A Case of Mistaken Identity: Identifying Rheumatology Mimics

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13 February 2016
Disclosure of Financial Relationships

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Has no relationships with any entity producing, marketing, re-selling, or distributing health care goods or services consumed by, or used on, patients.
Case 1.

• A 50 year old woman presents with 3 months of progressively worsening hand puffiness and the abrupt appearance of a painful lesion on the tip of the right second finger extruding chalky material

• PMH:
  – GERD x 10 years
  – digital blanching with cold exposure x 2 years
Case 1.
Case 1. Skin Biopsy

Normal Skin Biopsy

- Epidermis
- Dermis

H&E

Collagen Stains Pink

Case 1. Skin Biopsy

- Epidermis
- Thickened dermis
- Increased collagen
- Collagen replaces fat

H&E

Collagen Stains Pink

Gomori trichrome

Collagen Stains Blue
The most likely diagnosis is

1. Scleroderma
2. Scleredema
3. Scleromyxedema
4. Limited Joint Mobility Syndrome
5. Eosinophilic Fasciitis
Scleroderma

• Idiopathic autoimmune disorder characterized by vasospasm, vascular damage and excessive fibrosis in skin and internal organs

• Almost all scleroderma patients have
  – Raynaud’s Phenomenon
  – + ANA
  – Abnormal Nailfold Capillaroscopy
Nailfold Capillaroscopy

Normal

Scleroderma: Capillary Dilatation at Cuticles

American College of Rheumatology Image Bank; Wikipedia; Microvision
Scleroderma Nomenclature

• Clinical types:
  – Limited cutaneous systemic sclerosis (formerly CREST)
    • Limited skin involvement
    • Pulmonary hypertension
    • Centromere pattern +ANA
  – Diffuse cutaneous systemic sclerosis
    • Rapidly progressive skin involvement
    • Interstitial lung disease
    • Renal crisis
    • Nucleolar pattern +ANA
    • Anti-topoisomerase (anti-scl-70) antibody
Limited Cutaneous Systemic Sclerosis

- Calcinosis
- Raynaud’s
- Sclerodactyly
- Telangectasias

Esophageal Dysmotility
Diffuse Cutaneous Systemic Sclerosis

Rapid, Severe Skin Involvement

Interstitial Lung Disease

Scleroderma Renal Crisis

American College of Rheumatology Image Bank
Case 2.

- A 29 year old woman presents with diffuse brawny thickening and tightening of the skin of the face over the last 5 days

- PMH:
  - Resolving upper respiratory tract infection
Case 2.

Woody induration of forehead skin
Case 2. Skin Biopsy

Thickened dermis,
Swollen appearance of collagen bundles,
clear spaces between collagen bundles

Spaces between collagen bundles filled with mucopolysaccharides

H & E
Collagen Stains Pink

Alcian Blue
Mucin Stains Blue

American College of Rheumatology Image Bank
The most likely diagnosis is
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2. Scleredema
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Scleredema

- Diffuse, woody induration most often neck, upper back, shoulders
- Hands and feet are spared
- 3 clinical types:
  - Following influenza, measles, mumps, varicella, *streptococcus* (scleredema adultorum); abrupt onset; children, young adults; **self limited**
  - Associated with **paraproteinemia**; slow onset
  - Associated with poorly controlled, middle aged, obese **DM** (scleredema diabeticorum); slow onset
- **Labs:** ASO, SPEP, HgbA1c
- **Rx:** Phototherapy, PT
Case 3.

- A 46 year old man presents with a papular, pruritic skin eruption with marked skin thickening of his face, chest, arms, legs progressing over the last year

- PMH:
  - plasma cell dyscrasia
Case 3.
Case 3. Skin Biopsy

1. Thickened, irregular collagen bundles
2. Wide intercollagenous spaces
3. Interstitial dermal mucin
4. Increased number fibroblasts
The most likely diagnosis is
1. Scleroderma
2. Scleredema
3. Scleromyxedema
4. Limited Joint Mobility Syndrome
5. Eosinophilic Fasciitis
Scleromyxedema

