Dermatology of Systemic Disease

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Boca Raton, Florida
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1. Defining the need for Skin Cancer Education. JACOBSEN B.A., Claudina Canaan LACHAPELE M.D., CB WOHL M.P.H., Robert KIRSNER M.D., PhD. John Strasswimmer M.D., PhD. JAMA Dermatol In press
7. Strasswimmer J. Potential Synergy of Radiation Therapy with Vismodegib for BCC. JAMA Dermatol. 2015 Sep
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Boca Raton, Florida
Biopsy proven squamous cell carcinoma

- History of SLE
- Scheduled for plastic surgery excisions and also for radiation therapy
Mohs Surgeon and Systemic Dermatology

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Biopsy proven squamous cell carcinoma

- History of SLE
- Scheduled for plastic surgery excisions and also for radiation therapy
- Re-pbiopsed 4 times
- Diagnosis: “subacute” lupus with lichen plants features
- Treatment: **photoprotection**
CASE 1

Dr. Pamela Sheridan
CASE 1

- **HPI:** 51 yo male
- HTN and diverticulitis
- presented with worsening myalgias and arthralgias over the past few weeks
- severe lethargy and fevers over the last week.
- admitted for hyponatremia
- Dermatology consulted for a lesion on the abdomen.
OBJECTIVE

- **VSS**: T 98.3, P 88, RR 20, BP 140/73, O₂ 98
- **PE**: Gen: NAD, AAOx3
  - Ø cervical lymphadenopathy, Ø conjunctival, mucosal or other cutaneous lesions
- **Labs**
  - **CBC**: WBC 19.1, HB 12.3, HCT 35.2, PLT 330, EOS 0.6%,
  - **CMP**: Na 119, K 3.1, Cl 86, CO₂ 30, BUN 16, Cr 0.7, CRP 16.8
  - Viral panel (-), HIV (-), pending blood cultures
Tender erythematous, purpuric papules on third digit of L hand
Tender erythematous purpuric papule at the on the R foot
Tender erythematous purpuric papule at the on the L foot
DIFFERENTIAL

- Osler nodes or Janeway lesions
  - Secondary to:
    - **Most likely:** Subacute/acute endocarditis
    - **Other:**
      - Systemic lupus erythematosus
      - Gonococcemia
      - Hemolytic anemia
Dense dermal inflammatory pattern
Thrombi within vessel wall

Endothelial cell obliteration

Thrombi within vessel wall
Dermal Abscess
RADIOLOGIC AND MICROBIOLOGIC WORKUP

• Blood cultures (+) MSSA x 4 → (-) x 3.
• MRI back: Epidural abscess of L5-S1.
• TEE: (12/23/15): small vegetation of the aortic valve, large vegetation of the mitral valve with a large perforation. EF: 65%.
CLASSIC CUTANEOUS FINDINGS IN ENDOCARDITIS

• >50% of patients have a finding”
  – Petechiae:
  – **Subungual (splinter) hemorrhages**: Dark-red, linear lesions in the nail beds
  – **Osler nodes**: Tender subcutaneous nodules usually found on the distal pads of the digits
  – **Janeway lesions**: Non-tender maculae on the palms and soles
  – **Roth spots**: Retinal hemorrhages with small, clear centers; rare
CUTANEOUS FINDINGS IN ENDOCARDITIS

- Subconjunctival Hemorrhage (2-5%)
- Roth spots < 5%
- Osler nodes (5%)
- Mucosal Petechiae 20-30%
- Clubbing 10%, long standing only
- Splinter hemorrhages 10%
- Janeway lesions
- Petechial rash (40-50%)
- Loss of pulses
- Pallor
OSLER NODES AND JANEWAY LESIONS

OVERVIEW

• **Cutaneous manifestations of bacterial endocarditis.**
  – Also rarely described in systemic lupus erythematosus, gonococcemia, hemolytic anemia and typhoid fever.

