Chronic paronychia with subungual purpura

A 24-year-old man was apparently well till 4 years ago, when he developed sudden onset of breathlessness and was diagnosed to have bilateral pneumothorax. Six months after the episode, he developed skin lesions which started as itchy red papules and pustules on the scalp, axilla and perianal region. The nails were involved 1 year after the skin lesions. The affection started as painful swelling of nail folds associated with nail destruction [Figure 1]. The nail changes including plate thickening, distal splitting, elkonyxis, onycholysis and subungual hemorrhage, were confined to the finger nails sparing the toe nails. The patient also had history of recurrent painful oral erosions of 2 years duration with loss of the lower teeth. He also had intermittent watery discharge from both ears. There was elbow and knee pain of 6 months duration. There was no history suggestive of bowel disturbances, weight loss, eye complaints or diabetes insipidus.

Dermatoscopy was performed in all nail units using a handheld dermatoscope which gives a 10 × magnification (Dermlite DL3, 3Gen Inc., USA) in polarized mode using ultrasound gel as interface fluid and images were taken with a Sony Cybershot DSC-W800 20.1 MP digital camera. Subungual purpura [Figure 2] was present only in the left middle finger along with the above nail findings. The left little finger showed nail plate thickening and elkonyxis. The rest of the nails showed subungual hemorrhages, onycholysis, distal splitting and longitudinal grooving [Figure 3]. Nail matrix and nail bed biopsy was performed from the left middle finger nail unit which showed CD-1a positive clusters of large cells with indented reniform nuclei with eosinophilic cytoplasm along with lymphocytes and few plasma cells [Figure 4].

What is Your Diagnosis?

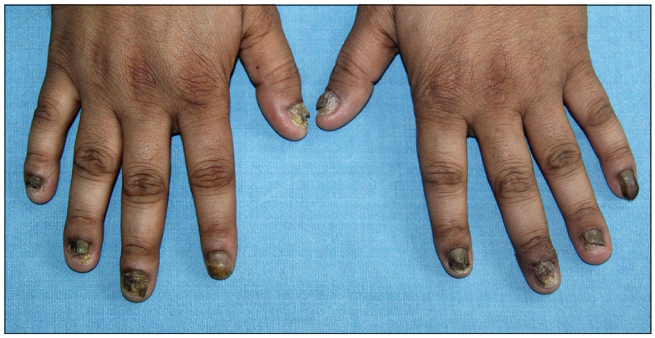

Figure 1a: Finger nails

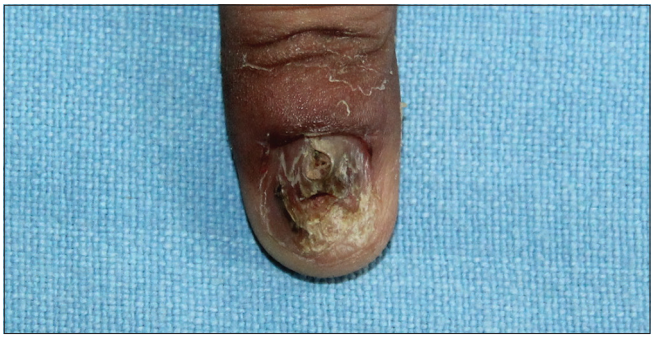

Figure 1b: Biopsied left middle finger nail

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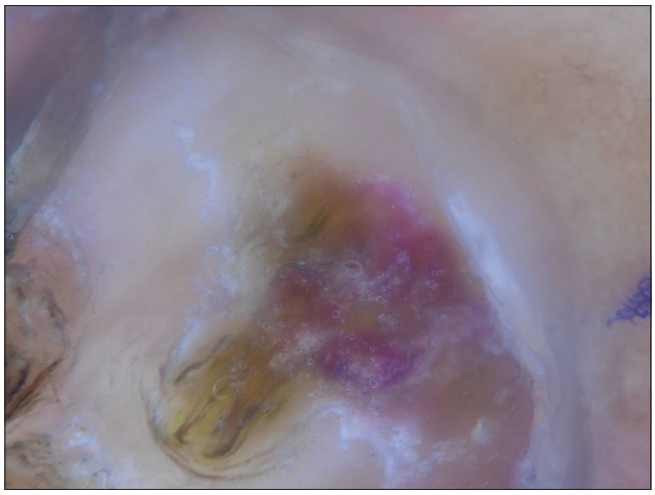

Figure 2: Dermatoscopy of left middle finger nail unit showing subungual purpura (polarized, ×10)