- Primary cutaneous mucinosis → Thickened skin with clusters of waxy papules that coalesce
- Very rare, middle age
- Face, neck, upper trunk, arms, hands
- Thickened skin forms deep furrows → leonine (lion-like) facies
- Extensive skin involvement → joint pain, ↓ mobility, joint contractures
- Systemic involvement → disabling neurologic, myopathic, pulmonary disease; esophageal dysmotility, dysphagia
- Labs: 80% of patients have a paraprotein (IgG lambda); low titer +ANA can be seen
- 10% develop multiple myeloma
Leonine Facies of Scleromyxedema

Indian J Dermatol Venereol Leprol 2010;76:592
Myxedema Facies

Clinical Consequences:
• Puffy Eyes
• Broad Nose
• Thickened Lips
• Macroglossia

Pathology:
Accumulation of matrix glycosaminoglycans in the interstitial spaces of many tissues with associated tissue water retention

J Crows.com; Clinics in Dermatology (2006) 24, 247–255
Case 4.

• A 63 year old man presents with tight, waxy skin over the dorsum of the hands. He complains of decreased grip strength and can no longer use a screwdriver for minor home repairs. He had noticed an area of cord-like, painless thickening of the skin in his right palm over the last several years.

• On physical examination, he is unable to bring the palms of his hands completely together.

• PMH:
  – poorly controlled non-insulin dependent diabetes mellitus x 25 year
Prayer Sign

Duputryyn’s Contracture

Case 4. Skin biopsy
Increased collagen
Reduced skin appendages
The most likely diagnosis is
1. Scleroderma
2. Scleredema
3. Scleromyxedema
4. Limited Joint Mobility Syndrome
5. Eosinophilic Fasciitis
Limited Joint Mobility Syndrome (formerly Diabetic Cheiroarthropathy)

- Occurs in type 1 and type 2 diabetes
- More common with more longstanding disease
- ↑Glycosylated collagen → ↑stiffness, ↓ degradation, ↑accumulation
- Commonly affects joints of upper extremities and feet, esp hands
- Limited range of motion on physical examination
- May be at greater risk for microangiopathy elsewhere
What does Cheiro- Mean?

Cheiro = Hand

**Cheiro**
William John Warner,
(November 1, 1866 – October 8, 1936)
Irish astrologer

**Palm reader:**
Thomas Edison, Mark Twain,
Sarah Bernhardt, Mata Hari,
Oscar Wilde, Grover Cleveland...
Limited Joint Mobility Syndrome
(formerly Diabetic Cheiroarthropathy)

- Occurs in type 1 and type 2 diabetes
- More common with more longstanding disease
- $\uparrow$ Glycosylated collagen $\rightarrow$ $\uparrow$ stiffness, $\downarrow$ degradation, $\uparrow$ accumulation
- Commonly affects joints of upper extremities and feet, esp hands
- Limited range of motion on physical examination
- May be at greater risk for microangiopathy elsewhere
Case 5.

• A 47 year old woman presents with diffuse thickening of the skin of the arms and legs after beginning a rigorous program of daily running and weight lifting.

• On physical examination, the skin of the arms appears grossly normal at rest; when the arms are elevated, deep grooves become evident along the course of superficial veins.

• The skin of the calves is indurated and has a peau d’orange appearance.
Case 5.

Groove Sign

Peau d’Orange
Case 5. Skin Biopsy

A. Normal epidermis and dermis
B. Normal subcutaneous fat
C. Normal skeletal muscle
D. Markedly thickened fascia
E. Inflammatory infiltrates in fascia
The most likely diagnosis is
1. Scleroderma
2. Scleredema
3. Scleromyxedema
4. Limited Joint Mobility Syndrome
5. Eosinophilic Fasciitis
Eosinophilic Fasciitis

• Middle age (40s, 50s)
• Described after **extreme exercise, trauma**
• Woody skin, edema and induration of **extremities**
• Face, fingers spared
• **Limited range of motion** at wrists and ankles with fascial involvement
• No internal organ involvement
• **Labs**: eosinophilia, ↑ ESR/CRP, hypergammaglobulinemia, ↑ aldolase
• **Rx**: steroids, immunosuppressives

*Curr Rheumatol Rep (2012) 14:39–46*
MRI in Eosinophilic Fasciitis

Gadolinium enhanced images of proximal thighs
a. T1 weighted
b. T2 weighted

Normal

Case 6.

• A 57 year old man presents with rapidly progressive, painful skin thickening in the feet and distal calves; at first, the skin seemed red, puffy and itchy; over time, the thickening moved up to the thighs then involved the hands, then forearms, then upper arms; eventually, shiny, brawny hyperpigmentation developed, epidermal atrophy and hair loss occurred in affected skin and flexion contractures of the digits began to appear.

