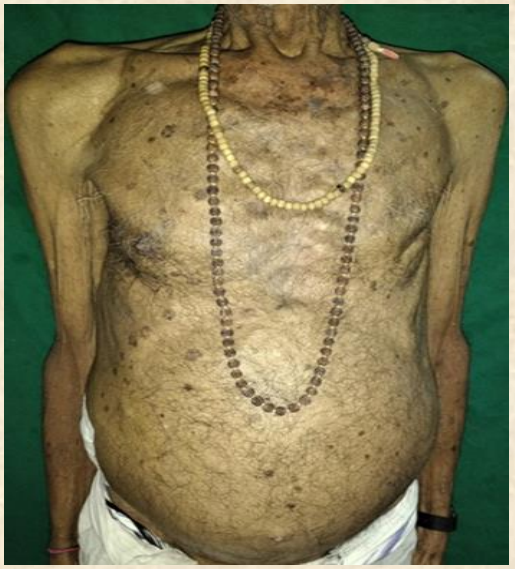
Sign of Leser-Trélat & cholangiocarcinoma: A rare association

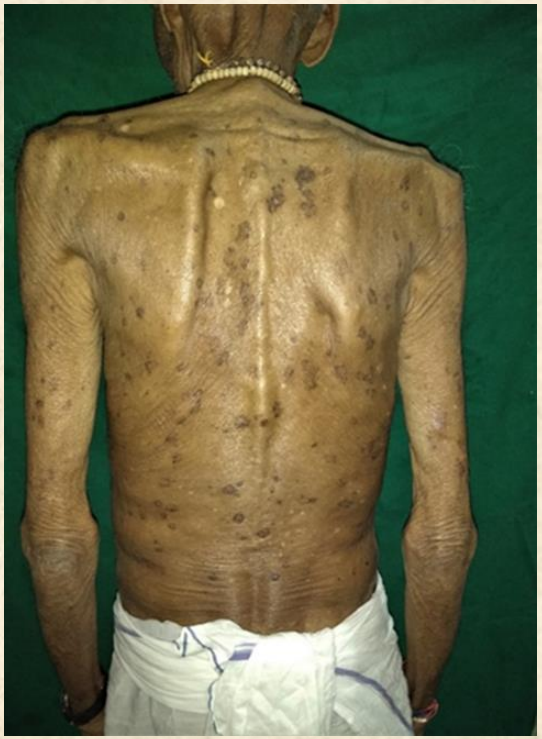
-Dr. Kalyani Deshmukh

Ganubhau/ 75/M /Farmer/ref;Med II

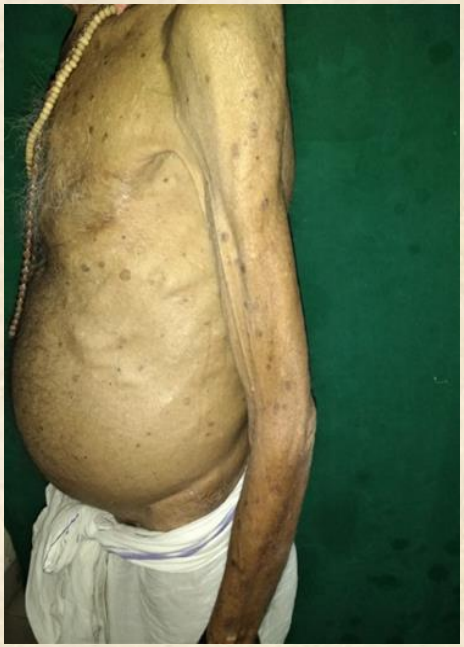


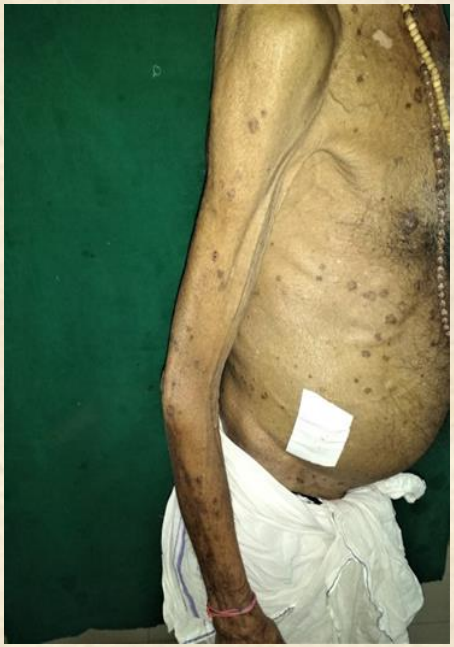
- Abdominal distention
- Swelling of legs
- Generalised weakness
- Warty eruptions over trunk