• **Osler nodes**: red-purple, slightly raised **tender** nodules often with a pale center. Average diameter 1 to 1.5mm.
  – Can occur at any time during the course of endocarditis (usually late in subacute)

• **Janeway lesions**: **non-tender**, hemorrhagic
  • palms and soles
  – More commonly see in **acute** endocarditis
PATHOGENESIS

• Two disparate theories
  – Circulating immune complex mediated vasculitis – Gutman et al.
  – Microembolization
CASE 2

Dr. Pamela Sheridan
CASE 3

- 40 yo Caucasian otherwise healthy male
- Chronic “rash” x 9 months in groin
  - not responsive to antifungals, antibiotics, corticosteroids
- admitted for altered mental status
DDX:

• Atopic dermatitis
• Necrolytic Migratory erythema (glucagonoma syndrome, pseudoglucagonoma syndrome)
• Acrodermatitis enteropathica (zinc deficiency)
• Pellagra (niacin deficiency)
Confluent parakeratosis

Buckshot dyskeratosis
DIAGNOSIS?

• Necrolytic Migratory Erythema
• Secondary to Neuroendocrine carcinoma
• (glucagonoma)
LABORATORY STUDIES & DATA

• Labs: CBC – microcytic anemia, CMP – hypoglycemia, elevated LFTs (alkp 340, AST 97, ALT 146), TSH 38

• Specialty labs:
  – Zinc level wnl, Vitamin C wnl
  – Insulin- 41.2 (2.0-19.6), c-peptide- 4.48 ng/ml (0.8-3.85)
  – Chromogranin A- 682 (1.9-15)
  – AFP 55.22, Ca 19-9 89
  – 5HIAA wnl

• Radiologic studies:
  – Portable abd x-ray: hepatomegaly with no obstruction
  – CT abd w/contrast: massively enlarged liver with numerous hepatic masses.
Needle core biopsy - liver

insulin-secreting tumor
NECROLYTIC MIGRATORY ERYTHEMA (GLUCAGONOMA SYNDROME)

α2-glucagon producing islet cell pancreatic carcinoma

insulin- producing tumor —> reactive hyper-glocogon state —> NME
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Case 1

55 year old man with two month history of pruritic papule on the wrists and ankles
Flexor Wrist

Buccal Mucosa
LICHEN PLANUS

Flexor Wrist

Buccal Mucosa
LICHEN PLANUS

• Purple, pruritic, polygonal papules
• Wickham’s striae - are fine white lines on top of papules
• Koebner phenomenon -
  • in linear groups due to trauma of scratching
LICHEN PLANUS

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• Usually self limiting 2-3 years.
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• Wickham’s striae - are fine white lines on top of papules
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• Management?
LICHEN PLANUS

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• Koebner phenomenon - they grow in linear groups due to trauma of scratching
• Usually self limiting 2-3 years.

Rx:
• Topicals> IM triamcinolone

Two complications
• Variable association of 0.1% to 35% with Hepatitis C
• Evolution to mucosal SCC
SUMMARY OF CUTANEOUS MANIFESTATIONS OF HEPATITIS C VIRUS

- Lichen Planus
- Porphyria Cutanea Tarda
- Polyarteritis Nodosa
- Necrolytic Acral Erythema
- Cryoglobulinemia
- Pruritus
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CASE 2

57 year old man with itchy hands for 2 years
57 year old man with itchy hands for 2 years
- Vesicles and bullae on sun-exposed areas, scarring with milia
- Hypertrichosis
• Vesicles and bullae on sun-exposed areas, scarring with milia
• Hypertrichosis
• Fragile skin with sclerodermoid changes
PORPHYRIA CUTANEA TARDA

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• Vesicles and bullae on sun-exposed areas, scarring with milia
• Hypertrichosis
• Fragile skin with sclerodermoid changes
• DDx: pseudoporphyria due to NSAIDS, OCP, etc.
PORPHYRIA CUTANEA TARDA

- Vesicles and bullae on sun-exposed areas, scarring with milia
- Hypertrichosis
- Fragile skin with sclerodermoid changes
- DDx: pseudoporphyria due to NSAIDS, OCP, etc.
- HCV+ in 60%
• **Pathogenesis** of HCV-related PCT:
  • Decompartmentalization of iron stores
  • formation of free iron radicals that oxidize UROD
  • Decrease UROD activity - photosensitizer
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VASCULAR DISEASES

