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ICHTHYOSIS AND ICHTHYOSIFORM DISORDERS

- Ichthyoses are a heterogeneous group of dermatoses characterized by the presence of fishlike scales (ICHTHYS)

- Scaling is generally worse in winter.

- Ichthyotic disorders are usually inherited but may be acquired

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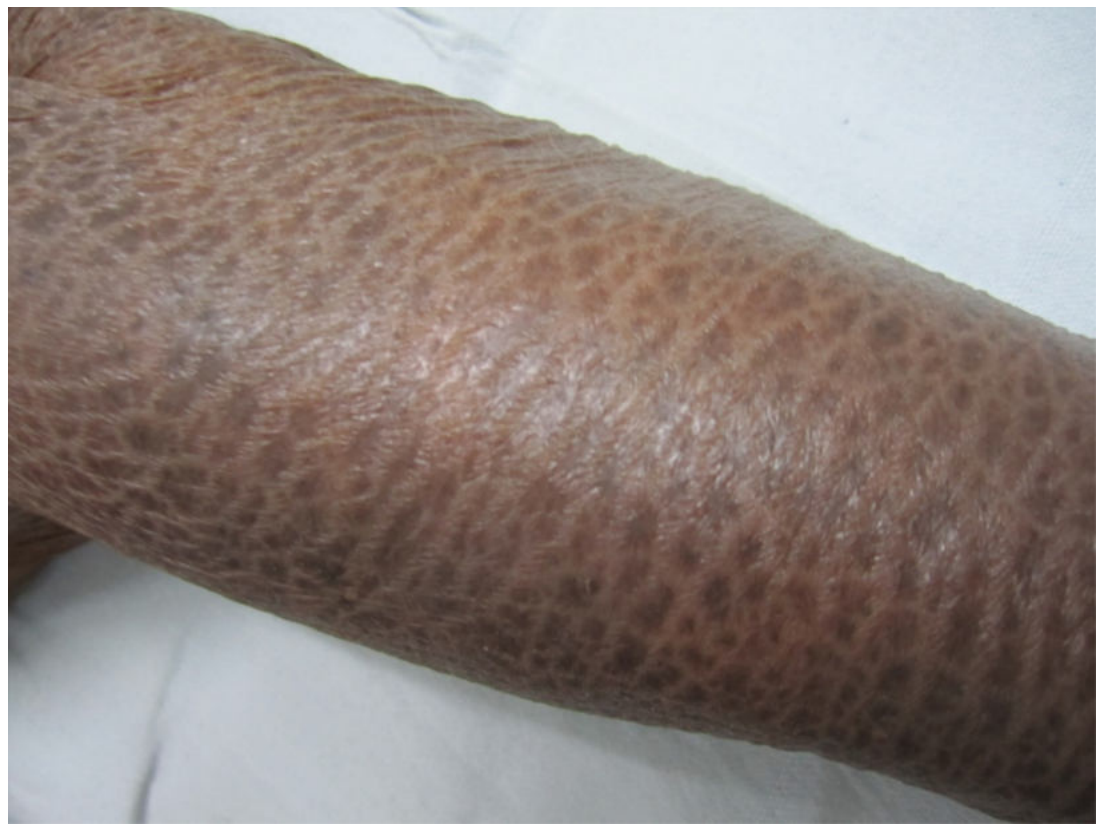

- Scale: flat plate or flake of stratum corneum

Congenital
Ichthyosis vulgaris
X-linked ichthyosis
Lamellar ichthyosis
Nonbullous ichthyosiform erythroderma
Epidermolytic hyperkeratosis

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ICHTHYOSIS VULGARIS

- Etiology

✓Inheritance: AD.

✓Molecular defect: Reduced or absent filaggrin3.

- Epidemiology

✓Prevalence: Common disorder (incidence of 4/1000).

✓Age of onset:3–12 months.

✓Gender predilection: Males = Females.

- Character of scales: On most parts of body: Fine, white scales.
- On extensors of lower extremities (most severely affected parts): Large scales, attached (pasted) at center and turned up at the edge, making skin rough. Superficial fissuring may develop on shins in winter.

- Distribution: Extensors of limbs, lower back. Flexures spared

- Associated features: Keratosis pilaris, hyperlinear palms and soles, atopic diathesis

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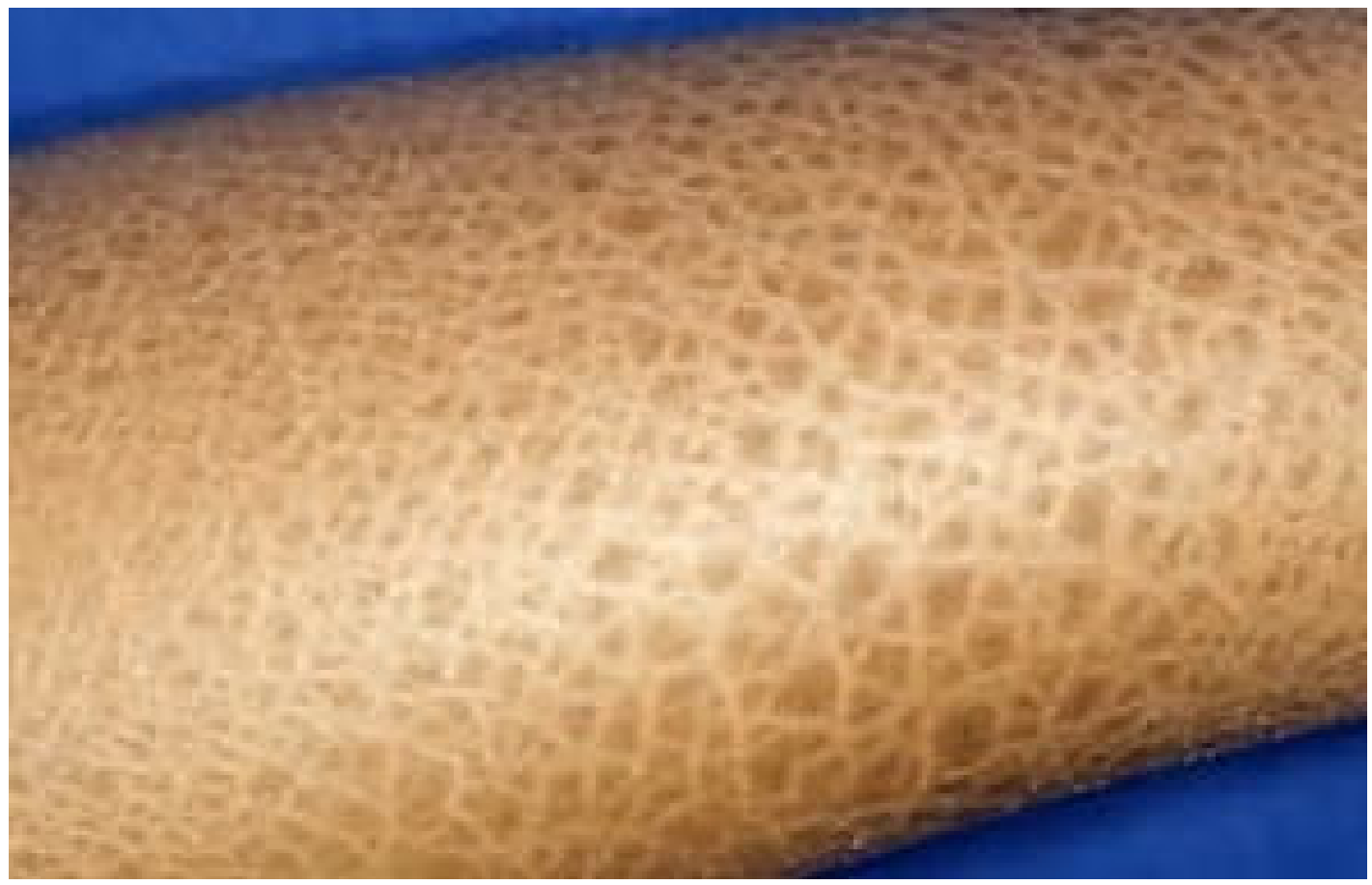