3 months



- Referred for dermatological consultation for multiple, pruritic, tan to black warty eruptions of sudden onset





- Similar lesions over flanks & arms

H/O

- Weight-loss (~ 7 kgs)
- Loss of appetite

} 6 mths

- Fever off & on
- **Consuming alcohol daily since 30 years (120 ml/day)**

General examination

- Conscious, cooperative, well-oriented
- Febrile
- Cachectic with BMI- 17
- Pallor - +++
- Pedal edema
- Icterus ++
- No lymphadenopathy , cyanosis, clubbing

Systemic examination

- **Per abdomen findings:**

✓ Ascites & distended veins

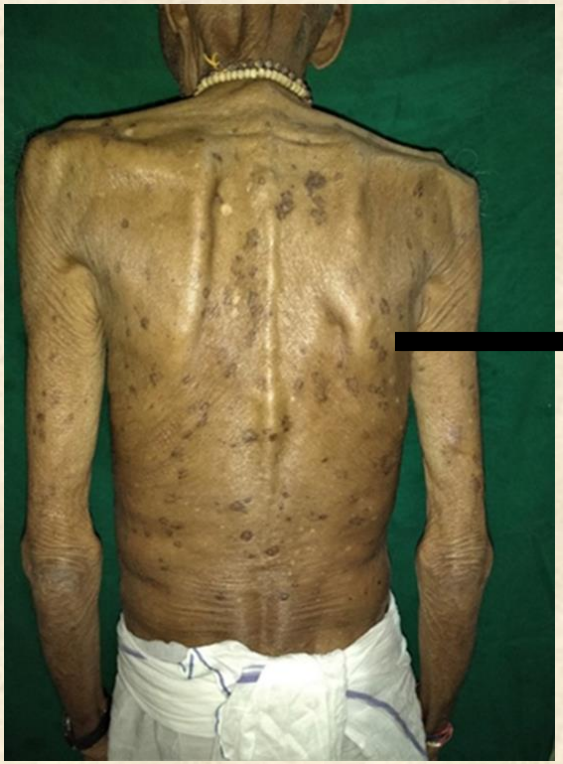
✓ Splenomegaly

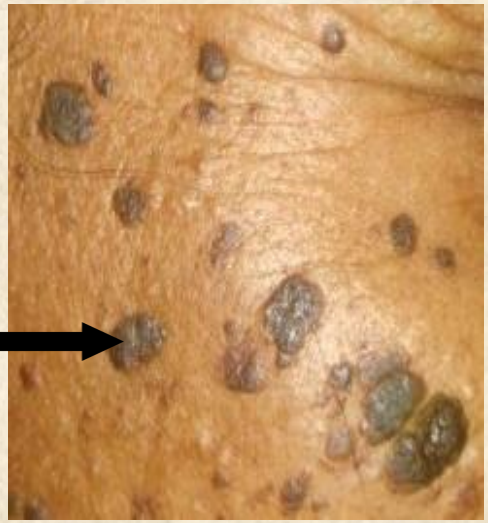
✓ Shifting dullness

✓ Absent bowel sounds

Dermatological examination

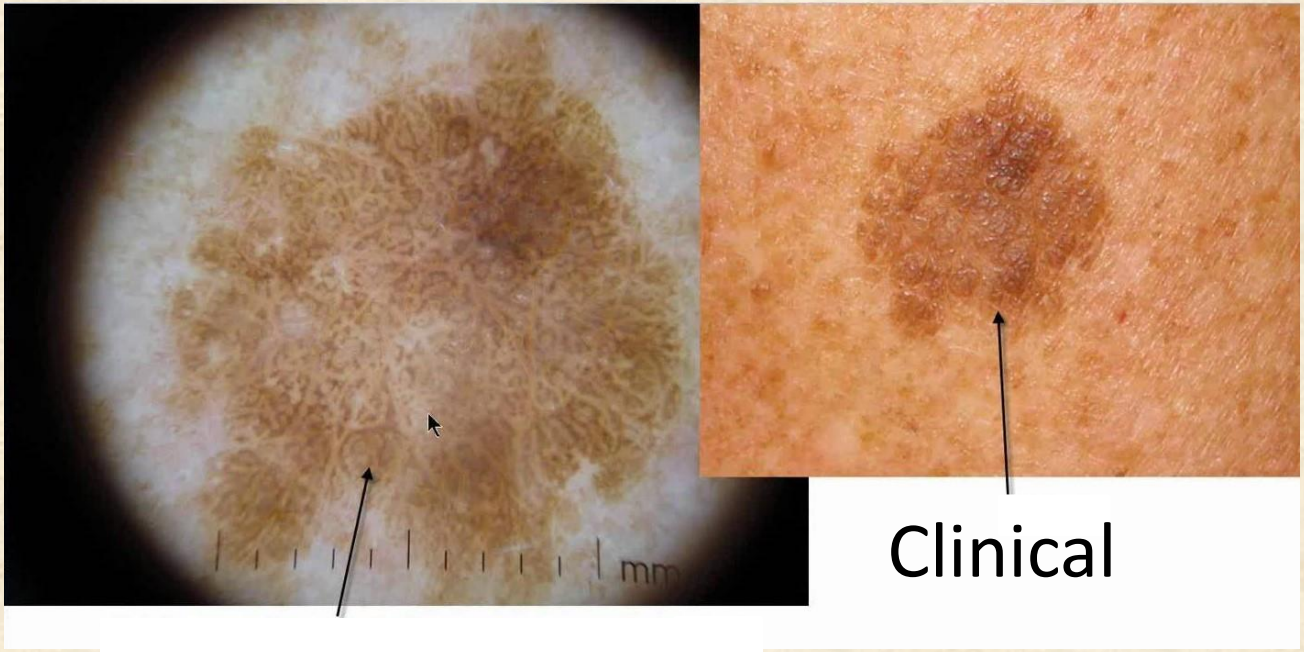
- Brown-black macules, papules, and plaques ; present on trunk