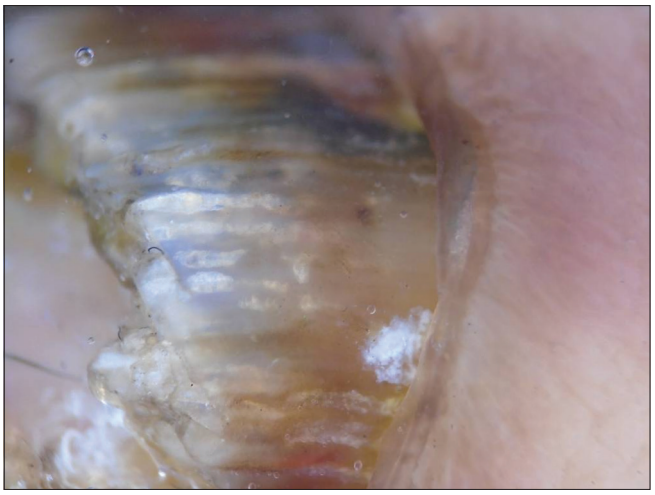

Figure 3: Dermatoscopy of nail unit showing onychodystrophy, nail plate discoloration and longitudinal ridging(polarized, ×10)

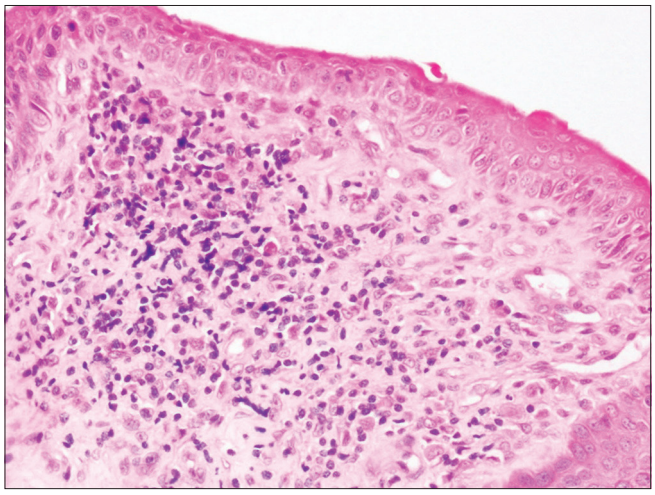

Figure 4a: Histopathology of nail matrix showing focal dense superficial infiltrate consisting of histiocytes with abundant eosinophilic cytoplasm admixed with lymphocytes (hematoxylin and eosin, ×200)

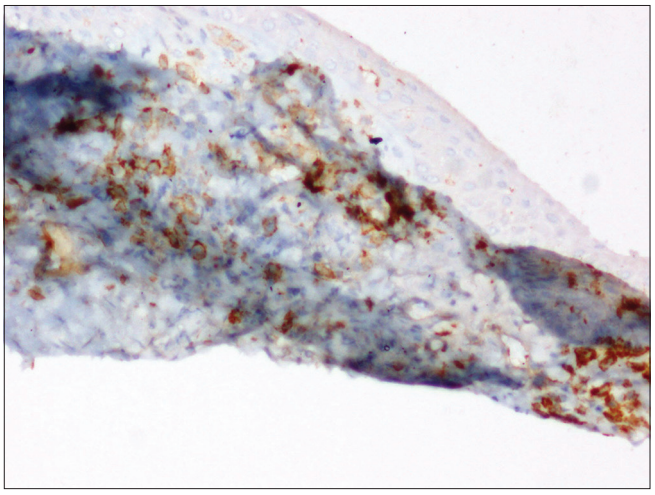

Figure 4b: Histopathology of nail matrix showing CD-1a positivity (CD-1a, ×200)

Answer

Langerhans cell histiocytosis.

Langerhans cell histiocytosis is a reactive multisystem disorder involving skin, bone, lymph nodes, pituitary, lung, liver, gastrointestinal tract and central nervous system.[1] Nail involvement is rare and is a possible pointer toward unfavorable prognosis and multisystem disease.[2]

Nail involvement in Langerhans cell histiocytosis includes:

- Nail plate: onychogryphosis, nail pitting, elkonyxis, longitudinal grooving, onychoschizia, onychorrhexis, onychomadesis, onycholysis
- Nail fold: chronic paronychia, paronychial erythema
- Nail bed: subungual purpura, subungual hemorrhage, subungual pustules, subungual hyperkeratosis.[3]

Subungual purpura along with paronychia are considered to be specific for Langerhans cell histiocytosis.[4] Polarized dermatoscopy confers its utility in detecting specific nail bed changes of Langerhans cell histiocytosis-like subungual purpura which can be missed by naked eye examination and are evident only with the aid of a dermatoscope. It is also useful to choose the nail unit that shows these changes for nail bed and matrix involvement and hence increases the yield of the biopsy in detecting Langerhans cells. In our patient, we did the biopsy of the left middle finger after finding subungual purpura with the dermatoscope, which is considered quite specific for Langerhans cell histiocytosis along with other features. This case is reported as dermatoscopy of nail unit in Langerhans cell histiocytosis and its utility is yet to be published in the literature.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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