UNILATERAL SELF-REGRESSING BLISTERING ADULT LCH (Langerhans Cell Histiocytosis)

- Chaidemenos Georgios, MD, Dermatology, Thessaloniki, Greece
- Kontochristopoulos Georgios, MD, Dermatology, Athens, Greece
- Zarabukas Thomas, MD, Histopathology, Thessaloniki, Greece
- Spiliopoulos Theofanis, MD, Dermatology, Patras, Greece

NO CONFLICT OF INTEREST EXISTS FOR THIS PRESENTATION
CASE REPORT

• A 53 year-old woman was presented with a one-week history of a slightly itchy eruption of the left body side from the axilla to the shin (Fig. 1). It was consisted of red-to-violaceous papules coalescing in plaques at some areas. Small round and digitiform blisters prevailed on left belly area (Fig. 2). A few punched-out ulcers were covered with hemorrhagic crusts in axilla and sub-mammary region (Figs 1, 2). On the left leg and shin, the lesions had a ringed, purpuric appearance (Fig. 3). Contact non polarized dermoscopy disclosed oval lesions with clear center and amorphous orange-red periphery, manifesting small mainly dotted, vessels (Fig. 4). The eruption was unresponsive to IM injection of Betamethasone.
Fig. 1. Involvement of left fronto-lateral torso.
Fig. 2. Papules and vesicles of diverse size and shape, coalescing in plaques. Hemorrhagic crusts cover round and oblong ulcers.
Fig. 3 Purpuric, lichenoid, ringed lesions on left shin
Fig. 4. Non-Polarized Dermoscopy: Red to orange-red amorphous borders of oval lesion. Small dotted vessels are noticed, mainly on left area of the picture.
EXAMINATIONS & COURSE

• Histology showed a spongiotic epidermis with lymphocytic exocytosis (Fig. 5). CD1a positive Langerhans cells were present in the blisters and the papillary dermis (Fig. 6). Chest, Skull and long bones X-ray was normal. Hematological investigation values were in normal range.

• The eruption reached its peak in 30 days to follow a gradual disappearance thereafter. At nine months FU, the patient remains in excellent general health without any residual skin lesion (Fig. 7).
Fig. 5. Small crust with few neutrophils (H+EX100)
Fig. 6 Immunostain CD1a reveals Langerhan’s cells and small intraepidermal vesicle with Langerhan’s cells (Immunostain X200)
Fig. 7. Only the scar of biopsy is seen at two months’ FU
DISCUSSION

• The spectrum of Langerhans’ cell histiocytosis (LCH) refers to a group of rare disorders of unknown origin with diverse outcomes and clinical presentations. The disorder is characterized by Langerhans cells typified by Birbeck’s granules with EM or CD1 antigen labelling(1).

• Although atypical forms of LCH exist, blistering eruption is very rare in adults(2,3). A case of LCH presenting with brown lichenoid patches on one foot, closely similar to the purpuric appearance of the lesions of our patient’s left shin, has been reported(4).
DISCUSSION (Contd)

• Self-healing reticulohistiocytosis exist in children but to our knowledge, no case of unilateral blistering disease that self-regressed in an otherwise healthy adult, has so far been reported. However, cutaneous involvement may exist for many years in adults before the onset of visceral disease(5).

• This case illustrates the wide clinical spectrum and variable course of LCH
REFERENCES