• PMH:
  – Hypertension x 20 years
  – Chronic kidney disease x 10 years
  – Hemodialysis x 5 years
Case 6.
Case 6. Skin Biopsy

Increased mucin and collagen
Little inflammation

Nephrogenic Systemic Fibrosis

- Chronic kidney disease + gadolinium exposure
- Never involves face;
- No Raynaud’s, no periungual capillary dilatation, no telangectasias

- Unique eye finding:
slightly raised, **yellow plaques** on the sclera, adjacent to the iris, often accompanied by conjunctival injection
Yellow scleral plaque with conjunctival injection
Thickened Skin?

• Perform a skin biopsy
• Evaluate for diabetes
  – Scleredema (face)
  – Limited Joint Mobility Syndrome (hands)
• Evaluate for paraproteins
  – Scleredema (face)
  – Scleromyxedema (face & hands)
• Evaluate for eosinophilia
  – Eosinophilic fasciitis (extremities)
• Consider gadolinium exposure if chronic kidney disease is present
  – Nephrogenic systemic fibrosis (diffuse)
Case 7.

• A 20 year old woman presents with knee and back pain that has progressively worsened over the last 2 years

• On physical examination, she has no warmth, erythema, swelling of any joints; no knee effusions

• She is able to touch her thumbs to her forearms; she is able to extend her fingers to 90° at the MCPs; her elbows and knees extend beyond 180°; and she is able to touch her palms flat against the floor
Case 7.
The most likely diagnosis is
1. Marfan Syndrome
2. Ehlers Danlos Syndrome
3. Benign Joint Hypermobility Syndrome
Hypermobility

• Single Hypermobile Joint (15-20% of population)
• Generalized Hypermobile Joints (Beighton >4)
  – Asymptomatic
    • 10-30% of population
    • Large heritable component: monozygotic>>dizygotic
    • Increased in ballet dancers, gymnasts
Beighton score for joint laxity*

<table>
<thead>
<tr>
<th>Specific joint laxity</th>
<th>Left</th>
<th>Right</th>
</tr>
</thead>
<tbody>
<tr>
<td>Passive apposition of thumb to forearm</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Passive hyperextension of fingers</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Active hyperextension of elbow &gt;10 degrees</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Active hyperextension of knee &gt;10 degrees</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Ability to flex spine and place palms to floor without bending knees</td>
<td></td>
<td>1</td>
</tr>
</tbody>
</table>

* This score is based upon joint laxity of the above nine anatomic sites. It is calculated by adding all points, with nine being the highest total possible score. A score of four or higher is generally considered an indication of generalized joint laxity.

Beighton > 4
“Generalized Hypermobile Joints” or “Benign Joint Hypermobility Syndrome” or “Double Jointed”
Beighton Criteria  (score of 4/9 or more)

The Three Graces (1639)
Peter Paul Rubens

Trendelenburg Sign:
when R hip and R knee are flexed
R hip drops
due to L hip abductor weakness

Lurch toward affected side
Contralateral hemi-pelvis drop

Hyperlordosis
Scoliosis
Hip Abductor Weakness “Trendelenburg Sign”

Flat arches

Trendelenburg gait
Hypermobility

• **Benign Joint Hypermobility Syndrome**
  – Arthralgia, use-related joint pain, myalgia, dislocations
  – ? ↑ Risk early OA
  – Association with MVP, back pain, FM, TMJ, CTS, anxiety, cognitive impairment, palpitations, tremulousness, fatigue
  – “Minor criteria” of “Marfanoid habitus”, “abnormal skin”, “eye abnormalities”, uterine or rectal prolapse make diagnosis difficult
Differential Diagnosis: Hypermobility

- Marfan Syndrome
- Ehlers Danlos Syndrome
Marfan Syndrome

- One of the most common inherited disorders of connective tissue, incidence of 1:3000-5000
- Autosomal dominant but 25% represent new gene mutations
- Fibrillin-1 (essential to normal elastic fibrillogenesis) gene mutations (~100) in most
- Transforming growth factor β receptor mutation in 10% (premature fusion of the skull, widely spaced eyes, cleft palate, split uvula, tortuous arteries, aortic rupture, structural heart disease)