Texture - waxy to velvety
‘Stuck-on’ appearance

Dermoscopy



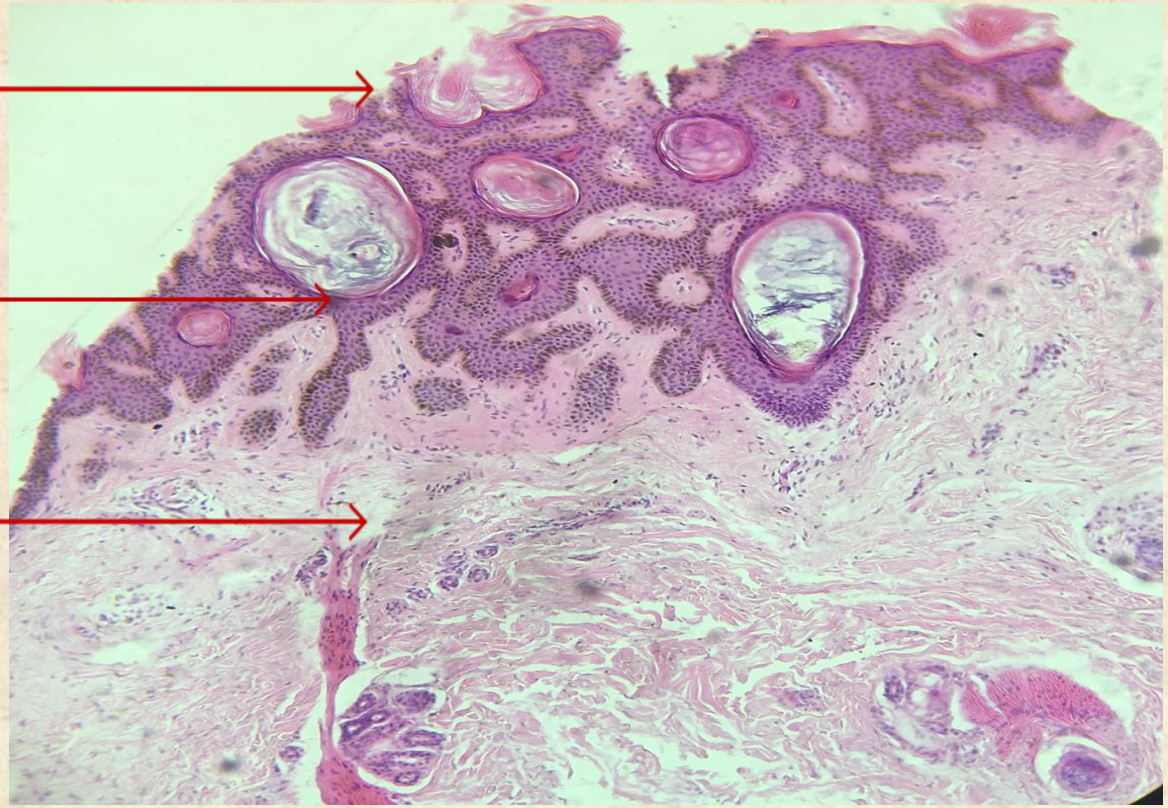
Cerebriform pattern

HPE

keratotic invaginations ("pseudohorn ") cysts

Intraepithelial keratin ("true horn")cysts

Dermis - moderate inflammatory infiltrate



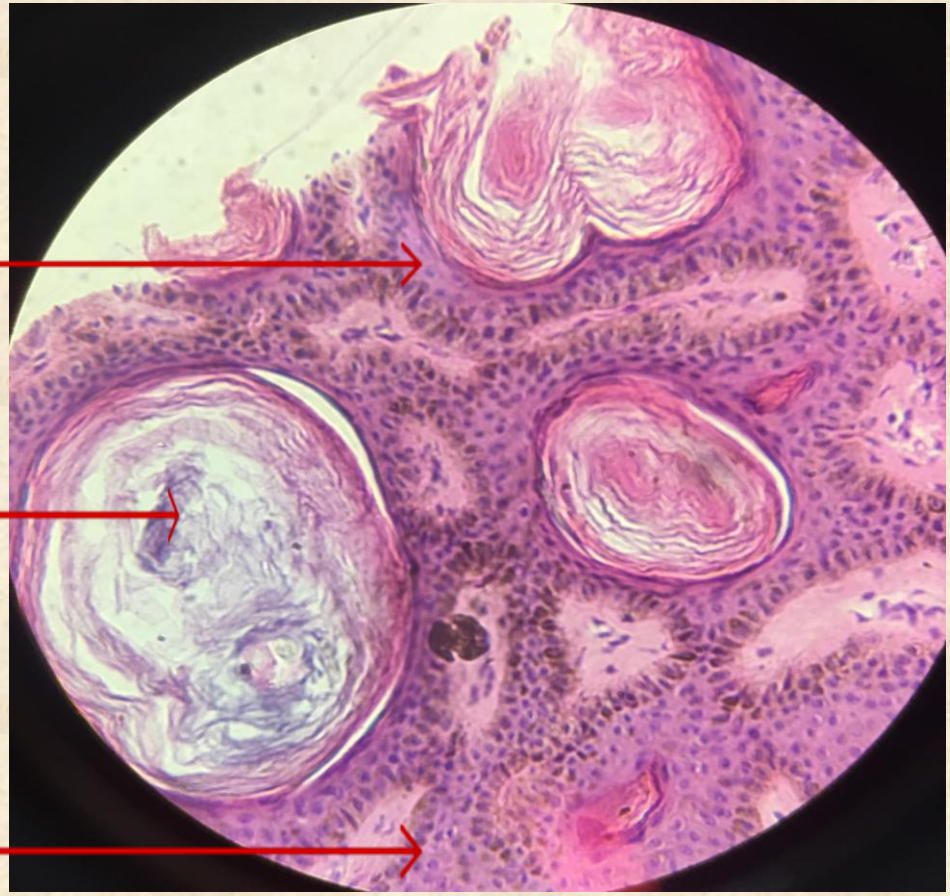
HPE- 100X H&E

Papillomatosis, parakeratosis, acanthosis

keratotic invaginations (“pseudohorn ”) cysts

Intraepithelial keratin (“true horn”)cysts

Melanin incontinence in the basal layer



HPE 400X H&E

In view of appearance of

- Sudden , multiple , pruritic **seborrhoic keratosis**
- In an aged cachectic, febrile icteric, pale patient
- With significant loss of weight & appetite

- Dermatological opinion was of **Leser-Trélat sign;** most likely paraneoplastic;

- Appropriate work-up to rule out underlying malignancy was advised

Lab investigations

- CBC : **Hb- 8.8 g/dl**
- TLC - **3600/ml**
- **ESR – 32mm**
- RBC indices suggests-

