

September 2000

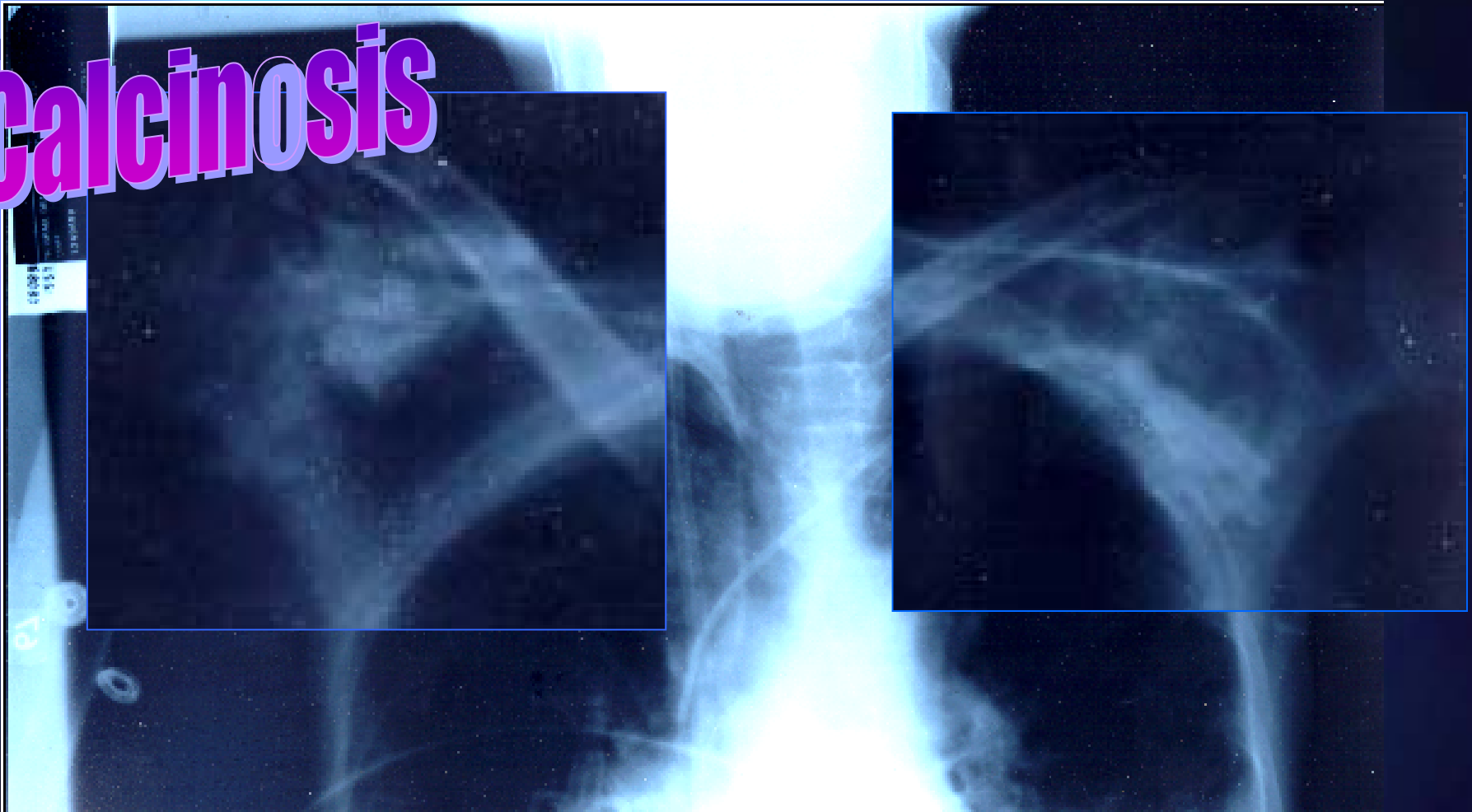
Calcinosis Cutis: Emerging Trends in Dermatoradiology

Peter Lio, Harvard Medical School Year- IV

Gillian Lieberman, MD

"Routine CXR"...

Calcinosis



Special Thanks to Dr. Hochman for this image.

What is Calcinosis?

