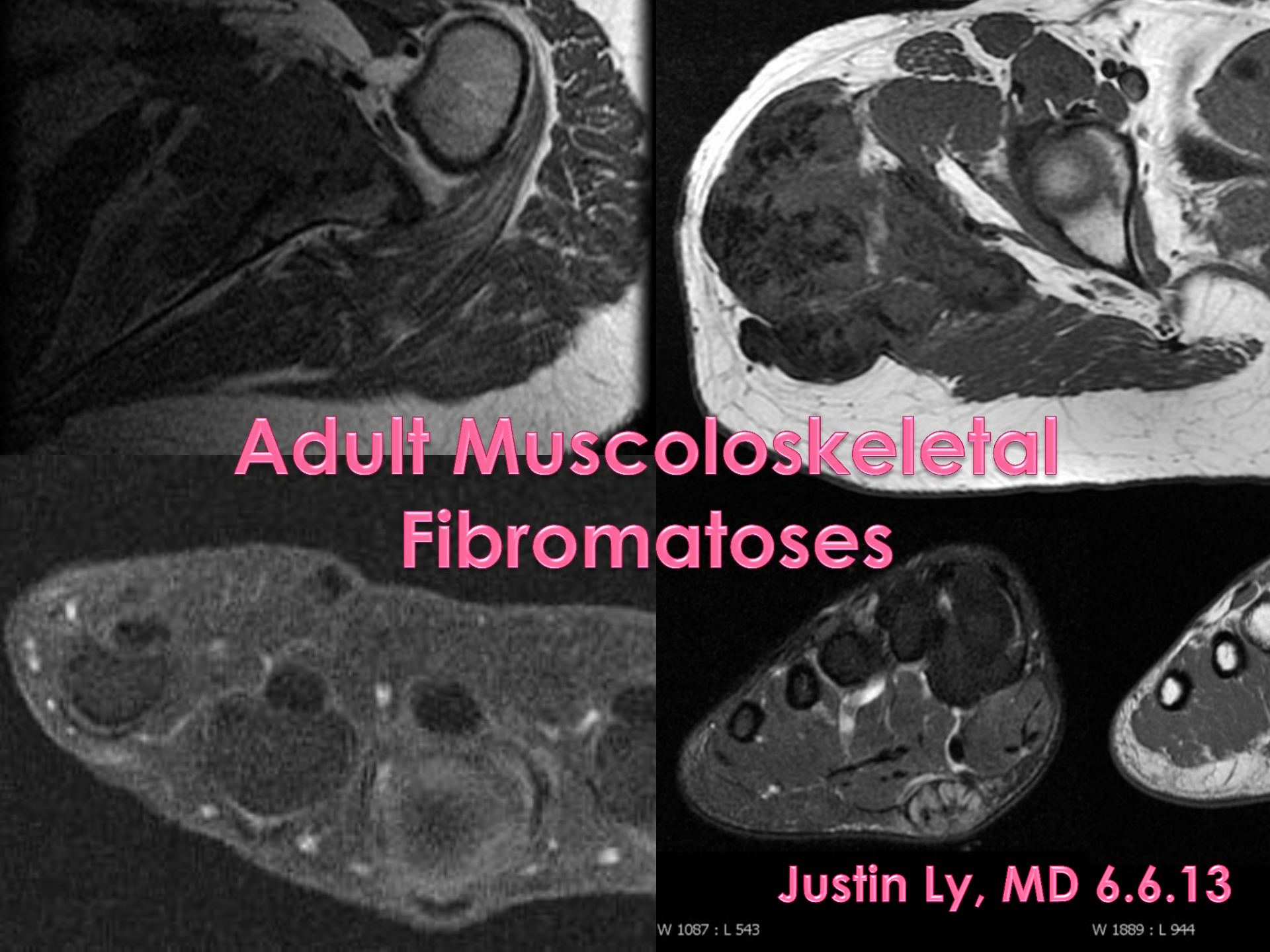
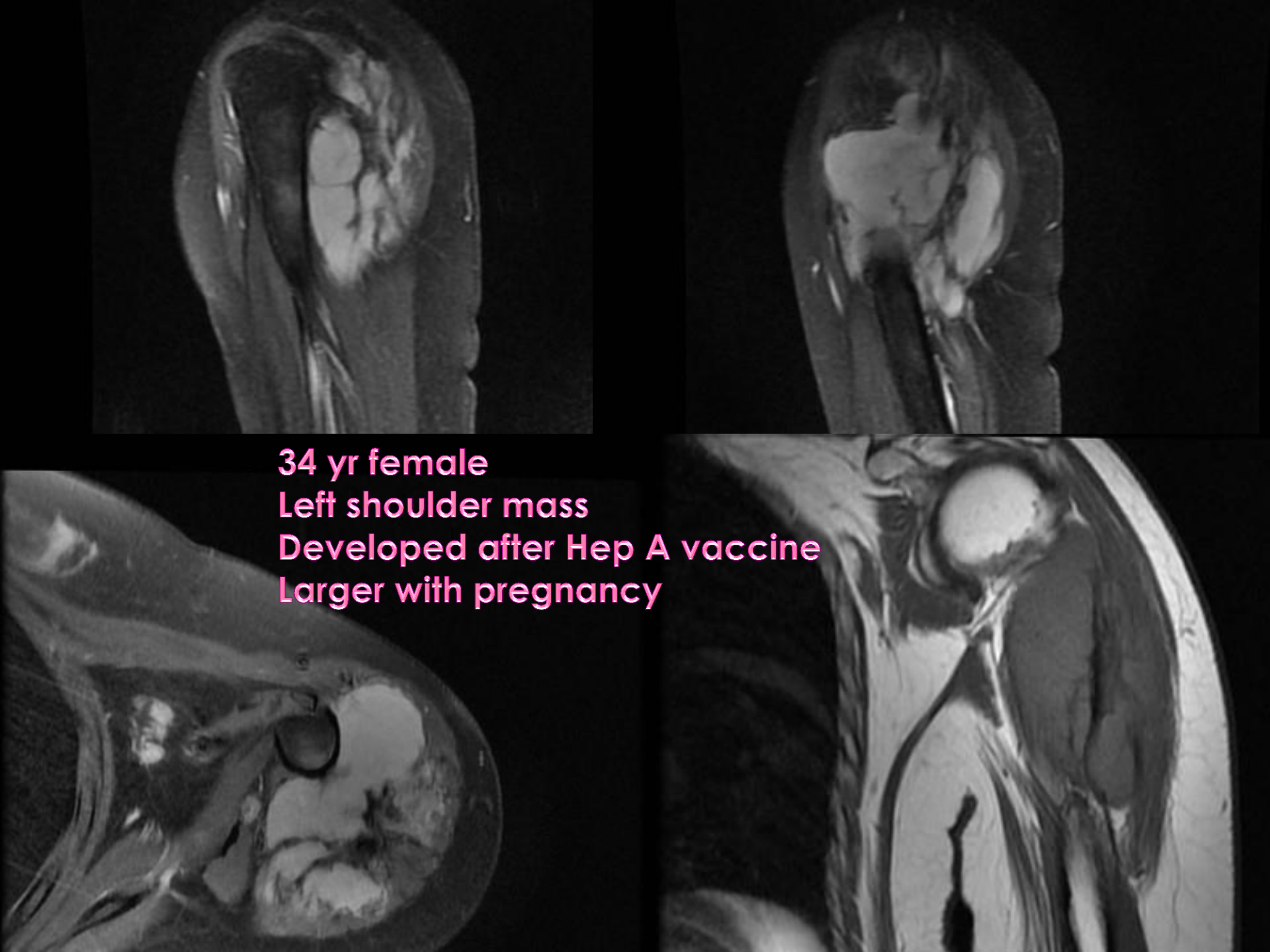

Adult Muscoloskeletal Fibromatoses
Justin Ly, MD 6.6.13
W 1087 : L 543
W 1889 : L 944


34 yr female
Left shoulder mass
Developed after Hep A vaccine
Larger with pregnancy

Objectives

- Review and correlate clinical, pathologic, and radiologic features of various types of Adult MSK fibromatoses
  - Review pertinent anatomy
- Discuss treatment & prognosis

Add yet another lecture to bonepit.com

UCSD Musculoskeletal Radiology

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Available Talks

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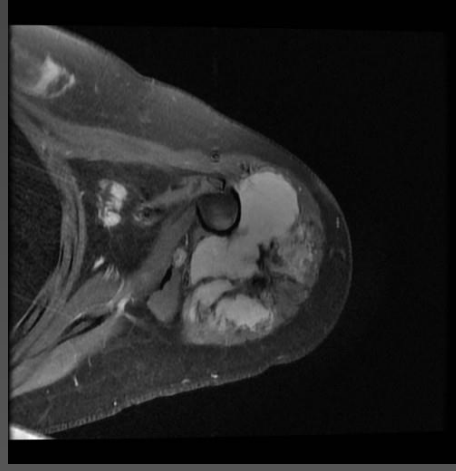
Title	Author
Throwing Shoulder	Joshua Franklin
Brachial Plexus	Jason Barksdale
Assessing bone viability MR	Rick Bhullar
TFCC	Adelaine Wong
Appendicular Skeletal Trauma	Tudor Hughes
UTE qMRI	Michael Im
THA	Clark Brixey
Sports related injuries of the fingers	Luke Scalcione
Acute pediatric elbow injury	Michelle Omura
Wrist Extensor Retinaculum	Jeffrey Tan
Hip trauma fracture classification	Alex Ho
Bone Dysplasia	Tudor Hughes
Arthrography	Tudor Hughes
MR of MSK Neoplasms	Tudor Hughes
Nerve entrapment	Tudor Hughes
Sports injuries	Tudor Hughes
DEXA	Tudor Hughes
Foot alignment adult	Tudor Hughes
Signs	Tudor Hughes
Soft Tissue Calcification	Tudor Hughes
Patella Tracking	Tudor Hughes
Commonly missed injuries of the extremities	Tudor Hughes
FAI	Tudor Hughes
How useful is pre-Achilles fat pad	Ankit Patel
Imaging of Peripheral Neuropathies	Dorota Linda
Osseous Pathology of the Mandible	Fernando J Rodriguez (Chino)
The temporal bone	Stephen Haltom
MSK MRI Essential Principles and Practical Applications	Eric Chang
Coalition	Julie L. Rutledge
Elbow injuries of the throwing athlete	Jimmy C. Wang
Carpal instability	Caroline Yang
Bone Malignancy	Suzanne Shepherd
Spine hardware	Arvin Hariri
Synovial Plicae of the Knee	James Koenig
MRI Protocols	Maya Borsu
AC Jt	Kamran Ahmad
Intra-articular pathology	Amin Matin
Radiographic Evaluation of Calcaneal Fractures	Kali Luker
Lumps and Bumps Soft Tissue and Bony Masses of the Wrist	Brady Huang
Spine Trauma	James Buratto
Articular diseases of the cervical spine	Binh-To Tran
Elbow instability	Matthew James Sharp
Radiology Business Management A Crash Course	Scot Yochim
Muscles	Kavita Gorantla
Ulna sided wrist pain	Federico Discepola
Pubalgia	Catherina Fu
Nerve Entrapment Lower Extremity	Andrew Knoll
Kienbock	Orrin Franko
Lisfranc fracture dislocation	Tara Robbins
Osteochondritis dissecans	Becca Rodriguez
Slipped upper femoral epiphysis	Amy Leu
Knee Chondral Repair	Rakersh Patel
Sports Related Injuries of the Elbow	James Wolff
The Growing Physis	John Stassen
Injury to the Extensor Mechanism	Romulo Baltazar
The Temporomandibular Joint	Jeffrey Hirata
Maintenance of Certification	Anne Roberts


HAMEL'S


Mission Beach

INTRODUCTION



- Musculoskeletal fibromatoses
  - ***Wide range of fibroblastic to myofibroblastic proliferations***
  - Grouped together because of their **similar pathologic appearances**

- Clinical behavior INTERMEDIATE between benign and malignant fibrous lesions
- Commonly demonstrating **infiltrative growth**, resulting in frequent **local recurrence** but *lacking metastatic potential*

- World Health Organization (WHO) Committee for Classification of Soft Tissue Tumors in 2002 categorized these lesions as **superficial or deep**, based on their anatomic location

MSK FIBROMATOSES

- **Superficial (fascial)**
  - Adult: palmar, knuckle pad, plantar
  - Pediatric: calcifying aponeurotic fibroma, lipofibromatosis, inclusion body fibromatosis
- **Deep (musculo-aponeurotic)**
  - Adult: desmoid type, abdominal wall
  - Pediatric: fibromatosis colli, myofibroma and myofibromatosis

MSK FIBROMATOSES

- **Superficial (fascial)**
  - Adult: palmar, knuckle pad, plantar
  - Pediatric [illegible] fibroma, lipofibro[illegible]fibromatosis
- **Deep (musculo-aponeurotic)**
  - Adult: desmoid type, abdominal wall
  - Pediatric [illegible] fibroma and myofibro[illegible]

MSK FIBROMATOSES

- **Superficial (fascial)**
  - Adults: palmar, knuckle pad, plantar
  - Small, grow slowly
  - Dx suggested by location

MSK FIBROMATOSES

- **Deep (musculo-aponeurotic)**
  - Adults: desmoid type, abdominal wall
  - **OFTEN LARGE, MORE RAPIDLY ENLARGING**

Outline

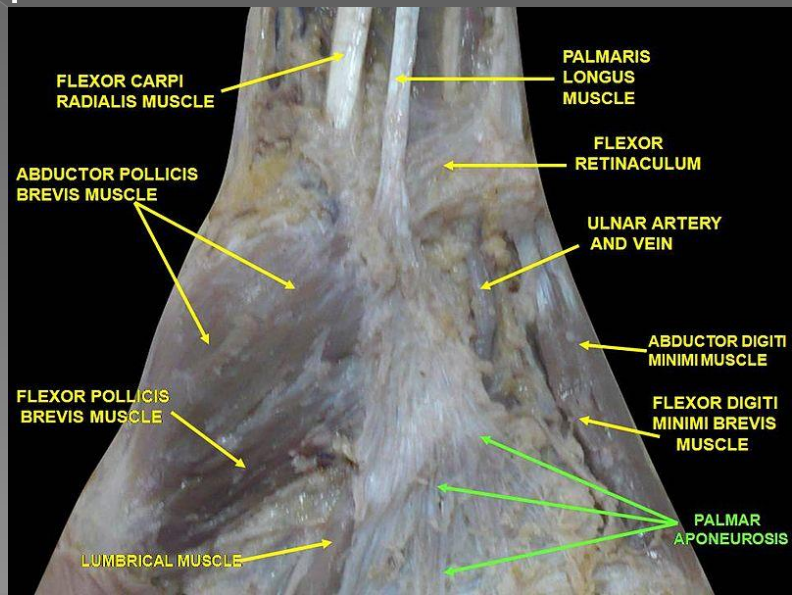
- **Superficial MSK Fibromatoses**
  - Palmar Fibromatosis
  - Knuckle Pad Fibromatosis
  - Plantar Fibromatosis
- **Deep MSK Fibromatoses**
  - Desmoid Type Fibromatosis
  - Abdominal Wall Fibromatosis

Outline

- **Superficial MSK Fibromatoses**
  - Palmar Fibromatosis
  - Knuckle Pad Fibromatosis
  - Plantar Fibromatosis
- **Deep MSK Fibromatoses**
  - Desmoid Type Fibromatosis
  - Abdominal Wall Fibromatosis

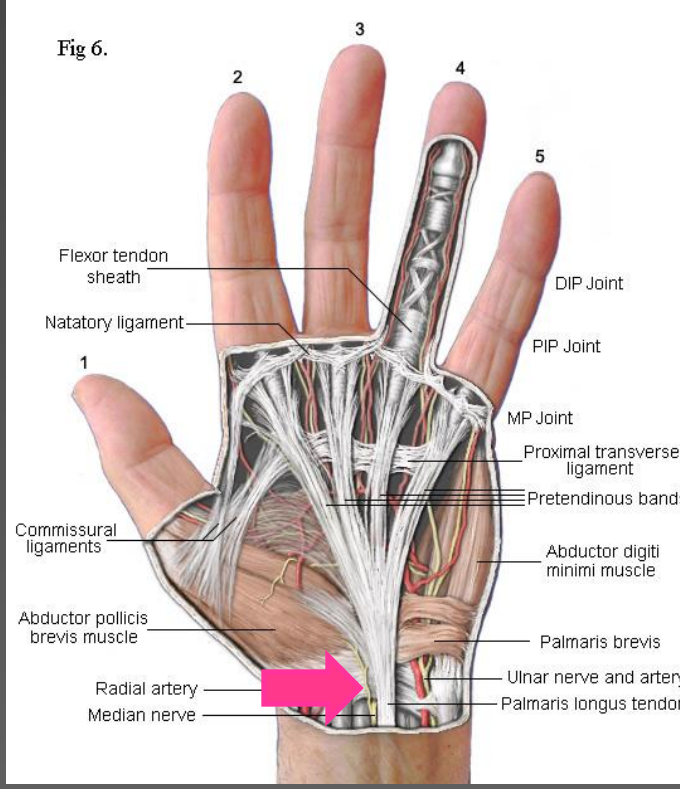
Palmar Aponeurosis

- Flexible fibrotendinous/ *collagenous* **scaffolding**
  - provides support, holding together different components of hand/skin
  - allows considerable flexibility



Palmar Aponeurosis

- Collagenous complex
- Tendinous extension of palmaris longus (arrow)
- Fibers spread out like **fan** across palm

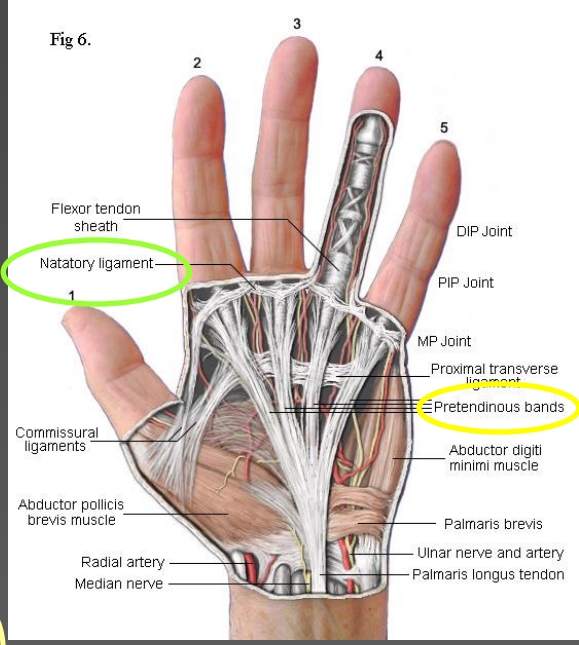


http://www.dupuytrens.me.uk/dupuytrens.html

Palmar Aponeurosis

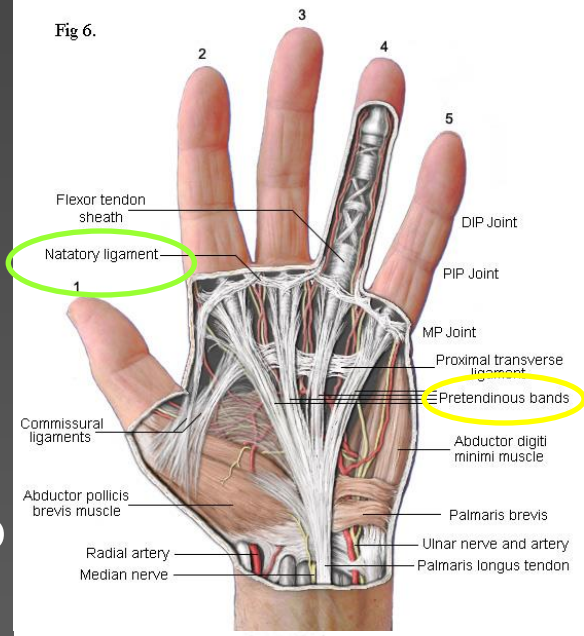
- Fibers bunch together into 4 groups - **pretendinous bands**, each aligned with a finger
  - central cord (MCP contracture)

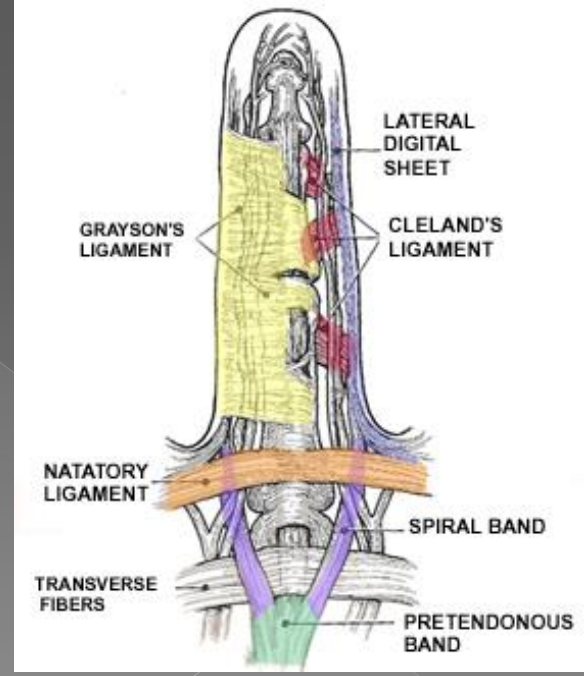
- Several collagenous transverse ligaments (superficial and deep)
  - ***natatory ligaments*** @ base of finger (superifical palmar transverse lig)



Palmar Aponeurosis

- ****Spiral cord** (cause of PIP contracture) involves several structures:
  - pretendinous band
  - spiral band
  - natatory ligament
  - lateral digital sheet
  - Grayson's ligament





Digital Fascia

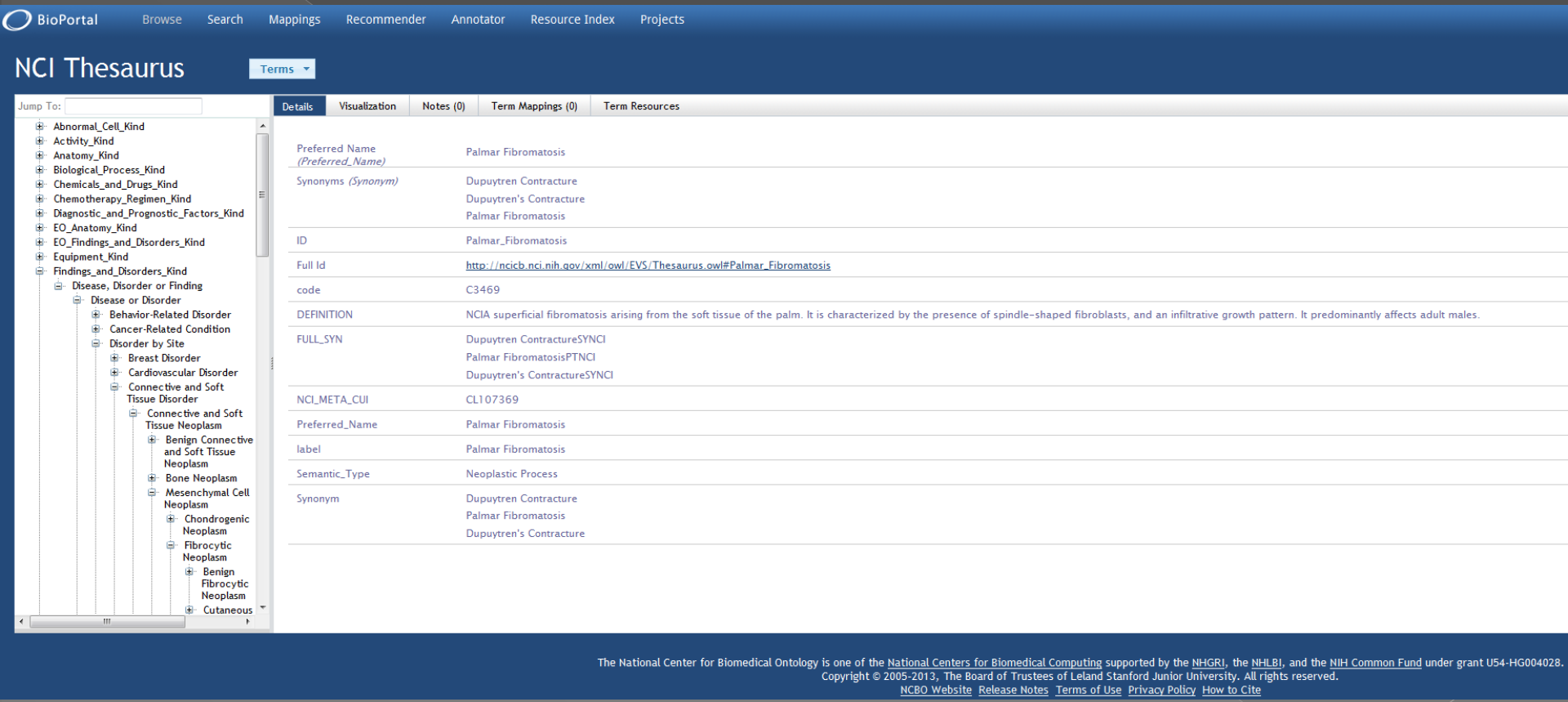
http://www.dupuytrens.me.uk/dupuytrens.html

PATHOLOGY
Palmar Fibromatosis

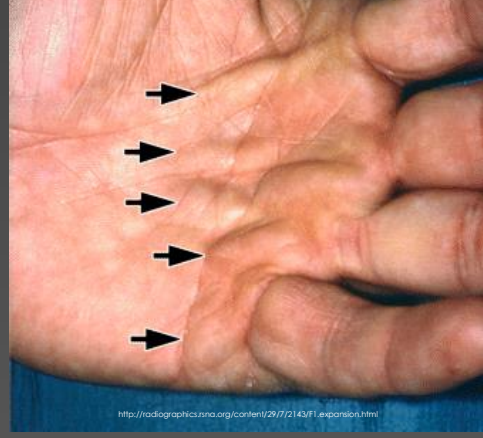
- Baron Guillaume Dupuytren, surgeon who described an operation correct the affliction in the *Lancet* in 1831
- AKA
  - **Dupuytren disease**
  - **Dupuytren contracture**
  - **Morbus Dupuytren**

What is the most current terminology? Dupuytren's contracture or palmar fibromatosis?