  Macrocytic normochromic

  anaemia

- LFT – **Serum bilirubin & transaminases** ↑
- RFT – Normal
- Urine - RE – normal;

  - ME – **Pus cells: 6-8,**

    Epithelial cells : 1-2 ;

    RBCs: 6-8

Ascitic fluid –

- **Gross**: Volume-2-3ml; yellowish; clear
- **Microscopy**: nucleated cells – 150/mm3

  (lymphocytes 70%)

- **Biochemistry parameters** :

-Sugar
-Protein
-LDH – 39 SU

} Normal

-Albumin – 0.6 mg%
-ADA – 35 u/l (normal - <30/l)

- HIV
- Hbs Ag — Negative
- HCV

- Sr. PSA – 0.4 ng/ml

USG abdomen

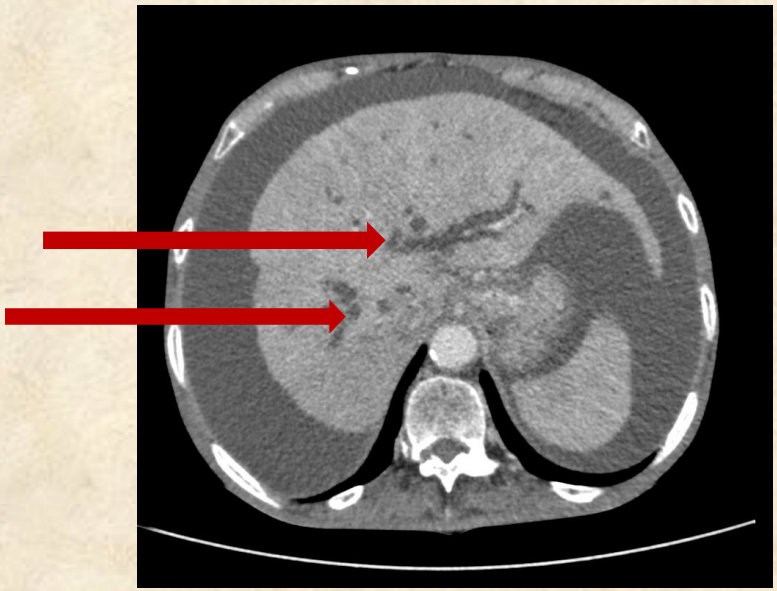
- Liver parenchymal disease
- Gross ascites
- Splenomegaly
- Changes of cystitis
- Minimal pleural effusion
- Borderline prostatomegaly

OGD scopy – grade II oesophageal varices with **portal hypertensive gastropathy** with **GAVE duodenitis**

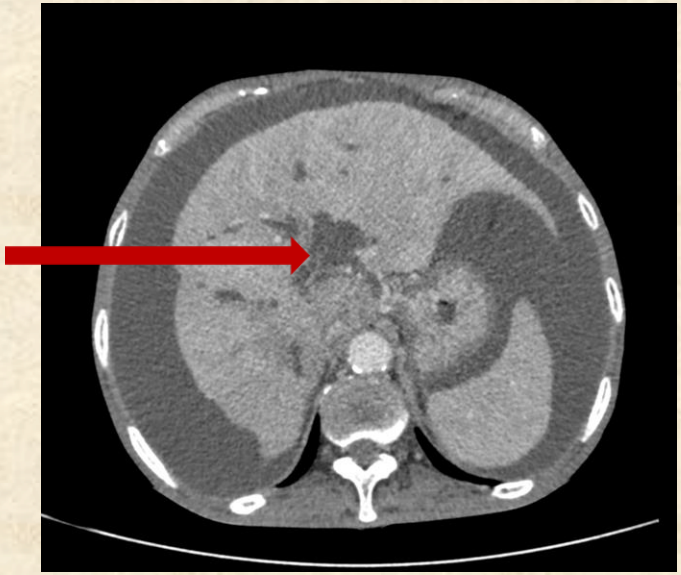
2D ECHO – EF – 60% , **mitral valve annular calcification, aortic valve sclerosed**

CECT Abdomen

- Mild central dilatation of intrahepatic biliary radicals in right and left lobes