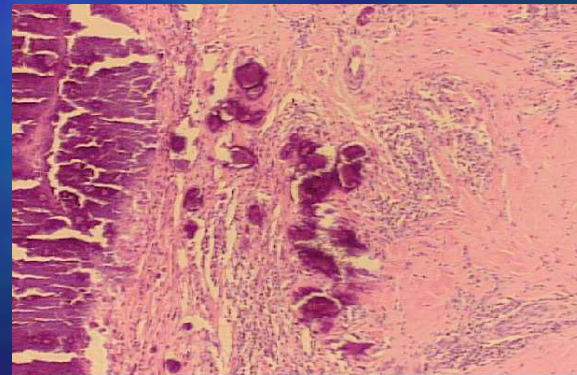
- **Calcinosis** refers to pathologic calcification of the skin and soft tissues.
- It occurs in a variety of systemic and localized conditions.
- The deposits are generally composed of calcium phosphate crystals which is why we can see them on radiographic imaging.

Classical Dermatology

- In classical dermatology, the skin lesion is correlated with pathologic findings in the skin.



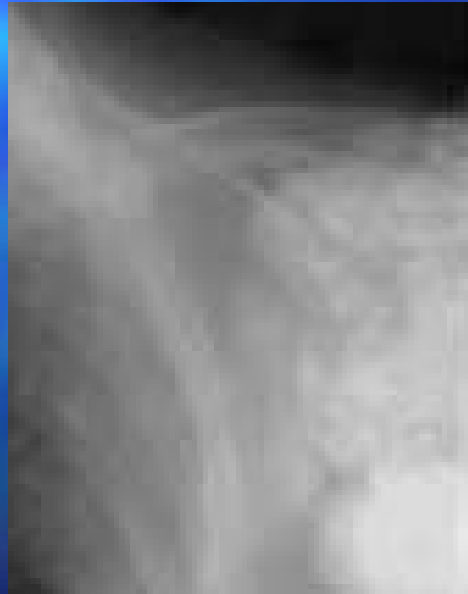
<http://www.dermis.net/bilddb/diagnose/englisch/i709330.htm>



<http://www.autopsydb.org/dermpath.htm>

"Dermatoradiology"

- In dermatoradiology, the nature of the heavy metal Calcium and its salts allow for radiographic visualization.



Classic Dermatology

- 13 year old female with firm, white-yellow nodules on an erythematous base in her thumb.



From: <http://www.dermis.net/bilddb/diagnose/englisch/i709330.htm>

Dermatoradiology

- Note the areas of subcutaneous calcifications, especially at the first and third phalanges.
- This radiograph is from a different patient but you get the idea...



From: <http://www.rad.washington.edu/maintf/cases/unk39/answers.html>

Calcinosis: The Breakdown

■ Ectopic calcification (i.e. calcification outside of bone) is generally classified into two groups:

Metastatic calcifications

- Result from persistent hypercalcemia or hyperphosphatemia
- $\text{Ca} \times \text{PO}_4 > 70$

Dystrophic calcifications

- Follow a local metabolic or degenerative tissue abnormality

Descriptors

- Calcinosis may be isolated to a small area:

*Calcinosis
Circumscripta*

- Calcinosis may be diffuse:

*Calcinosis
Universalis*

Pathophysiology

- Poorly understood...
- Theories include:
 - Local elevations in alkaline phosphatase activity which lead to hydrolysis of extracellular pyrophosphates that normally inhibit calcium deposition.
 - Local tissue injury which increases cell membrane permeability, thus allowing cytosolic influx of sufficient calcium to exceed the capacity of mitochondria to sequester calcium and phosphate, and leading to the precipitation of cytosolic CaPO_4 .

Differential Diagnosis (Ddx)

- Dystrophic (95-98%)
- Metastatic Calcification (1-2%)
- Idiopathic Tumoral Calcinosis (<1%)
- Metastatic Osteosarcoma (<<1%)
- 1° Soft Tissue Osteosarcoma (<<1%)
- Heterotopic Ossification
- Calciophylaxis
- Calcium Pyrophosphate Dihydrate Deposition Disease
CPPD(1-2%)

Ddx Dystrophic Calcification

■ (pneumonic) VINDICATE:

- Vascular
 - Venous insufficiency
- Infection
 - Parasites*
- Neoplasm
 - Primary bone-forming tumor
 - Tumor necrosis
- Drugs
 - Vitamin D
- Autoimmune
 - Dermatomyositis
 - Scleroderma
 - SLE
- Trauma