NCIt Browser is a web-based terminology browser



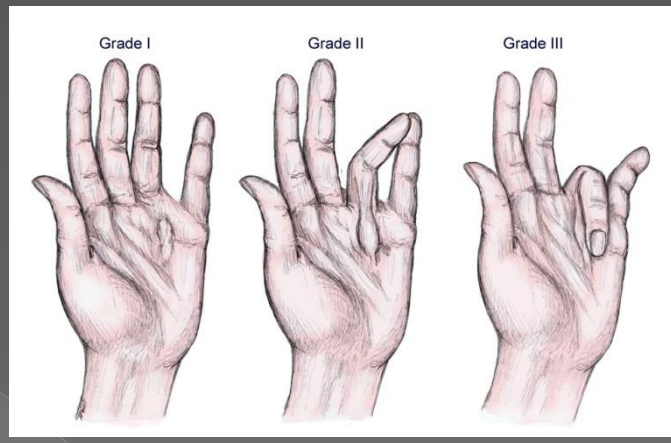
Palmar Fibromatosis



- **Clinical Features**
- **Most common** superficial fibromatoses, affecting 1%–2% of general population
- Almost exclusively Caucasians, particularly frequent in those of Northern European ancestry *(highest prevalence northern Scotland, Iceland, Norway, Australia)*
- Most common > 65 yrs
- Men 3-4X more likely to be affected
- Bilateral 40%–60%

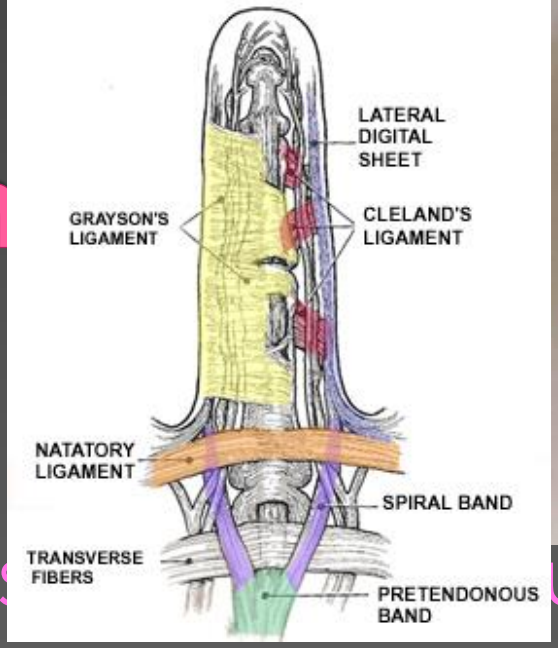
Palmar Fibromatosis

- **Clinical Features**
- ETIOLOGY not completely understood
  - Thought to be multifactorial:
    - ***Trauma***
    - Microvascular injury
    - **Immunologic processes**
    - *Genetic factors (up to 68% have family history of msk fibromatoses)*



http://emedicine.medscape.com/article/329414-overview

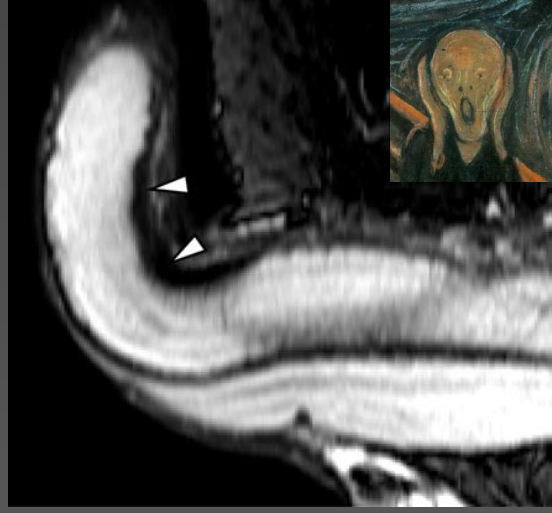
Palmar Fibron



- **Clinical Features**
- Present with painless[illegible]us nodules, progress slowly (months to yrs) to fibrous cords *that attach to* & cause traction on underlying flexor tendons, resulting in flexion contractures
- Ulnar-sided rays, 4th/5th most common

http://www.patient.co.uk/doctor/Dupuytren%27s-Contracture.htm

Palmar Fibromatosis

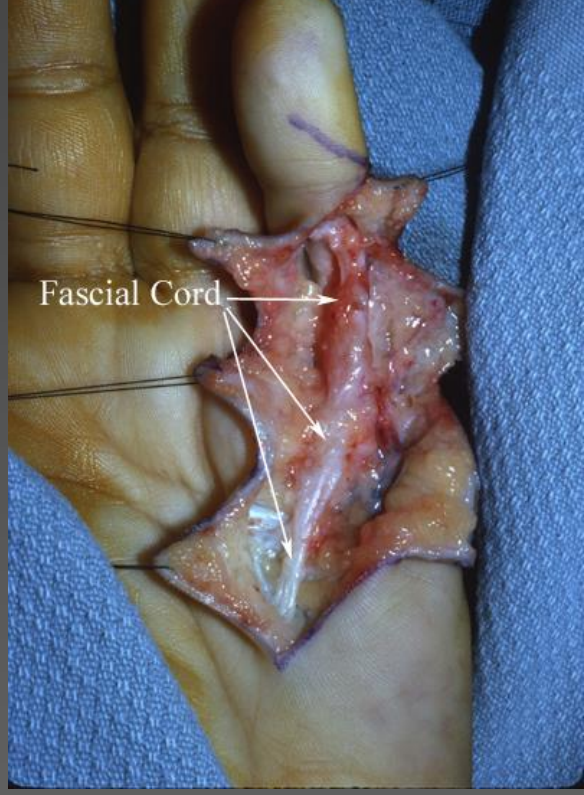


http://radiographics.rsna.org/content/29/2/477/F16.expansion.html

- **Clinical Features**
- Commonly have other types of fibromatoses, including plantar fibromatosis (5%–20% of cases), Peyronie disease, knuckle pads
- Additional associations: diabetes mellitus (20% of pts), alcoholism, keloids

http://radiographics.rsna.org/content/29/7/2143/F7.expansion.html

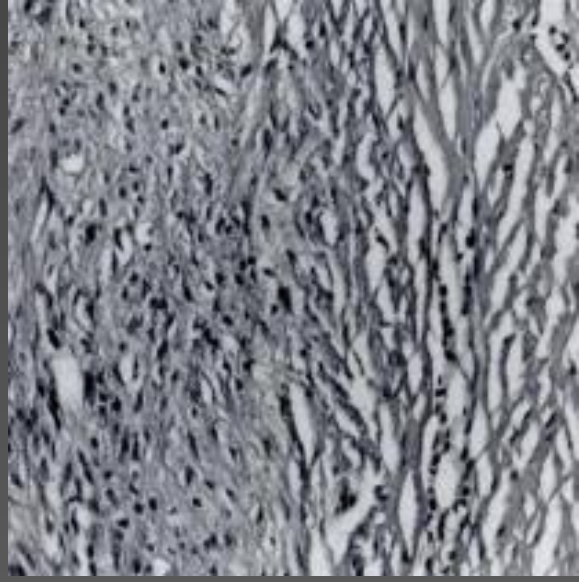
Palmar Fibromatosis

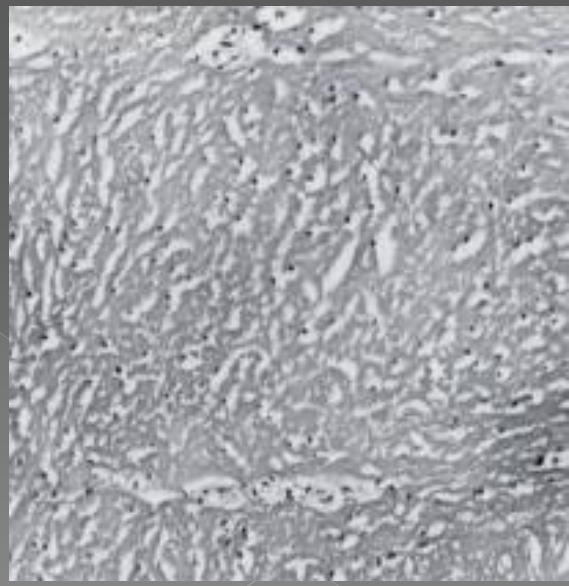


- **Pathologic Features**
- At gross pathologic examination: gray-white or gray-yellow
- Nodules typically very small (<1 cm), often coalescent
- Intimate with palmar aponeurosis, may be *adherent to skin, causing puckering or dimpling*

Palmar Fibromatosis

- **Pathologic Features**
- **Histologic analysis**: uniform fibroblastic-myofibroblastic proliferation of spindle-shaped cells with variably prominent vascularity, although vascularity typically less than in desmoid type fibromatosis

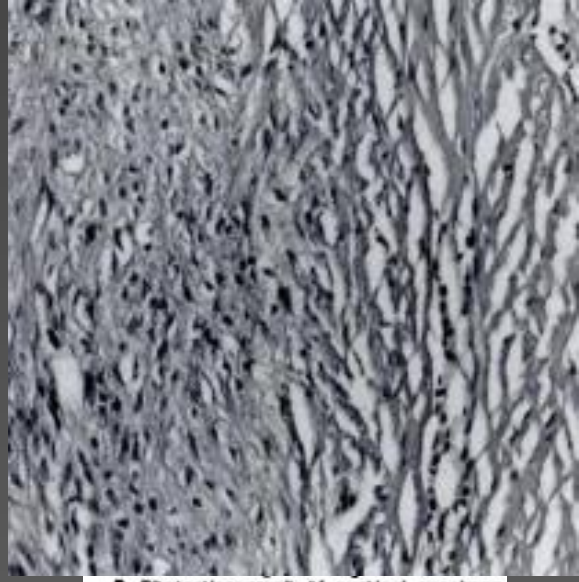




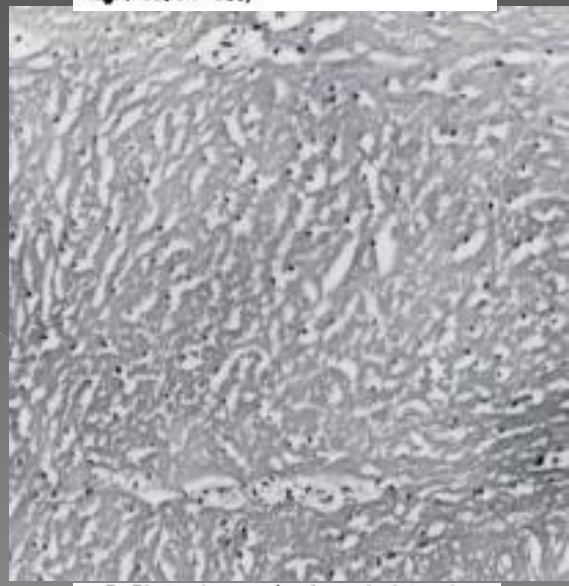
http://radiographics.rsna.org/content/29/7/2143.full

Palmar Fibromatosis

- **Pathologic Features**
  - Degree of cellularity depends on age of lesion, with **younger lesions** (proliferative phase) showing hypercellularity
  - Older, more mature lesions - less cellularity, more collagen



D, Photomicrograph of surgical specimen shows nodule has heterogeneous composition, with cellular regions intermixed with bundles of collagen fibers. (H and E; original magnification ×300)

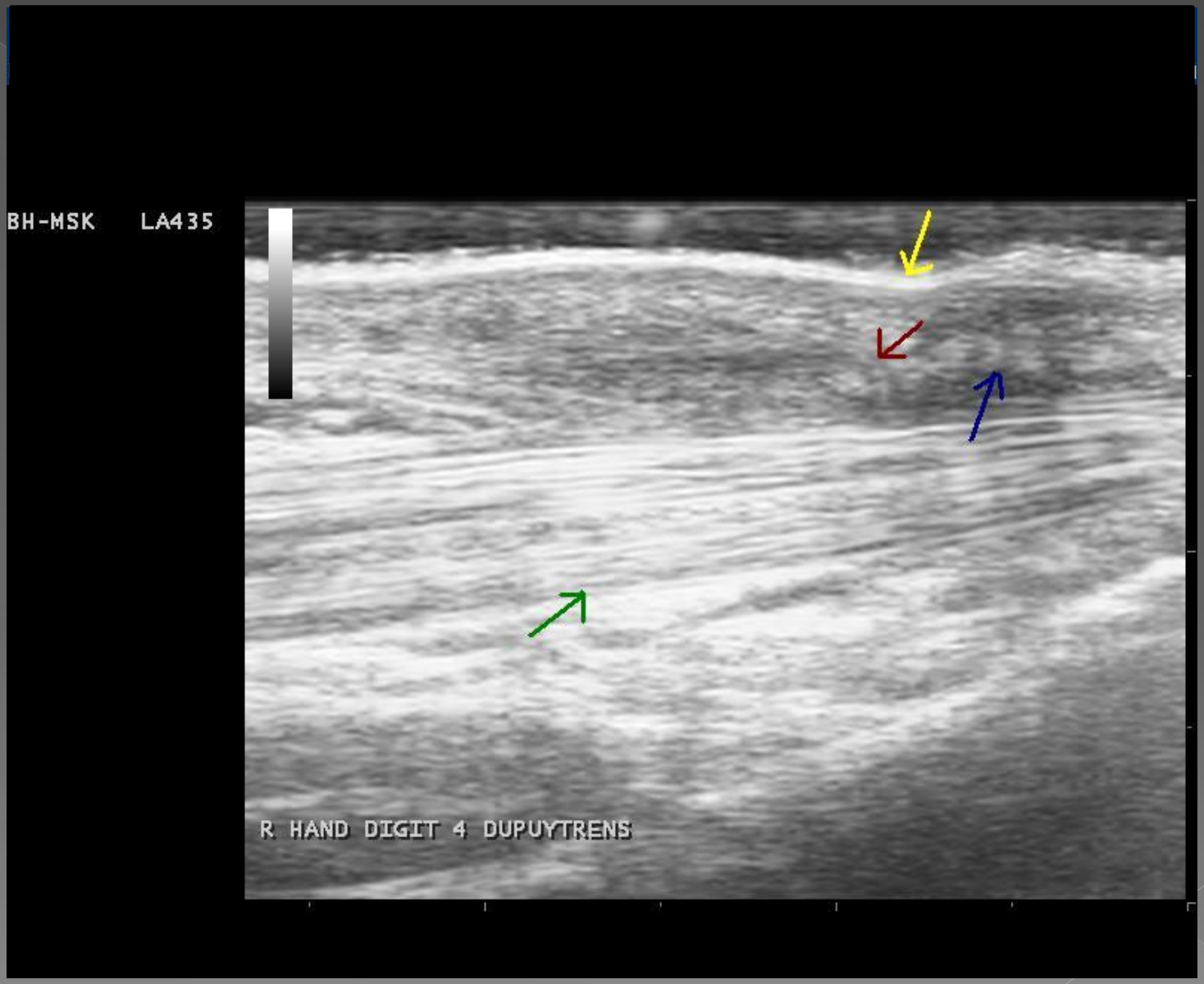


D, Photomicrograph of surgical specimen shows cord is hypocellular, composed mostly of collagen. (H and E; original magnification ×300)

http://radiographics.rsna.org/content/29/7/2143.full

Palmar Fibromatosis

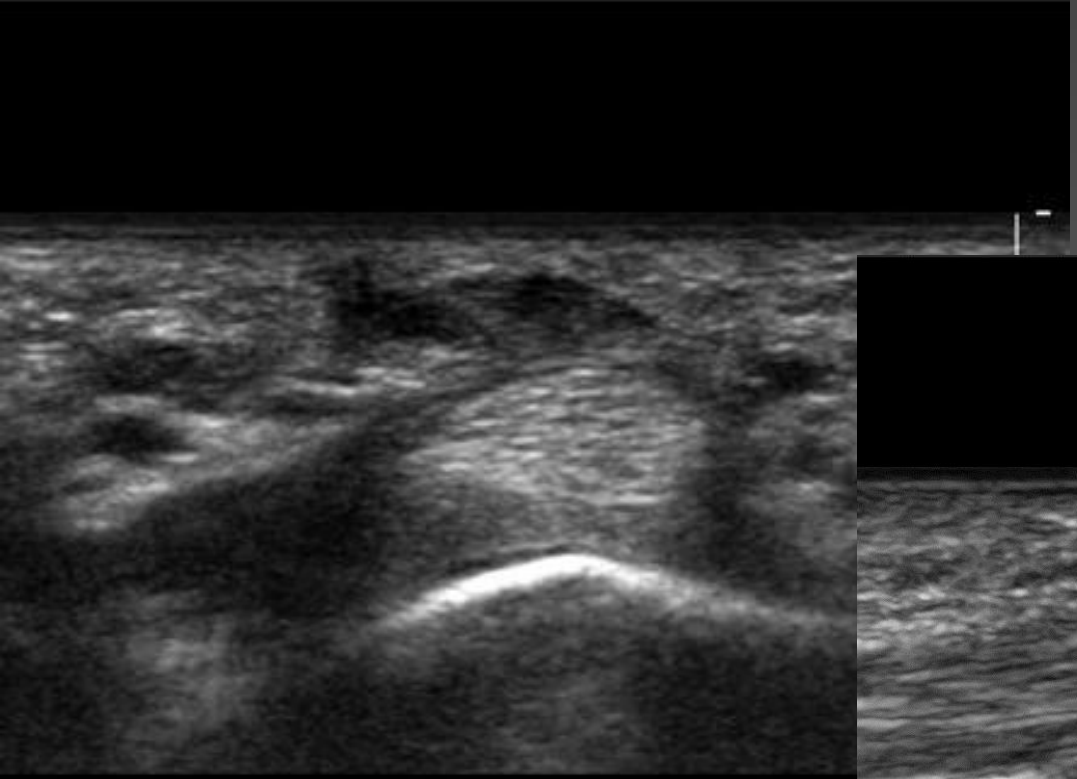
- **Imaging Features**
- Radiography: normal to flexion contractures
- CT shows nonspecific, nodular regions of thickening with attenuation similar to or slightly higher than muscle
- US: hypervascular, hypoechoic nodules in subcutaneous tissues, superficial to flexor tendons
- US also allows real-time dynamic assessment of integrity of flexor mechanism of digit(s)

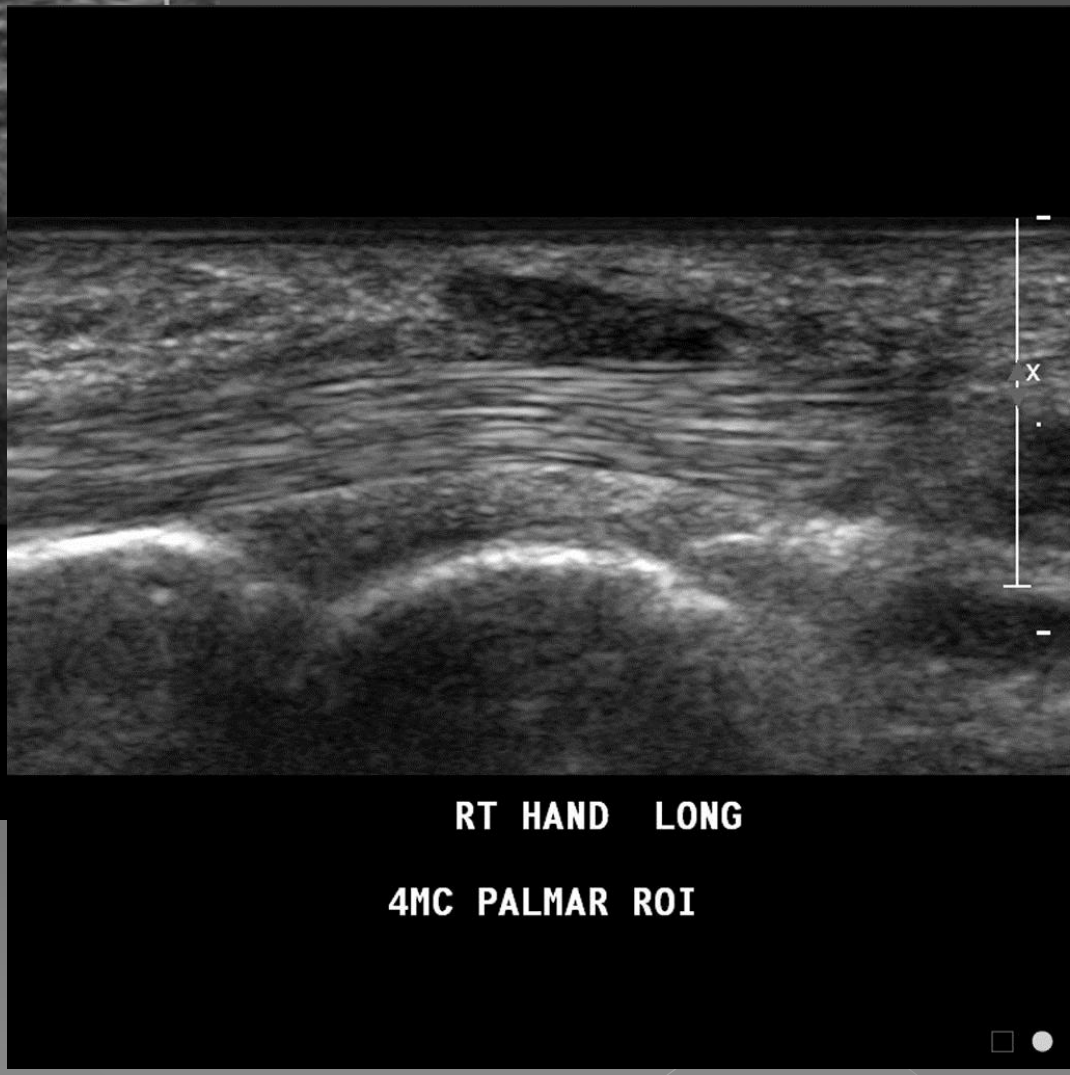


Red arrow: Dupuytren nodule

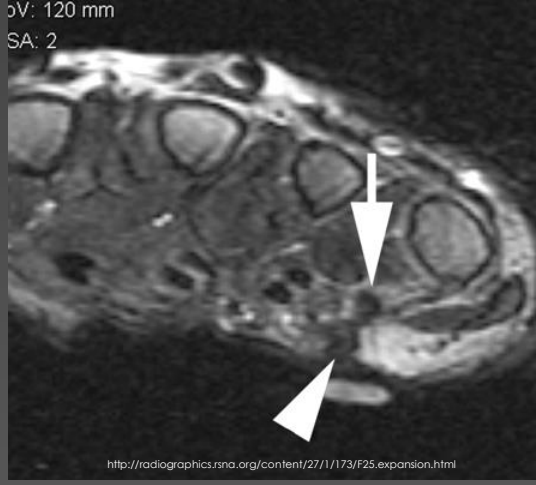
Blue arrow: Thickened palmar fascia

http://emedicine.medscape.com/article/329414-worku



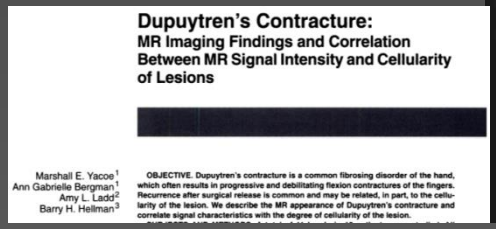


Palmar Fibromatosis



- **MRI**
  - Detect & define extent of dz
  - Nodules or cords
    - May terminate in branching or nodular configuration at level of distal metacarpal
    - Intimate with palmar aponeurosis
    - Extend superficially in parallel with flexor tendons

Palmar Fibromatosis

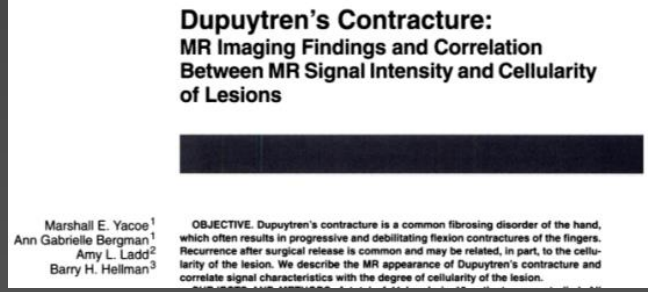

Dupuytren's Contracture:
MR Imaging Findings and Correlation Between MR Signal Intensity and Cellularity of Lesions

Marshall E. Yacoe[1]
Ann Gabrielle Bergman[1]
Amy L. Ladd[2]
Barry H. Hellman[3]

OBJECTIVE. Dupuytren's contracture is a common fibrosing disorder of the hand, which often results in progressive and debilitating flexion contractures of the fingers. Recurrence after surgical release is common and may be related, in part, to the cellularity of the lesion. We describe the MR appearance of Dupuytren's contracture and correlate signal characteristics with the degree of cellularity of the lesion.

- **MRI**
- **Signal Characteristics**
  - **Yacoe et al** evaluated **MRI appearance of palmar fibromatosis in 35 lesions-correlated findings with histologic cellularity**
    - **Nodular (early)** masses: 85% intermediate signal intensity (T1/T2WI)
    - **Cordlike (more mature)** masses: 82% signal intensity (T1/T2WI) predominantly hypointense (similar to tendon)
    - Enhance, degree variable, heterogenously or diffusely

Palmar Fibromatosis



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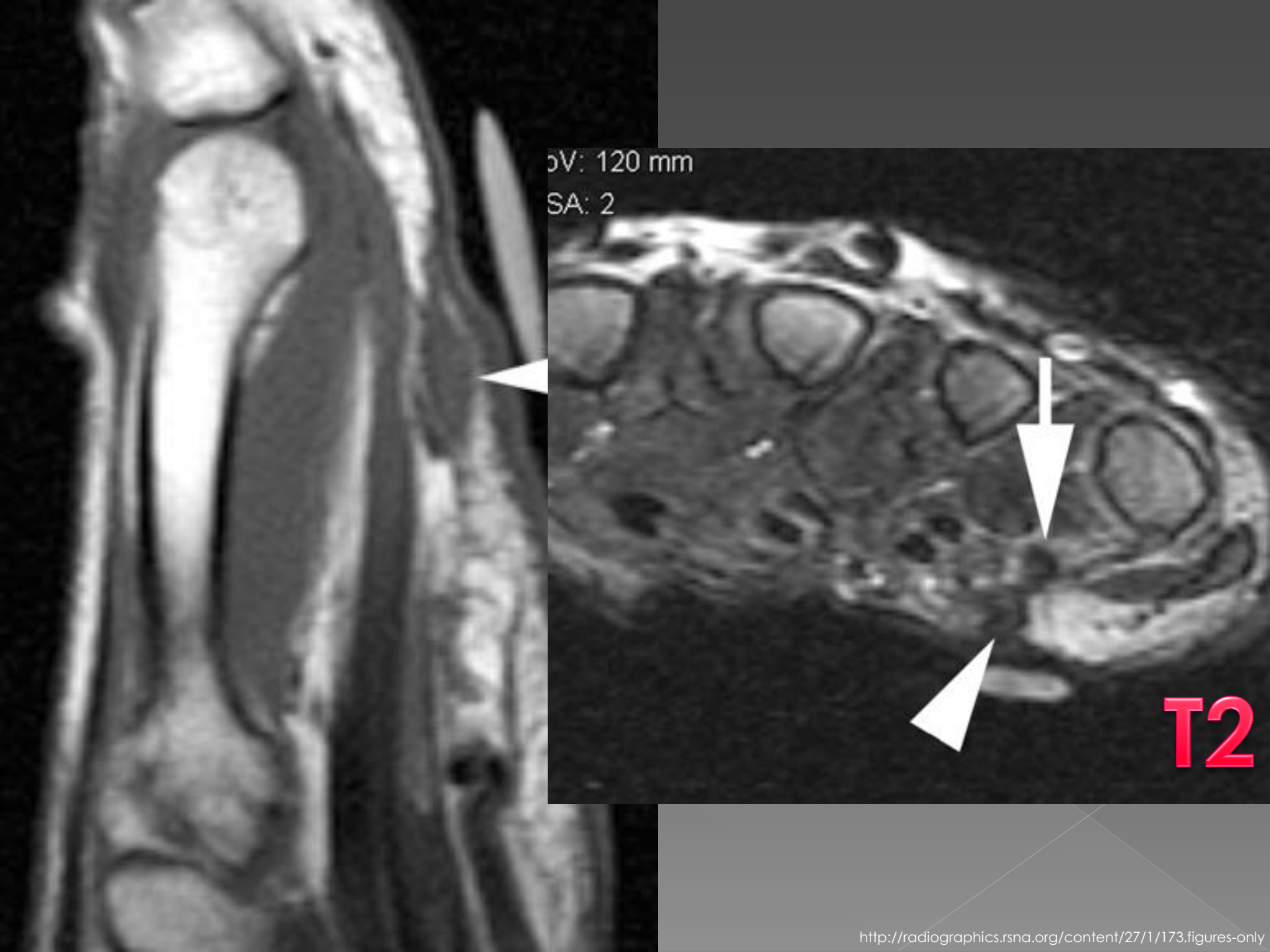
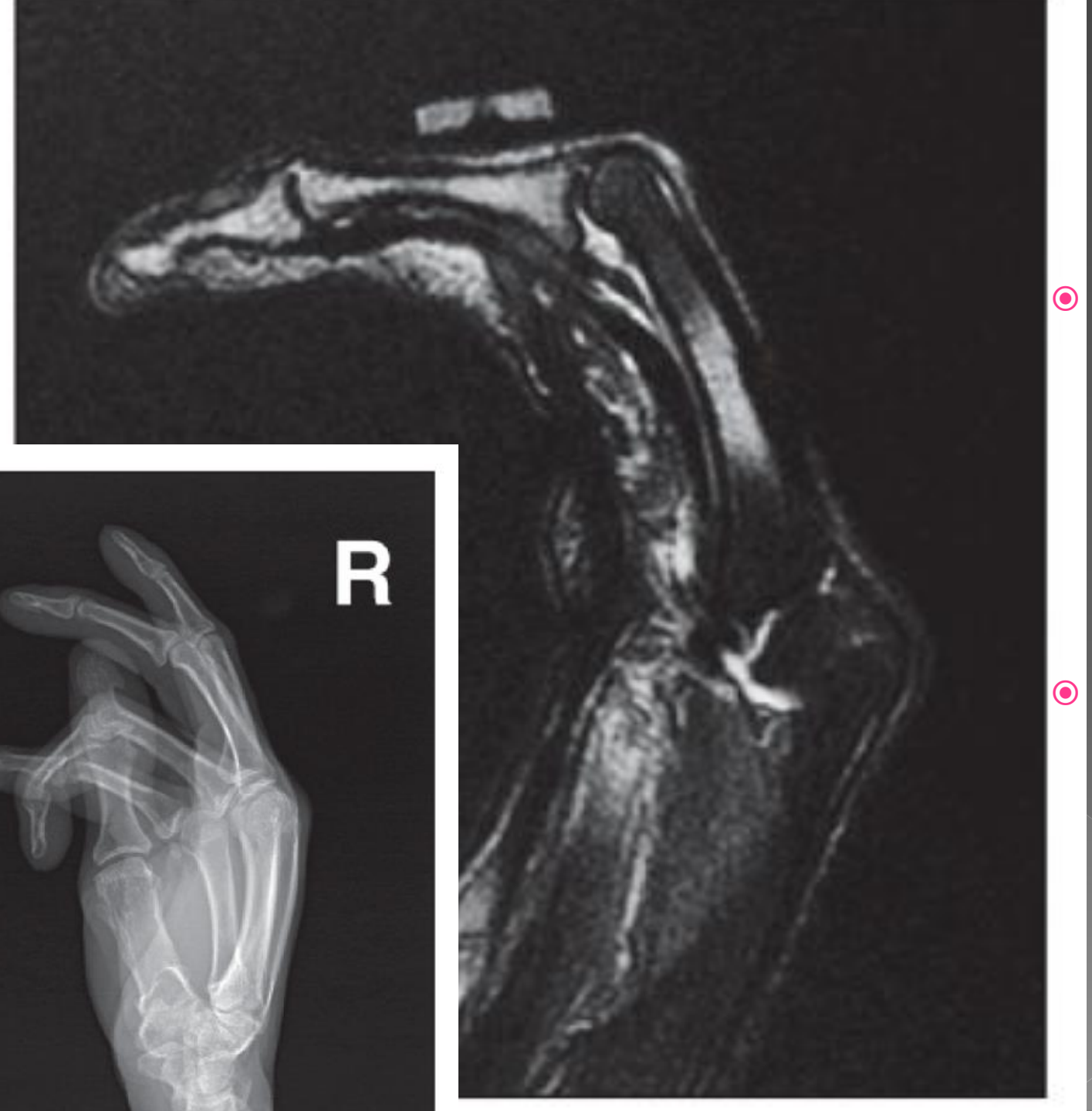
Rombouts J-J, Noel H, Legrain Y, Munting E. Prediction of recurrence in the treatment of Dupuytren's disease: valuation of a histologic classification. *J Hand Surg [Am]* **1989**;14-A:644–652

- LOWER SIGNAL = LOWER CELLULARITY (MORE COLLAGEN) = LESS LIKELY TO RECUR
- INTERMEDIATE SIGNAL = HIGHER CELLULARITY = MORE LIKELY TO RECUR
- Preoperative MRI can assists surgeon in determining risk of recurrence/appropriate timing for excision



http://radiographics.rsna.org/content/27/1/173.figures-only



- 58-year-old African American man with history of long-standing flexion deformity of right ring finger that was worsening.
- 70° of flexion at MCP joint, 90° of flexion at PIP joint

http://www.musculoskeletalnetwork.com/display/article/1145622/1786658

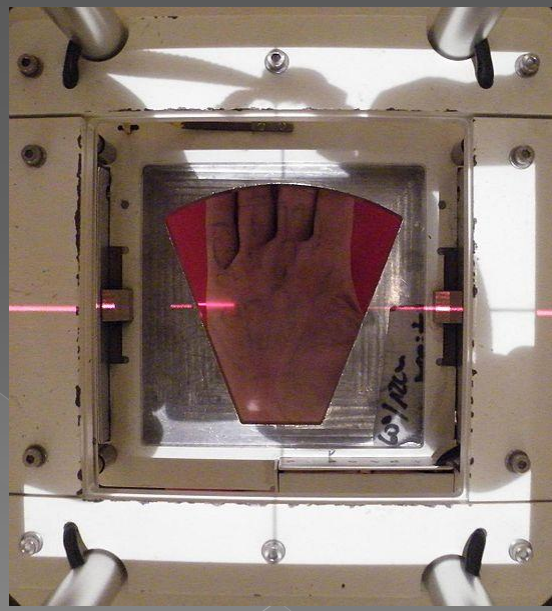
Palmar Fibromatosis

- **Treatment routes**
- Might involve one or more different types of treatments; sometimes need repeated tx
- Main categories listed by International Dupuytren Society in order of stage of disease:
  - Radiation Therapy
  - Needle Aponeurotomy(NA)
  - Collagenase Injection (Xiaflex)
  - Hand Surgery