• Polyarteritis Nodosa
  • Small vessel vasculitis (LCV = palpable purpura)
  • Medium vessel vasculitis (nodules on lower extremities)
  • Multi system disease
  • Associated with HCV or HBV
HEPATITIS B OR C

- About 30% may have Urticaria or present a serum sickness like picture (because of circulating immune complexes)
- Associated with 5-7% cases of Polyarteritis nodosa
  - Classical PAN
  - Renal vasculitis present
  - ANCA negative
SUMMARY OF CUTANEOUS MANIFESTATIONS OF HEPATITIS C VIRUS

• Lichen Planus
• Porphyria Cutanea Tarda
• Polyarteritis Nodosa
• **Cryoglobulinemia**
• Necrolytic Acral Erythema
• Pruritus
VASCULAR DISEASES

• 62 year old woman
VASCULAR DISEASES

- 62 year old woman
  - Arthralgias,
  - Elevated LFT
  - Glomerulonephritis
VASCULAR DISEASES

Cryoglobulinenemia

- Small vessel vasculitis (LCV = palpable purpura)
- Clotting in vessels: livedo reticularis
VASCULAR DISEASES

Cryoglobulinenemia

- Small vessel vasculitis (LCV = palpable purpura)
- Clotting in vessels: livedo reticularis

- Systemic symptoms:
  - Arthralgias,
  - Elevated LFT
  - Glomerulonephritis
VASCULAR DISEASES

Cryoglobulinemia

- Small vessel vasculitis (LCV = palpable purpura)
- Clotting in vessels: livedo reticularis

- Systemic symptoms:
  - Arthralgias,
  - Elevated LFT
  - Glomerulonephritis

- Due to:
  - IgG reversibly precipitate in cold
  - Mixed (type 3) polyclonal IgG/IgM

- Associated with
  - HCV
  - Multiple myeloma
NECROLYTIC ERYTHEMAS (ACRAL OR MIGRATORY)
NECROLYTIC ERYTHEMNAS (ACRAL OR MIGRATORY)

NECROLYTIC Acral ERYTHEMA

- Coalescing, arcuate papule and vesicles
- Chronic, more scale
- HCV

NECROLYTIC Migratory ERYTHEMA

- Coalescing, arcuate papule and vesicles
- Chronic, more scale
- Involves flavor surfaces
- Glucogonemia
PRURITUS

Pruritus

- lesions where can reach
- often in linear arraignment
- multiple phases of healing
- hemmoragic crust: largely unique to trauma (and Herpes infections)
Pruritus

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- often in linear arraignment
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- hemmoragic crust: largely unique to trauma (and Herpes infections)
Pruritus

- lesions where can reach
- often in linear arrangement
- multiple phases of healing
- hemorrhagic crust: largely unique to trauma (and Herpes infections)

- Skin findings:
  - linear erosions
  - lichenification
  - Prurigo nodules
PRURITUS

Causes:

- Iron deficiency
- Liver disease
- Malignancy (e.g. Hodgkin’s lymphoma)
- Neurological disorders
- Polycythemia
- Renal failure
- Thyroid dysfunction

Work-up:

CBC, LFT, BUN/Cr, TSH
Chest x-ray
HBV, HCV, HIV
Dermatitis herpetiformis:

- Symmetric, grouped vesicles on extensors
- Very pruritic
- Associated with Hashimoto’s thyroiditis, lymphoma, DM
- Due to IgA antibodies against epidermal transglutaminase-3
- GI gluten sensitivity demonstrated in 20%
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CUTANEOUS MANIFESTATIONS OF HEPATITIS C VIRUS

“a great mimic” of the 21st century
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• Lichen Planus
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• Polyarteritis Nodosa
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• Cryoglobulinemia
• **Pruritus**
CUTANEOUS ERUPTIONS OF HEPATITIS C VIRUS TREATMENT

• IFN:
  • alopecia, lichenoid (lichen-planus like) eruption, eczema, malar erythema and local cutaneous necrosis
CUTANEOUS FINDINGS OF HEPATIC DISEASE

• Palmar erythema: hypothenar erythema that later spreads to fingers and rest of the palm

• Gynecomastia (hyperestrogenemia)