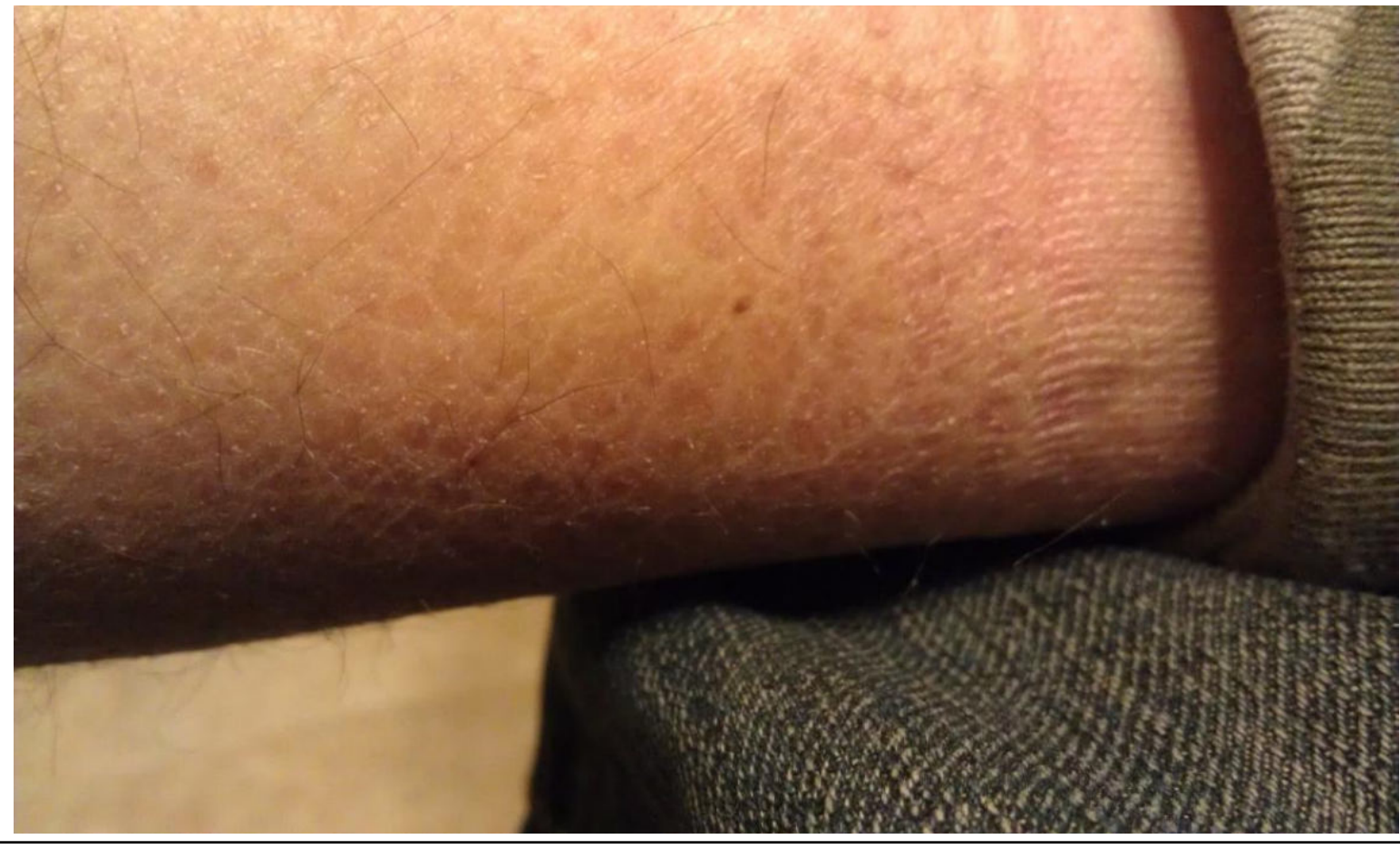

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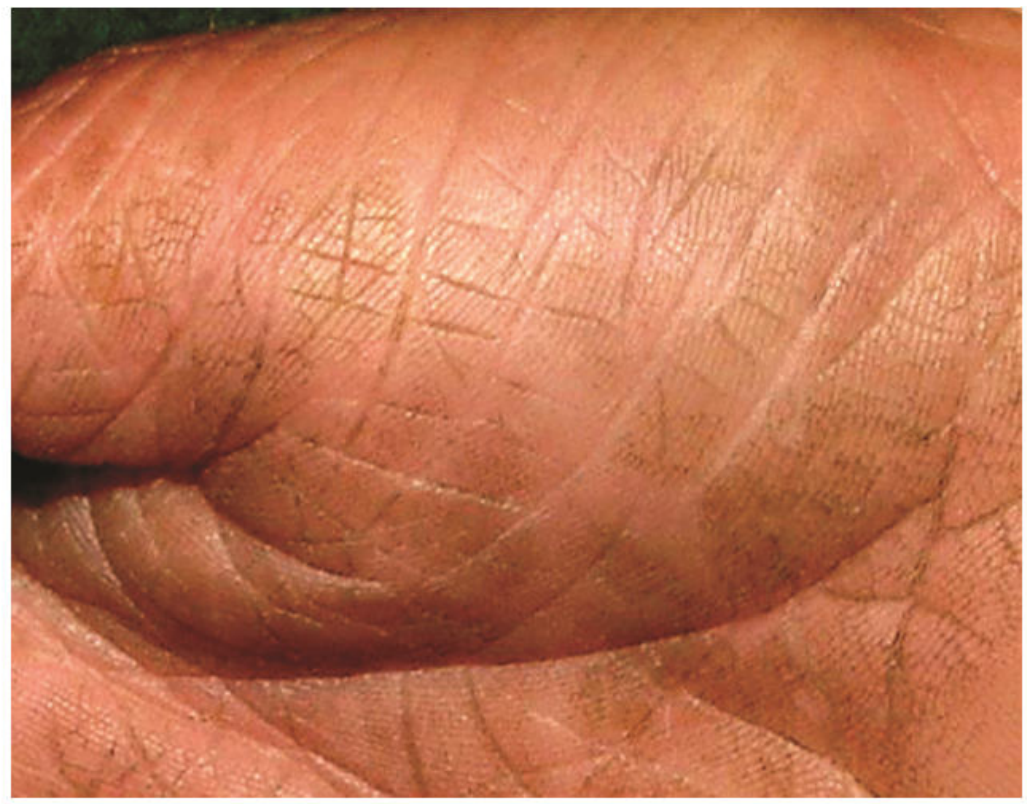

Fig. 3.4. Hyperlinear palms: accentuated skin creases on the palms.

- Investigations- None needed
- Skin biopsy:
- Hyperkeratosis with Hypogranulosis or absent granular layer

- Diagnosis : CLINICAL

➢Points for diagnosis : Diagnosis of IV is based on the presence of: Scales, which are generally fine (white) but are larger and pasted on the shins. Characteristic distribution on extensors with conspicuous sparing of major flexures. Associations: Hyperlinear palms, keratosis pilaris and atopic diathesis.

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- Treatment: USUALLY REQUIRED IN WINTERS
- Hydration (by immersing in water) and immediate lubrication (petrolatum; urea+glycerine+water) of skin.