- Considerable variability in phenotype
Major Criteria for Marfan Syndrome

• Skeletal system features (NEED 4):
  – Reduced upper to lower body segment ratio or arm span exceeding height
  – Arachnodactyly of fingers and toes, with positive thumb and wrist signs
  – Scoliosis >20º or spondylolisthesis
  – Medial displacement of the medial malleolus causing pes planus
  – Reduced extension at the elbows (<170º)
  – Pectus carinatum
  – Pectus excavatum requiring surgery
  – Protrusio acetabuli of any degree (ascertained on radiographs)
Arm Span Exceeding Height

Wingspan
Michael Phelps is 6 feet 4 and 195 pounds, but his wingspan measures nearly 6-7. That length means that he needs fewer strokes to cover a 50-meter pool than a 5-10 man.

Lung capacity
Physiologists measure lactate levels in the blood to determine how efficiently swimmers produce energy. No tester has lactate levels as low as Phelps.

Long trunk, short legs
Phelps has a long trunk and relatively short legs. “That allows me to plane in the water,” he said.

Flexibility
Phelps can hyperextend his elbows, knees and ankles. That range of motion allows him to explore angles in his stroke technique that few other swimmers can.

Illustrative of the finding; no implication regarding genetics should be drawn!
Major Criteria for Marfan Syndrome

• Skeletal system features (NEED 4):
  – Reduced upper to lower body segment ratio or arm span exceeding height
  – *Arachnodactyly of fingers and toes, with positive thumb and wrist signs*
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Arachnodactyly

Positive Thumb Sign

Positive Wrist Sign

Med J Austral 2006;184:627-631
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Medial Displacement of Medial Malleolus → Pes Planus (“flat feet”)
Major Criteria for Marfan Syndrome

• Skeletal system features (NEED 4):
  – Reduced upper to lower body segment ratio or arm span exceeding height
  – Arachnodactyly of fingers and toes, with positive thumb and wrist signs
  – Scoliosis >20º or spondylolisthesis
  – Medial displacement of the medial malleolus causing pes planus
  – **Reduced extension at the elbows (<170º)**
    – Pectus carinatum
    – Pectus excavatum requiring surgery
    – Protrusio acetabuli of any degree (ascertained on radiographs)
Reduced Extension of the Elbow

The opposite of Benign Hypermobility Syndrome......
Major Criteria for Marfan Syndrome

• Skeletal system features (NEED 4):
  – Reduced upper to lower body segment ratio or arm span exceeding height
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  – Pectus excavatum requiring surgery
  – Protrusio acetabuli of any degree (ascertained on radiographs)
Protrusio Acetabuli
Major Criteria for Marfan Syndrome

• **Cardiovascular system feature** (NEED 1):
  – dilation of the aorta involving the sinuses of Valsalva with or without aortic regurgitation
  – ascending aortic dissection

• **Ocular system feature**: Ectopia lentis (detected by slit lamp examination)

• **Dura feature**: Dural ectasia (detected by CT or MRI)
Major Criteria for Marfan Syndrome

• **Family or genetic history (NEED 1):**
  1st degree relative who meets diagnostic criteria
  
  OR presence of FBN1 mutation known to cause the Marfan syndrome
  
  OR presence of a haplotype around FBN1
Ehlers Danlos Syndrome

• Group of genetic diseases characterized by joint hypermobility, skin hyperextensibility, tissue fragility
• Incidence 1 in 5000-20,000
• Mutations in collagen genes affecting synthesis and processing
• Multiple subtypes
Classic EDS

- Hypermobility by Beighton Score (≥4/9)
- Recurrent joint dislocations
- Subluxations of shoulder, patella, and temporomandibular joints
- Joint effusions, pain
- Scoliosis
- Early OA
  (same as in Hypermobility EDS)

- Doughy, hyperextensible skin
Vascular EDS

• Absence of large joint hypermobility
• Mild skin hyperextensibility
• Risk of vascular or visceral rupture is 80% by age 40
• Can mimic large or medium vessel vasculitis
Cervical artery dissections and type A aortic dissection in a family with a novel missense COL3A1 mutation of vascular type Ehlers–Danlos syndrome

European Journal of Medical Genetics, Volume 58, Issue 11, 2015, 634–636

http://dx.doi.org/10.1016/j.ejmg.2015.10.009
Fig 3. Celiac artery aneurysm. Preoperative selective arteriogram of (A) a poststenotic celiac artery aneurysm and (B) a postoperative computed tomographic arteriography image after resection and primary end to end anastomosis of the proximal hepatic to proxim...