• Xanthomas
CHRONIC LIVER DISEASES

- Clubbing
- Longitudinal ridging
- Thickening
- Britleness
- Total leuconychia
- Terry’s nails
- (whitening of the entire nail plate except for a narrow pink band distally)
- Muehrcke’s nails

Multiple parallel transverse white bands

Terry’s nails

Muehrcke’s nails
DIAGNOSIS? MOBILE NODULE
THYROGLOSSAL DUCT CYST

- Embryonic duct remnant
- Midline anterior neck; mobile
THYROID HORMONE AND THE SKIN

• Pivotal role in hair and sebum

• Thyroid hormone regulates epidermal metabolism —> determination of epidermal thickness
HYPERTHYROIDISM AND THE SKIN

• Skin is usually warm, moist, and smooth
HYPERTHYROIDISM AND THE SKIN

• Skin is usually warm, moist, and smooth (best assessed on the inner aspect of arm and over the chest)
HYPERTHYROIDISM AND THE SKIN

- Skin is usually warm, moist, and smooth (best assessed on the inner aspect of arm and over the chest)
- Facial flushing
- Palmar erythema
- Hyperpigmentation, esp. creases of palms and soles
- Hair is fine and friable — > hair loss
- Hyperhidrosis, particularly of palms and soles
HYPERTHYROIDISM AND THE SKIN

**Thyroid dermopathy**
(pretibial myxedema)

- Coalescing, waxy papule and vesicles
- Increased hyaluronic acid
HYPERTHYROIDISM AND THE SKIN

Scleromyxedema

- Coalescing, waxy papule and vesicles
- Increased hyaluronic acid
- Increased fibroblasts
HYPERTHYROIDISM AND THE SKIN

Auto-immune diseases

- Vitiligo, alopecia
- Pernicious anemia
- Connective tissue diseases
Generalized Myxedema

- diffuse deposition of hyaluronic acid, chondroitin
- non-pitting
- Characteristic facies: swollen lips, broad nose, macroglossia, and puffy eyelids, hands, feet
- Nerve entrapment: carpal tunnel, facial palsy
HYPOTHYROIDISM AND THE SKIN

Nonspecific changes

• Xerosis
HYPOTHYROIDISM AND THE SKIN

Nonspecific changes

- Xerosis
- palmoplantar keratoderma
Hypothyroidism and the Skin

Nonspecific changes

- Xerosis
- Palmoplantar keratoderma
- Madarosis: loss of lateral 1/3 of brows
- Carotieimia, poor wound healing, clotting
HYPOTHYROIDISM AND THE SKIN

Nonspecific changes

- Xerosis
- Palmoplantar keratoderma
- Madarosis: loss of lateral 1/3 of brows
- Caroteimia, poor wound healing, clotting
CUTANEOUS MANIFESTATIONS OF DIABETES

• 30% of patients with DM develop skin lesions
  • Type I patients get more autoimmune-type lesions
  • Type II patients get more cutaneous infections
• May be the first presenting sign
• Approach:
  • Skin diseases associated with DM
  • Cutaneous infections
  • Cutaneous manifestations of diabetic complications
  • Skin reactions to diabetic treatment
CUTANEOUS MANIFESTATIONS OF DIABETES

• 30% of patients with DM develop skin lesions
  • Type I patients: more autoimmune-type lesions (vitiligo)
  • Type II patients: more cutaneous infections
• May be the first presenting sign
• Approach:
  • Skin diseases associated with DM
  • Cutaneous infections
  • Cutaneous manifestations of diabetic complications
  • Skin reactions to diabetic treatment
Diabetic Dermopathy “shin spots”

- Most common skin finding in diabetes
- Lesions are predominantly situated on the shins, forearms, thighs and over bony prominences
- The color is due to hemosiderin in histiocytes near the vessels
- Trauma and microvascular disease may play a role
Diabetic Bullae

- Painless bullae on non-inflamed base
- Contain clear, sterile fluid
- Trauma and microvascular disease may play a role
CUTANEOUS MANIFESTATIONS OF DIABETES

Acanthosis nigricans
Acanthosis nigricans

- **Mechanism:** Insulin binds to Insulin-like growth factor -> growth of keratinocytes, fibroblasts
- Incidental finding in obesity
- Associated with gastric CA
- Secondary to medications (nicotinic acid, estrogen, or corticosteroids)
- Pineal tumors
- Other endocrine syndromes (PCOS, acromegaly, Cushing’s disease, hypothyroidism)
Scleredema diabeticorum