- Use keratolytic agents (hydroxy acids, propylene glycol, and salicylic acid) when severe.

X-LINKED ICHTHYOSIS

- Etiology
- Inheritance: XLRI .
- Molecular defect: Deficiency of enzyme steroid sulfatase.
- Epidemiology Prevalence : 1: 5000 males. Age of onset: At birth. Gender predilection:Affects only males

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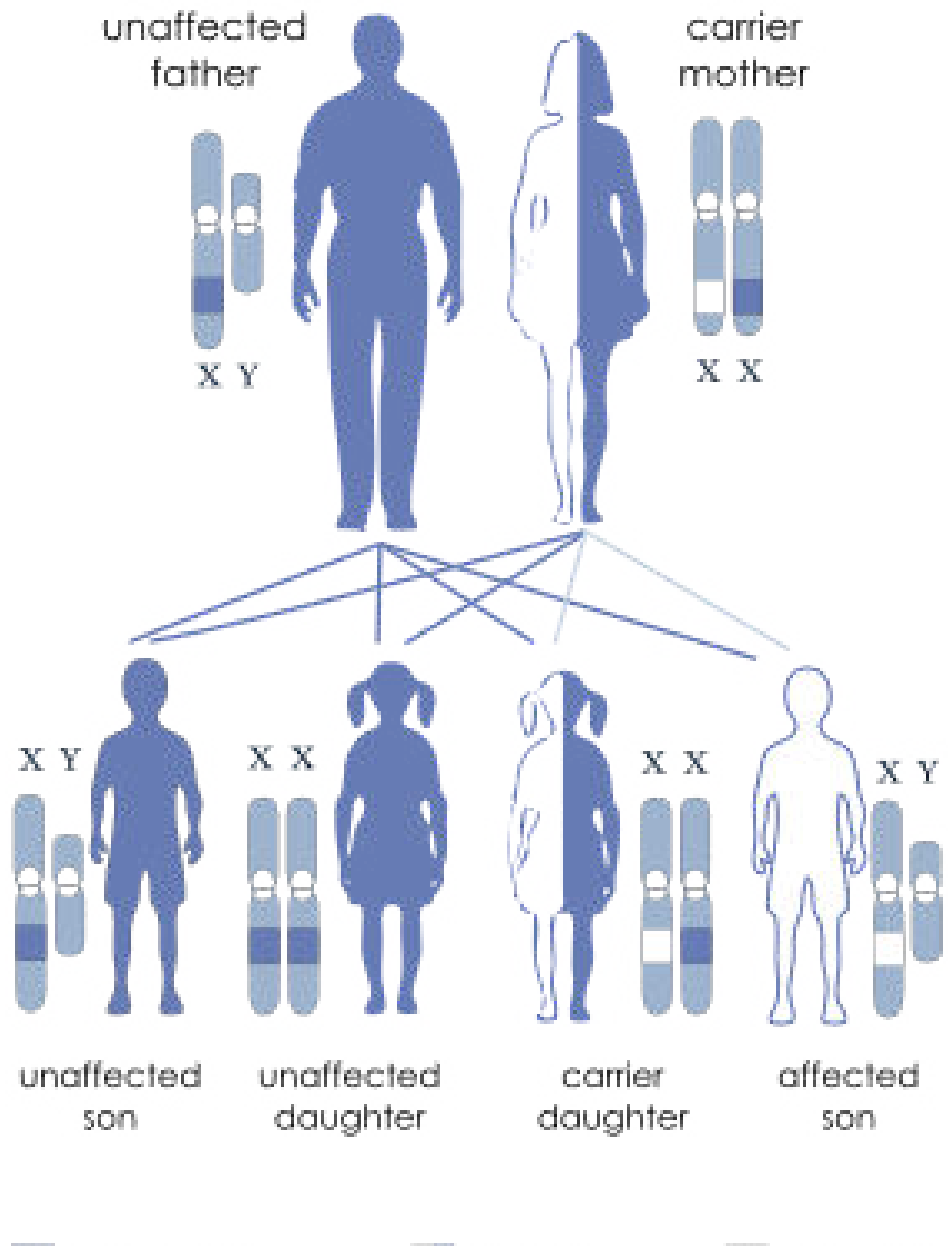
X-linked recessive inheritance



- Character of scales: Large, adherent and brown (sometimes almost black

- Sites of predilection:

➢ Generalized involvement with no (only minimal) sparing of the body flexures.

➢ Scales most pronounced on the posterior aspect of neck, extensors of arms and legs encroaching cubital and popliteal fossa.