Palmar Fibromatosis

- Radiation Therapy is effective at early nodules and cords stage "Stage N“; also used at N/I stage of 10 degrees or less of deformation

outer line demonstrates radiotherapy portal outline



Beam's eye view of radiotherapy portal on hand's surface with lead shield cut-out placed in machine's gantry

http://en.wikipedia.org/wiki/Dupuytren%27s_contracture

Palmar Fibromatosis

- Needle Aponeurotomy (Fasciotomy) most effective at "Stage I" of 6-45 degrees of deformation
  - quick
  - requires only local anesthesia
  - minimal or no scarring
  - fast recovery
  - less expensive than surgery
  - can be repeated
  - typically fairly painless

Sampson S et al. J Am Osteopath Assoc 2011;111:113-116

Palmar Fibromatosis

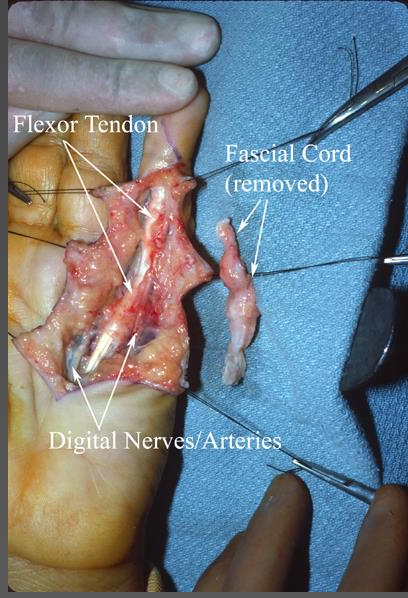
- Collagenase injection most effective at "Stage I"; also used at "Stage II" of 46-90 degrees of deformation
- XIAFLEX® contains combination of 2 collagen enzymes, isolated and purified from fermentation of Clostridium histolyticum bacteria When injected directly into cord, these 2 types of collagenase work synergistically to enzymatically disrupt collagen.

before, next day, two weeks after first tx

http://en.wikipedia.org/wiki/Dupuytren%27s_contracture

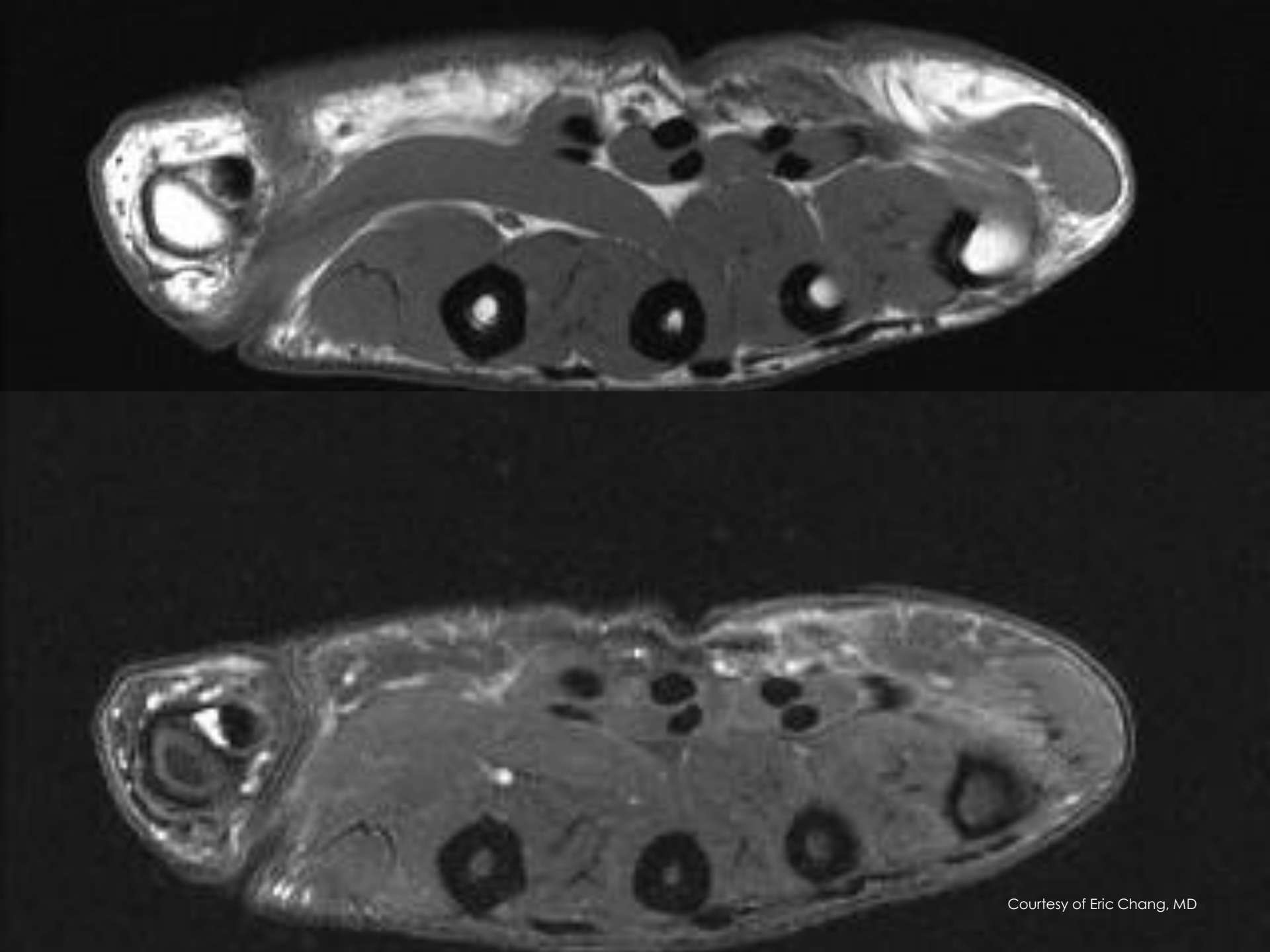
Palmar Fibromatosis

- **Treatment & Prognosis**
- ***Surgery*** effective at all stages
- Dependent on symptoms
- Guidelines: flexion contracture > 20° @ MCP jt or > 30° at PIP jt
- **Current surgical** treatment consists of selective fasciectomy of only diseased locations
- Local recurrence common (30%–40%)

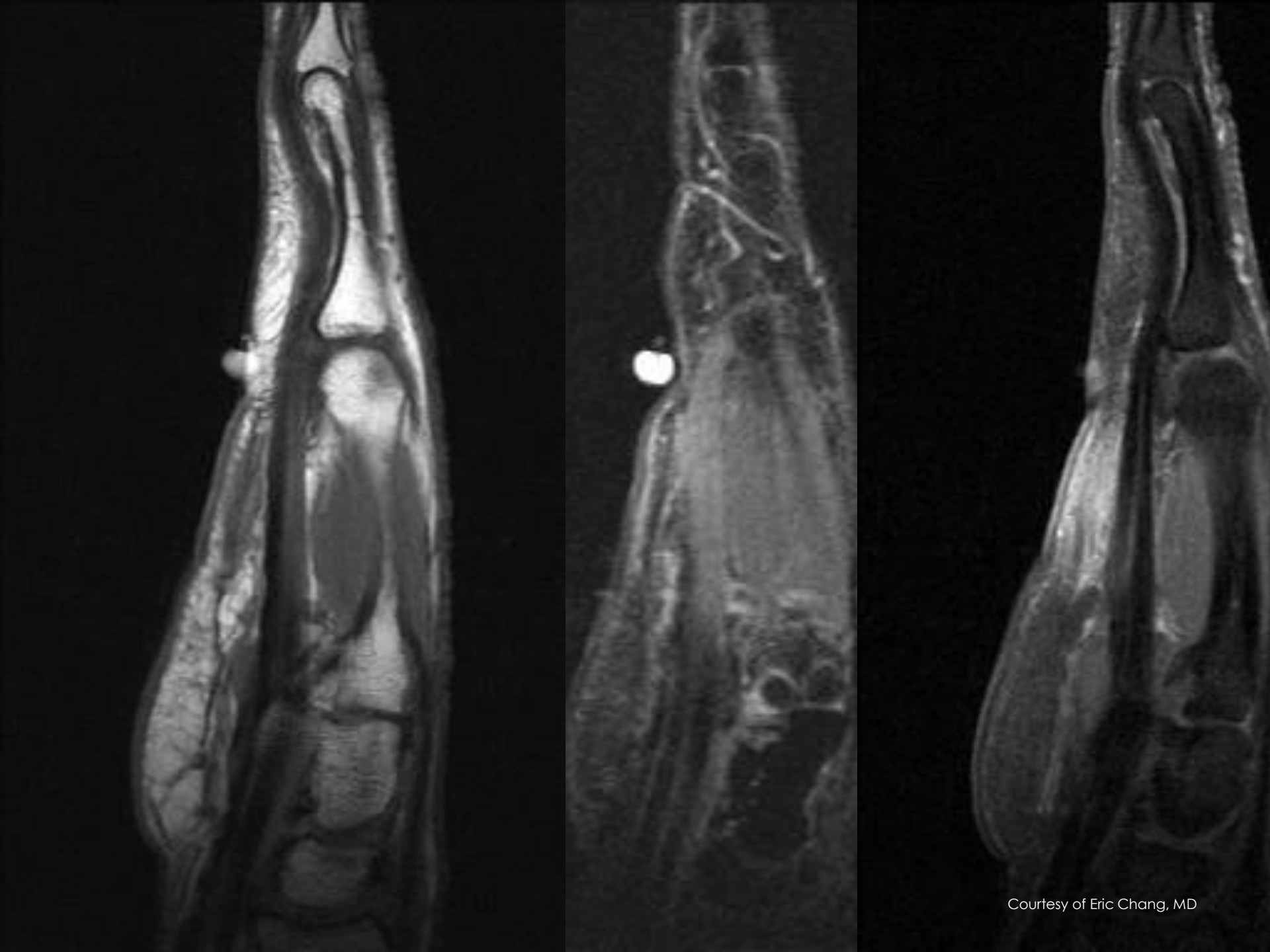


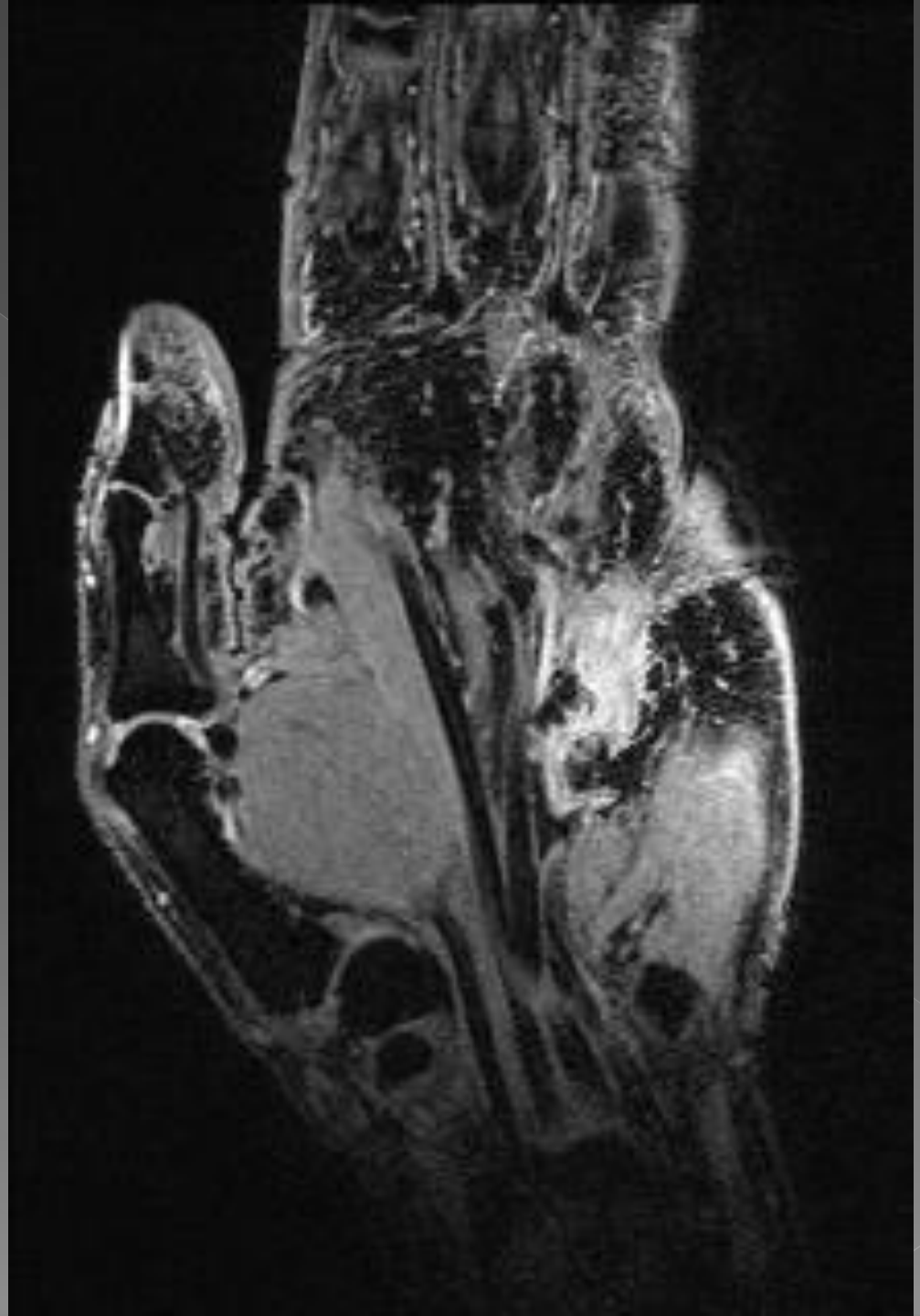
http://radiographics.rsna.org/content/29/7/2143.full

http://en.wikipedia.org/wiki/Dupuytren%27s_contracture

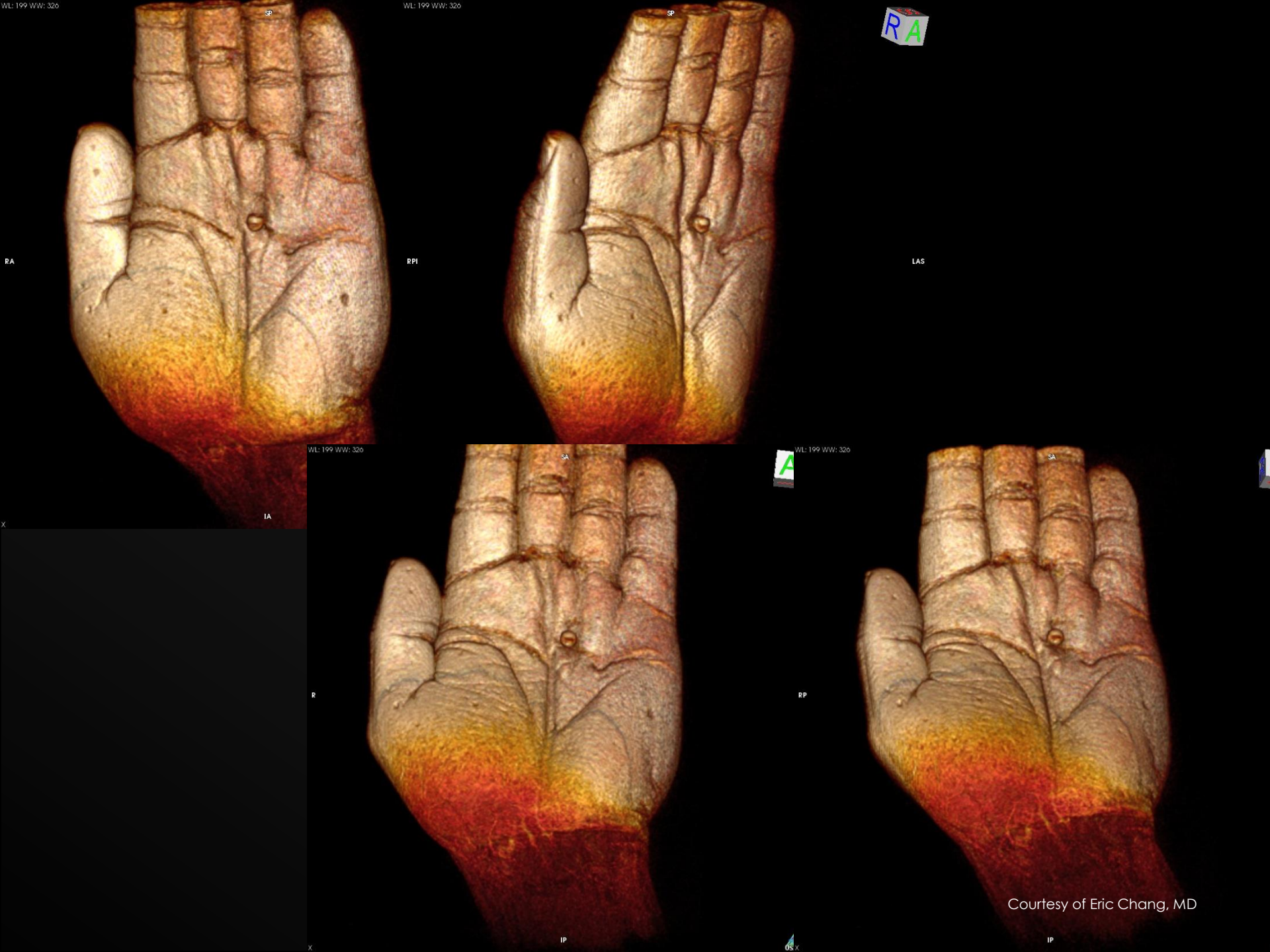


Courtesy of Eric Chang, MD


Courtesy of Eric Chang, MD



Courtesy of Eric Chang, MD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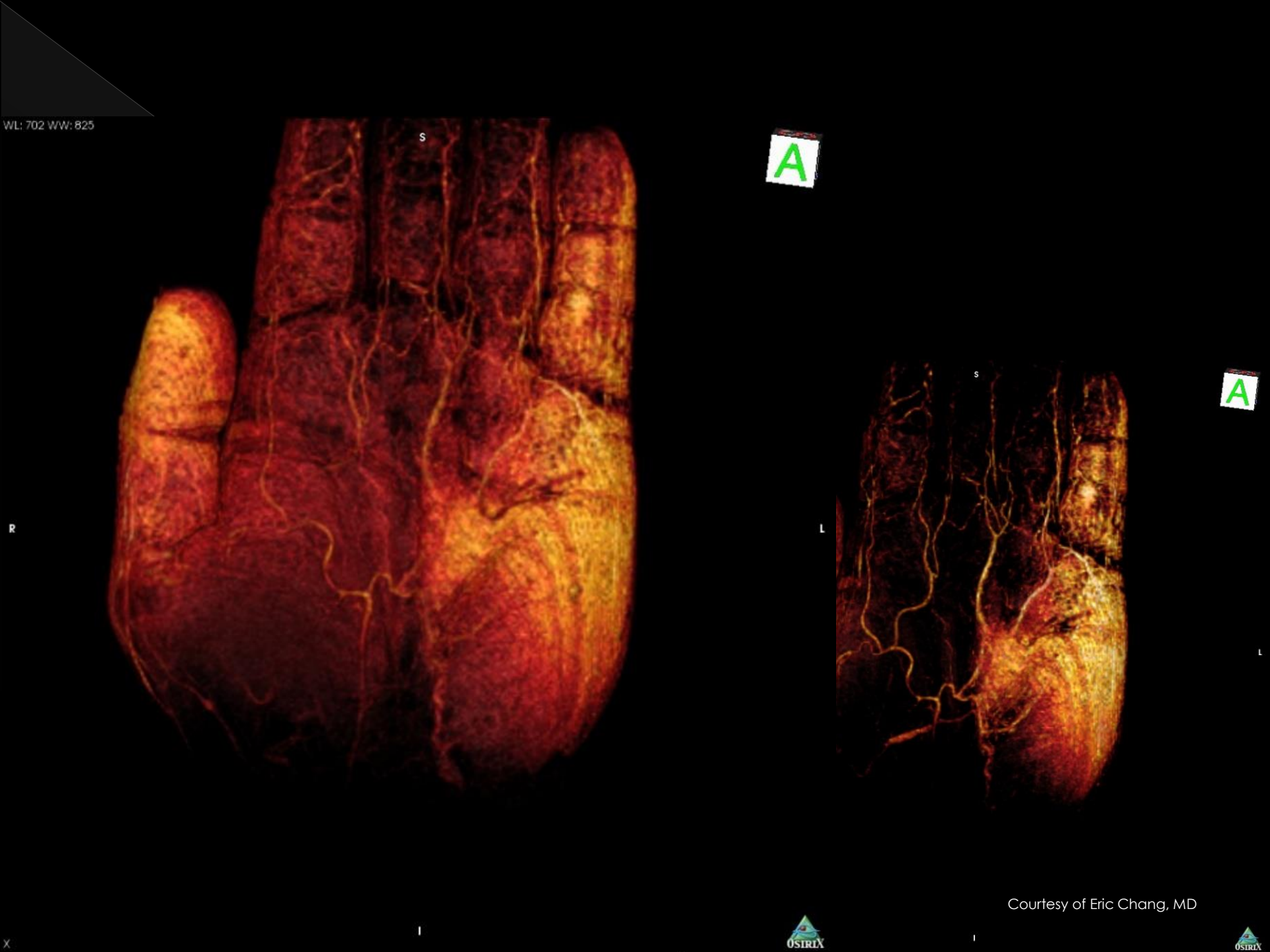
Courtesy of Eric Chang, MD


WL: 702 WW: 825
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Courtesy of Eric Chang, MD


DEL MAR

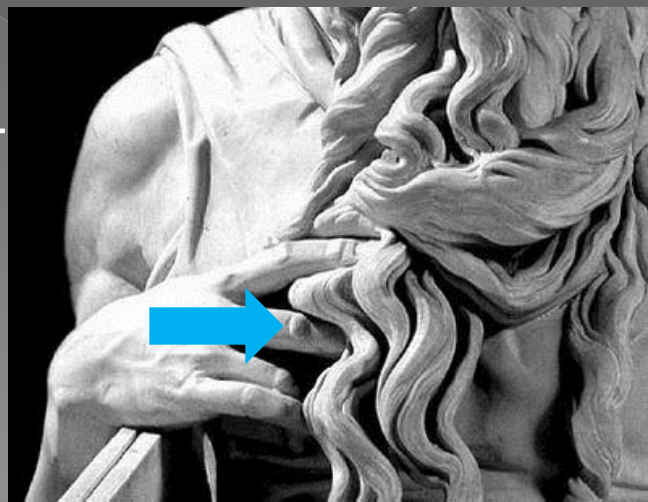
Outline

- **Superficial MSK Fibromatoses**
  - Palmar Fibromatosis
  - Knuckle Pad Fibromatosis
  - Plantar Fibromatosis
- **Deep MSK Fibromatoses**
  - Desmoid Type Fibromatosis
  - Abdominal Wall Fibromatosis

Knuckle pad (fibromatosis)

- Garrod first described in 1893 (aka Garrod's pads, dorsal pads, holoderma)
- Benign, asymptomatic
- Located in skin over dorsal aspects of MCP & PIP jts
- ? MISNOMER because most occur over PIPs jt, not over knuckles

Michelangelo's Moses, carved early 16th century with visible knuckle pads



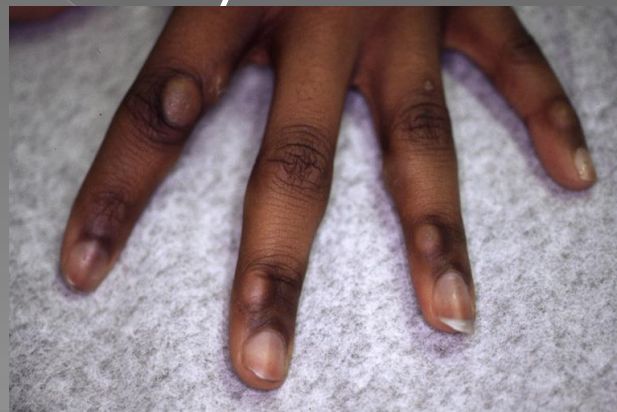
Knuckle pad (fibromatosis)

- **Frequency**
  - Common, up to 9%
  - Pts with palmer fibromatosis 4X as likely to have them as general population
  - May precede development of palmar or plantar fibromatosis

http://emedicine.medscape.com/article/1074379-clinical#a0217

Knuckle pad (fibromatosis)

- **Race/Sex/Age**
- No racial predilection
- Affects males & females equally
- Present any age, reported in young children who bite and suck their fingers; more commonly seen adults > 40 yrs



http://emedicine.medscape.com/article/1074379-clinical#a0217

Knuckle pad (fibromatosis)

- Etiology ?
  - Idiopathic
  - Genetic
  - Acquired as a response to *repetitive trauma* (sports or occupation)

http://emedicine.medscape.com/article/1074379-clinical#a0217

Knuckle pad (fibromatosis)

- **Mortality/Morbidity**
- Little morbidity
- Typically asymptomatic but can cause pain/tenderness and difficulty with hand functioning

http://www.dupuytren-online.info/knuckle_pads.html

Knuckle pad (fibromatosis)

- Well-circumscribed, smooth, firm, skin-colored dermal papules, nodules, or plaques,
- 0.5-3 cm
- Located on extensor aspect of PIP (more common) or MCP jts

http://emedicine.medscape.com/article/1074379-clinical#a0217

Knuckle pad (fibromatosis)

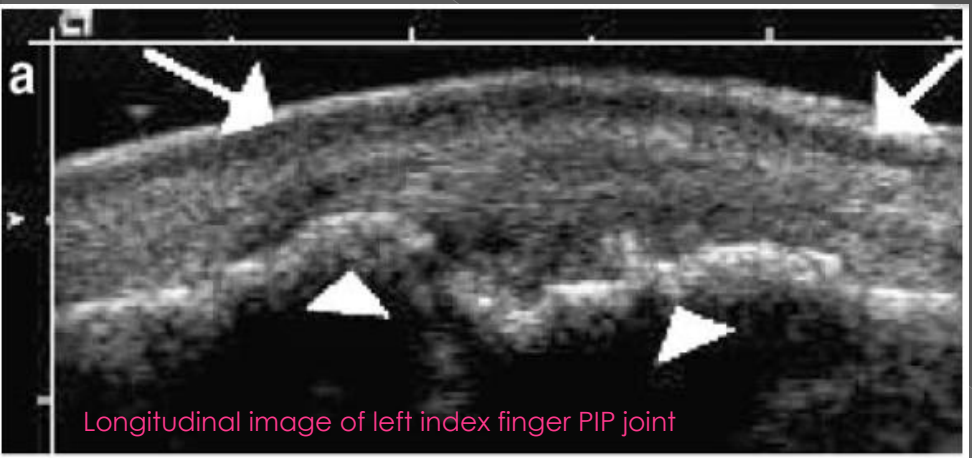
- Work-up
  - Imaging
    - XR:
      - Dorsal soft tissue thickening
      - No calcifications
      - Exclude inflammatory arthritis
    - US:
      - ***Increased dorsal subcutaneous thickening***
      - Either diffuse or focal hypoechoic areas
      - Absence of synovial proliferation
  - Biopsy if dx in doubt

http://radiographics.rsna.org/content/29/7/2143.figures-only

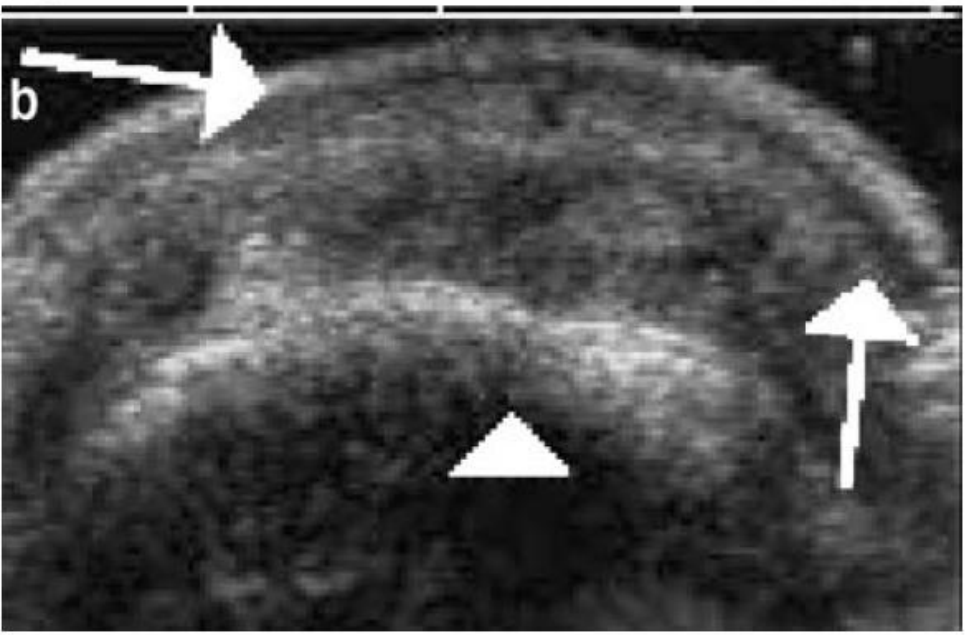
Fig. 1 Middle-aged male with bilateral knuckle pads. Note the findings are predominantly involving the PIP joints, although the MCP joints of left hand are also involved