- Painless, symmetric, woody “peau d’orange” induration
- Upper back and neck
NLD: Necrobiosis lipoidica diabeticorum

- atrophic, telangiectatic plaques
- yellow-brown
- 20% of NLD patients have diabetes
Eruptive Xanthomas

- Sudden crops on firm, non-tender yellow papules with a red rim on extensors
- Slowly resolve when the diabetes is properly managed
- Hypertriglyceridemia >2000mg/dl
- Secondary to EtOH, estrogens
CUTANEOUS MANIFESTATIONS OF DIABETES

Candidiasis in DM

- Intertriginous areas
  - “satellite” lesions
- Angular cheilitis:
  - White, curdlike material adherent to erythematous, fissured
  - oral commisure;
- Median rhomboid glossitis
  - middle of tongue
- Chronic paronychia
  - fingernails
- Erosio interdigitale blastomycetia
  - fissures in finger web spaces
CUTANEOUS MANIFESTATIONS OF DIABETES

Erytrasma

- *Corynebacterium minutissimum*
- Well demarcated red or brown patches
- Topical clindamycin
Rhinocerebral mucormycosis
• Uncontrolled diabetics with ketosis
• Involves the turbinates, septum, palate, maxillary and ethmoid sinuses
• Headache, fever, lethargy, nasal congestion and facial ocular pain
• Treatment:
  • Correction of ketosis
  • Debridement
  • IV antifungal agents
• Mortality ranges from 15-34%
CUTANEOUS REACTIONS TO INSULIN

Lipoatrophy and lipodystrophy

- **Lipoatrophy**
  - Circumscribed depressed areas of skin at the insulin injection site 6-24 months after starting insulin

- **Lipodystrophy**
  - Soft dermal nodules that resemble lipomas at sites of frequent injection
  - May be a response to the lipogenic action of insulin
  - Treat and prevent by rotating sites of injection
SKIN FINDINGS OF RENAL FAILURE

Findings

• General
  • Xerosis, Pruritus
  • Pigmentary alteration
  • Nail Changes, Hair Changes
SKIN FINDINGS OF RENAL FAILURE

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“Uremic Frost”

• Very Rare
  • blood urea nitrogen level of more than 250-300 mg/dl
  • frequent in the pre-dialysis era
Findings of Renal Failure

Findings

- General
  - Xerosis, Pruritus
  - Pigmentary alteration
  - Nail Changes, Hair Changes
SKIN FINDINGS OF RENAL FAILURE

Findings

- General
  - Xerosis, Pruritus
  - Pigmentary alteration
  - Nail Changes, Hair Changes
- Acquired perforating disorder
- Bullous disease of dialysis
- Calcinosis cutis (metastatic)
- Calciphylaxis
- Nephrogenic systemic fibrosis
SKIN FINDINGS OF RENAL FAILURE

Findings

• General
  • Xerosis, Pruritus
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- Calcinosi
  - Cutis (metastatic)
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SKIN FINDINGS OF RENAL FAILURE

Perforating dermatosis

- Primary diseases: rare (Kyrlies, Elastosis Perforans serpiginosa)
- Secondary to:
  - Renal Failure (worse in diabetics)
SKIN FINDINGS OF RENAL FAILURE

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*Hands of a transfusion-dependent patient on long-term hemodialysis. Several uramic-related cutaneous disorders are visible. The pigmentary alteration results from retained urochromes and hemosiderin deposition. The large bullae are consistent with either porphyria cutanea tarda or the bullous disease of dialysis. All nails show the distal brown-red and proximal white coloring of half-and-half nails.*
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- **Calciphylaxis**
- Nephrogenic systemic fibrosis
Metastatic dermal calcification

- Arteriopathy —> gangrene
- Findings:
  - Angular ulcerations
  - Very painful
  - Elevated PTH
  - High mortality
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• Nephrogenic systemic fibrosis
Scleroderma + Contractures

- Peau d’orange
- Very firm skin
  - advancing arcuate edges develop on limbs and trunk
  - relative sparing of head neck
- Specific to radiography contrast