*“Rice grain” = cysticercosis

* Small crecentic = dracunculiasis

Differential Diagnosis: A Radiologist's Perspective

- Idiopathic Tumoral Calcinosis
 - Mass-like calcific deposits about joints
 - Usually normal or slightly elevated serum calcium and phosphate
- Calcinosis universalis
 - Associated with scleroderma or dermatomyositis
 - Calcification is usually thin, plaque-like and in skin and subcutaneous tissues
 - No large, lobular masses
- Calcinosis circumscripta
 - 40% are associated with scleroderma, dermatomyositis or Raynaud's
 - Calcifications are thin, and occur in fingertips and "toe-tips"
- Milk-alkali syndrome
 - Large, calcified periarticular masses
 - Calcification also of lung, kidneys, vessels
- Metastatic calcification (hyperparathyroidism and chronic renal disease)
 - Serum PTH and calcium levels abnormal
 - Soft tissue calcifications are usually fine and punctate
- Heterotopic ossification
 - Often associated with some predisposing condition
 - Prior musculoskeletal trauma
 - Prior surgery
- Scleroderma
 - Usually thin calcifications of hands, feet
- Dermatomyositis
 - Usually associated with fine, reticular calcifications

Menu of radiologic tests for imaging ectopic calcifications

- Radiographic plain film
- CT scan- most sensitive
- RN bone scan
- MRI- sometimes

PLAIN FILMS

Polymyositis

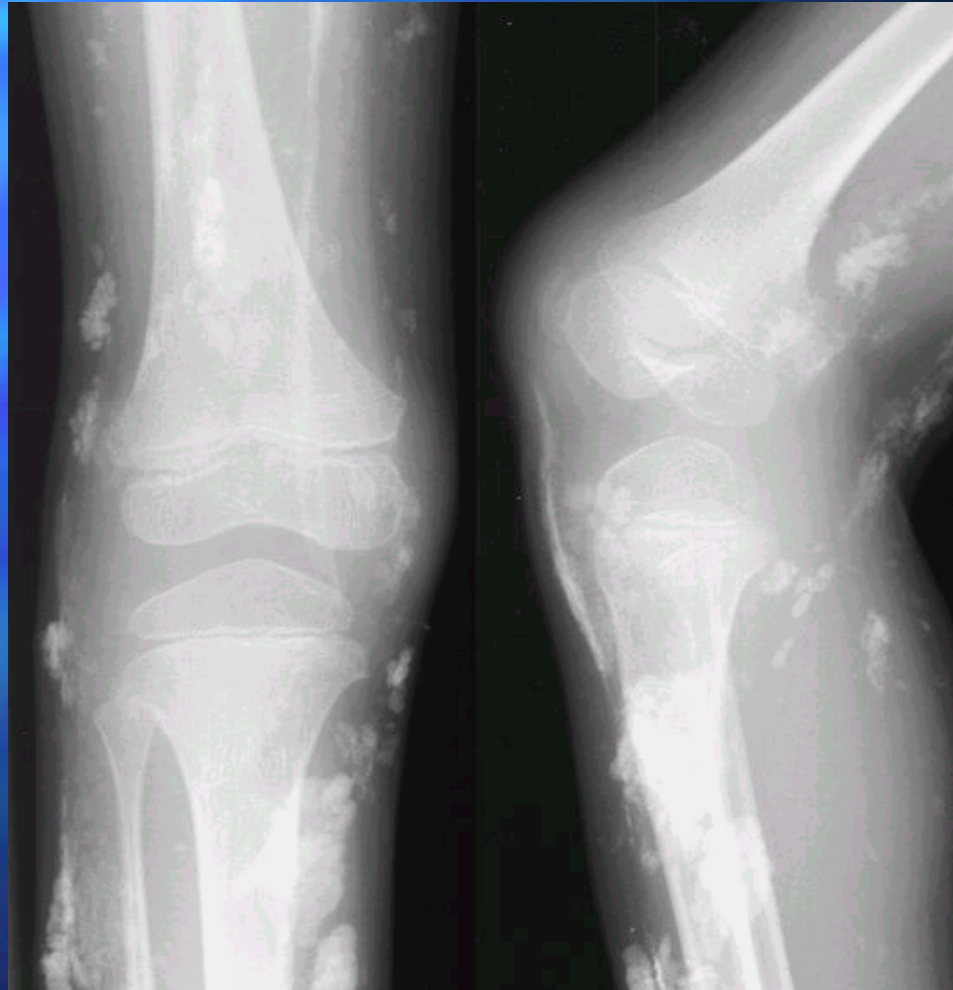
- Dystrophic calcinosis:
Plain film of the hand of a woman with polymyositis.
- Note extensive calcinosis involving both subcutaneous tissues and tendon sheaths.