➢ Palms and soles spared

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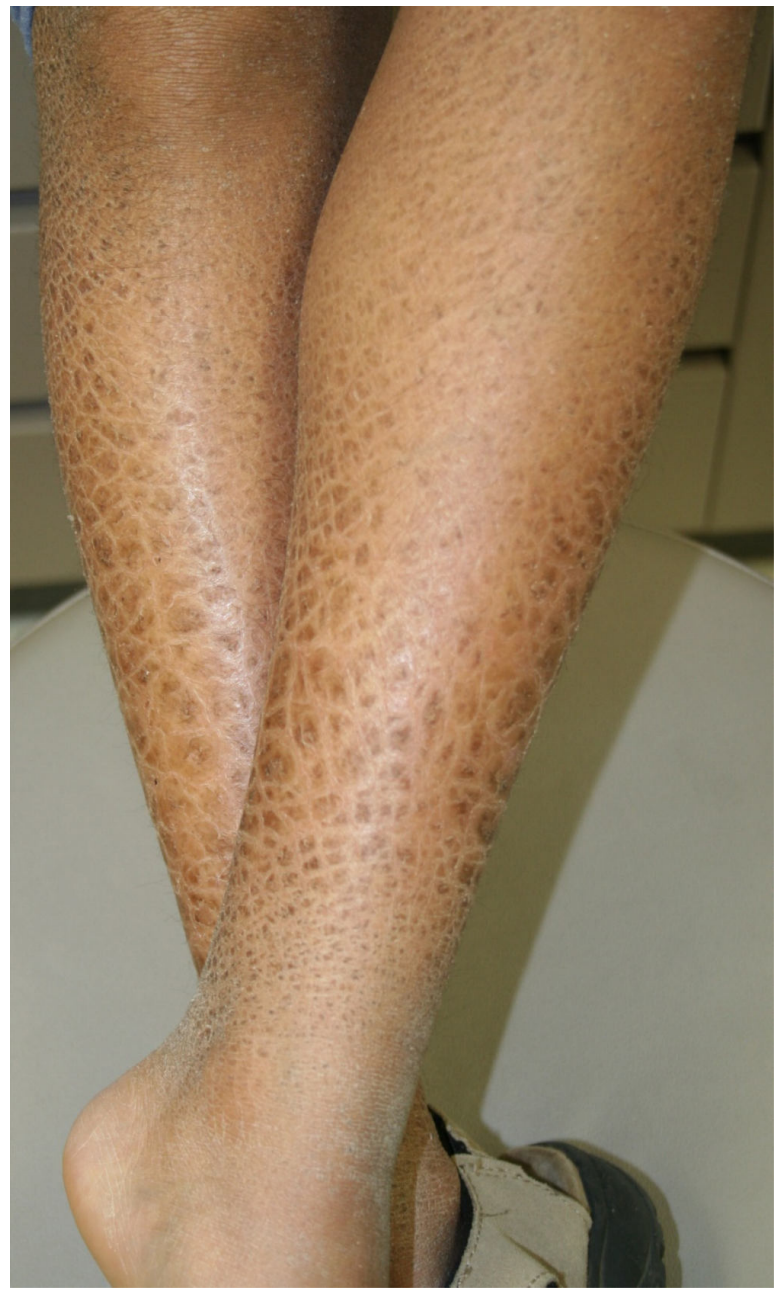

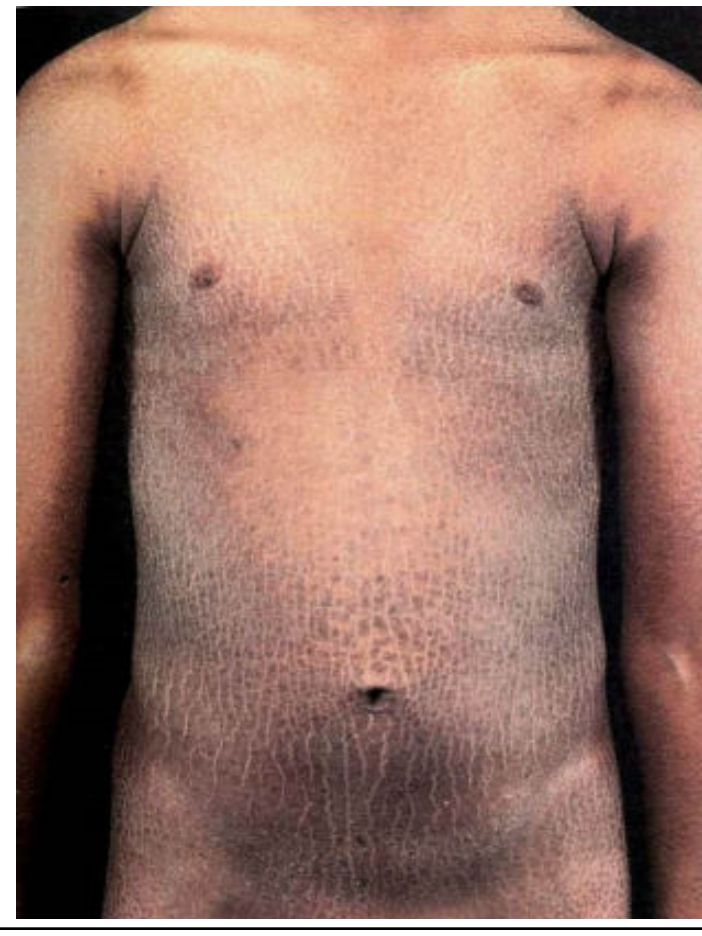

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- Associated features

➢Comma-shaped corneal opacities (do not interfere with vision).

➢Cryptorchidism

- Investigations
- Skin biopsy (if done) shows: Hyperkeratosis
- Hypergranulosis (cf., IV, where granular layer is absent or thin).
- Elevated serum cholesterol sulfate.
- Lowered steroid sulfatase in fibroblasts cultured from a skin biopsy (done for research purposes).