Skeletal Radiol (2006) 35: 823–827

Knuckle pads in a patient with Palmer Fibromatosis

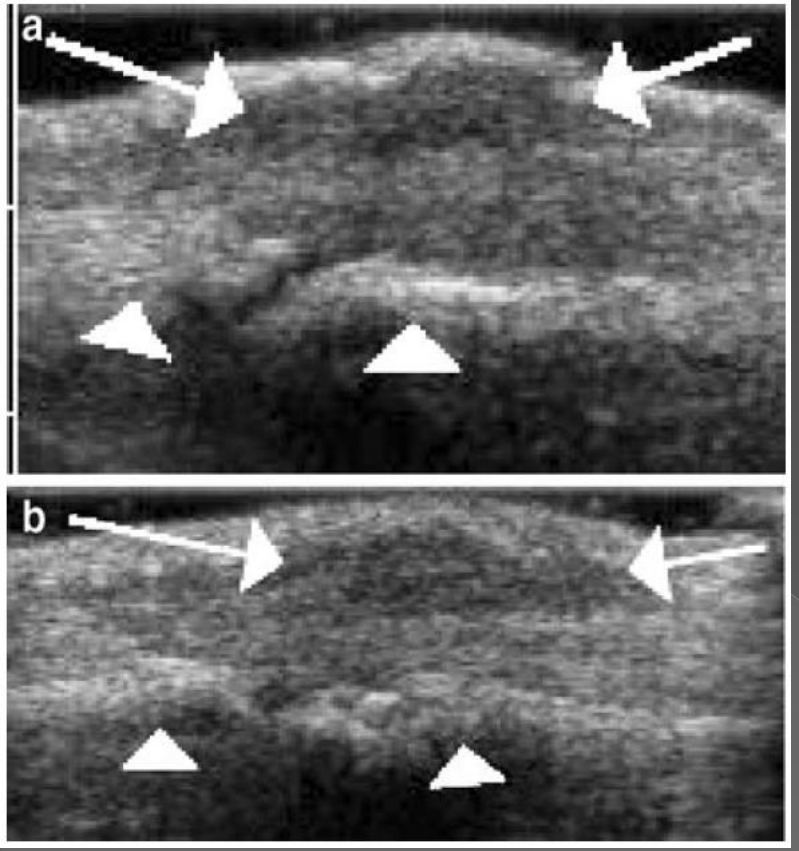


Longitudinal image of left index finger PIP joint



Diffuse thickening of the subcutaneous tissues with linear hypoechogenicity (arrows) noted in superficial subcutaneous layers

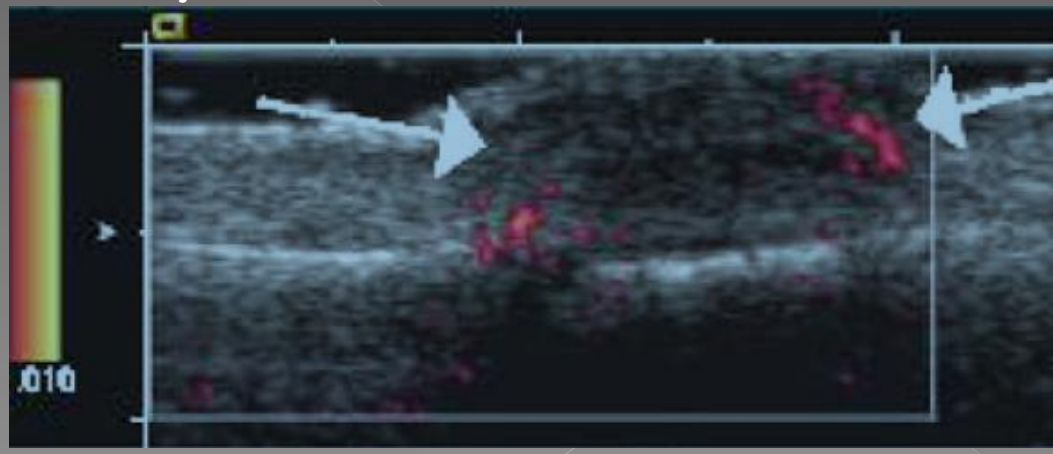
Skeletal Radiol (2006) 35: 823–827



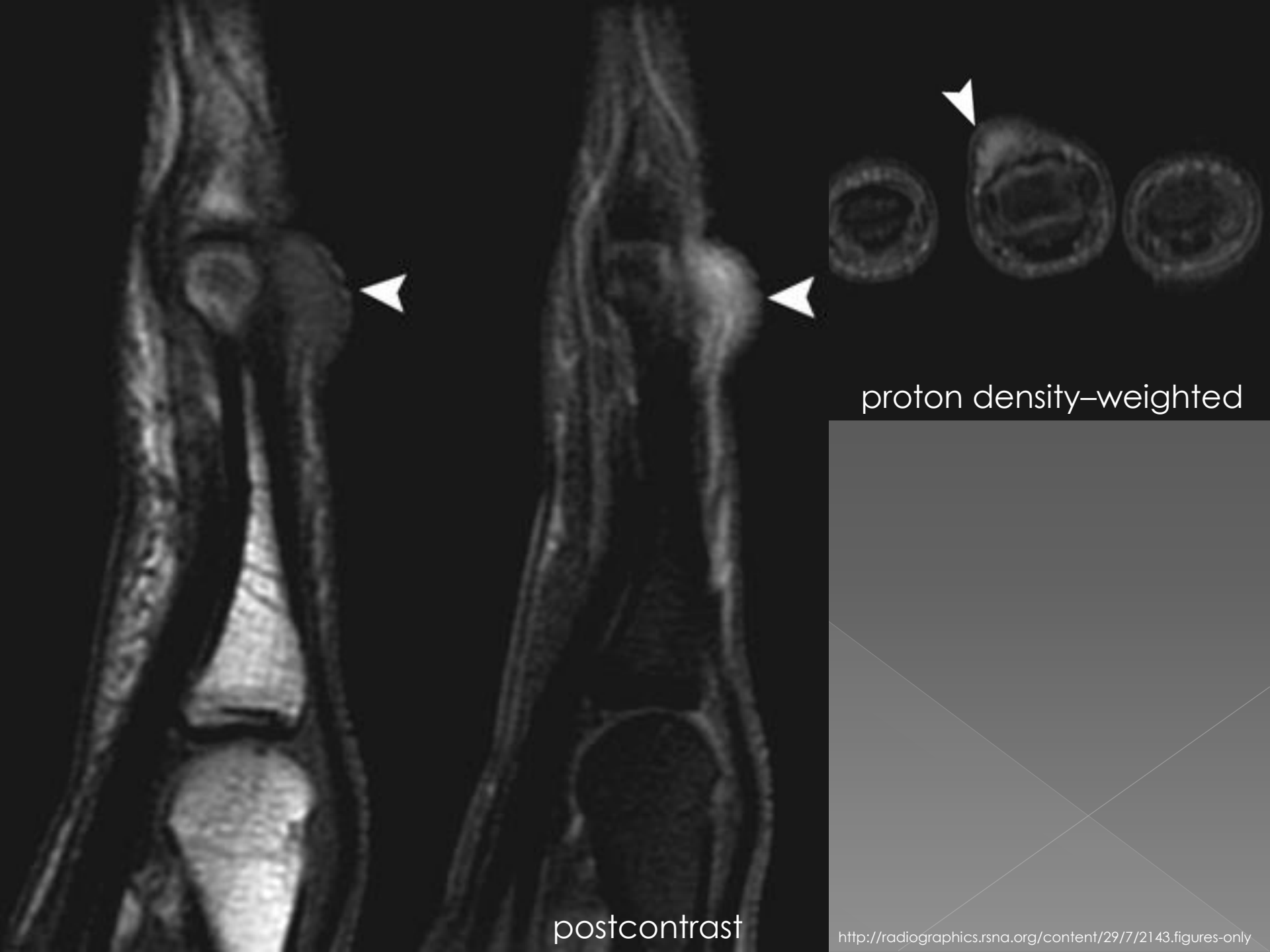
Longitudinal: rt long-finger PIP jt

Thickening of subcutaneous tissues with a focal hypoechogenic area (arrows) noted in superficial subcutaneous layers

US with power Doppler (PD) can help quantify inflammatory activity in joints



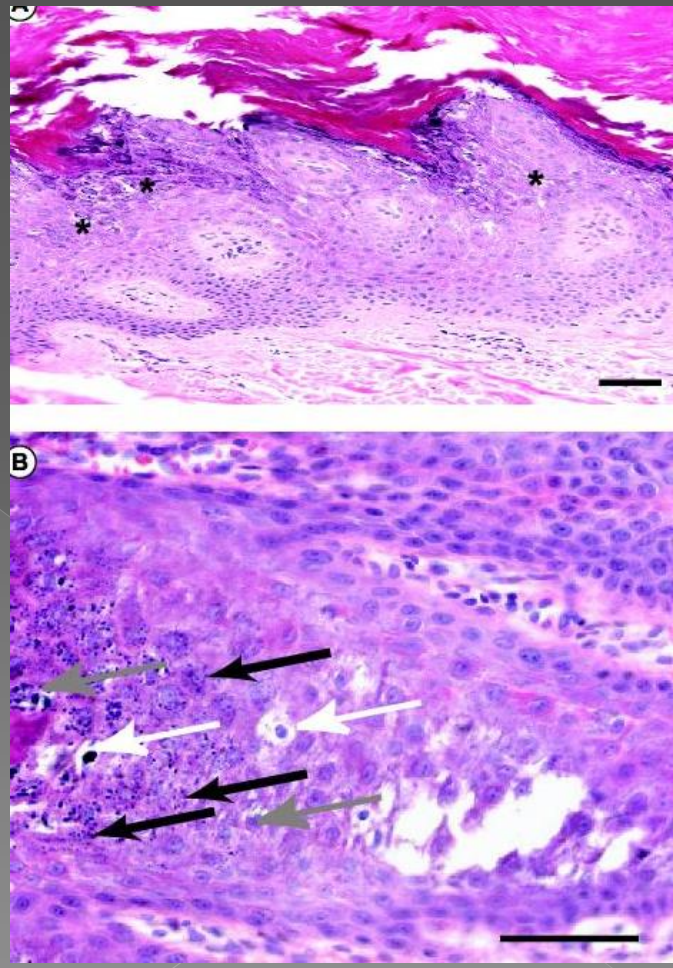
Skeletal Radiol (2006) 35: 823–827



http://radiographics.rsna.org/content/29/7/2143.figures-only

Knuckle pad (fibromatosis)

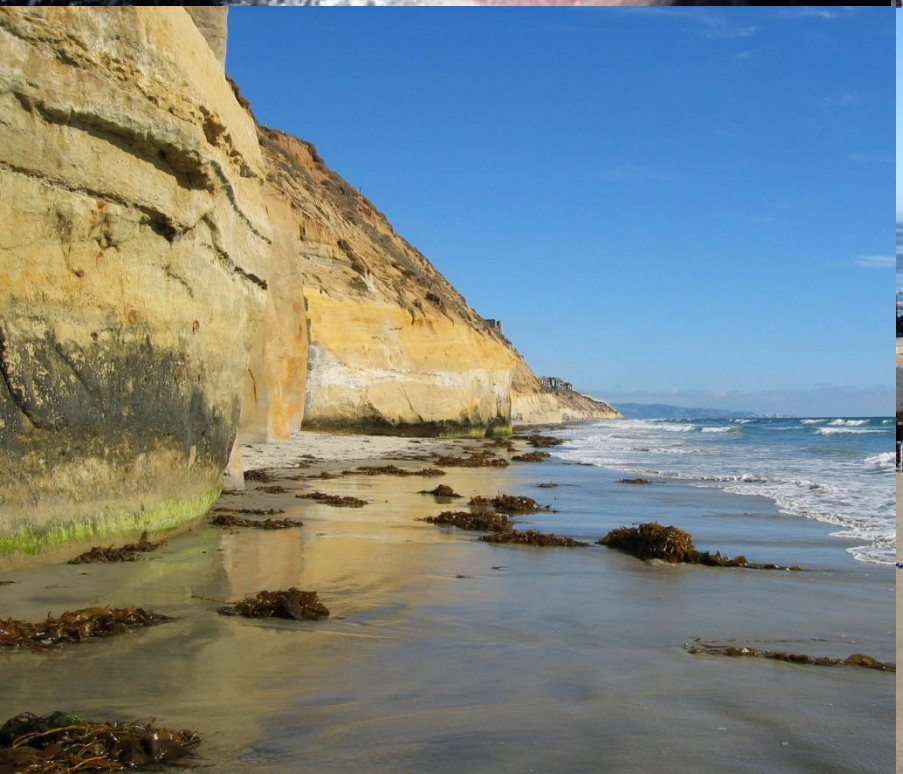
- Histology
  - **Epidermal abnormalities** include hyperkeratosis and mild acanthosis
  - Dermal changes include:
    - Slight proliferation of fibroblasts & capillaries in papillary dermis
    - Thickened, irregular collagen bundles are present, but little accompanying inflammation
    - Changes **resemble palmar fibromatosis**



http://www.jle.com/e-docs/00/04/46/DE/article.phtml?fichier=images.htm

Knuckle pad (fibromatosis)

- **Treatment & Prognosis**
  - Most asymptomatic, require no tx
  - Neither medical nor surgical interventions very effective (corticosteroid inj, radiation)
  - Surgical intervention if functional problem
    - Risk of recurrence
  - Spontaneous resolution can occur, especially if inciting repetitive injury is eliminated
  - ***Most persist indefinitely*** with little change




SHARK WARNING!
On 04/25/08 at approximately 7:40AM. A swimmer was attacked by what is believed to be a Shark in Solana Beach
Swimming, Surfing, and other Water Sports are not recommended until MONDAY, APRIL 28, 2008.


SOLANA BEACH


IMPERIAL BEACH

TOP 10 SAN DIEGO BEACHES

1. Blacks Beach + CO
2. La Jolla Cove +
3. Windansea Beach +
4. Tourmaline Beach +
5. Mission Beach + + + +
6. Ocean Beach + + + +
7. Sunset Cliffs + + TP
8. Coronado Beach + +
9. Silver Strand State Beach +
10. Imperial Beach + + +

N W E S

- Lifeguard Service
- Dog Friendly
- Sports
- Rocky Cliffs
- Surfing
- Wind Surfing
- Shops & Restaurants
- Great for Kids & Infants
- Amusements
- CO Clothing Optional
- Great for Walking
- TP Tide Pools

Outline

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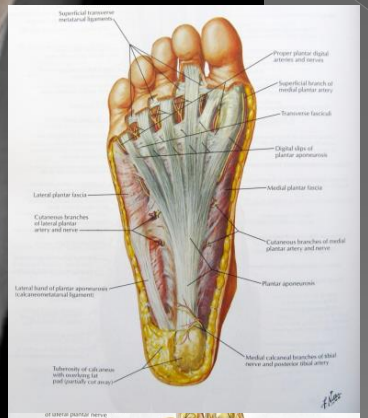
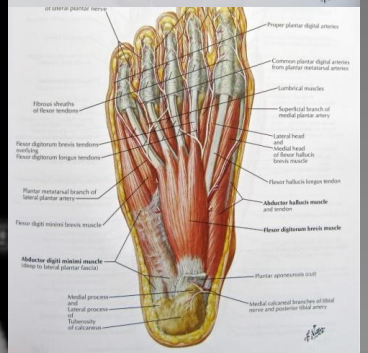
PLANTAR APONEUROSIS





ANATOMY

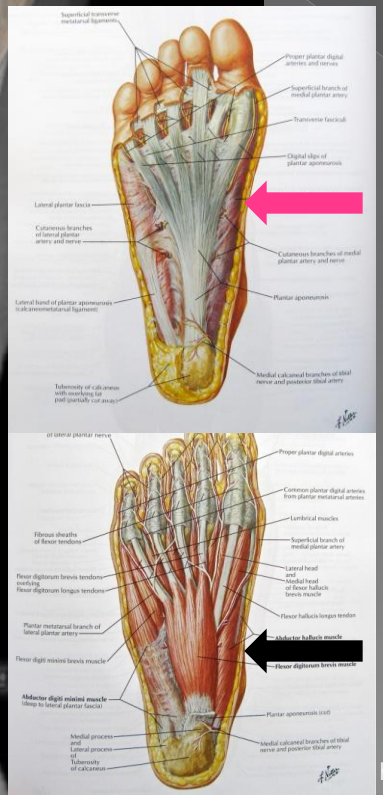
- fibrous aponeurosis composed of central, medial, lateral segments

-base attached to calcaneus, has fibers continuous with Achilles tendon

-plays significant role in **longitudinal arch support**

http://roentgenrayreader.blogspot.com/2011/10/plantar-aponeurosis-anatomy.html

PLANTAR APONEUROSIS



ANATOMY

Medial segment (purple): Arises from central segment and attaches to inferior portion of abductor hallucis muscle

http://roentgenrayreader.blogspot.com/2011/10/plantar-aponeurosis-anatomy.html

PLANTAR APONEUROSIS

ANATOMY

- **Central segment** (tan):
  - thickest component
  - *proximal attachment* to posterior aspect of medial calcaneal tuberosity (posterior to origin of FDB tendon)

http://roentgenrayreader.blogspot.com/2011/10/plantar-aponeurosis-anatomy.html

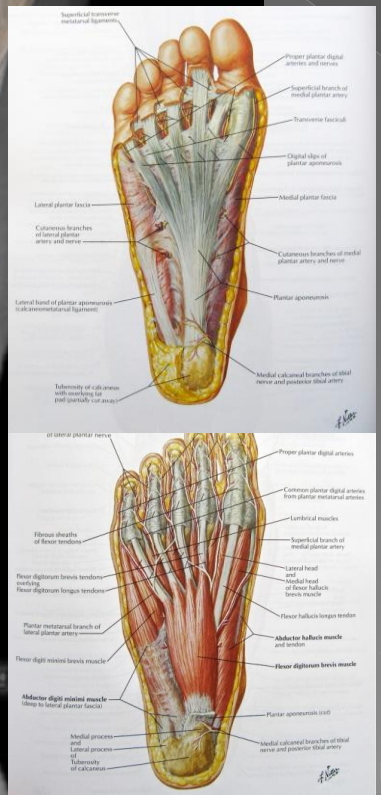
PLANTAR APONEUROSIS

ANATOMY

- **Central segment** (tan):
  - ***distal attachments*** are at level of MTP jts, dividing into <u>5 pairs</u> of superficial & deep fasicles
    - deep branches (**blue**) insert onto MTP jts
    - ***superficial branches bifurcate into sagittal septa,*** which attach onto plantar plates (**red**), interosseous ligament, and deep transverse metatarsal ligaments of 2nd-5th digits and *plantar plate/sesamoid bones* (**white**) of great toe

http://roentgenrayreader.blogspot.com/2011/10/plantar-aponeurosis-anatomy.html

PLANTAR APONEUROSIS

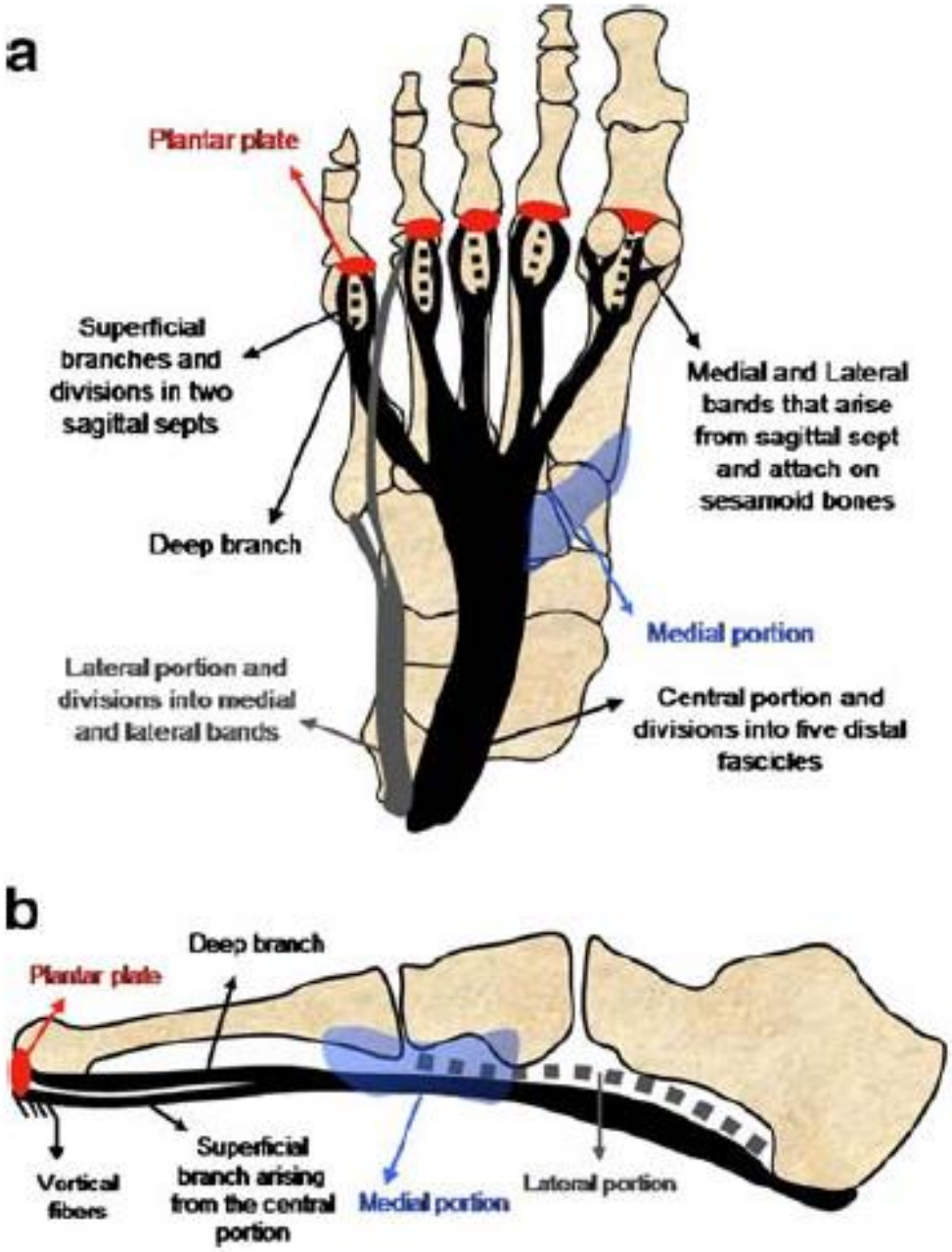


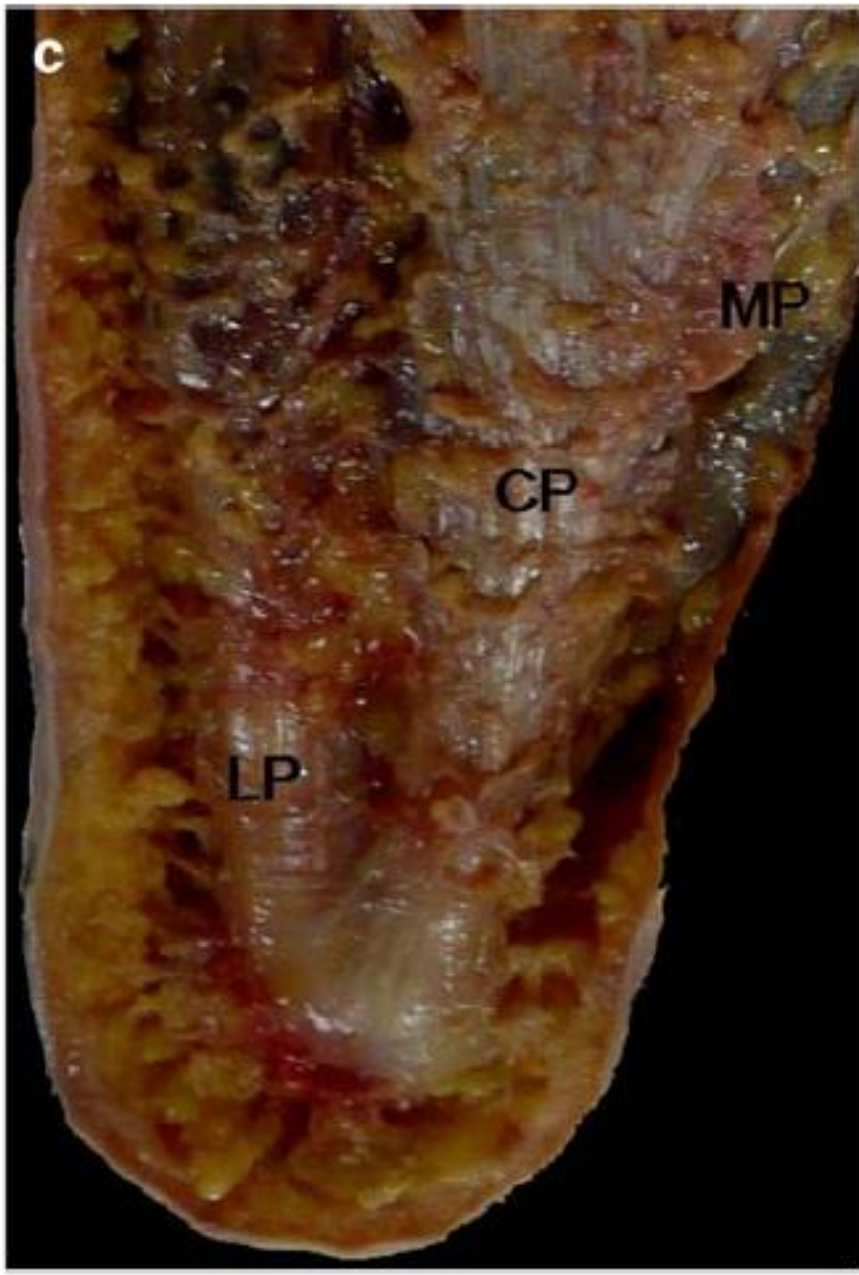
ANATOMY

Lateral segment (green):
*attaches proximally to lateral aspect of medial process of calcaneal tuberosity
*continuous medially with central segment

Distally, medial band (**arrows**) inserts onto **plantar plate of 4th** and sometimes third MTPS jts; ***lateral band attaches to base of 5th metatarsal***

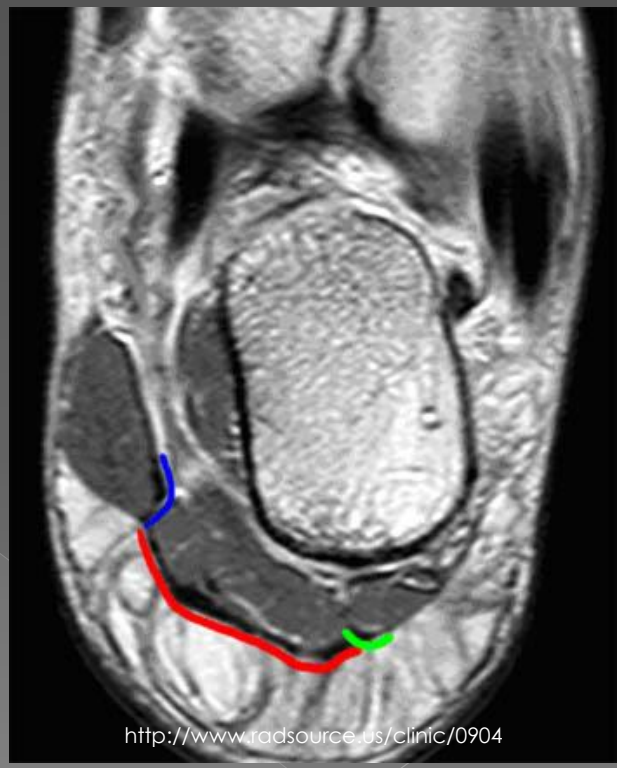
http://roentgenrayreader.blogspot.com/2011/10/plantar-aponeurosis-anatomy.html

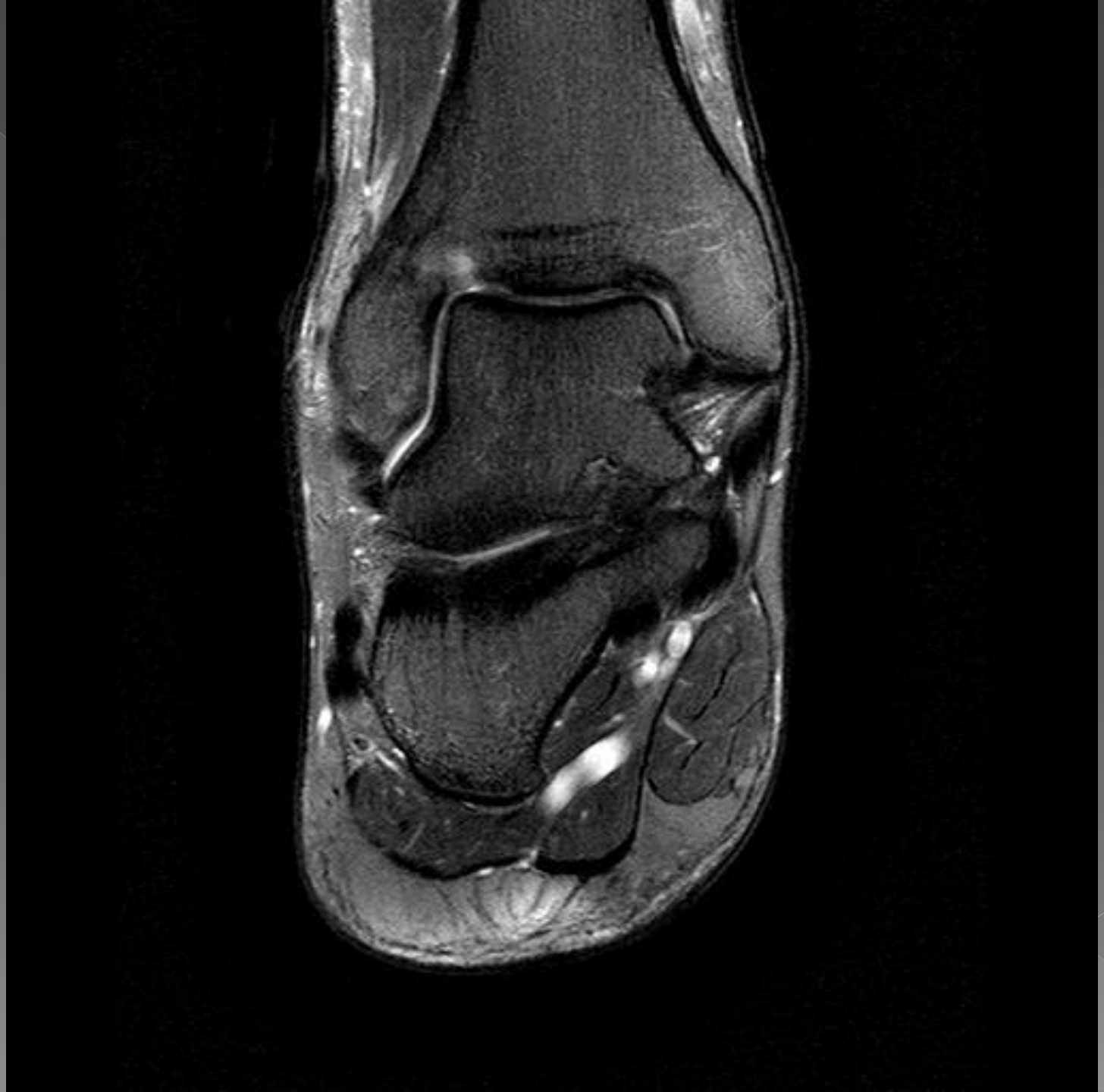

a
Plantar plate
Superficial branches and divisions in two sagittal septs
Deep branch
Lateral portion and divisions into medial and lateral bands
Medial and Lateral bands that arise from sagittal sept and attach on sesamoid bones
Medial portion
Central portion and divisions into five distal fascicles
b
Deep branch
Plantar plate
Vertical fibers
Superficial branch arising from the central portion
Medial portion
Lateral portion