PLAIN FILMS

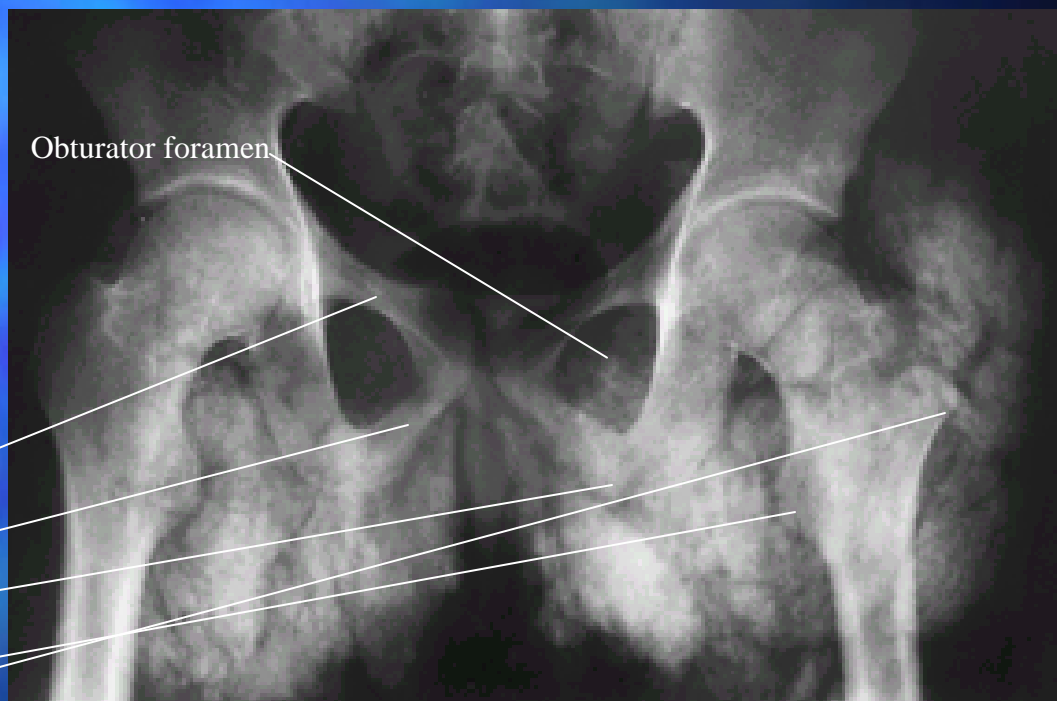
Juvenile Dermatomyositis

- 5-year old female with an 18 month history of difficulty walking.
- Note extensive soft tissue calcifications:
 - Not in a Vascular Pattern
 - Not attached to bone



Juvenile Dermatomyositis

- 18 year old boy with Dermatomyositis: extensive deep calcified deposits are seen about each hip and extending into the thighs.



Superior ramus of pubis

Inferior ramus of pubis

Ischial tuberosity

Lesser trochanter

Greater trochanter

Eddy MC, Leelawattana R, McAlister WH, Whyte MP. Calcinosis universalis complicating juvenile dermatomyositis: resolution during probenecid therapy. *J Clin Endocrinol Metab.* 1997 Nov;82(11):3536-42.

Juvenile Dermatomyositis

- Same 18 year old boy with Dermatomyositis: extensive deep calcified deposits are seen about the shoulder extending into the arm.
- The clustering around joints mimics tumoral calcinosis.



Idiopathic Tumoral Calcinosis

- Note extensive soft tissue calcifications.
- Multiple lobulated, dense calcifications are noted around joints in the hands and wrist and in other soft tissues.
- Linear collection of calcification tracking along a tendon sheath.



Idiopathic Tumoral Calcinosis

- The soft tissue calcifications may be single or multiple, lobulated, cystic or solid.
- Often occurs at hips, elbows and shoulders.
- The masses may range in size from 1 to 20 cm in diameter.
- Family history in 30 - 40 % of cases.
- Onset during childhood or adolescence.
- May be increased prevalence in patients of African descent.

RADIONUCLIDE BONE SCAN

Idiopathic Tumoral Calcinosis

- Bone scan demonstrates prominent uptake in soft tissues, especially around the pelvis and shoulders



Idiopathic Tumoral Calcinosis

- What type of study is this?

- MRI...

Why?...