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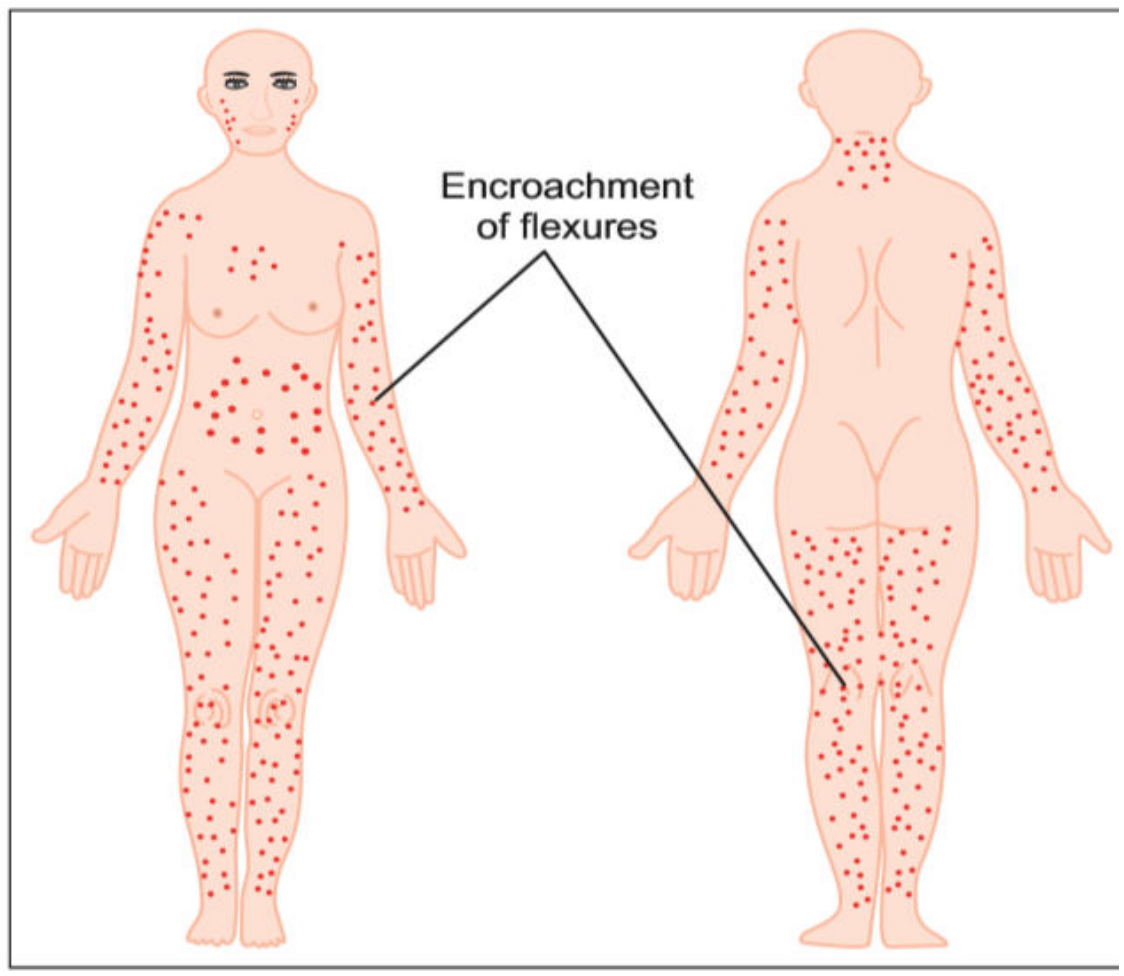
- Diagnosis
- Points for diagnosis:

✓ Patient being male

✓Presence of large, dark, adherent scales

✓Involvement of extensors with encroachment of flexures.



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IV	*XLI*
Inheritance: ADI	XLRI. Maternal uncle affected, but parents not affected
Gender: both males and females	Only males
Onset: 1st few years	At birth
Course: may improve in adolescence	Persists for life
Scales: small and branny except on shins where large and pasted	Large and dark (very!!)
Sites: extensors. Flexures spared.	Generalized. Flexures encroached
Associated features: ❖ Hyperlinear palms and soles ❖ Keratosis pilaris ❖ Atopic diathesis	❖ Corneal opacities ❖ Cryptorchidism

- Treatment Measures as for IV

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Lamellar Ichthyosis (LI)

- Epidemiology: Rare.
- Etiology: AR.
- Abnormality of gene encoding for transglutaminase.

- Onset: At birth, as collodion baby. ensheathed in a membrane; when the membrane sheds, typical scales become apparent.
- Character of scale: Large thick, brown, pasted scales.

- Distribution: Generalized, including flexures

- Treatment: Mild cases: Manage with hydration, lubrication and keratolytics.

- Severe cases: Acitretin

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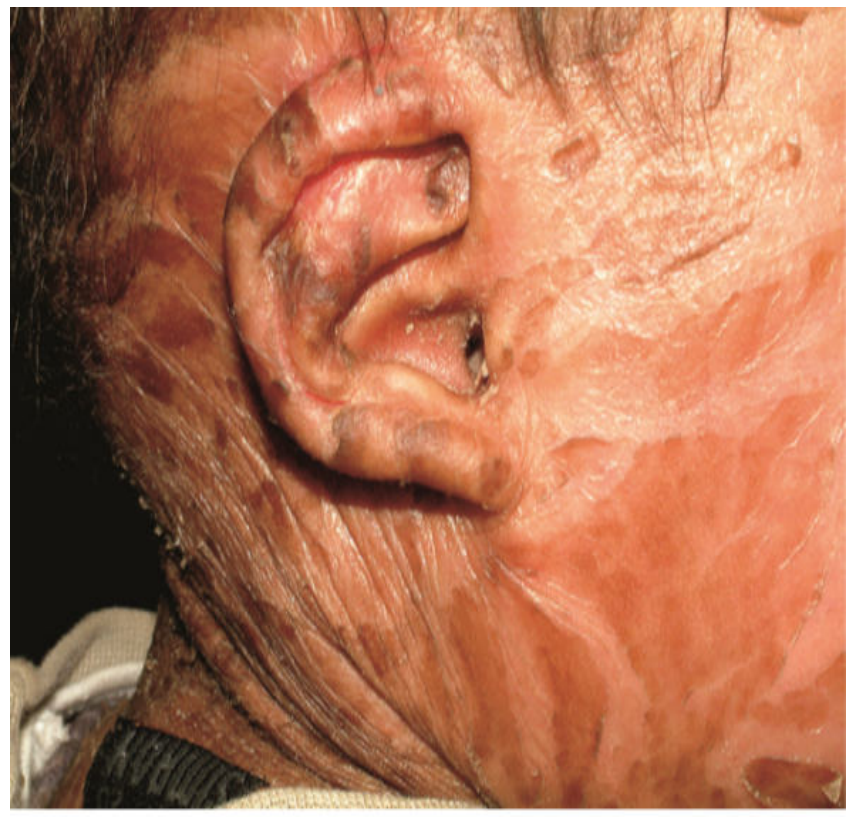