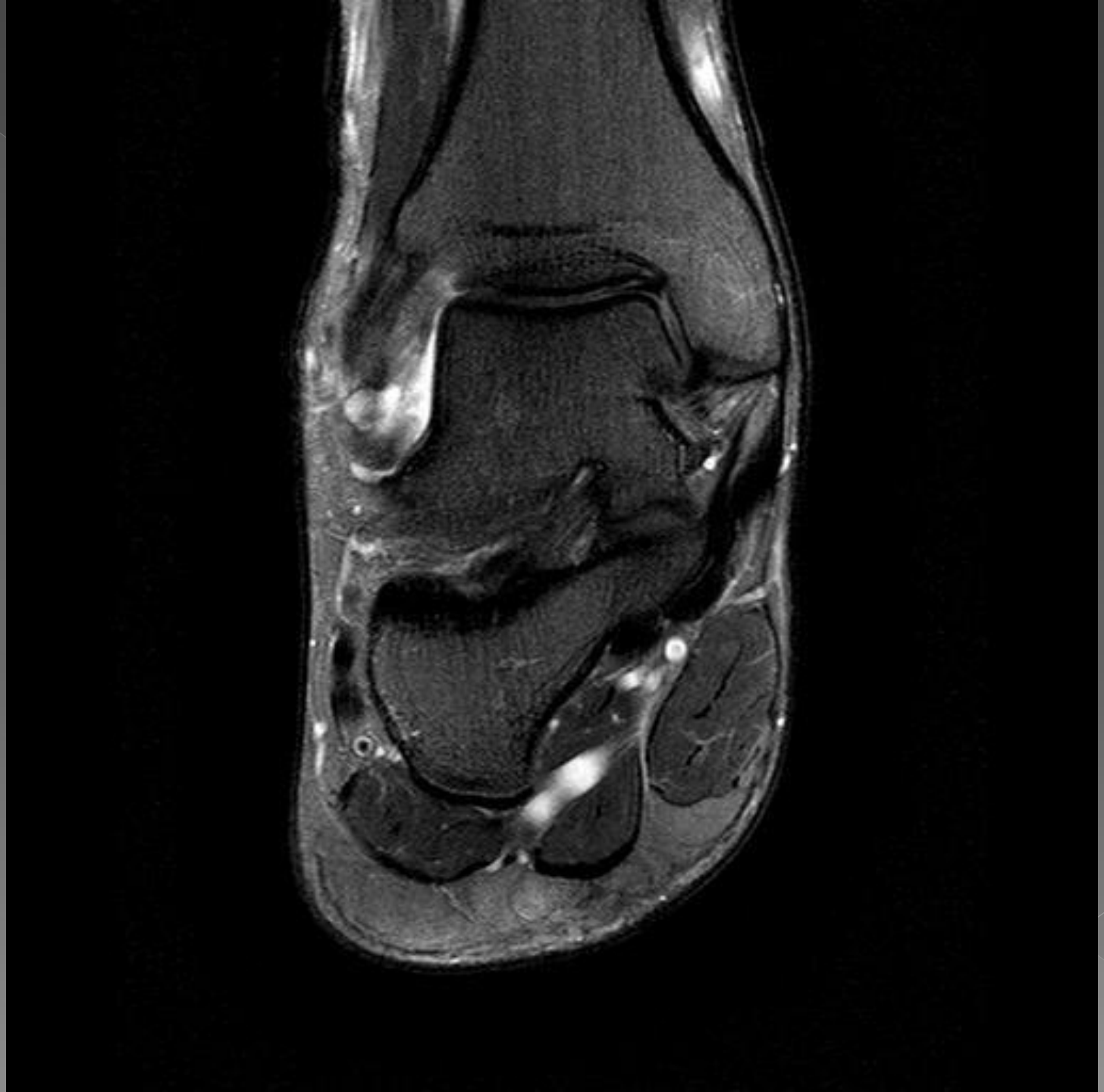

c
MP
CP
LP

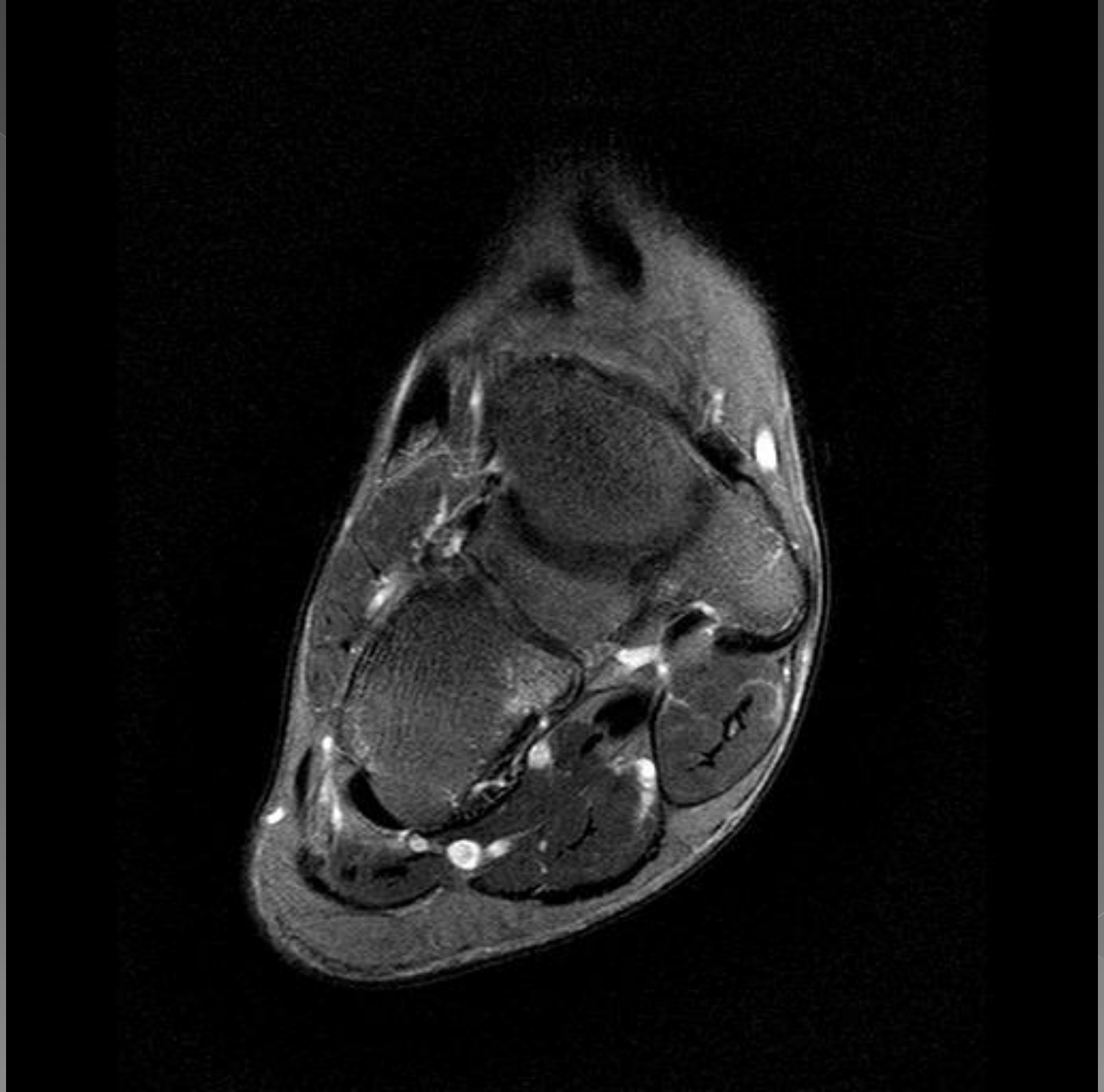
PLANTAR APONEUROSIS

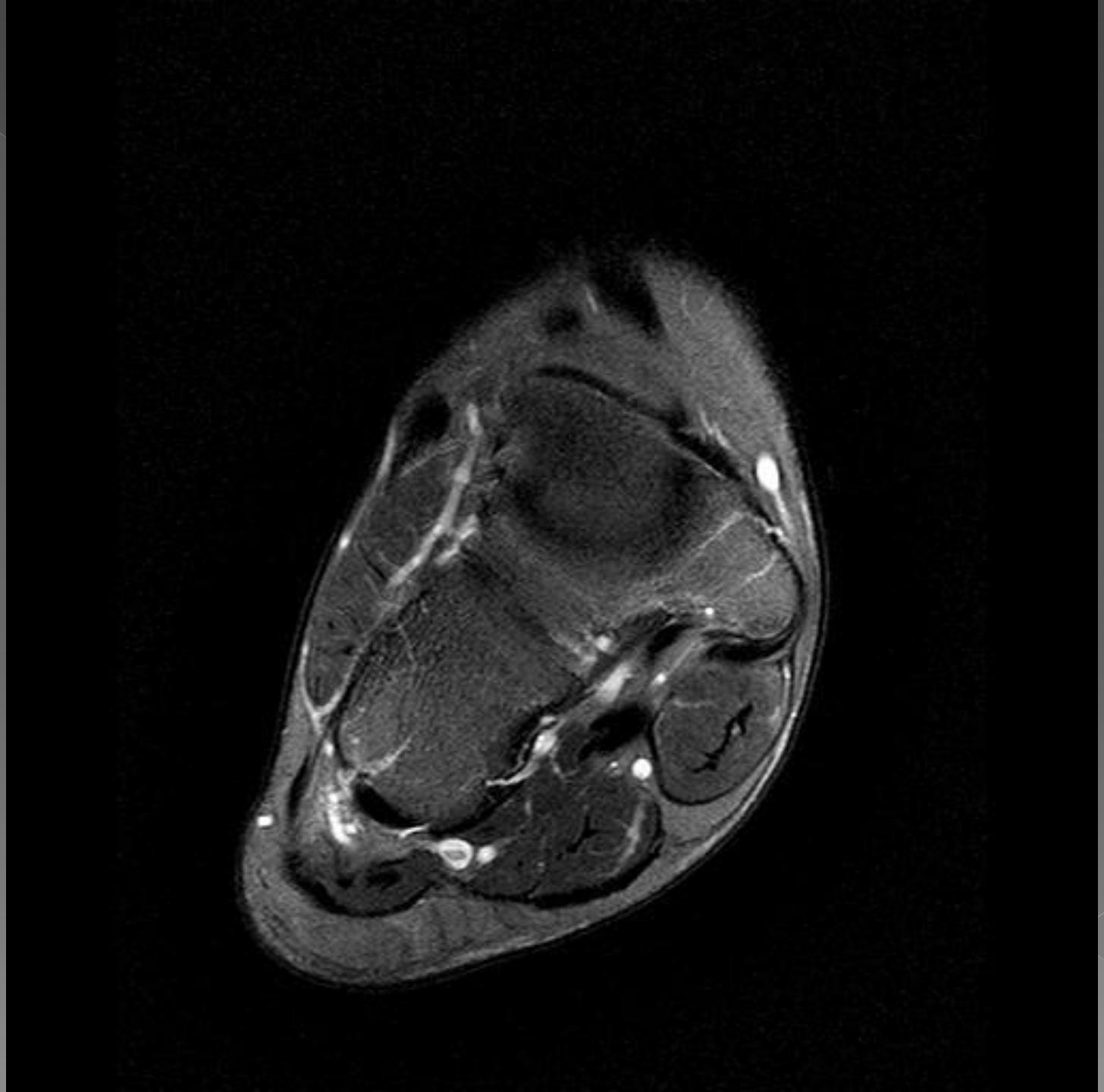
- Coronal image:
  - central portion overlying flexor digitorum brevis muscle (red)
  - medial portion beneath abductor hallucis muscle (blue)
  - lateral component overlying abductor digiti minimi muscle (green)