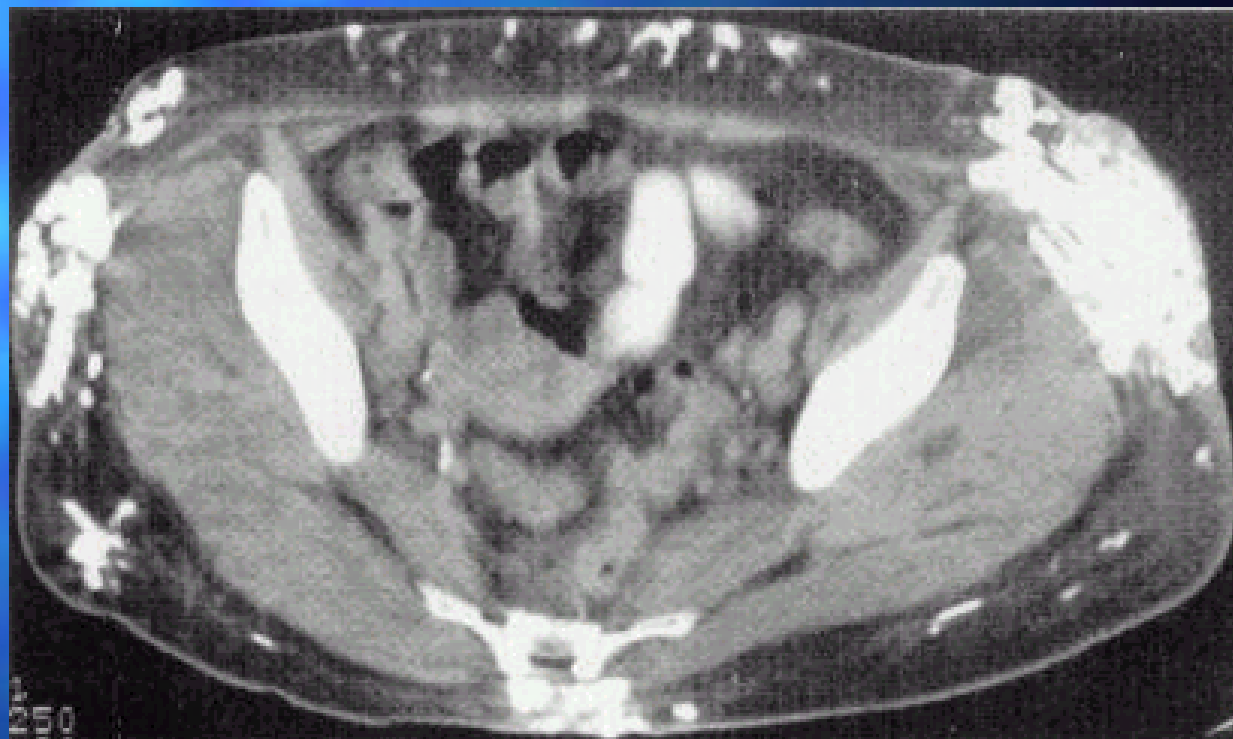
- That's right!
Because the bone cortex is black...
And the fat is white!



MRI (T2) of the hips reveals large cystic areas of **low signal** intensity (arrows). This patient had Idiopathic Tumoral Calcinosis.

Computed Tomography

- CT of the pelvis in a patient with SLE.
- Note extensive calcifications (contrast the high attenuation with that of MRI) in the subcutaneous tissue.



From: www.hawaii.edu/medicine/pediatrics/pemxray/v6c11.html7

Unknown...

- The patient is a 900 year old man from the Degobah System...



Master Yoda

? Scleroderma



Treatment of Calcinosis Cutis

- Medical treatments include:
 - Intralesional corticosteroid injection
 - Etidronate disodium (a bisphosphonate)
 - Aluminum hydroxide (phosphate-binder)
- However, these are not very effective.
- Surgical excision has been shown to be of benefit, as it can provide symptomatic relief.

Summary

- Soft tissue calcifications are relatively common radiographic findings.
- Dystrophic calcifications predominate.
- Often incidental finding, but can present with skin lesions or pain.
- Good history is key.
- Plain film to start; other imaging can be helpful if plain film is normal.
- Medical treatment has variable success; surgical removal of calcification is indicated if symptomatic.

Bibliography

- Koopman: Arthritis and Allied Conditions, 13th ed., 1997.
- Richardson, M.L. Approaches to differential diagnosis in musculoskeletal imaging: soft tissue calcifications.
<http://www.rad.washington.edu/mskbook/softtissueca.html>., 1994.
- Stewart, V.L. Herling, P., Dalinka, M.K. Calcification in soft tissues. JAMA 1983; 250 (1): 78-81.
<http://www.hawaii.edu/medicine/pediatrics/pemxray/v6c11.htm>
- <http://www.rad.washington.edu/maintf/cases/unk39/answers.html>
- Cousins, M.A. Surgical management of calcinosis cutis universalis in systemic lupus. Arthritis Rheum 1997 Mar; 40(3): 570-2..

Acknowledgments

Beverlee Turner

Larry Barbaras

Special thanks to Dr. Hochman for the chest X-Ray

The end.