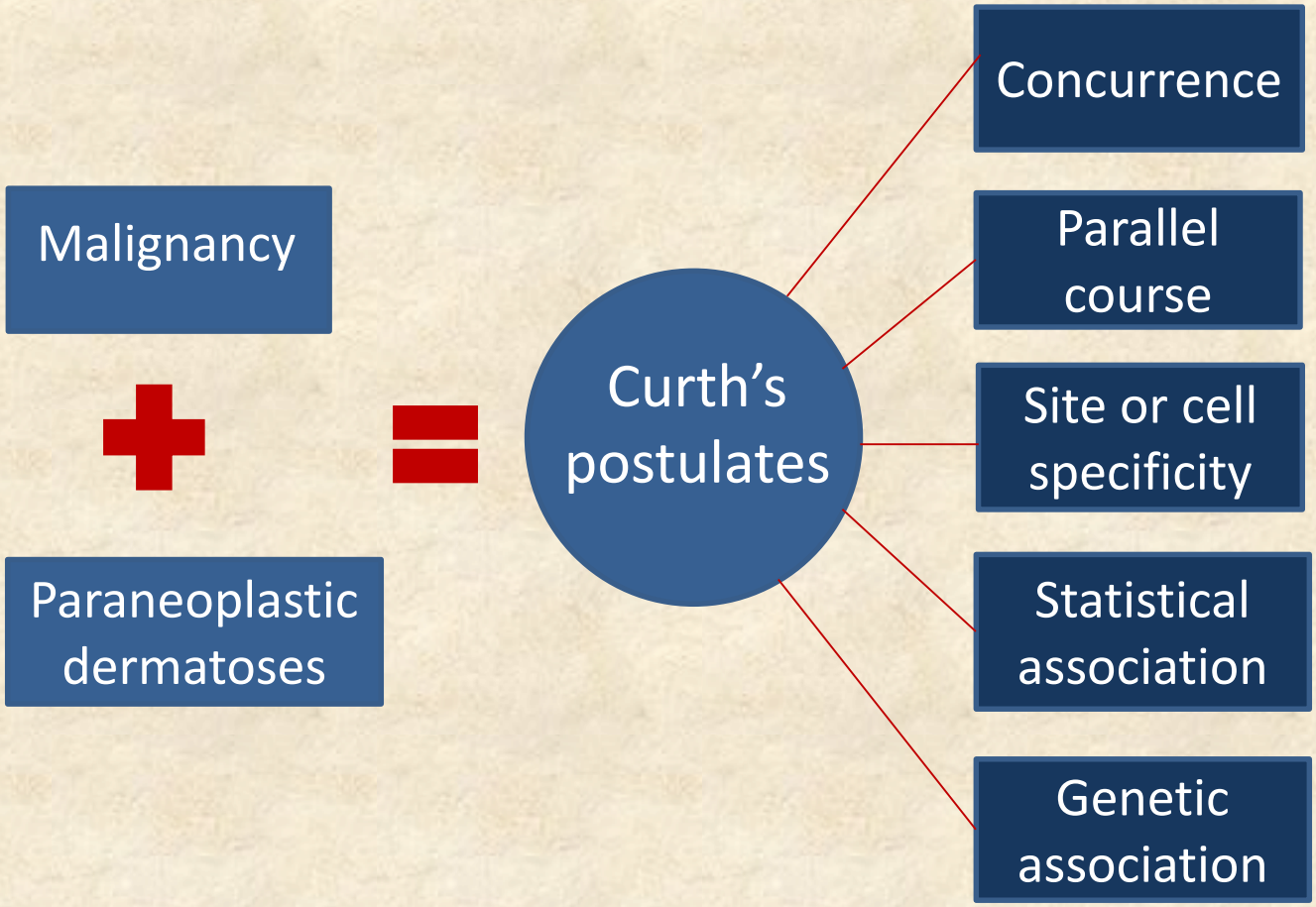


- Common hepatic duct-common bile duct junction at porta hepatis not well visualized and showed wall thickening –