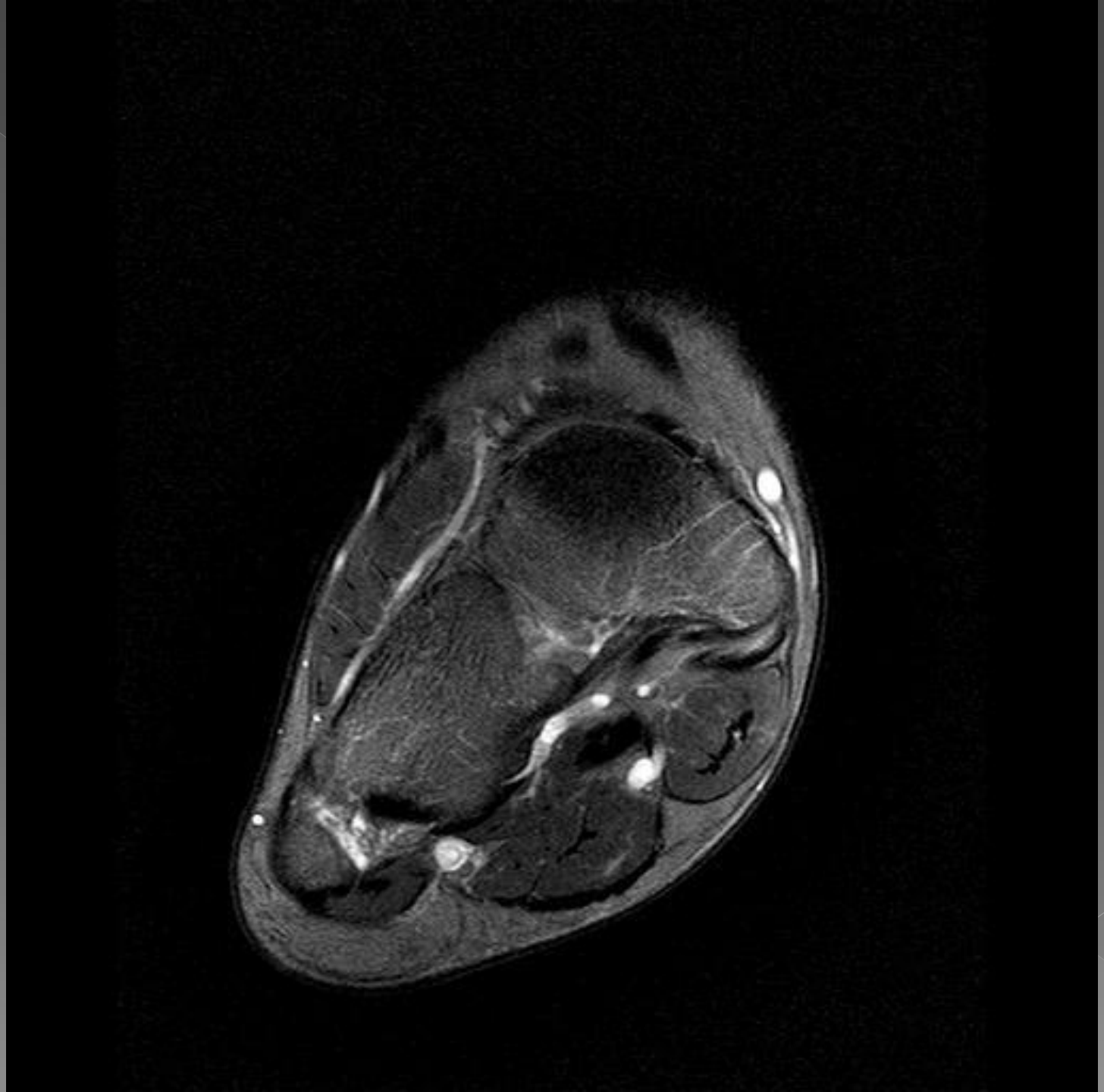


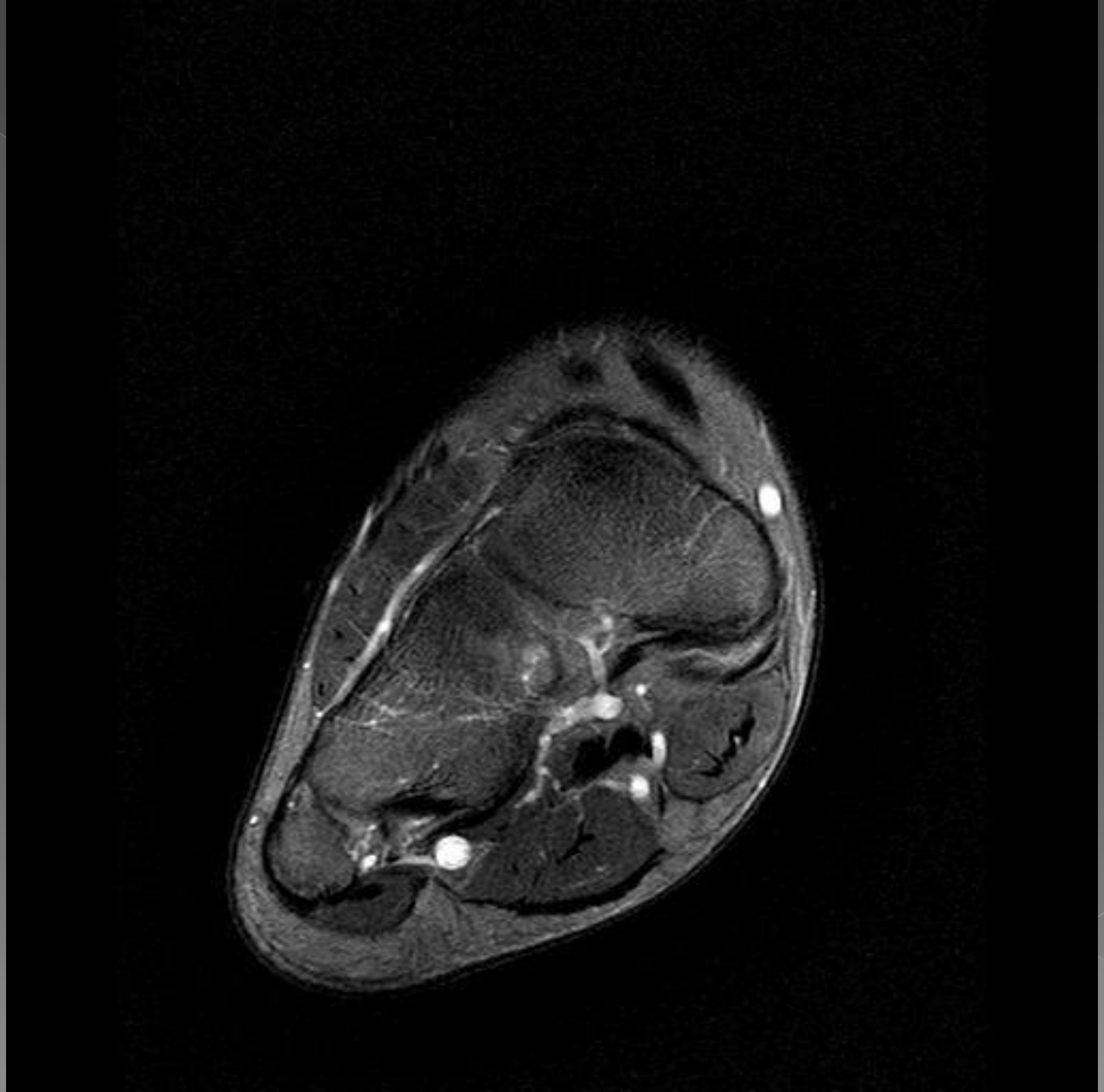


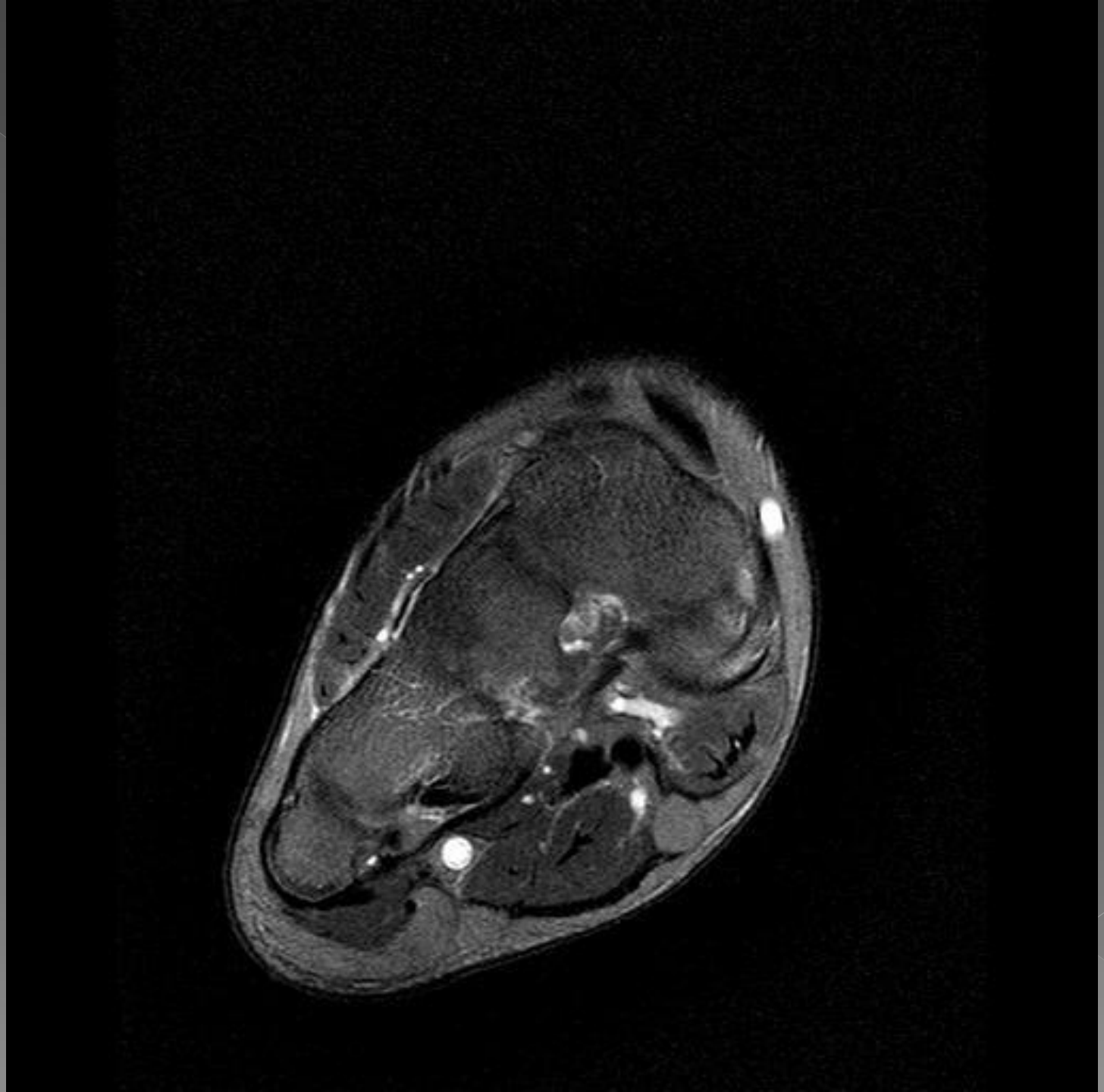


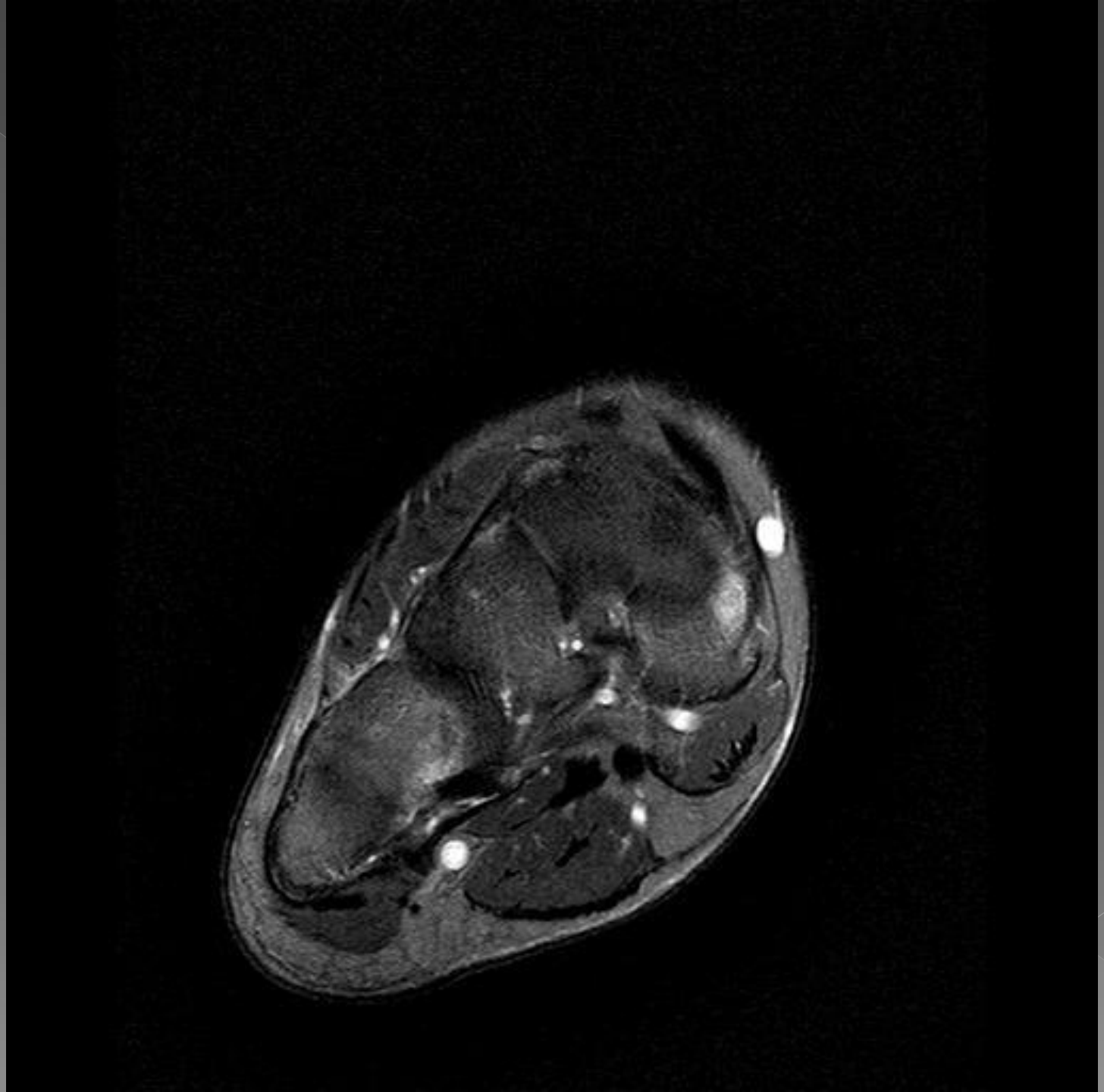


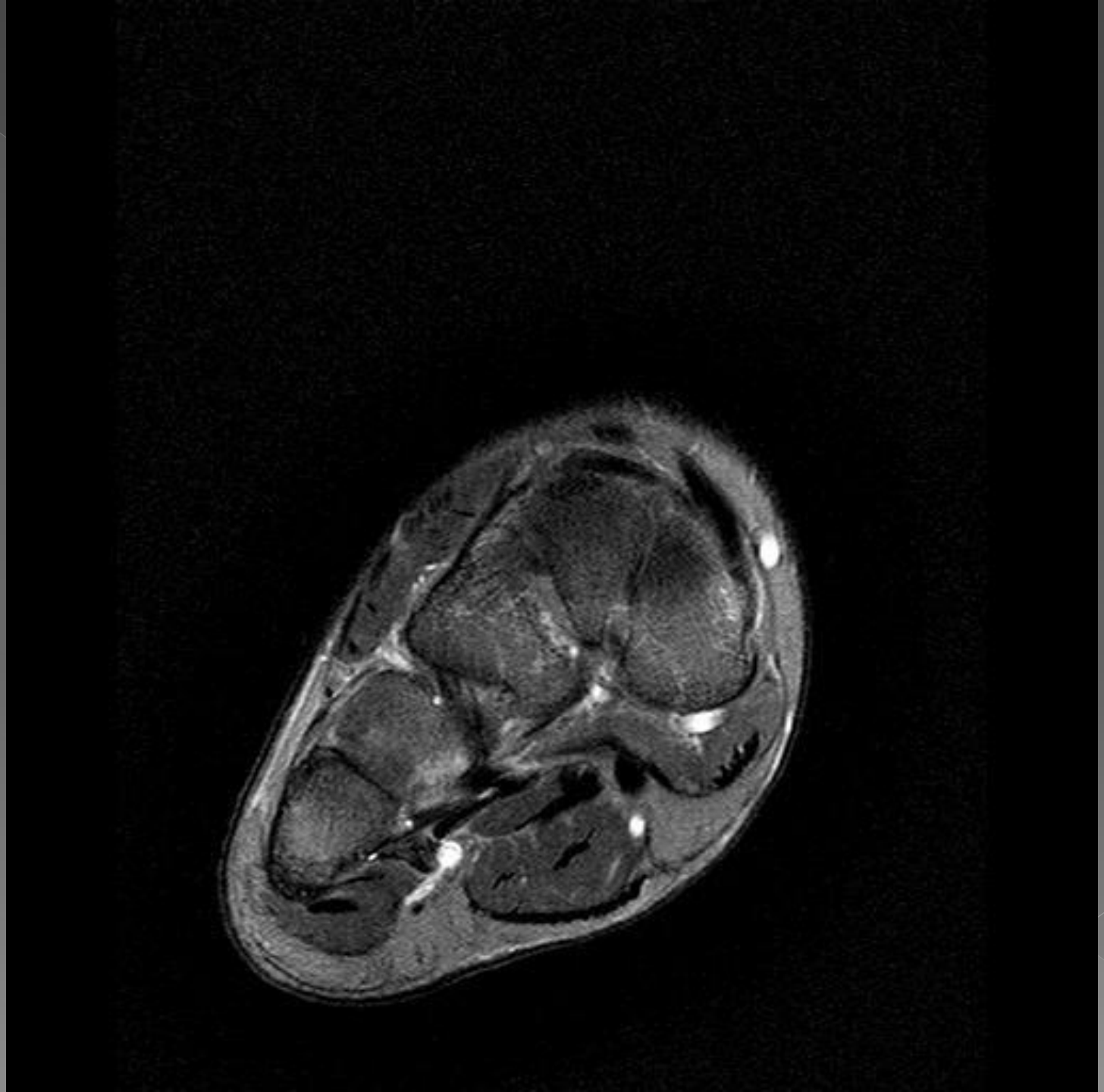


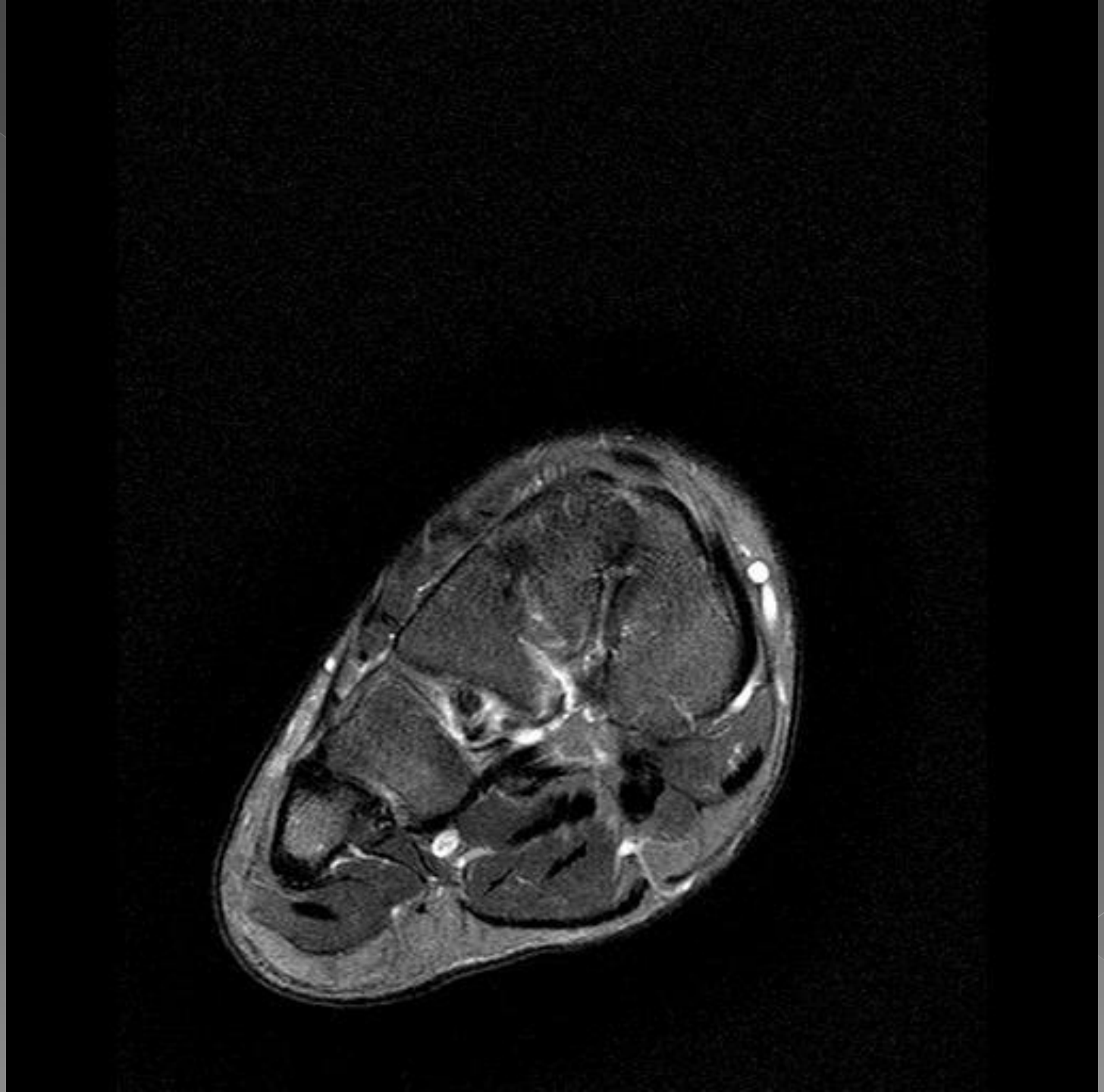












PLANTAR APONEUROSIS

- Coronal image:
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  - medial portion beneath ***abductor hallucis muscle*** (blue)
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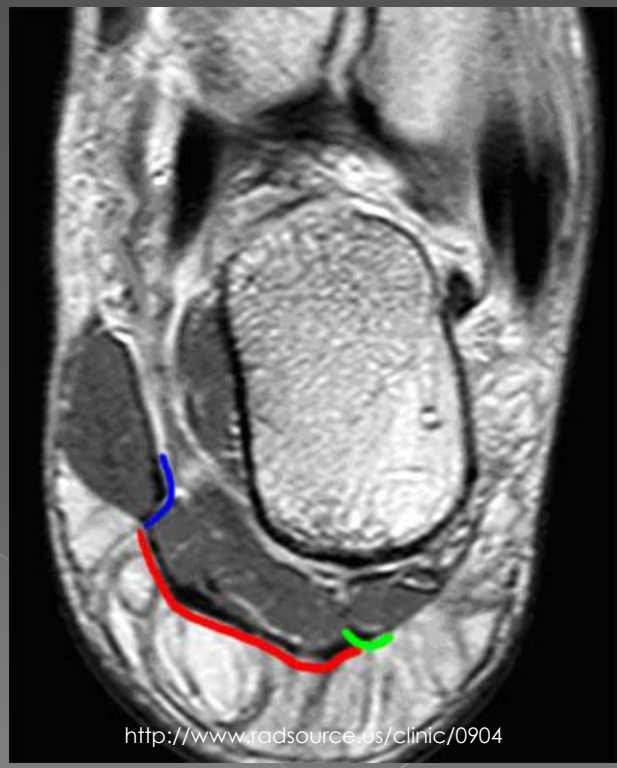


Plantar Fibromatosis

http://www.feetfixer.com/html/plantar_fibroma_surgery.html

- **Clinical Features**
- Benign nodular fibroblastic proliferative disorder of pl. aponeurosis
- Reported by Dupuytren1832, more extensively described by German Surgeon Dr. Ledderhose1897 (aka Ledderhose's disease, Morbus Ledderhose)
- Prevalence 0.23%
- Most common 30–50 yrs
- Men 2X

Plantar Fibromatosis

- **Clinical Features**
- Most solitary, unilateral; but multiple & b/l 20%–50% of pts (typically metachronous with 2–7 yr interval)
- Concomitant palmar disease in10%–65% of pts (usually metachronous with 5–40 yr interval; rarely synchronous)
- Knuckle pads seen in up to 42% of pts

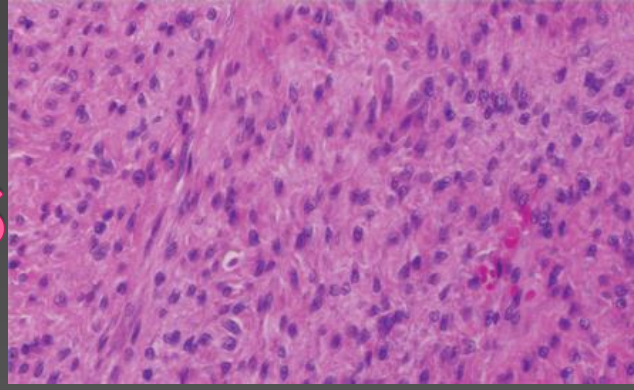
http://www.bonetumor.org/tumors-foot-and-ankle/plantar-fibroma-foot-and-ankle

Plantar Fibromatosis

- **Clinical Features**
- *Multifactorial etiology*, including genetic & traumatic causes
- **More common** in pts with diabetes, epilepsy, keloids, alcoholism with liver disease
- Present as firm soft-tissue mass on medial aspect sole of foot
- Multiple nodules 33%
- Frequently asymptomatic (can be tender or activity related pain)
- No contracture
- Rare cases, large lesions may invade adjacent muscles or neurovascular structures

http://www.bonetumor.org/tumors-foot-and-ankle/plantar-fibroma-foot-and-ankle

Plantar Fibromatosis



[H-E] stain) reveals relatively hypercellular tumor composed of fascicles of fibroblasts that represent more proliferative phase

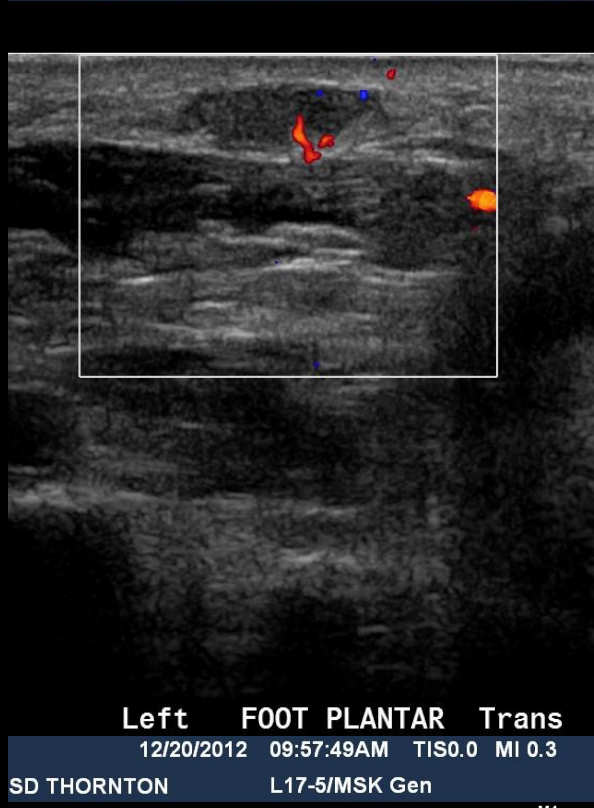
- **Pathologic Features**
- At gross pathologic / histologic examination: **identical to palmar fibromatosis**
  - **Three phases**
    - Proliferative phase: Nodular fibroblastic proliferation
    - **Active phase**: Collagen synthesis and deposition
    - Mature phase: Reduced fibroblastic proliferation, collagen maturation
- Forms larger masses (2–3 cm; often coalescent nodules) compared with palmar lesions
- May be adherent to overlying skin
- *Mitotic activity can be more prominent in larger lesions*

Plantar Fibromatosis

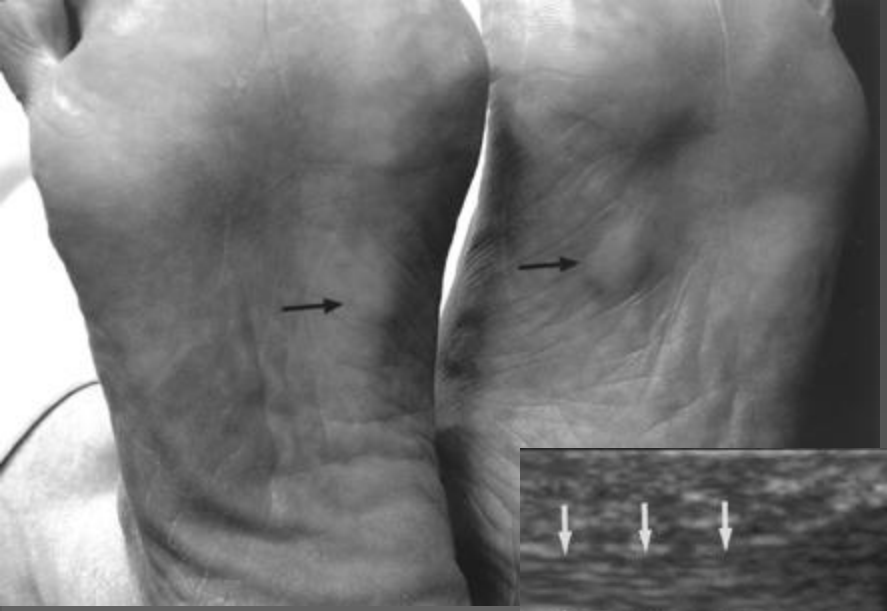
- **Imaging Features**
- Radiography almost always normal

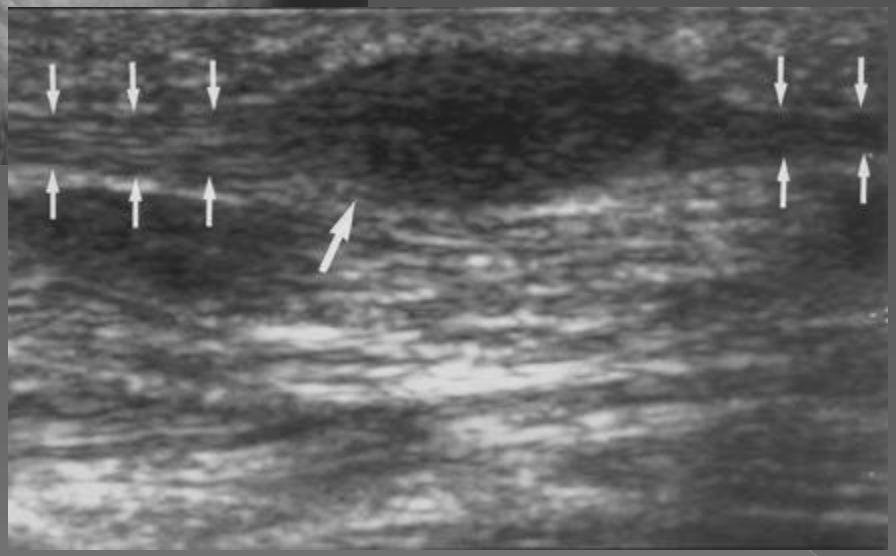
- US:
  - Typically **hypoechoic or mixed echogenic nodules** in subcutaneous tissues superficial to plantar aponeurosis (which is often thickened), either medially (60% of cases) or centrally (40%)
  - May be well defined (64%) or ill-defined (36%), frequently fusiform (76%)
  - Intrinsic vascularity (10%)
  - Posterior acoustic enhancement (20%)

http://radiographics.rsna.org/content/29/7/2143/F16.expansion.html



55 y/o with mass bottom foot



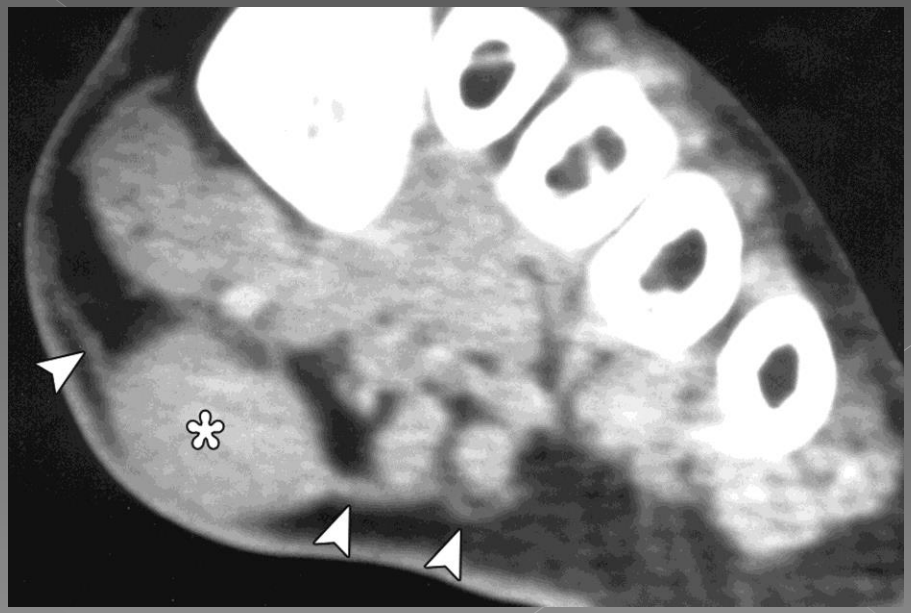


52-year-old man
well-defined fusiform hypoechoic nodule arising within plantar fascia

AJR November 2002 vol. 179 no. 5 1167-1172

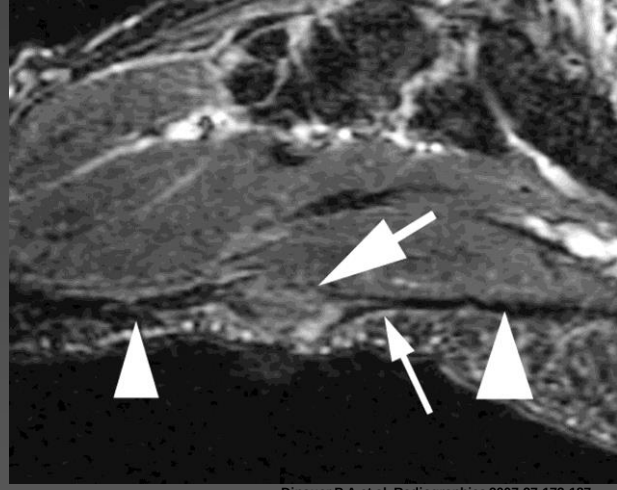
Plantar Fibromatosis

- **Imaging Features**
- CT: nonspecific soft-tissue mass with attenuation similar to or mildly higher than muscle



http://radiographics.rsna.org/content/29/7/2143/F11.expansion.html

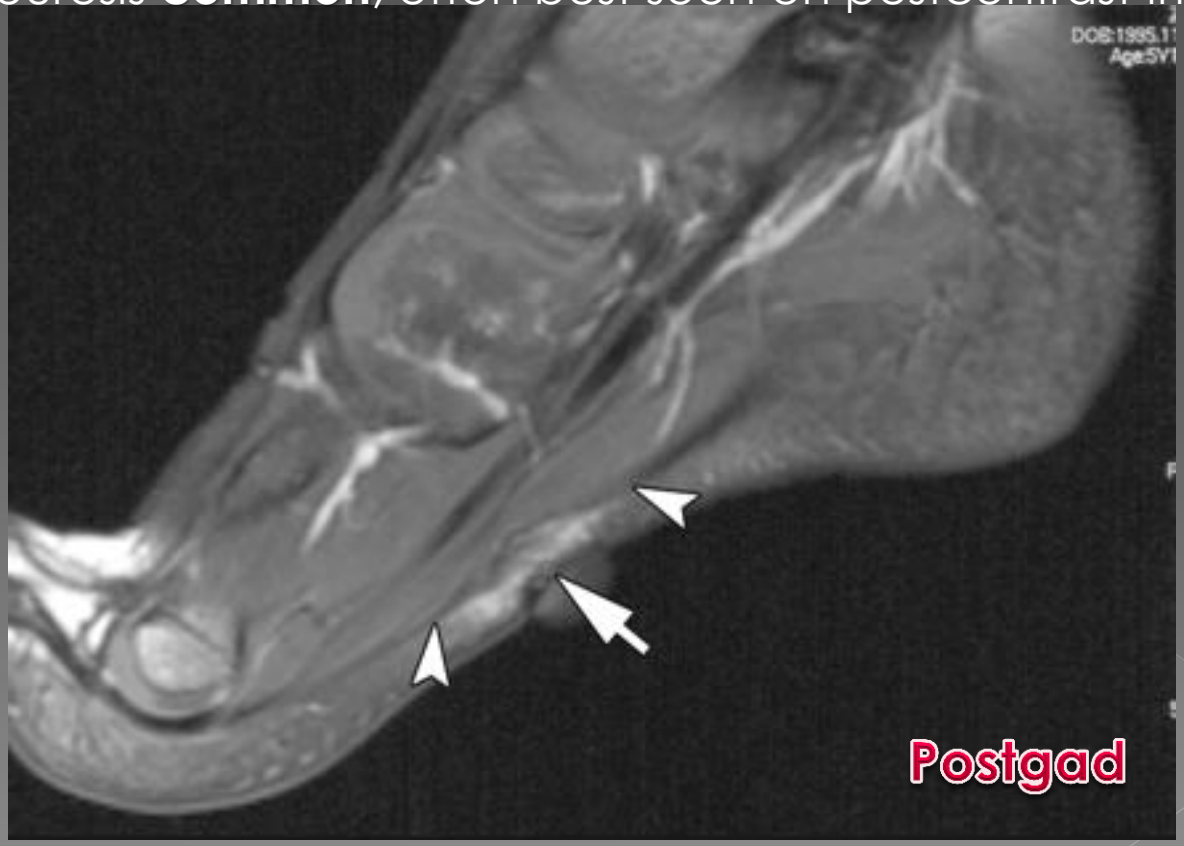
Plantar Fibromatosis


Dinauer P A et al. Radiographics 2007;27:173-187

- **Imaging Features**
- **MRI**: well- or ill-defined superficial soft-tissue mass occurring along deep pl. aponeurosis
- Often inseparable from adjacent plantar musculature
  - Deep invasion occurs in minority
- Typically: heterogeneous SI (92% of cases), predominantly low to intermediate SI (similar or equal to skeletal muscle) on T1WI (100%)/T2WI (78%)
- High T2 signal (22%)
- Enhancement common (93%)
- Degree of enhancement marked in 64%, mild in 36%

"Fascial Tail Sign"

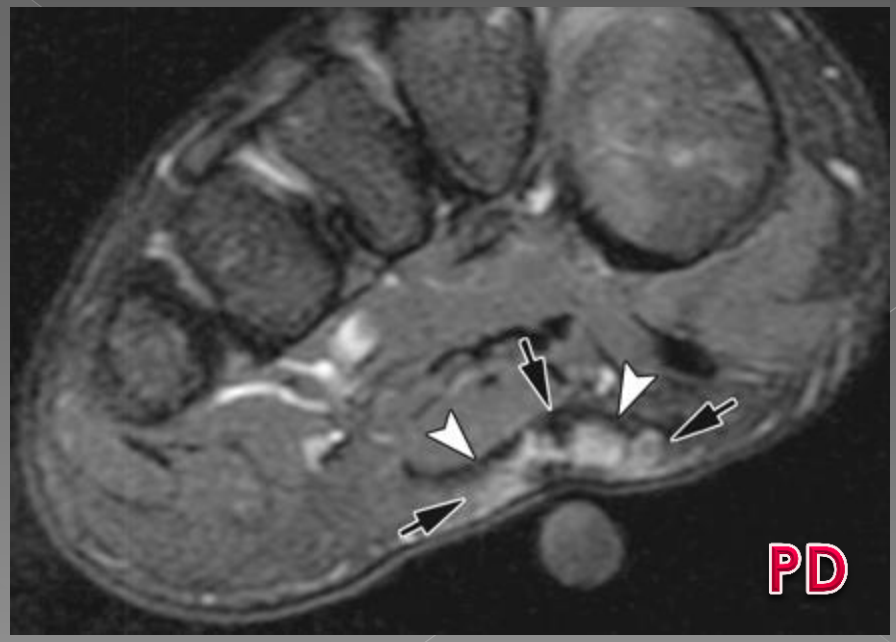
Linear tail of extension ("fascial tail" sign) along plantar aponeurosis **common**, often best seen on postcontrast images



http://radiographics.rsna.org/content/29/7/2143/F16.expansion.html

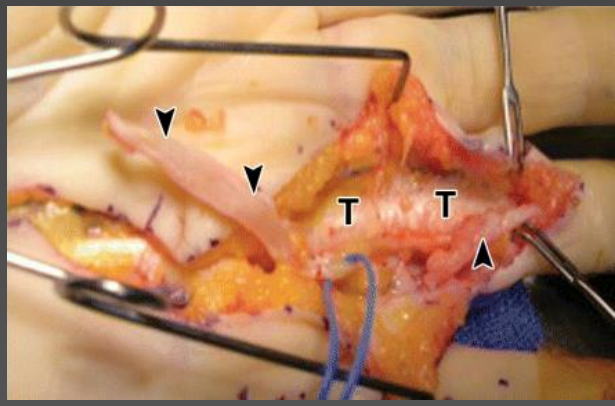
Plantar Fibromatosis

- **ON ANY FORM OF IMAGING…**
  - If nodule has indistinct ± infiltrative superficial or deep margin, consider aggressive plantar fibromatosis



Images Courtesy of Karen Chen, MD

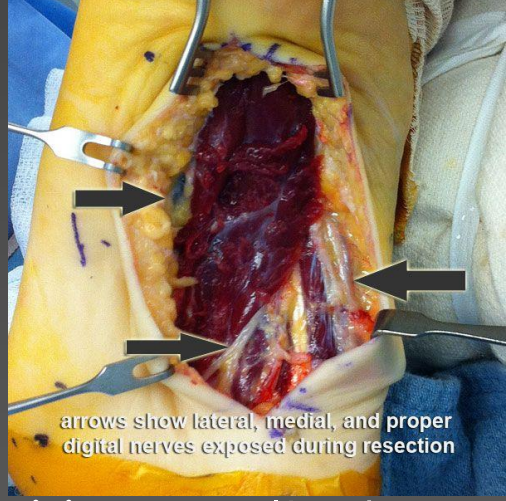
Plantar Fibromatosis



November 2009 RadioGraphics, 29, 2143-2183.

- **Treatment and Prognosis**
- In contradistinction to palmar fibromatosis, tx is **conservative in majority**
  - Nodules become smaller and lesser painful
  - Footwear modifications, pads, or orthotics aimed at relieving symptoms
  - Intralesional steroid injections successful in some
- **Surgical resection is reserved** for large, infiltrative lesions that cause significant disability and are refractory to nonoperative management

Plantar Fibromatosis



- **Treatment and Prognosis**
- Historically, surgical tx consisting of simple excision resulted in high rates of local recurrence (20%–40%)
- **Wide excision, including resection of *normal fascia* (ie, at surgical inspection) proximal and distal to lesion(s) <u>now advocated</u>**
- Radiation tx in foot often poorly tolerated, typically reserved for use in conjunction cases involving wide re-excision or for unresectable recurrent lesions
- Increased rate of local recurrence has been associated with multiple nodules, b/l lesions, postoperative neuromas, and + fam hx

http://www.bonetumor.org/tumors-foot-and-ankle/plantar-fibroma-foot-and-ankle

Outline

- **Superficial MSK Fibromatoses**
  - Palmar Fibromatosis
  - Knuckle Pad Fibromatosis
  - Plantar Fibromatosis
- **Deep MSK Fibromatoses**
  - Desmoid Type Fibromatosis
  - Abdominal Wall Fibromatosis


WELCOME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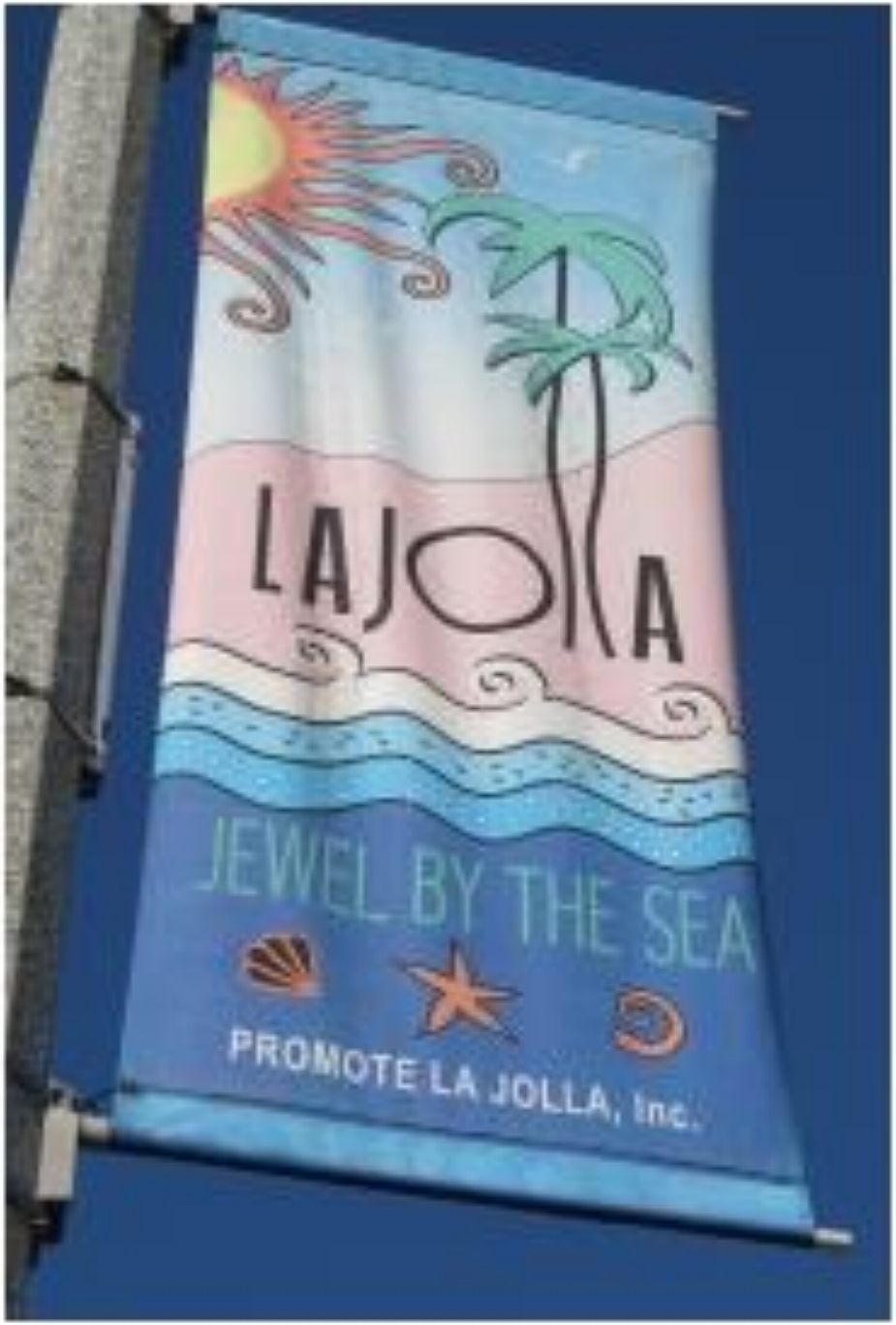
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Outline

