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PUZZLING SKIN AND NAIL MANIFESTATIONS IN A CHILD

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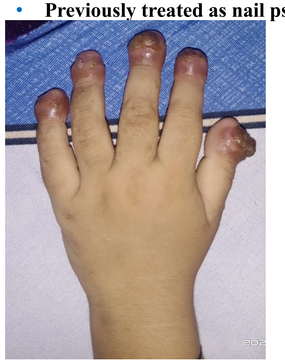
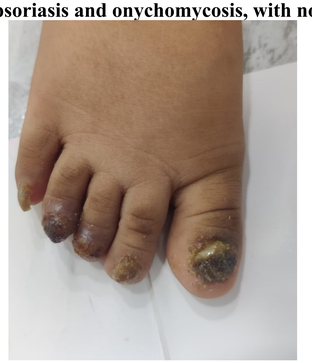
INTRODUCTION

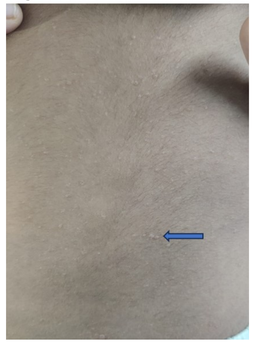
- Langerhans cell histiocytosis (LCH) is a spectrum of disorders classified by the number of organs involved and the presence of organ dysfunction. Children under 2 years with multisystem disease have significantly higher mortality.
- Low-risk organs include the skin, bones, lymph nodes, and pituitary gland, while high-risk organs include the bone marrow, liver, and spleen. Although lungs were once considered high-risk, recent studies no longer support this.
- Nail involvement in LCH is extremely rare, with only 22 cases reported worldwide and often associated with multisystem disease and poor prognosis

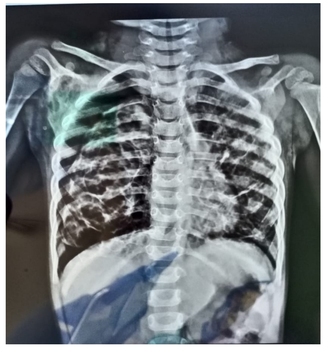
CASE REPORT

- 2-year-old male, non-consanguineous parents, full-term normal delivery, immunised as per schedule, normal developmental milestones.
- 7-month h/o painless swelling and nail plate loss of fingers and toes
- Started on right fingers → other hand in 15 days → toes after 1 month
- Associated with intermittent fever, breathlessness, failure to thrive and loss of appetite
- Treated multiple times for suspected pneumonia without lasting relief
- No significant family or past history
- Previously treated as nail psoriasis and onychomycosis, with no improvement









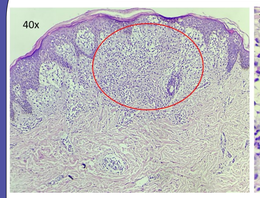
- Erythematous bulbous swelling of nail bed of all fingers with thick, adherent haemorrhagic & honey coloured crusting associated with onycholysis in few nails
- Palmar and plantar surface of all finger tips & toes appeared normal
- Multiple, discrete, skin coloured to hypopigmented, tiny papules noted over dorsal surface of B/L hands & anteroposterior trunk
- Systemic examination : Tachypnea, hepatomegaly +
- Differential diagnoses: Epidermolysis Bullosa with nail involvement, Langerhans cell histiocytosis, chronic persistent paronychia

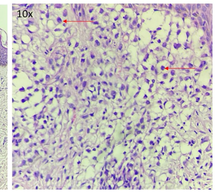
INVESTIGATIONS

- Hemogram : Anemia, leukocytosis, thrombocytosis
- ESR: 35 mm/hr, ALP: 569 IU/L, S. Albumin: 3.6 g/dL, TSH: 90 µIU/mL (↑), FT4: 0.6 ng/dL → Suggestive of hypothyroidism
- Chest X-ray: Right pneumothorax, bilateral cavitations (more on right), patchy fibrosis
- FDG-PET CT:

➢ Hypermetabolic skin/subcutaneous lesions (fingers, toes), Hepatomegaly, periportal thickening, Bulky thyroid with diffuse lesions, Right lung collapse, large pneumothorax, mediastinal shift, Bilateral cystic lung changes, Lytic bone lesions (skull base, right scapula)

HISTOPATHOLOGY

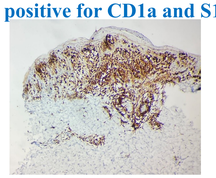




Histopathology from papule over back: Lichenoid infiltrate of histiocytes with large histiocytes with abundant amphopilic cytoplasm and kidney-shaped nuclei, epidermal hyperplasia, papillary dermal edema

Molecular: BRAF V600E mutation negative

IHC positive for CD1a and S100

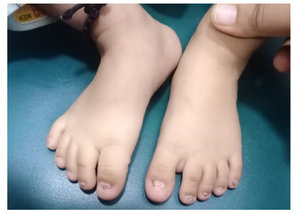


Final Diagnosis- Multisystem LCH with Twenty-nail dystrophy and LCH-induced Hypothyroidism

TREATMENT

- Right pneumothorax relieved with internal catheter.
- Started on High-risk LCH protocol:
  - IV Vinblastine (6 mg/m² weekly)
  - Oral Prednisolone (40 mg/m²/day x 4 weeks)
  - Oral Etoposide (50 mg/m²/day x 3 weeks)
- Pneumocystis pneumonia prophylaxis: Cotrimoxazole (5 mg/kg/day on weekends)
- Levothyroxine 50 mcg daily for hypothyroidism
- Developed fungal pneumonia → Treatment paused for 10 days
- Revised Treatment Approach (due to high risk):
- Considerations: Severe lung damage, multisystem involvement, fungal pneumonia, risk of DI
- Shifted to targeted therapy with:
  - Oral Trametinib (MEK inhibitor) at 0.025 mg/kg/day
- Outcome - Complete resolution by 6 months post-immunotherapy (as seen in photos)

POST 6 MONTHS TREATMENT



CONCLUSION

- Nail involvement in LCH is extremely rare and often associated with skin lesions and multisystem disease. It may serve as a poor prognostic indicator
- Common nail changes include onycholysis, subungual hyperkeratosis, nail dystrophy, paronychia, haemorrhagic crusts and purpuric striae
- Nail bed is the primary site of Langerhans cell infiltration. Histology mirrors cutaneous LCH with $CD1a^{+}/S100^{+}$ cells infiltrating the nail unit
- Nail involvement in LCH warrants careful evaluation as it reflects systemic disease burden and requires timely diagnosis and management

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- The authors declare that they have no conflict of interest

- References:

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