Frank M. Davis, Jonathan L. Eliason, Santhi K. Ganesh, Neal B. Blatt, James C. Stanley, Dawn M. Coleman

Pediatric nonaortic arterial aneurysms


http://dx.doi.org/10.1016/j.jvs.2015.08.099
Marfan vs EDS

Marfan
- Joint hypermobility
- Scoliosis
- Disproportionate tall stature
- Dislocation of the lens
- Pectus carinatum
- Progressive aortic dilatation

Classic or Hypermobility EDS
- Joint hypermobility
- Scoliosis
- Mild aortic dilatation
When to Seek Genetics Consultation

• Joint hypermobility, multiple joint dislocations
• Translucent skin, poor wound healing, easy bruising, and unusual scars
• Spontaneous rupture of an organ or dissection of a blood vessel
• Family history
Case 8.

43 year old man presents with a 3 week history of purpuric skin lesions on the extremities that progresses to ulceration and necrosis

ROS: No fever, conjunctival injection, visual symptoms, rhinorrhea, nasal congestion, sinus pain or discharge, epistaxis, ear fullness, cough, SOB, CP, hemoptysis

PMH: none
Case 8. Physical Examination: Lesion on Thigh
Case 8. Laboratory Studies

ESR 28
ANA negative
CRP 48.3
Cryoglobulins negative
C4 16
Normal C3, CBC, CK
pANCA + >1:640
Creatinine 1.28
MPO+ UA 3+ protein, 1+ blood

Skin biopsy: acute neutrophilic necrotizing vasculitis involving the small and medium-vessel with fibrinoid necrosis, immunofluorescence + C3 + IgM granular staining dermal vessels
Case 8. Course

- Started on prednisone 60 mg daily
- Tapered over a 2 week period
- 5 weeks later, “dark spots” appear on ears and resolve
- 6 weeks later, painful pink lesions appear on ears and lower extremities
- Lesions progress to purple then black, some form hemorrhagic bullae
Case 8. Physical Examination 6 Weeks Later: Lesions on Extremities, Ear
Case 8. Laboratory Studies Second Admission

ESR 43

pANCA+ 1:640  Creatinine 1.54

MPO+  UA 3+ protein, 1+ blood

Levamisole-Induced Vasculitis

- 70% of cocaine in US cut with levamisole
- Associated with ANCA+ vasculitis
- Most common manifestations: arthralgia, skin lesions
- Most have constitutional symptoms: fever, night sweats, weight loss, myalgias
- 100% ANCA+ MPO+, 70% ANCA + PR3+
- Leukopenia, hematuria, proteinuria seen
ANCA can be positive in other disorders

- Preeclampsia and eclampsia
- Acute infectious mononucleosis
- Acute parvovirus B19 infection
- Leprosy
- Malaria
- Subacute bacterial endocarditis
- Cocaine-induced osteochondral destruction (cocaine-induced midline destructive lesions)
- Buerger's disease
- Diffuse alveolar hemorrhage
- Chronic graft-versus-host disease
- Autoimmune hepatitis
- Crohn's disease, ulcerative colitis
Is It Large Vessel Vasculitis?

• Review family history:
  – Marfan, Vascular EDS

• Rule out infection:
  – Blood cultures, echocardiogram
  – PPD, HIV

• Adequately image the involved vasculature
  – MRI/MRA
Case 9.

- 59 year old man with prior history of drug and alcohol abuse presents with recurrent pancreatitis
- Following Whipple procedure developed postoperative gout
- One year later, diagnosed with retroperitoneal fibrosis
- Asked for re-review of surgical specimen
IgG4 Related Disease

• New category for poorly understood group of disorders previously thought unrelated

• Share tumor-like swelling of involved organs due to lymphoplasmacytic infiltrate enriched in IgG4+ plasma cells + variable degrees of fibrosis with characteristic “storiform” pattern

• ↑ serum IgG4
What does “storiform” mean?