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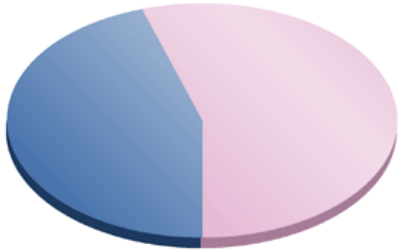
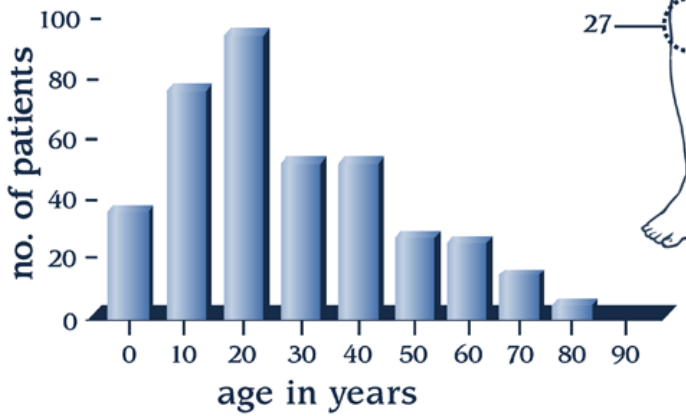
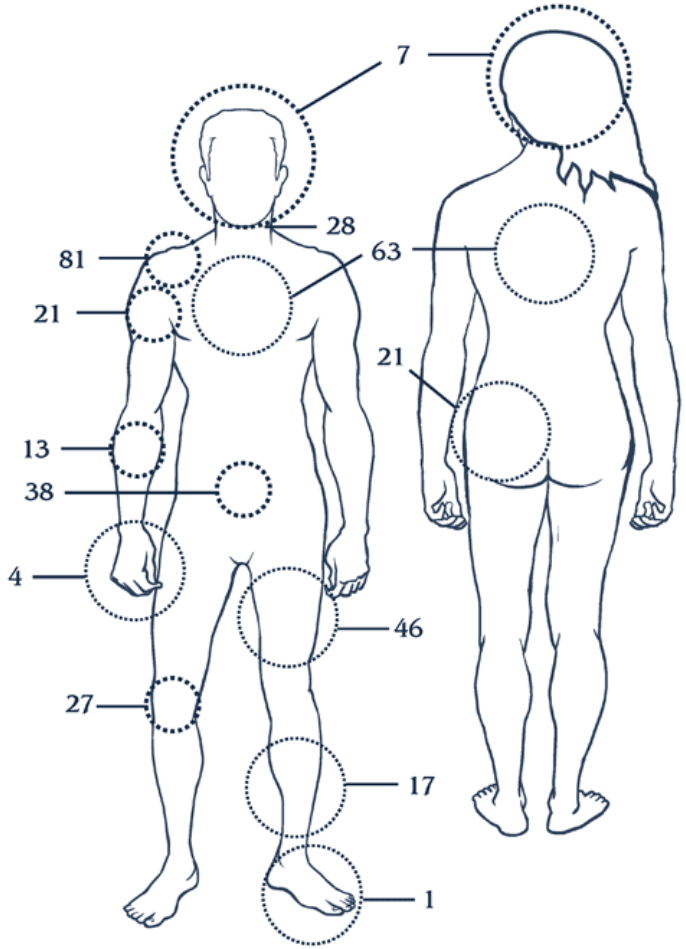
AGGRESSIVE FIBROMATOSIS

Data from ENZINGER & WEISS



♂ MALE < FEMALE ♀





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Desmoid-Type Fibromatosis (DTF)

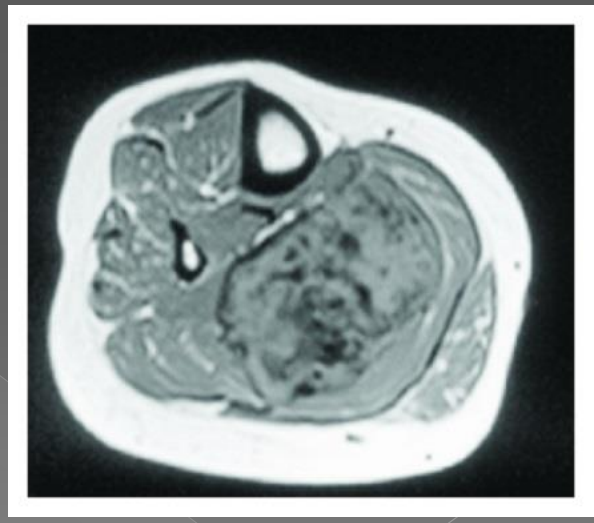
- **First described by McFarlane (1832)**
- **1838 Muller first coined term "desmoid"**

http://worddomination.com/fibromatosis.htm

- **Synonyms**
  - extraabdominal desmoid
  - desmoid tumor
  - aggressive fibromatosis
  - musculoaponeurotic fibromatosis
  - well-differentiated nonmetastasizing fibrosarcoma

Desmoid-Type Fibromatosis

- **Definitions**
- Benign, but locally aggressive, clonal fibroblastic proliferation



http://www.hindawi.com/journals/srcm/2012/578052/f

- **Etiology unknown**
  - Multifactorial pathogenesis
    - Genetic
    - Endocrine, as associated with pregnancy
    - Trauma

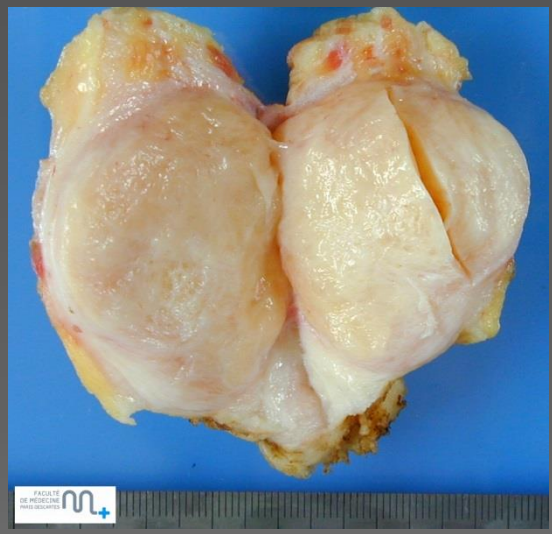
Clinical Features of DTF

- **Painless, firm, ill-defined deep** soft-tissue mass in extremities or head & neck
- Locally invasive
- Commonly recur locally
- Do not metastasize

May 2008 RadioGraphics, 28, 901

Pathologic Characteristics of DTF

- Macroscopic Examination:
  - Nonencapsulated gray-white tissue confined to musculature and overlying fascia
  - Gross specimens are firm / glistening white on cross section
  - Resembles scar tissue
  - Hemorrhage or necrosis not typical



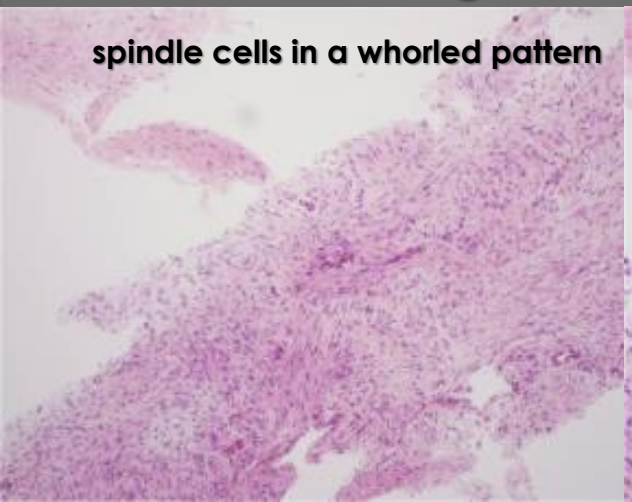
http://www.humpath.com/spip.php?article2440

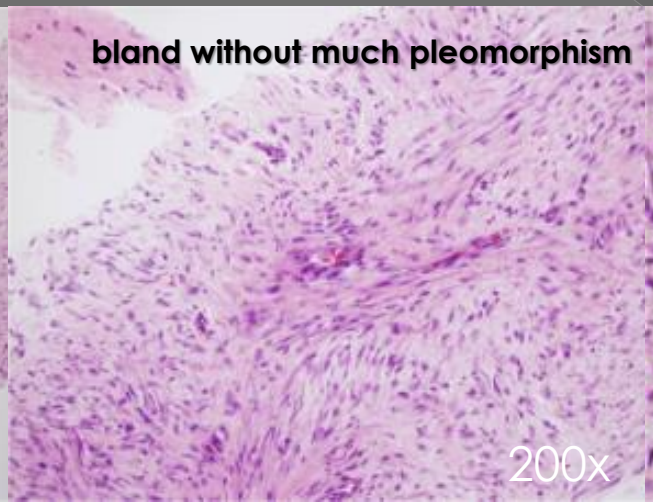
http://radiographics.rsna.org/content/21/3/585/F1.expansion.html

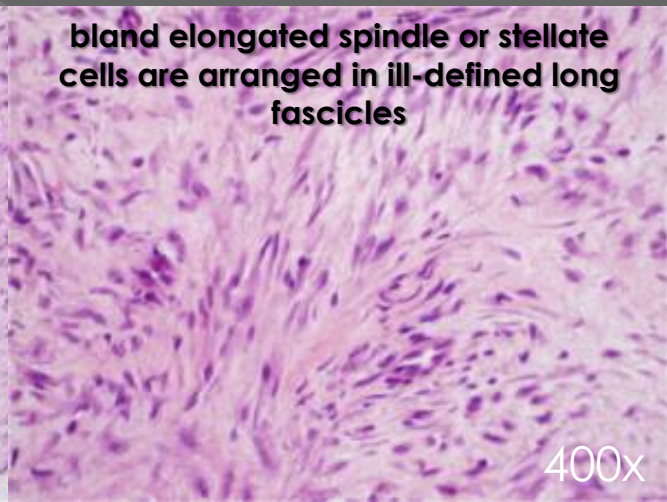
http://www.orthopaedicsone.com/display/PORT/Aggressive+Fibroma

Histologic Characteristics of DTF

- Microscopic examination:
  - Characteristic growth along fascial planes
  - May infiltrate adjacent subcutaneous tissue & muscle
  - Alternating bundles of locally infiltrating, monomorphic elongated, spindle-shaped fibroblast and myofibroblast bundles within collagenous stroma
  - Cellularity is low
  - Collagen interlaced between tumor cells

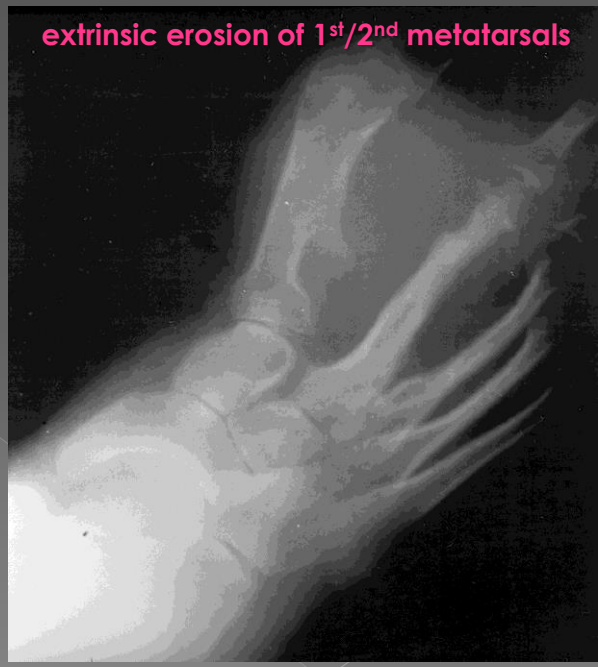






Radiographic Findings of DTF

- Usually normal
- Calcification or ossification is rare
- Bulging or puckering of overlying skin
- Bone involvement uncommon but can include *erosion or periosteal reaction*
  - Increased with recurrent tumors



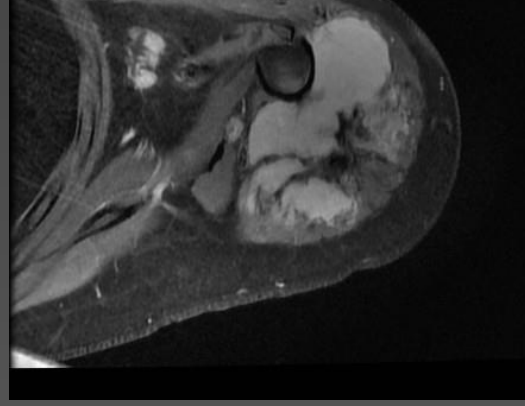
http://radiographics.rsna.org/content/21/3/585/F31.larg

CT Findings of DTF

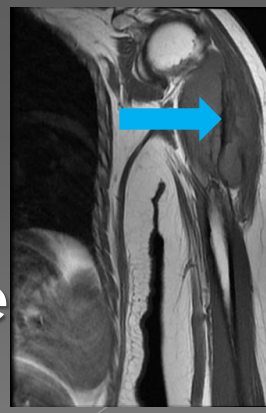
- Nonspecific, ill-defined soft tissue mass
- ***Variable attenuation***: higher, lower, or similar to muscle
- Mild, heterogeneous enhancement (typical)
  - Enhancement may be absent

http://www.amepc.org/qims/article/viewFile/677/732

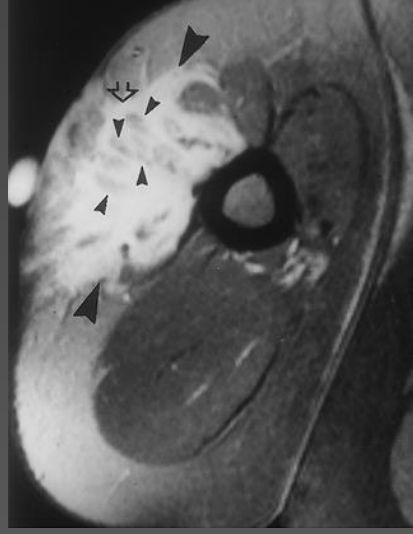
MRI Findings of DTF



- ***Heterogeneous*** soft tissue mass that may extend along fascial plane (displace or invade adjacent soft tissues/muscle)
- Variable signal intensity based on amount of collagen
  - Low to intermediate T1 signal
  - Intermediate to high T2 signal
  - Regions with **low T1/T2** signal suggest **mature collagen**
  - **Bandlike areas of low signal = highly suggestive of diagnosis**
    - More mature lesions have lower recurrence rates



MRI Findings of DTF



http://radiographics.rsna.org/content/21/3/585/F26.expansion

- **Variable enhancement**
  - Usually moderate to marked, but enhancement may be absent
  - More cellular regions avidly enchance
  - Lesions without CT enhancement may still show enhancement on MR
  - Myxoid lesions have least enhancement

Treatment of DTF

- Although standard first-line treatment is still wide-local surgical excision, experience shows that **risk of local relapse = high** even in presence of clear margins *(almost 30%)*

- Radiation therapy as adjunct or primary therapy, but recurrences can occur in 19 to 25% of these cases

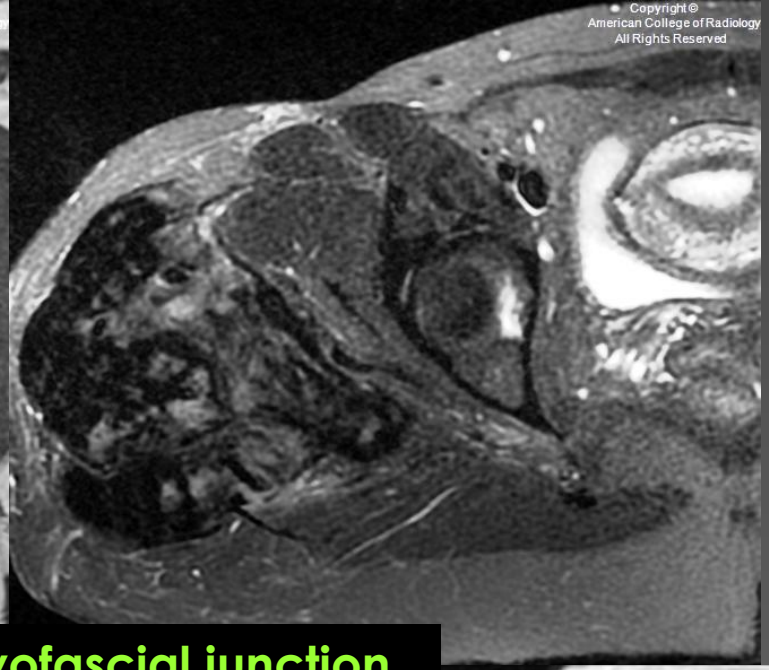
- Other nonsurgical approaches: chemotherapy, hormonal therapy, NSAIDs, radiofrequency ablation, cryoablation

Figure 2. Perioperative and reconstruction of the seventh postoperative day.

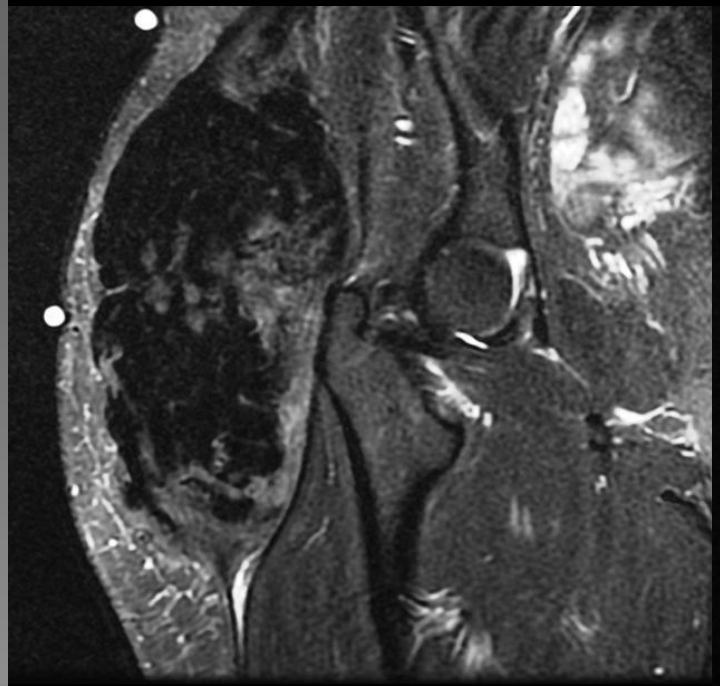
http://www.scielo.br/scielo.php?pid=S2237-93632012000300018&script=sci_arttext

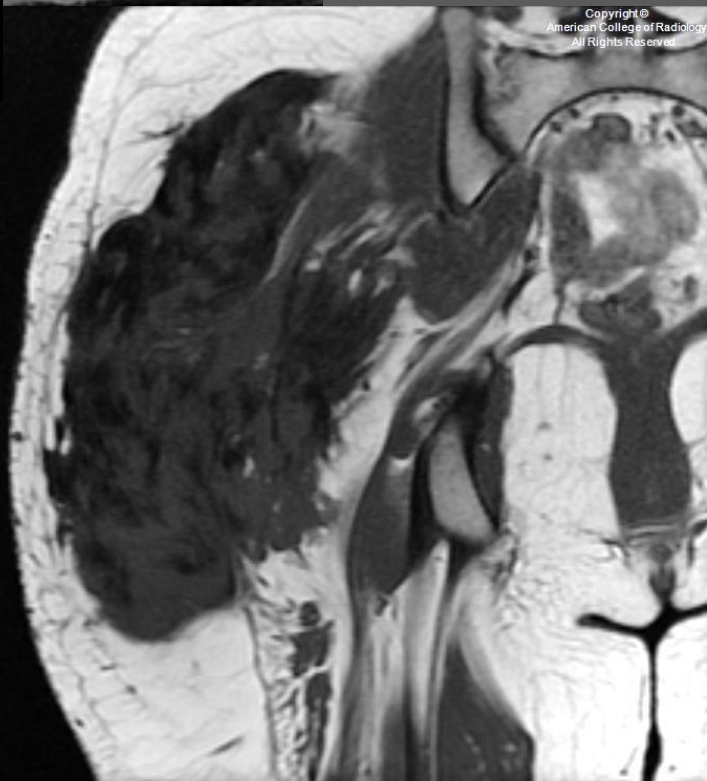
Treatment of DTF

- Due to high risk of relapse despite treatment, conservative management/followup recently proposed as first-line therapy (restricting surgery to patients with symptomatic disease or progression)



*Crosses myofascial junction
*Invades gluteal muscles





65 yr old woman with right lateral thigh mass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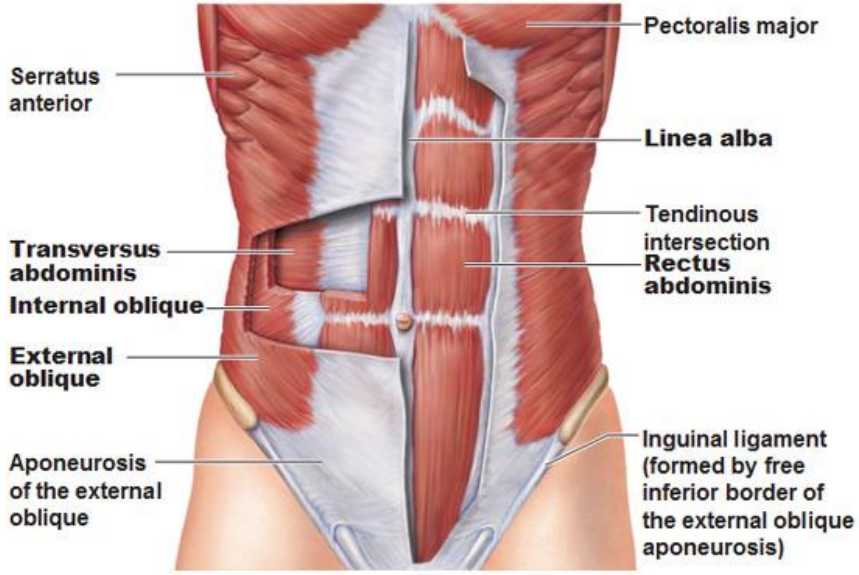
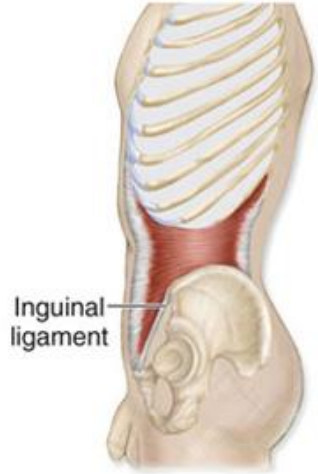
Anatomy (Abdominal Wall)

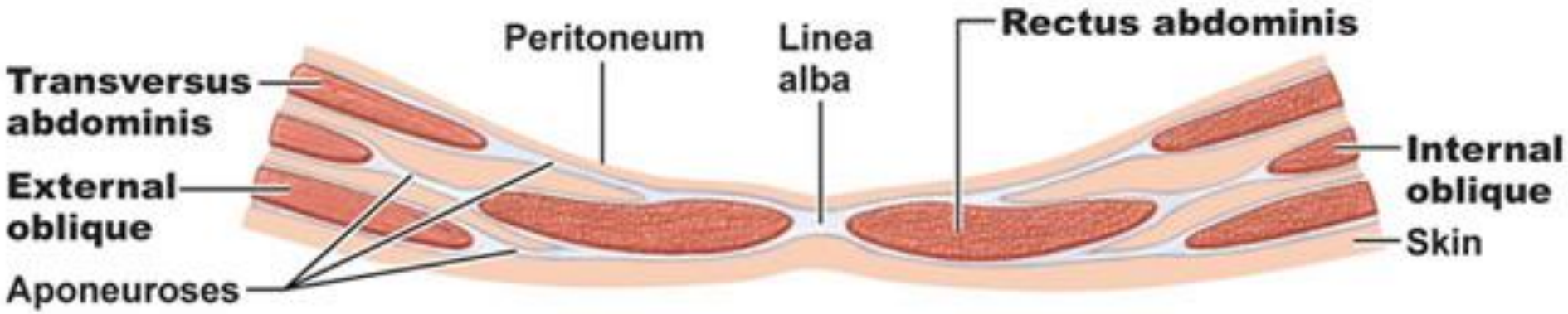



External oblique

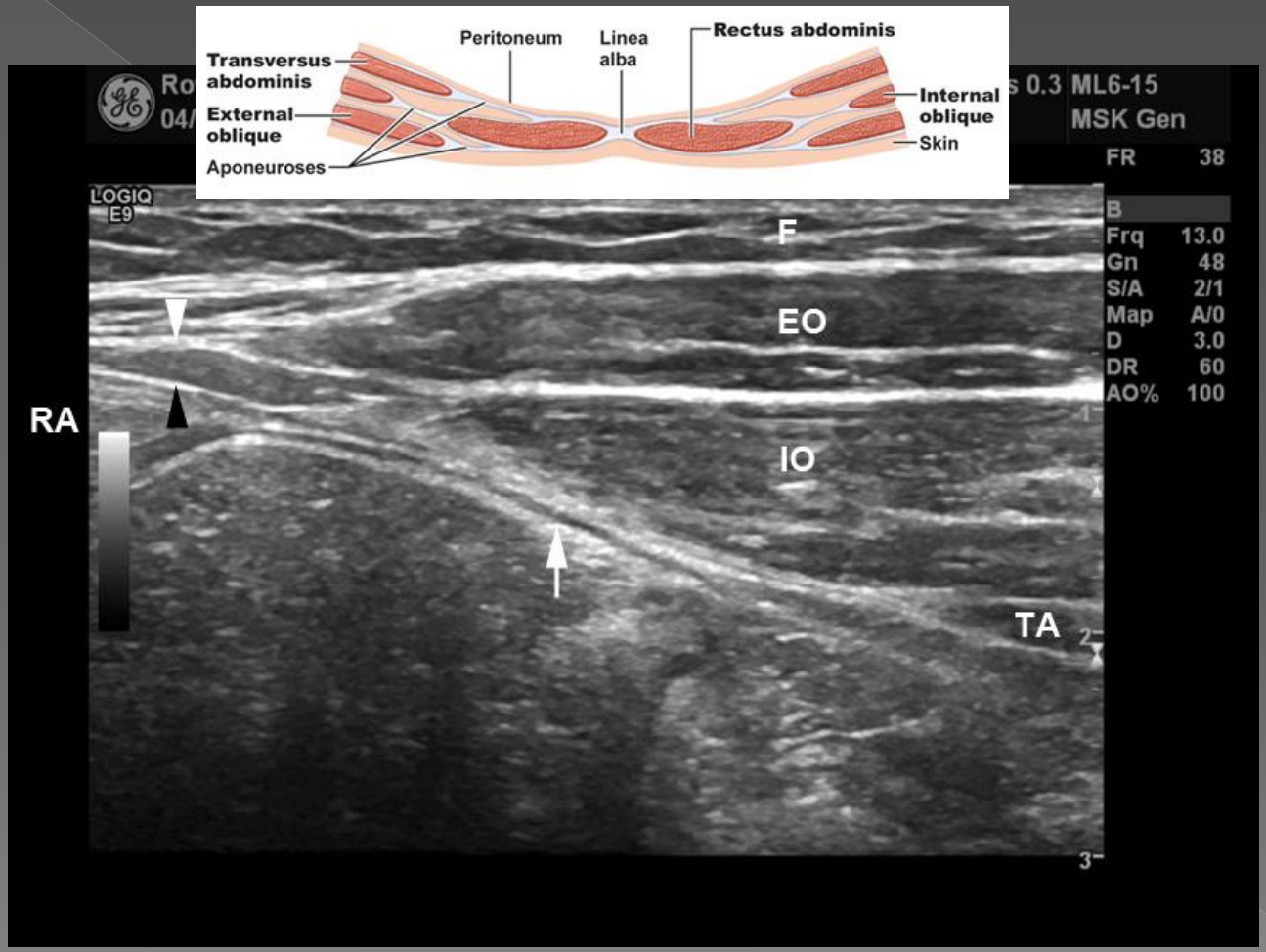

Internal oblique and rectus abdominis



Transversus abdominis

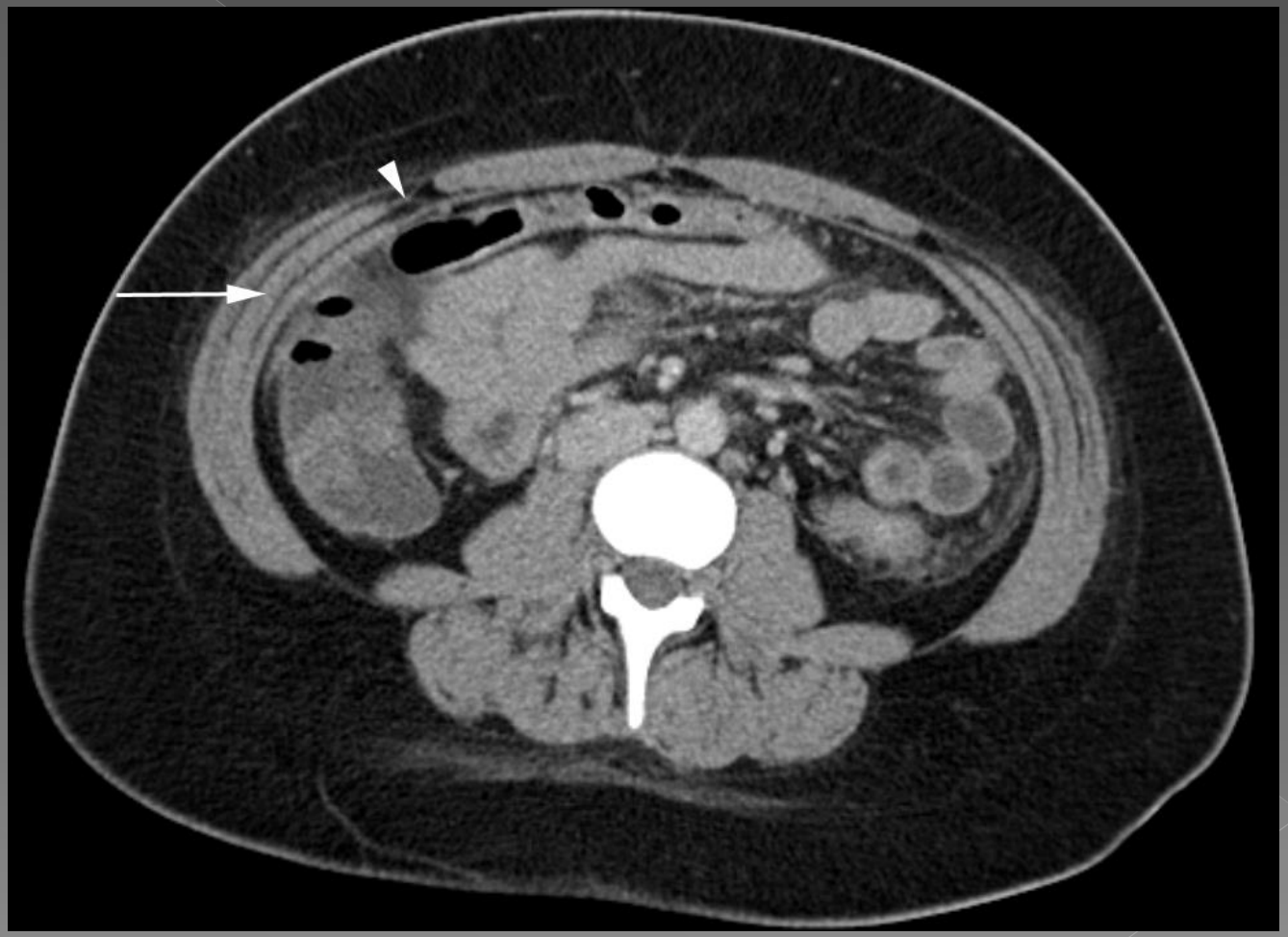


Anatomy (Abdominal Wall)