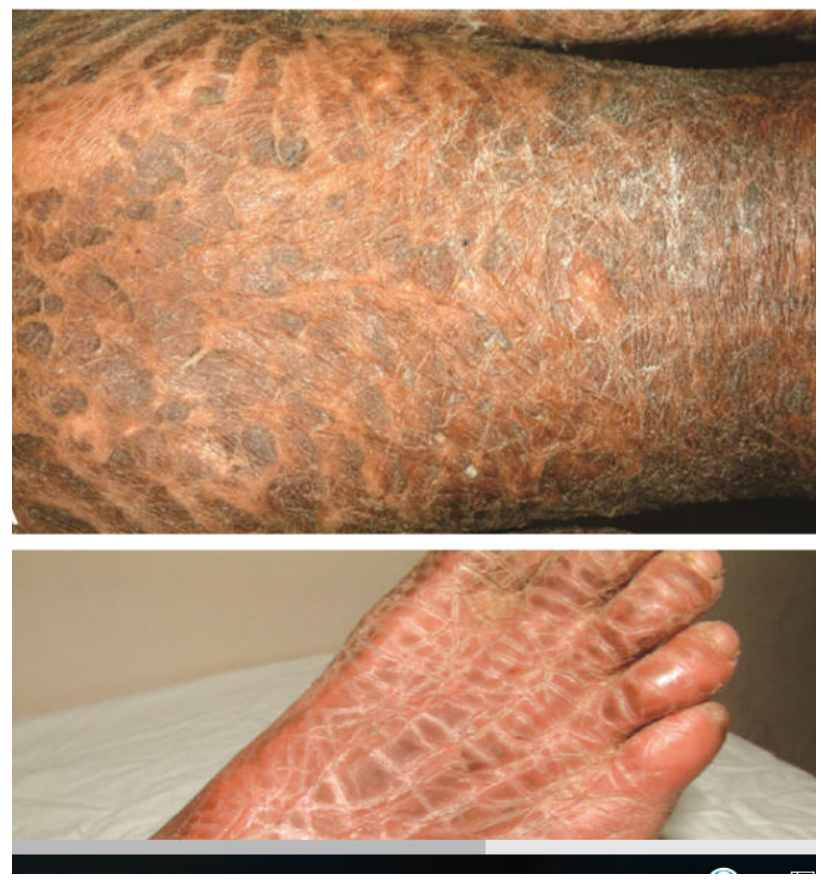

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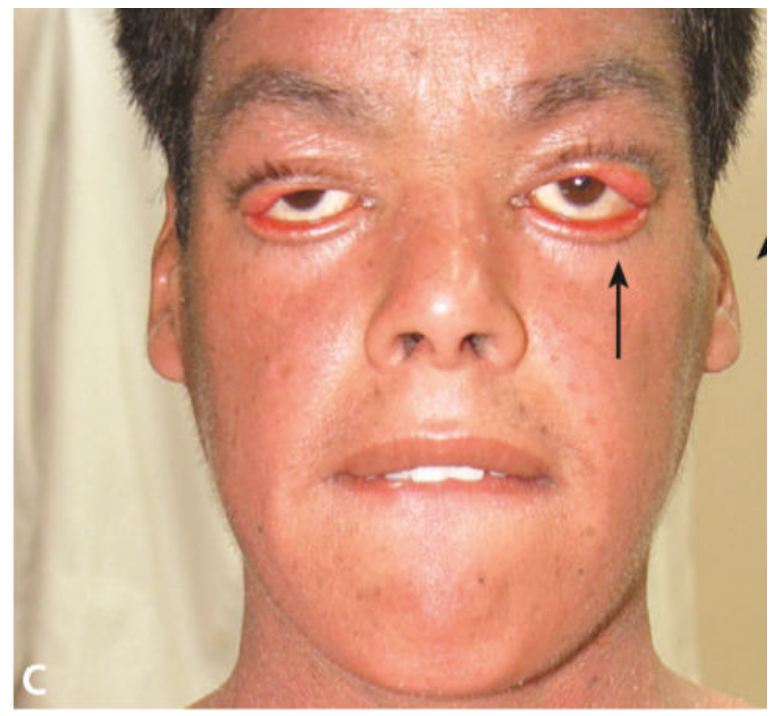
- Associated features
- Ectropion and eclabium
- Rippled hyperkeratosis around joints.
- Palmar and plantar keratoderma frequent.
- Crumpled ears



- Diagnosis

➢History of collodion membrane at birth

➢Characteristic thick, large, brown, pasted scales,especially on the shins.

➢Continuous rippling around joints.

➢Minimal erythema (except on face).

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- Differential diagnosis : NBIE

- Treatment
- Mild cases: Managed as patients with IV .
- Severe cases: Acitretin, under careful supervision.

Nonbullous Ichthyosiform Erythroderma (NBIE)

- Etiology: AR.

- Onset: At birth, as collodion membrane.

- Character of scales: On shedding of collodion membrane, there is fine diffuse scaling on background of erythema.

- Distribution: Generalized.

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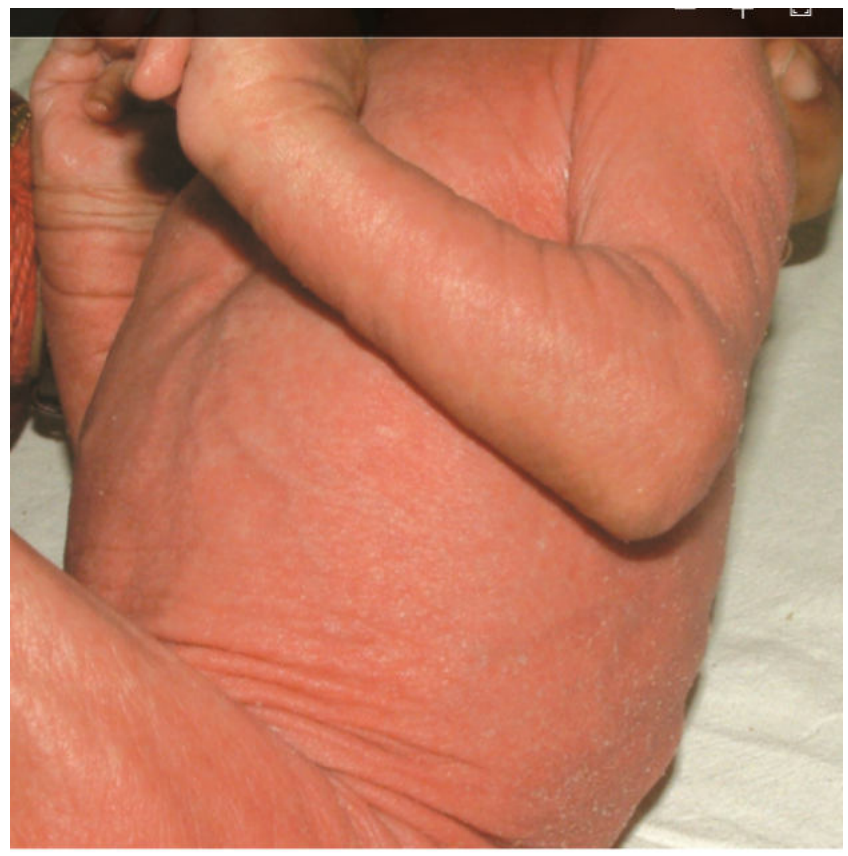

- Diagnosis
  - ➢Presence of collodion membrane at birth.
  - ➢Presence of small branny scales on a background of diffuse erythema.
  - ➢Generalized involvement.

