vesicular and papulo-nodular lesions may develop.[1,4,5] Our patient had haemorrhagic crusted vesicular lesions on the anterior of aspect the body and the palmoplantar region.

Brown-purplish papules can develop anywhere on the body in infants. The most common findings in the oral area are intraoral mass, gingivitis, mucosal ulcers, and tooth loss.[2,5] In our case, a gingival ulcer was seen in the oral mucosa. Erythematous vesiculopustules are the most common lesion in the neonatal period and can be confused with herpetic infection or chickenpox.[1,3] Our patient was also diagnosed with chickenpox in another centre.

The diagnosis of LCH can be confidently made with a combination of clinical signs, histological findings, and immunohistochemical staining for S100 protein, langerin and CD1a.[1] The diagnosis of our case was made by histopathological and immunohistochemical examination of the lesional skin.

The prognosis is generally very favourable in LCH with only skin involvement. Since the disease can turn into progressive disease in ~5% of babies with only skin involvement, this group of patients should be closely monitored.[2,5]



FIGURE 1 A, Crusting vesicular lesions, some of which are haemorrhagic, in the anterior aspect body, genital area, and (B, C) palmoplantar region and (D) gingival ulcer on the oral mucosa

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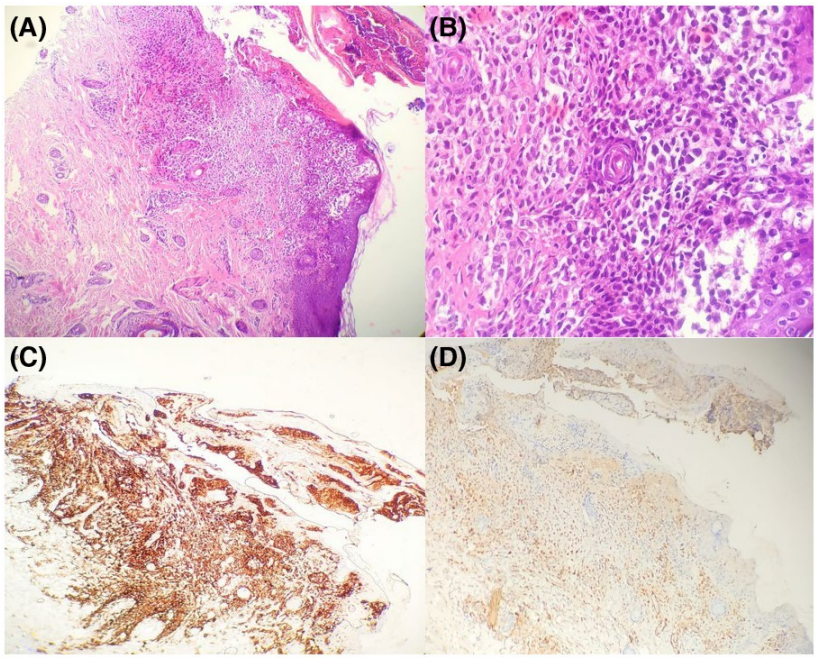


FIGURE 2 A,B Histopathological examination of the lesional skin revealed histiocytic cells with large eosinophilic cytoplasm forming slots in the diffuse pattern in the upper dermis, irregular and oval shape, some with double or lobulated nuclei were observed. (HEX10, HEX40) In immunohistochemical examination (C) CD1a surface antigen positive (X10) and (D) S100 protein-positive (X10) were detected

In conclusion, Langerhans cell histiocytosis, which is a differential diagnosis of herpetic infections or chickenpox, should be considered in children with crusting vesicular lesions on the body, and necessary tests should be performed in terms of systemic involvement.

DISCLOSURES

The authors declare no conflict of interest.

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