- **Infiltrating cholangiocarcinoma**

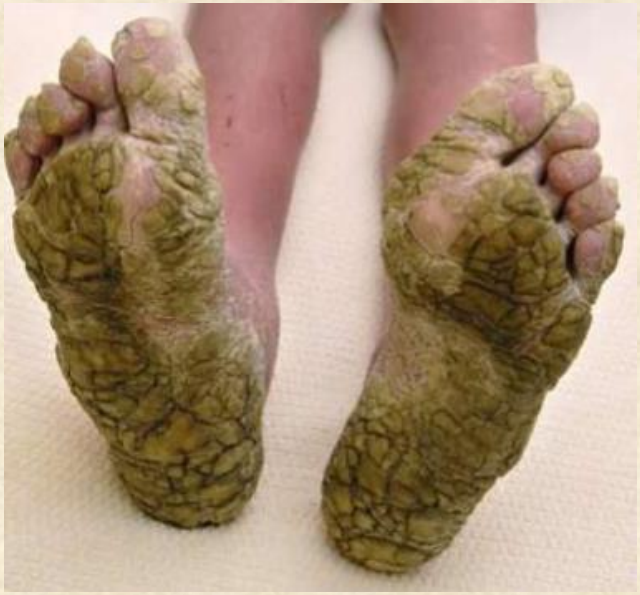
Discussion


Malignancy
Paraneoplastic dermatoses
Curth's postulates
Concurrence
Parallel course
Site or cell specificity
Statistical association
Genetic association

Strength of correlation

Strong	Bazex syndrome
	Acanthosis palmaris (tripe palms)
	Florid cutaneous papillomatosis
	Primary amyloidosis
	Acquired hypertrichosis lanuginosa
Moderate	Sweet syndrome , Pyoderma gangrenosum
Weak	Acanthosis nigricans in isolation
	Acquired ichthyosis
	Leser–Trélat sign

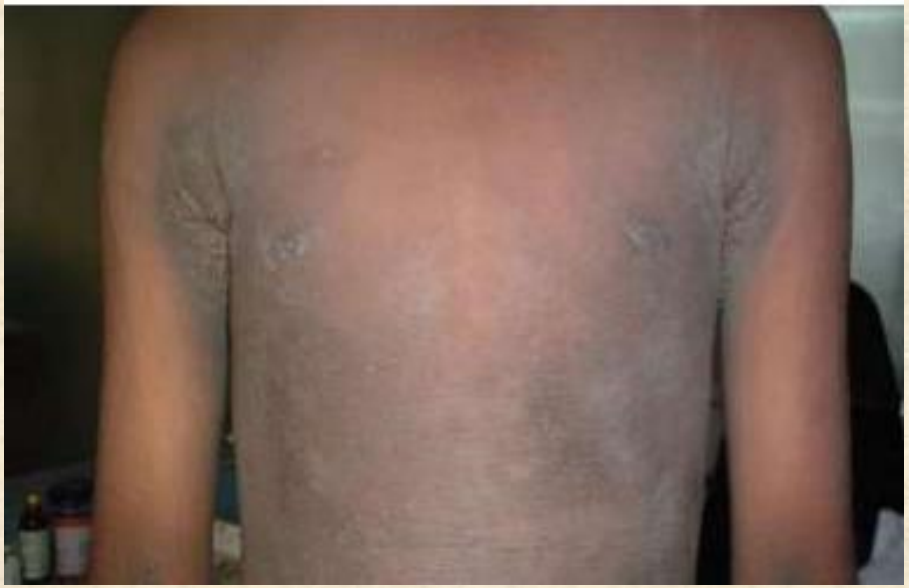
Bazex syndrome



SCC of upper aerodigestive tract

Malignant acanthosis nigricans

\



Adenocarcinomas: intraabdominal;

gastric (50%–60%)

Tripe palms(acanthosis palmaris)

Lung Ca. (most common)
Gastric Ca. (second)

Leser-Trélat sign

- Rare paraneoplastic cutaneous sign
- Refers to **sudden** onset and **dramatic increase** in the number and size of seborrheic keratoses

Associations

Non-malignant

- Elderly
- Eczema
- Acanthosis nigricans
- Erythroderma
- HIV
- Acromegaly

Malignant

- **Gastric, rectal, colon Ca**
- **Lymphoproliferative dis. (CTCL, pre B cell LL)**
- Breast ca.
- Lung ca.
- RCC , TCC-UB
- Malignant haemangiopericytoma

- However, concurrence of Leser-Trélat sign with **cholangiocarcinoma**, as in our patient, is very rare

- Previous solitary report –

  Scully et al ,British Journal of Dermatology , 2001,vol 145, 506-7

- **2nd case** to be noted in English literature till date
- **1st case in Indian scenario**

Prognosis and course

- It can appear 5 months **prior** to /10 months **after** the diagnosis of **malignancy**
- Once diagnosed, prognosis is poor with **average survival time** of **10.6 months**

Take home message !

Seborrheic keratoses are very common in the elderly

BUT

Sudden ,eruptive and pruritic ones

require further work-up