Storiform fibrosis
(collagen staining blue)

Mesenteric Biopsy

Renal Biopsy

“cartwheel”

“straw mat”
IgG4 Related Disease

• Autoimmune pancreatitis
• IgG4 Related
  – Orbital pseudolymphoma
  – Chronic sclerosing sialadenitis (Sjogren’s-like disease)
  – Mediastinitis
  – Cholangitis
  – Inflammatory bowel disease
  – Retroperitoneal fibrosis, periaortitis
  – Tubulointerstitial nephritis
IgG4 Related Autoimmune Pancreatitis

- Pancreatic mass
- Mild abdominal pain with or without attacks of acute pancreatitis and chronic pancreatitis
- Pancreatic duct strictures
- Peripancreatic vascular complications

Dx ("HISORt" Criteria):
- Diagnostic **Histology**
- Characteristic **Imaging** on computed tomography and/or pancreatography
- Elevated serum **IgG4 levels** on **SeroLogic testing**
- **Other organ** involvement (sialadenitis, retroperitoneal fibrosis)
- **Response of pancreatic and extrapancreatic manifestations to glucocorticoid therapy**
Gross specimen:
Pancreatic mass in pancreas affected by IgG4 autoimmune pancreatitis

Microscopic specimen:
Biliary obstruction due to periductal lymphoplasmacytic infiltrate
IgG4 Related Disease

- Autoimmune pancreatitis
- IgG4 Related
  - Chronic sclerosing sialadenitis (Sjogren’s-like disease)
  - Orbital pseudolymphoma
  - Mediastinitis
  - Cholangitis
  - Inflammatory bowel disease
  - Retroperitoneal fibrosis, periaortitis
  - Tubulointerstitial nephritis
IgG4 Related Chronic Sclerosing Sialadenitis

Salivary gland biopsy:
Large germinal centers (arrow), dense lymphoplasmacytic inflammation, **Storiform fibrosis**: cellular fibro-inflammatory stroma (arrowhead)
Case 9: Review of Surgical Pathology

- Pancreas: mild chronic inflammation, mostly normal
- CBD: chronic inflammation
- Nonspecific chronic cholangitis
- Peripancreatic lymph node: normal
- Portal lymph nodes: numerous non-caseating epithelioid granulomas suggestive of sarcoidosis
Sarcoidosis and Rheumatic Disease

• Multisystem disease, fatigue
• Lofgren’s (acute ankle periarthritis, erythema nodosa, hilar nodes)
• Hand tenosynovitis
• Sarcoid myopathy (acute and chronic)
• Osseous sarcoidosis (hands, feet)
• Co-occurrence with rheumatic disease (SLE, Sjogren’s, SS, PM, IBM, AS, vasculitis)
• Drug-induced (TNFα inhibitors, interferonα)
Evaluation for Sarcoidosis

- History, including occupational and environmental exposure
- Physical examination
- PA chest radiograph
- PFTs, including spirometry and DLCO
- CBC
- Serum chemistries, including creatinine, calcium, LFTs
- Urinalysis
- Electrocardiogram
- Routine ophthalmologic examination
- PPD
Transbronchial biopsy findings are consistent with the clinical diagnosis of sarcoidosis. The histopathologic specimen shows granulomatous inflammation.

*Courtesy of Talmadge E King Jr, MD.*
Summary

• Abnormal skin findings:

what is the distribution?
**Scleredema**
Neck, Upper Back, Shoulders

**Scleromyxedema**
Face, Neck, Upper Trunk, Arms, Hands

**Eosinophilic Fasciitis**
Extremities

**Nephrogenic Systemic Fibrosis**
Body other than Face
Summary

• Abnormal skin findings: get skin biopsy
  – Skin biopsy findings:
    • Collagen
      – Increased: scleroderma
      – Increased glycosylated collagen: limited joint mobility syndrome (diabetes)
    • Mucin
      – Increased: scleredema
      – With fibroblasts: scleromyxedema
      – With increased collagen: nephrogenic systemic fibrosis
    • Glycosaminoglycans
      – Increased: myxedema
    • Eosinophils
      – Increased: eosinophilic fasciitis
Summary

• Hypermobility: other organs systems?
  – Absence of internal organ involvement:
    • Hypermobile Joint
    • Benign Hypermobility Syndrome
  – Elongated limbs and digits:
    • Marfan
  – Skin hyperextensibility:
    • Ehlers Danlos
Summary

• Vasculitis Mimics
  – Rule out infection and malignancy
  – Tissue biopsy

• IgG 4 Related Disease
  – Serum IgG 4
  – Tissue biopsy
Thank You!
Questions?