- Treatment: As for LI.

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LI	*NBIE*
Prevalence: very rare	Rare
Erythema: minimal or absent	Marked
Scales: large, brown, adherent, scales especially on the shins	Branny scales
Palmoplantar keratoderma: frequent	Less frequent

Epidermolytic Hyperkeratosis (EHK)

- Etiology: AD.
- Onset: Self-limiting blistering stage.
- Character of scales: Brown hyperkeratotic (warty) waxy scales forming broad linear lesions with skip areas.
- Distribution: Generalized with accentuation in flexures.

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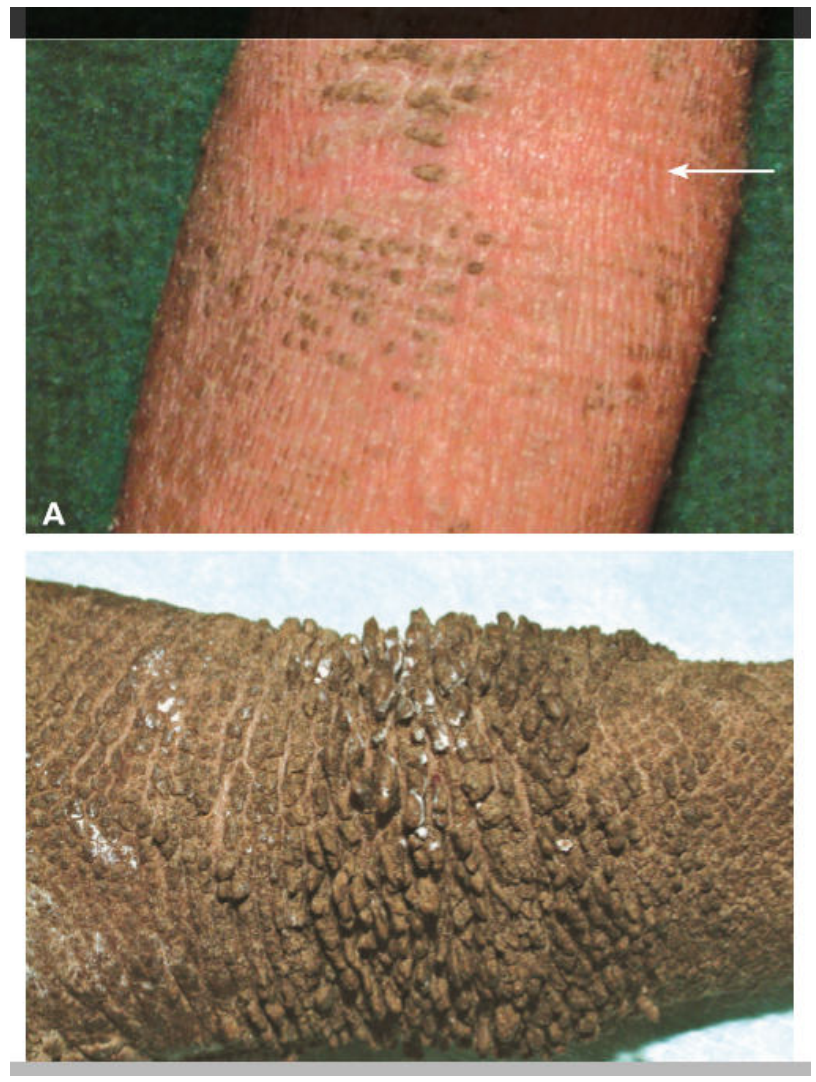

- Associations: Palmoplantar keratoderma.

- Treatment: Emollients. Mild disease: Topical retinoic acid (care in flexures!).

- Extensive disease: Systemic retinoids.

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LI	*EHK*
Onset: at birth as collodion baby	At birth with erythema with blistering
Scales: large brown and pasted	Warty hyperpigmented scales. May peel off leaving bald patches (skip areas)
Flexures: continuous rippling in flexures	Discontinuous rippling in flexures

Collodion Baby

- A morphological diagnosis.
- Most frequently associated with an underlying ichthyotic disorder:

✓Nonbullous ichthyosiform erythroderma

✓Lamellar ichthyosis.

