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Clinical Image

Multisystem involvement of Langerhans cell histiocytosis in an adult

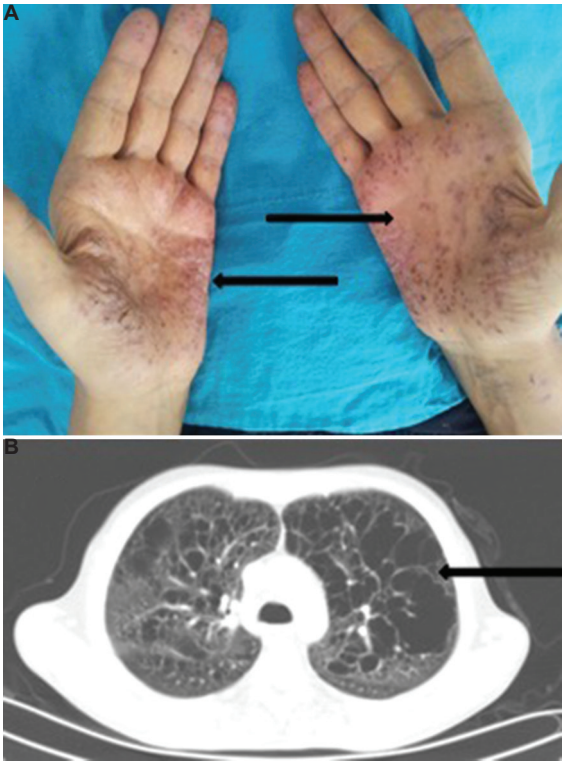

Fig. 1. (**A**) Erythematous-vesicle-squamous-crusty lesions on palms (arrows). (**B**) Computed tomography chest showing bullous emphysematous lesions in both lungs (arrows).

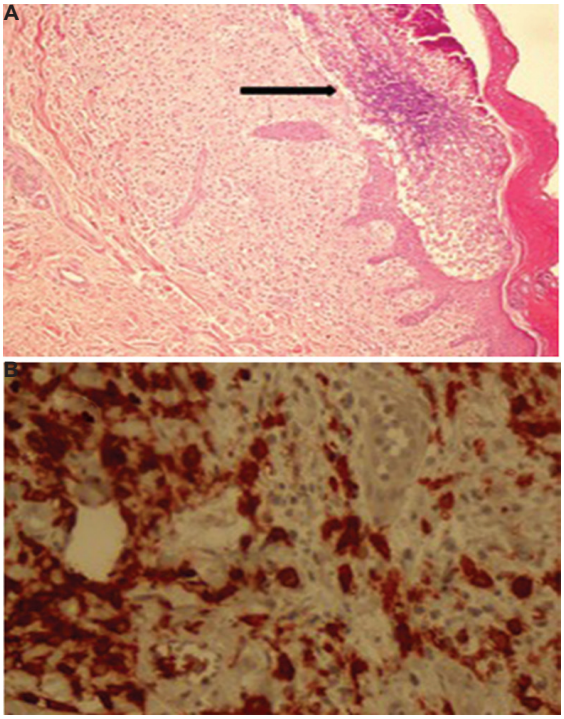

Fig. 2. (**A**) Epidermal hyperkeratosis with intra- and subepidermal pustules (arrow); and histiocytes, eosinophilic infiltration of inflammatory cells containing polymorphs and congested capillaries with extravasated red blood cell (H and E, ×100). (**B**) Positive immunohistochemistry for antigen CD1a (H and E, ×CD1a, ×400).

A 27 yr old man† admitted to the Internal Medicine department of Haseki Training and Research Hospital, Turkey, in January 2015 with complaints of painful, itchy, red papules on his palms and soles for the past four months. He had diabetes insipidus in his medical history. Physical examination revealed diminished breath sounds and hepatomegaly. On his palms and soles erythematous-vesicle-squamous-crusty, lesions were seen (Fig. 1A). Aspartate aminotransferase: 587 U/l, alanine transaminase: 291 U/l, alkaline phosphatase (ALP): 696 U/l, gamma-glutamyl transferase (GGT): 1004 U/l, total bilirubin: 26 mg/dl, direct bilirubin:13 mg/dl were measured in the laboratory. Computed tomography findings reveal pleural thickening in the right hemithorax and bullous emphysematous lesions in both lungs, the largest measured 8 cm (Fig. 1B). Magnetic resonance cholangiopancreatography findings were compatible with sclerosing cholangitis. A tube was inserted endoscopically to the bile duct. The punch biopsies were taken from petechial lesions on the hands. Biopsy sections revealed epidermal hyperkeratosis with intra- and subepidermal pustules and at the base of pustules, histiocytes, eosinophilic infiltration of inflammatory cells containing polymorphs were seen.

†Patient's consent obtained to publish clinical information and images.

Histiocytic cells had a positive reaction with CD1a and it was consistent with the Langerhans cell histiocytosis group disease (Fig. 2A and B). After ursodeoxycholic acid and bile duct tube treatment, the value of the ALP, GGT and bilirubin decreased. The treatment consisted of systemic chemotherapy with vinblastine and prednisone. The patient died because of the respiratory failure.

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***Conflicts of Interest*:** None.

Nilay Sengul Samanci[1,*] & Mesut Ayer[2]
[1]Department of Medical Oncology, Istanbul University Cerrahpasa Medical Faculty &
[2]Department of Hematology, Haseki Training & Research Hospital, Istanbul 34098, Turkey
[*]*For correspondence:*
nilaysengulsamanci@gmail.com

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