NEPHROGENIC FIBROSING DERMOPATHY (NFD)
Not true porphyria

- Normal blood urine testing
- Associated with renal disease
- Associated with medications
  - NSAIDS
  - Dapsone - Furosemide - Nalidixic Acid - Tetracycline
  - Pyridoxine
SLIDES OF MY LP/LE CASE

• Why is a Mohs surgeon son concerned about skin signs of systemic disease???
Not true porphyria
• Normal blood urine testing
• Associated with renal disease
• Associated with medications
  • NSAIDS
  • Dapsone - Furosemide - Nalidixic Acid - Tetracycline
  - Pyridoxine
MALIGNANCY- ASSOCIATED DERMATOSES

Findings

• General
  • Xerosis, Pruritus
  • Pigmentary alteration
  • Nail Changes, Hair Changes

• Acquired perforating disorder

• Bullous disease of dialysis

• Calcinosis cutis (metastatic)

• Calciphylaxis

• Nephrogenic systemic fibrosis
SKIN TUMORS THAT IDENTIFY UNDERLYING DISEASE

Angiokeratomas

- Multiple on the toes: Fabry’s disease (storae diease)
- very rare childhood disease
SKIN TUMORS THAT IDENTIFY UNDERLYING DISEASE

Muir-Torre Syndrome

- AD disorder
- sebaceous neoplasms
- multiple keratoacanthomas
- internal malignancy
- PTEN mutation
- colon CA screening age 25
Birt-Hogg-Dubee

- Specific benign skin tumors:
  - fibrofolliculomas, others
  1) Oncocytomas
  2) Chromophobe adenomas
  3) Papillary renal cell carcinoma
SKIN FINDINGS THAT IDENTIFY UNDERLYING MALIGNANCY

Malignant down

- Growth of fine lanugo hairs
- Soft non-pigmented hair on face
- With time, may become more coarse
- Exclude drugs, anorexia and endocrine disorders
- Associated with lung CA
SKIN FINDINGS THAT IDENTIFY UNDERLYING MALIGNANCY

**Acquired ichthyosis**

- If new onset in adulthood, consider: underlying malignancy
- Thick plaques of scale
- Hodgkins > breast CA
SKIN FINDINGS THAT IDENTIFY UNDERLYING MALIGNANCY

Malignant acanthosis nigricans

- VERY severe form of AN
- thick plaques of scale
- “tripe palms”
- associated “oral florid papillomatosis”
- Onset > age 40
- GI malignancies
SKIN FINDINGS THAT IDENTIFY UNDERLYING MALIGNANCY

Sweet’s syndrome

- Acute onset of
  - fever
  - leukocytosis
  - tender, non-pruritic, erythematous plaques or papules
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  - fever
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- Non-malignancy:
  - Associated with infection of the upper respiratory and/or gastrointestinal tract,
  - IBD
  - pregnancy
  - GM-CSF administration

- Malignancy-associated:
  - hematologic malignancies (10-20%)
  - also solid tumors
SKIN FINDINGS THAT IDENTIFY UNDERLING MALIGNANCY

Dermatomyositis

- Can be a marker for internal neoplasia (may predate the diagnosis of the cancer)
- 10-50% of pts
- OVARIAN, colorectal, lung, pancreatic, stomach, and lymphoma
- The diagnosis of malignancy is usually made within 1 year, but can be several years later
SKIN FINDINGS THAT IDENTIFY UNDERLYING MALIGNANCY

Scleromyxedema

- Chronic, progressive condition characterized by dermal fibrosis and mucinosis with **normal thyroid** function
- Usually associated with paraproteinemia
- Often MGUS, progression to multiple myeloma rare
SKIN TUMORS THAT IDENTIFY UNDERLYING DISEASE

AngioMyoLipomas

- Often multiple user extremity
- Common finding
- Underlying Renal disease

Leiomyomas

- Common finding
- Reed syndrome: uterine bleeding, renal cell carcinoma
PARANEOPLASTIC PEMPHIGUS

Extensive mucosal involvement

MC associated malignancy with paraneoplastic pemphigus

NHL
CLL

Ass. w/Thymoma & castleman's disease
Thank you

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