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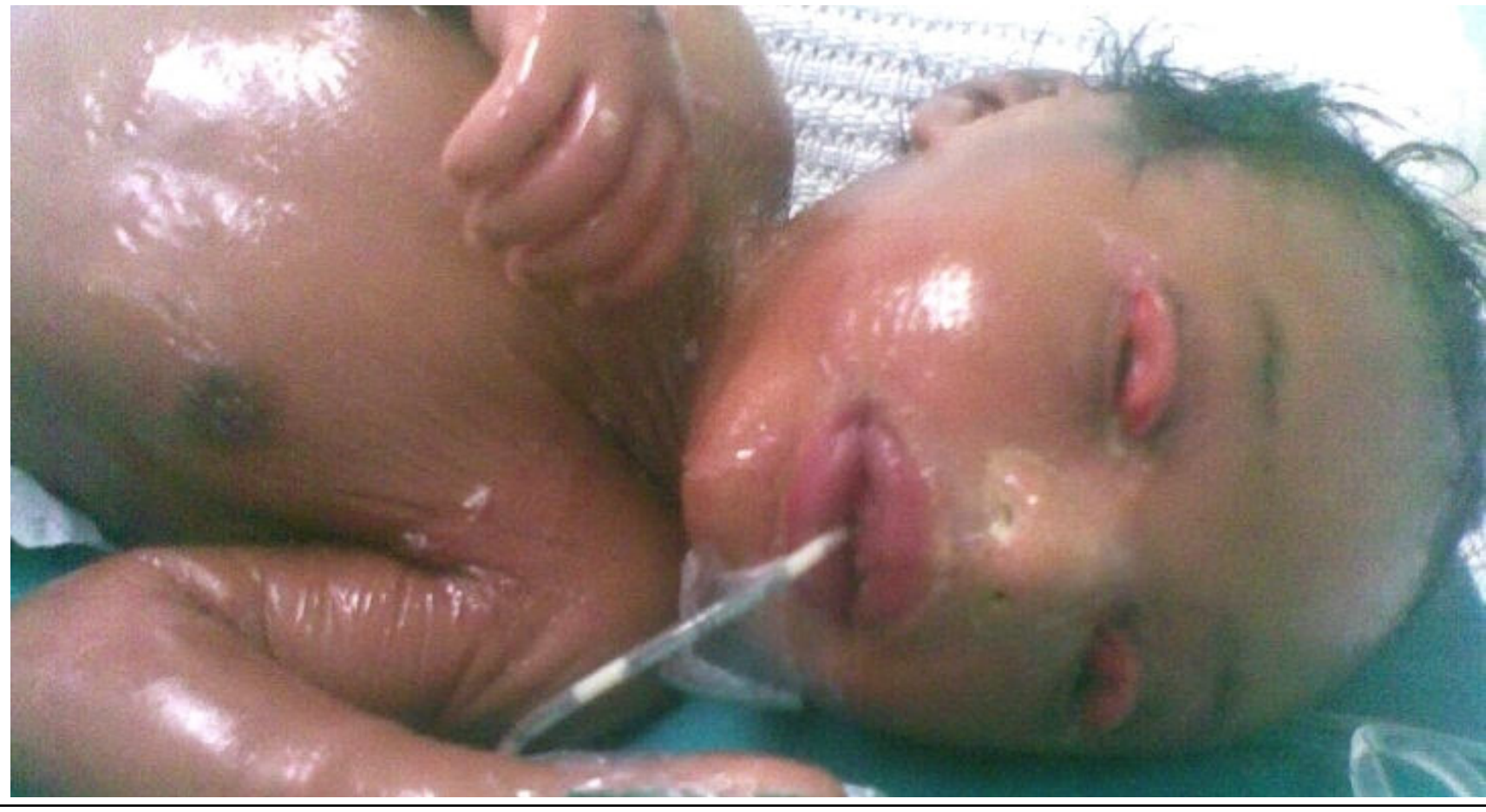
- Clinical Features:
- Neonate is born with a smooth and shiny skin(lacquered appearance), as if covered with cellophane or collodion . Tightness of the skin causes ectropion and eclabium.
- Outer cover (which is present all over the body) is eventually shed to reveal the underlying ichthyosis.



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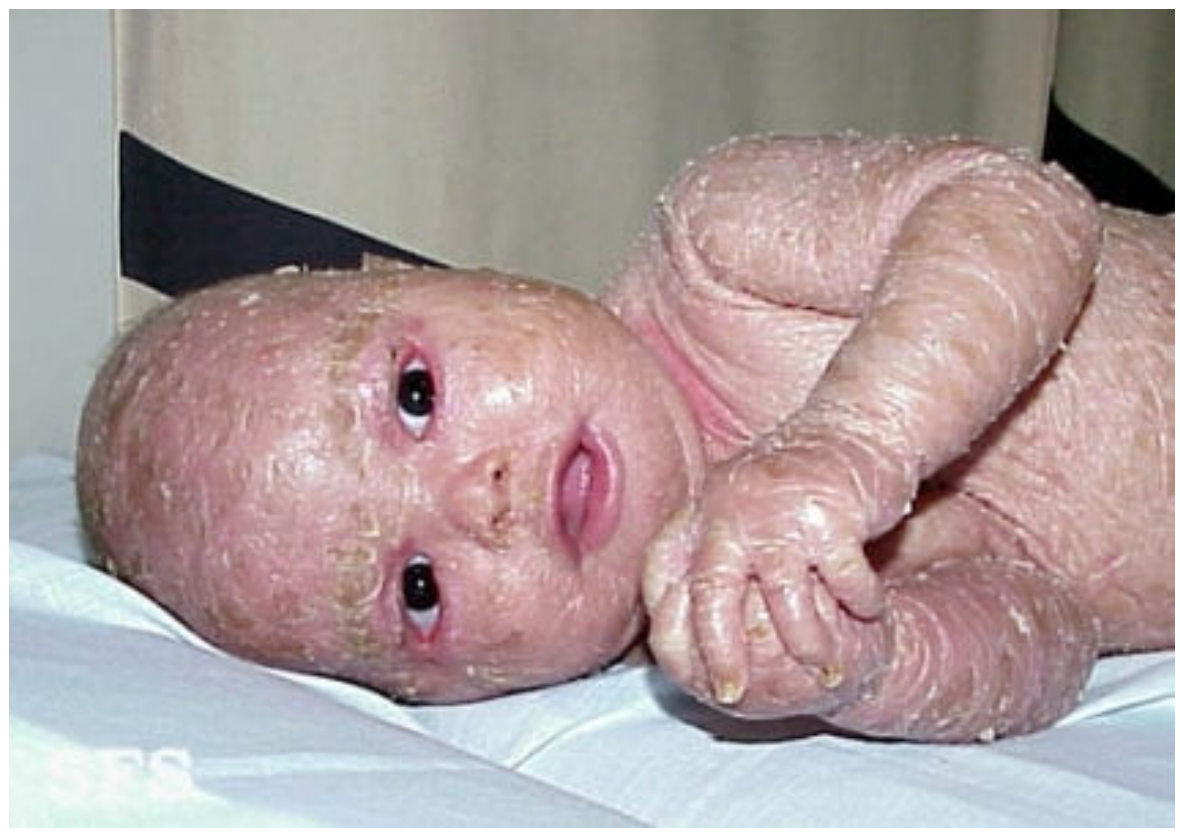

- Complications:

✓ Temperature dysregulation.

✓Feeding difficulties due to eclabium.

✓Water and electrolyte imbalance

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- Variants :
- Harlequin fetus:

➢Where the skin is covered with thick fissured skin resembling an armor.

➢Often fatal.

- Treatment

❖High-humidity incubator nursing necessary to maintain body temperature and to restrict water loss.

❖Regular application of emollients to make skin supple.

❖Short course of acitretin (oral retinoid) hastens shedding.

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Acquired Ichthyosis

Acquired
Infections: leprosy
Drugs: clofazimine
Malignancies: lymphomas
Endocrine disorders: hypothyroidism
Systemic diseases: sarcoidosis
Nutritional deficiencies

- If ichthyosis appears in adult life, suspect an underlying cause
- Clinical features resemble IV
- Symptomatic treatment with topical emollients after hydration.
- Treatment of underlying problem often reverses the ichthyosis.

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