Poster ID 32

Ichthyosiform Sarcoidosis with Thrombocytopenia in a Child

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Introduction

Sarcoidosis chronic granulomatous multi-system disease

- Pulmonary
- Ocular
- Cardiac
- Neurological
- **Cutaneous**
  - **Ichthyosiform lesions**
    - Rare
    - Specific
    - 95% systemic association
    - No reports in children
- Musculoskeletal
- Gastrointestinal
- Renal
- Hematological
  - **Thrombocytopenia**
    - Extremely uncommon
    - <100 cases reported

Discussion

Cutaneous involvement :25-30% cases. Many specific manifestations:

- Lupus pernio
- Infiltrated plaques
- Macular
- Papular
- Ichthyosiform
  - Precedes systemic involvement
  - Prognosis unaffected
- Subcutaneous nodules
- Hypopigmented patches
- Alopecia
- Verrucous lesions

Thrombocytopenia in sarcoidosis: 3 mechanisms hypothesized

- Hypersplenism
- Granulomas in bone marrow
- Auto-immune thrombocytopenic purpura (ITP).
  - Occur after onset of systemic disease
  - More common in dark-skinned patients
  - Poorer prognosis

Systemic corticosteroids ± IVIg: first-line treatment in systemic sarcoidosis, with significant response as seen in our case.

- Ichthyotic lesions improved significantly: unlike previous reported cases
- Hematologic : gradual and modest improvement, similar to previous reports

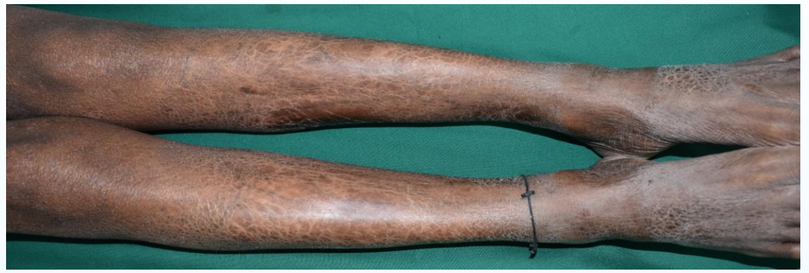
Conclusion

- All cases of acquired ichthyosis should be seriously evaluated
- Extensive systemic evaluation of all cases of ichthyosiform sarcoidosis is warranted

Case Report

Clinical

11 year-old boy presented with generalised cutaneous atrophy and hyperpigmented scaling over the face, trunk and distal extremities for last 6 months



Recurrent high-grade fever with arthralgia, myalgia, thrombocytopenia x 2 years

Generalised lymphadenopathy

Hepatosplenomegaly

Panuveitis

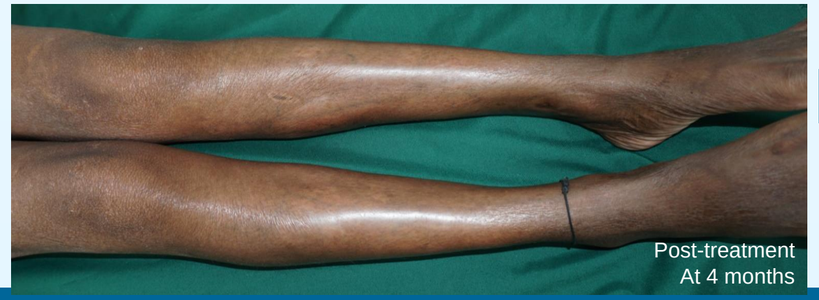
No response to ATT

Laboratory Investigations

- **CBC: Hb: 9.6g/dL, Platelet: 42,000/µL**
- **ACE : 316 IU/L (8-52)**
- **Mantoux test- Negative**
- **LN biopsy: granulomatous lymphadenitis**
- **BM biopsy: normal**
- **CECT Chest, Abdomen: ground glass opacification, septal thickening, cardiomeglly, hepatosplenomegaly, lymphadenopathy**
- **PET-CT: diffuse sytemic uptake, periarticular uptake**

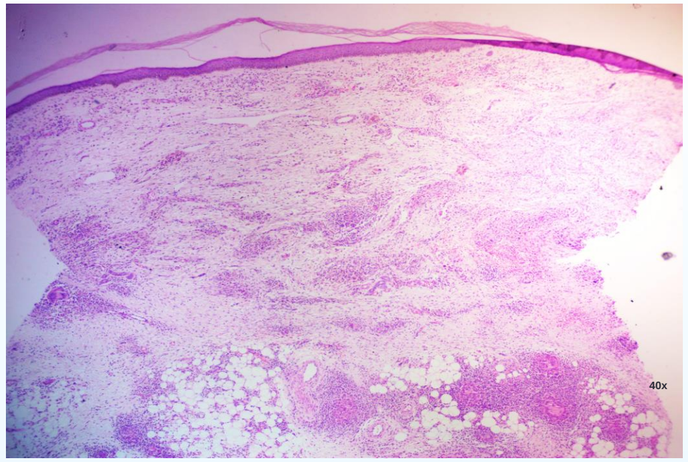
Treatment

Oral prednisolone 1mg/kg, with IVIg 2g/kg

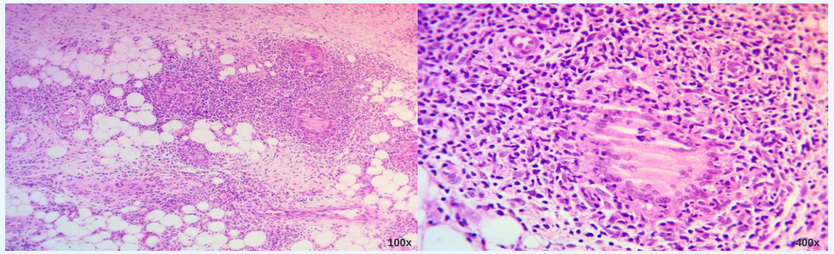


Histopathology

Skin biopsy from ichthyotic region over leg



Atrophic epidermis and multiple granulomas in the lower dermis and subcutis



Epithelioid-cell granulomas with giant cells (Langhans and foreign body type) with lymphocytic cuff

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