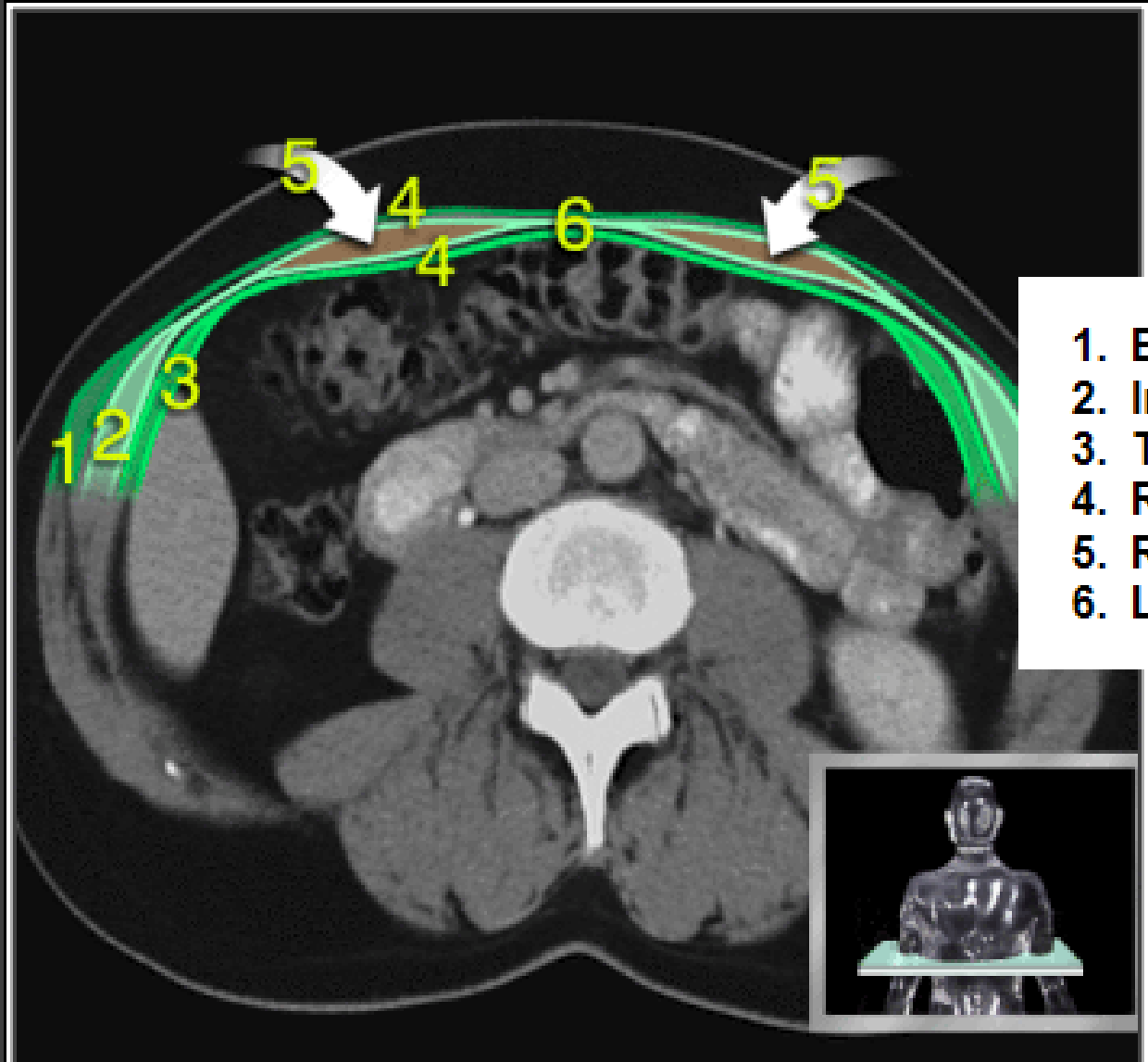


Layers of the abdominal wall as seen on high frequency ultrasound. Fat (F); external oblique muscle (EO); internal oblique muscle (IO); transversus abdominis muscle (TA); rectus abdominis (RA); peritoneum (arrow); aponeurosis of external oblique (white arrowhead) and anterior part of the aponeurosis of internal oblique contributing to the rectus sheath (black arrowhead).

Anatomy (Internal Oblique)



Anatomy (Abdominal Wall)



1. External oblique
2. Internal oblique
3. Transversus abdominis
4. Rectus sheath
5. Rectus abdominis
6. Linea alba

Anatomy (Tranversus Abdominus)

Anatomy (Rectus Sheath)

A midline transverse section US view of the linea alba (arrow) and the rectus abdominis muscles on either side (RA), the anterior (black arrowhead) and posterior (white arrowhead) components of the

Anatomy (Abdominal Wall)

MR anatomy of the abdominal wall demonstrating the three flat muscles (short arrow); the linea semilunaris (open arrow); rectus abdominis (black arrowhead); the linea alba (open arrowhead); the epigastric vessels (long arrow); the quadratus lumborum muscle (black arrow) and the erector spinae

Abdominal Wall Fibromatosis

- **Clinical Features**
- Aka Abdominal wall desmoid
- **Most common soft-tissue neoplasm of abdominal wall**
- Initially described1832 by McFarlane in a young woman after delivery
- Frequency similar to desmoid type fibromatosis

Abdominal Wall Fibromatosis

Figure 1. Preoperative.

http://www.scielo.br/scielo.php?pid=S2237-93632012000300018&script=sci_arttext

- **Clinical Features**
- Lesions distinguished from other deep musculoaponeurotic fibromatoses by ***location*** and ***predilection to develop in women of childbearing age***
- 87% occur in women, 95% develop in women who have had at least 1 child (usually occur during 1st yr after childbirth)
- Peak prevalence 3rd decade

Abdominal Wall Fibromatosis

- **Clinical Features**
- Etiology uncertain
- Majority idiopathic
- Estrogenic hormones, ***trauma*** (including surgery), and genetic abnormalities have also been implicated as potential causative factors

Abdominal Wall Fibromatosis

- **Clinical Features**
- Often history of birth control pill use and more frequent in third trimester
- Tend to regress after menopause or oophorectomy
- ***Antiestrogen agents*** such as tamoxifen have also been shown to have inhibitory effects (findings support role of hormonal factors in development of disease)

Abdominal Wall Fibromatosis

- **Clinical Features**
- Lesions occur after surgery, 20% of cases
- Predilection to develop near areas of postop scarring at prior incision sites, particularly c-section
- Reported at sites of *colostomies, laparoscopic trocar placements, catheter insertions for peritoneal dialysis*

http://pregnancy.about.com/od/cesareansection/ss/c-section-scars_9

Abdominal Wall Fibromatosis

- **Clinical Features**
- Prevalence of desmoid tumors in Familial Adenomatous Polyposis (FAP): 3.6% to 34%
- Pts with FAP have1000-fold increased risk of developing this lesion compared with GEN population
- Pts with phenotypic variant of FAP known as Gardner's syndrome may develop extraabdominal, abdominal wall, and intraabdominal desmoid tumors, in addition to polyposis coli/colon ca, osteomas, sebaceous cysts

Abdominal Wall Fibromatosis

- **Clinical Features**
- Progressive, locally infiltrative, and aggressive behavior
- Typically solitary

http://www.readcube.com/articles/10.1186/1477-7819-9-35

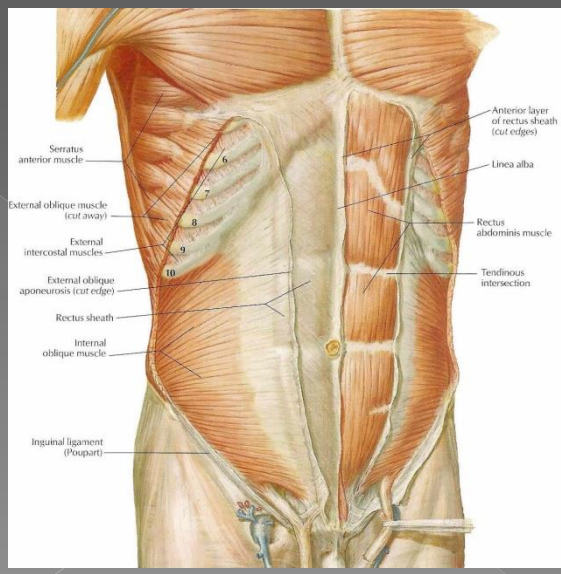
Abdominal Wall Fibromatosis

- **Clinical Features**

- Arise from musculoaponeurotic structures of abdominal wall, ***most frequently rectus abdominis, internal oblique muscles, and fascial coverings***

- Occasionally ***cross midline*** to involve both rectus abdominis muscles

- Rectus and fascial desmoids may be associated with ***intra-abdominal extension***



http://is.muni.cz/th/102436/fsps_m_a2/DIPLOMKA-_2_.pdf

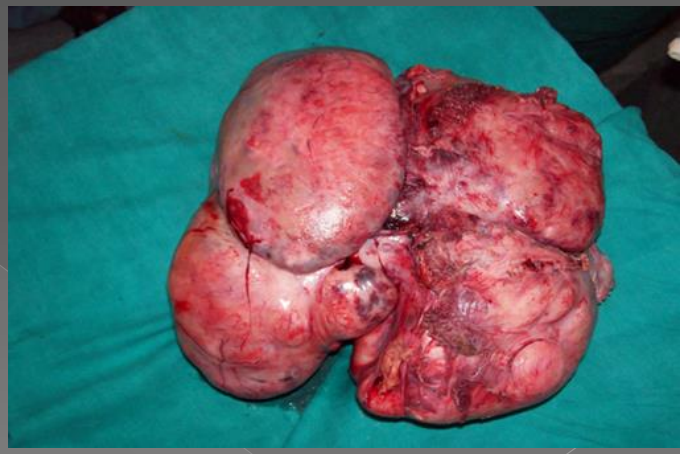
Abdominal Wall Fibromatosis

- **Clinical Features**
- Manifest as palpable, firm, soft-tissue mass
- Deep-seated
- Cause little or no focal symptoms initially (thus manifest late)

http://www.jcasonline.com/article.asp?issn=0974-2077;year=2011;volume=4;issue=2;spage=141;epage=143;aul

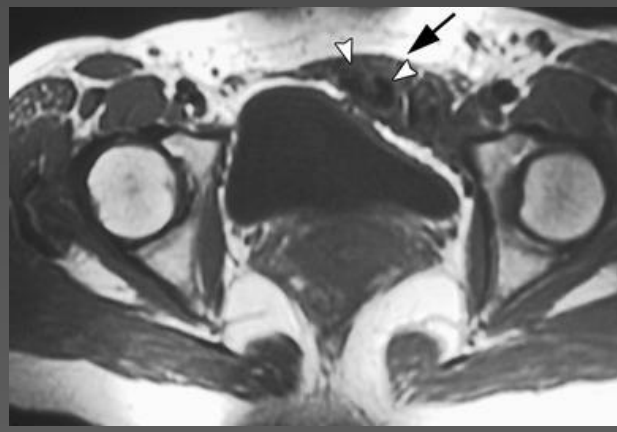
Abdominal Wall Fibromatosis

- **Pathologic Features**
  - Pathologic appearance, both gross & microscopic ***virtually identical to DTF***
  - Solid, firm, whitish masses, often have infiltrative, spiculated margin to skeletal muscle & subcutis (avg size 3–7 cm, often smaller at detection than other DTF)
  - ***Estrogen receptors*** common in abdominal wall desmoids (79% of lesions)



http://archive.ispub.com/journal/the-internet-journal-of-surgery/volume-10-number-2/large-desmoid-tumor-of-the-anterior-abdominal-wall-a-case-report-of-a-4-6kg-desmoid-tumor.article-g01.fs.jpg

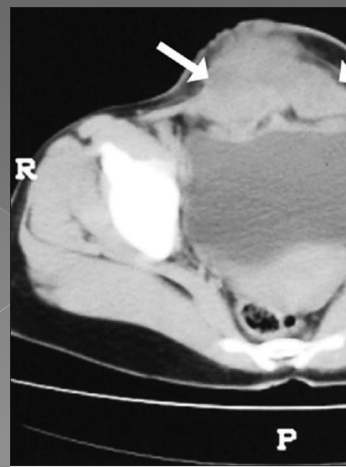
Abdominal Wall Fibromatosis



http://radiology.rsna.org/content/236/1/81/F2.expansion.html

Imaging Features

- Essentially identical to DTF
- Predominantly low to intermediate SI abdominal wall mass with linear extensions ("fascial tail sign") along superficial fascia at margins and **low SI bands** that do not enhance = "nearly pathognomonic"
- MRI optimal for detecting deep intraabdominal extension (unusual) to guide complete surgical resection



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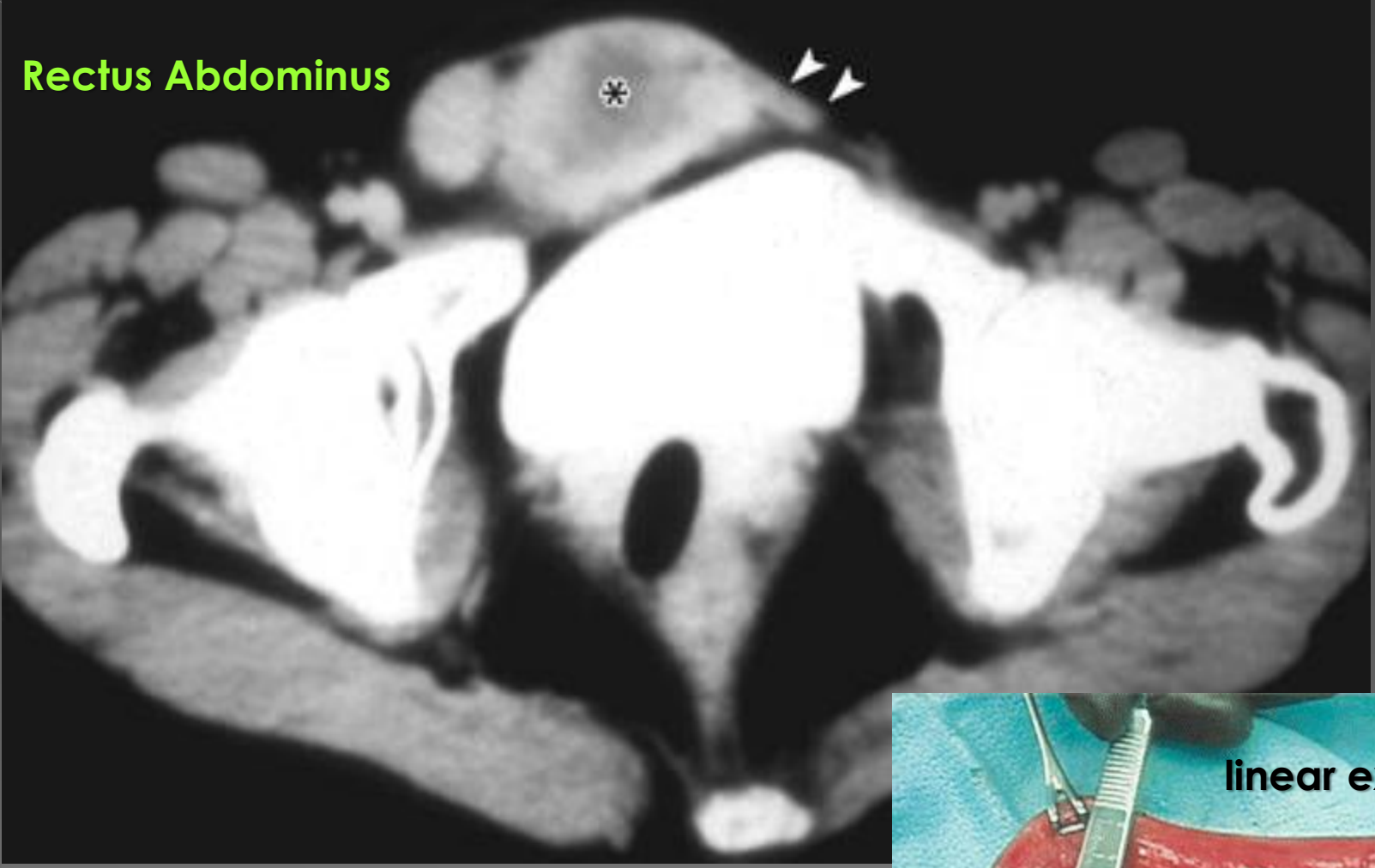
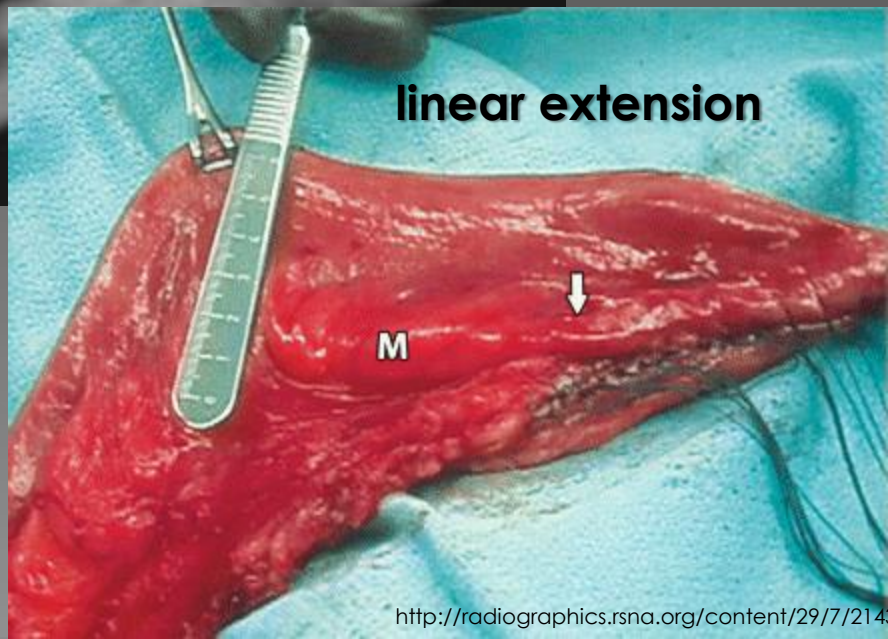
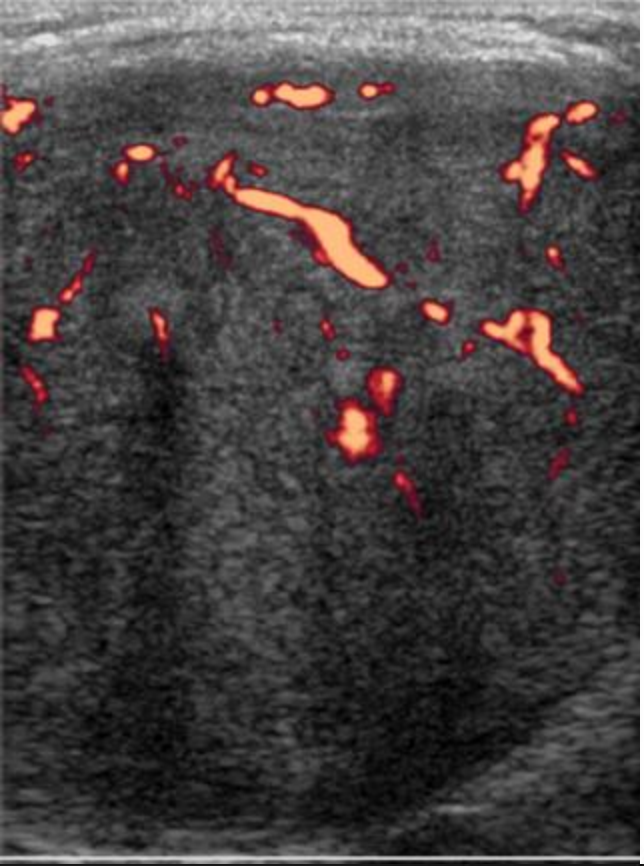
Rectus Abdominus



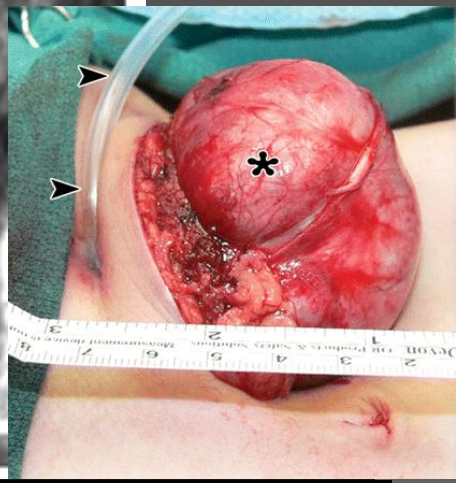


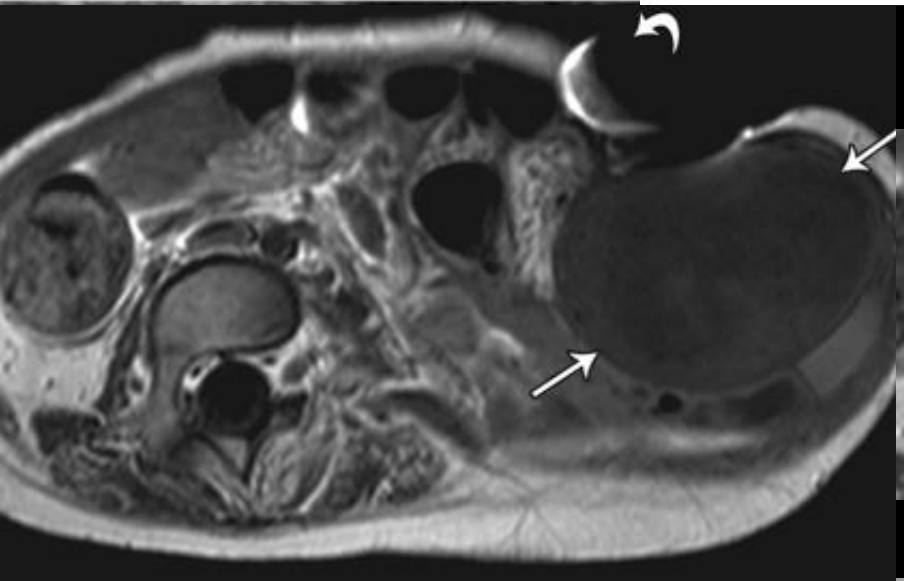
Higher attenuation (relative to adjacent muscle), owing to high collagen content

http://radiographics.rsna.org/content/29/7/214

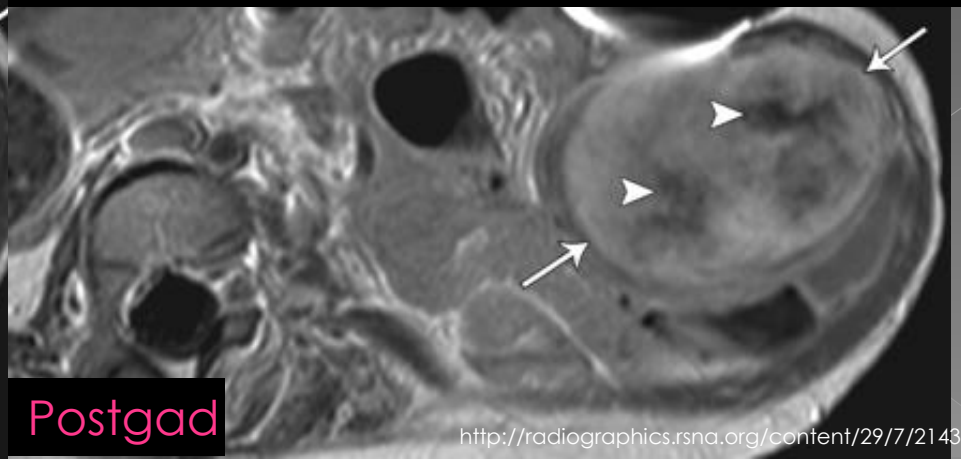


After linsertion of PEG tube





Diffuse enhancement excludes hematoma
Nonenhancing fibrous bands (arrowheads)



Postgad

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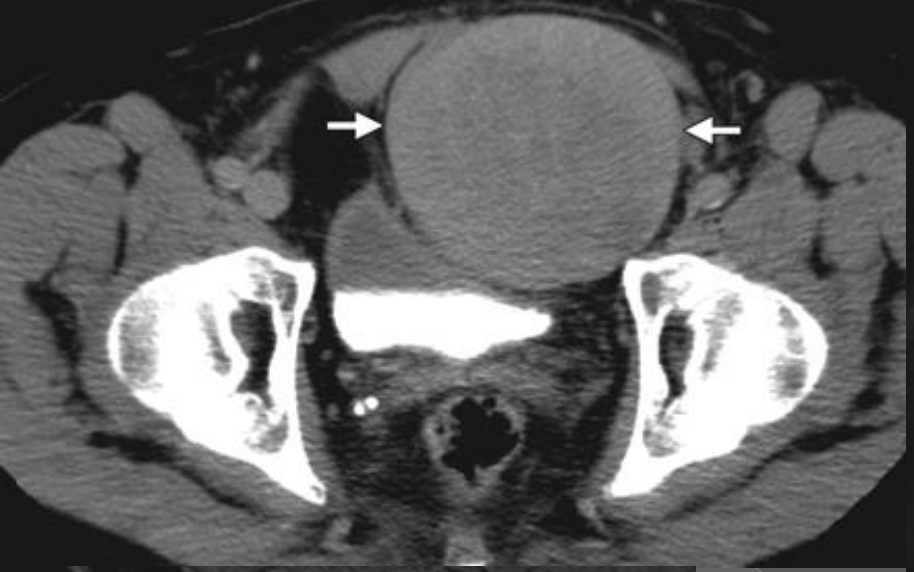

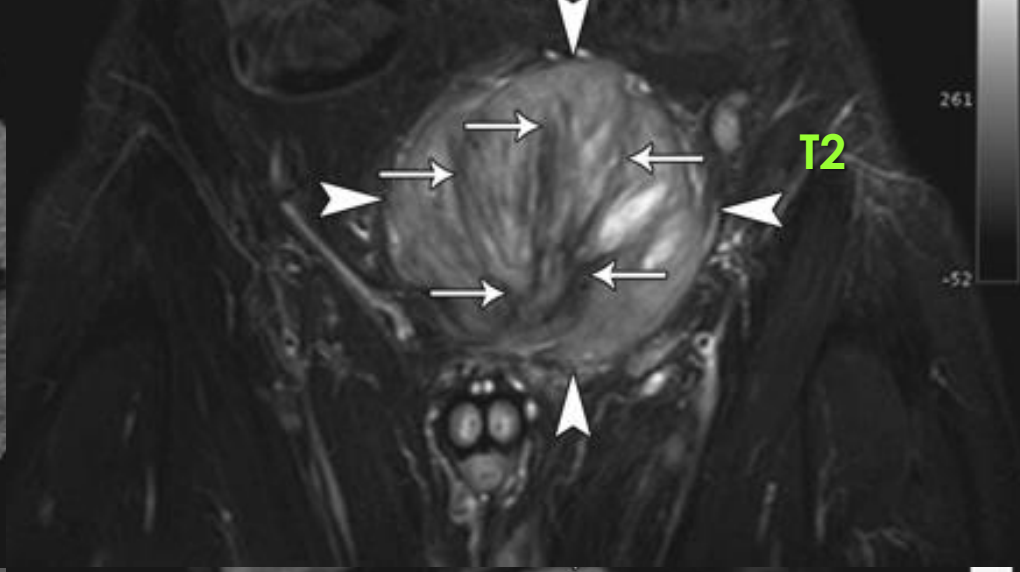

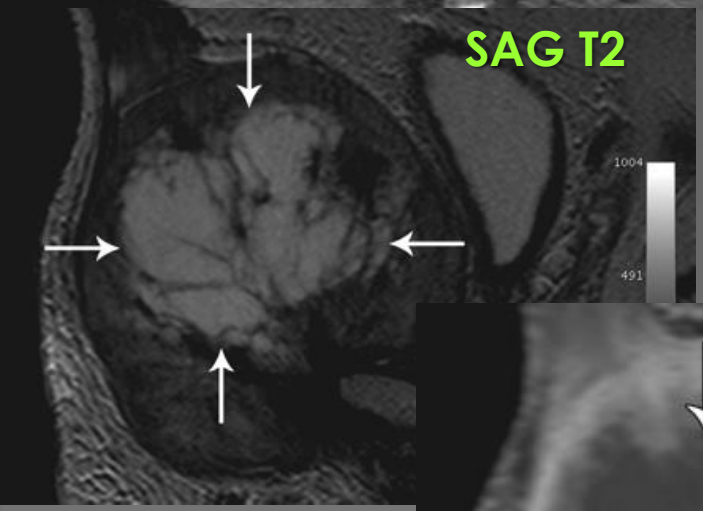

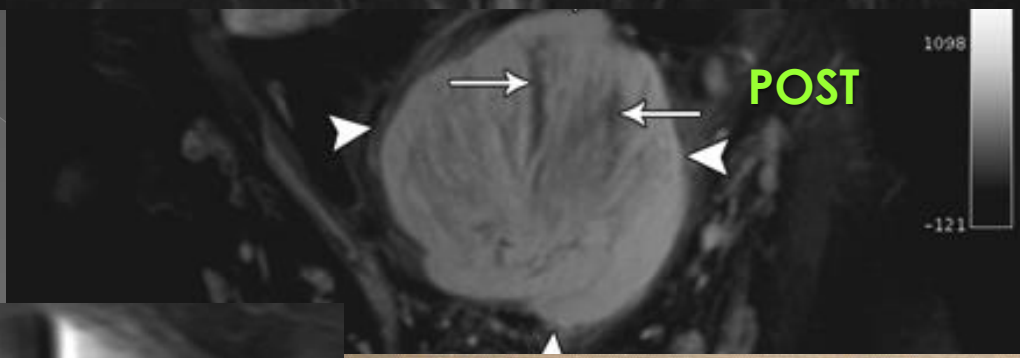

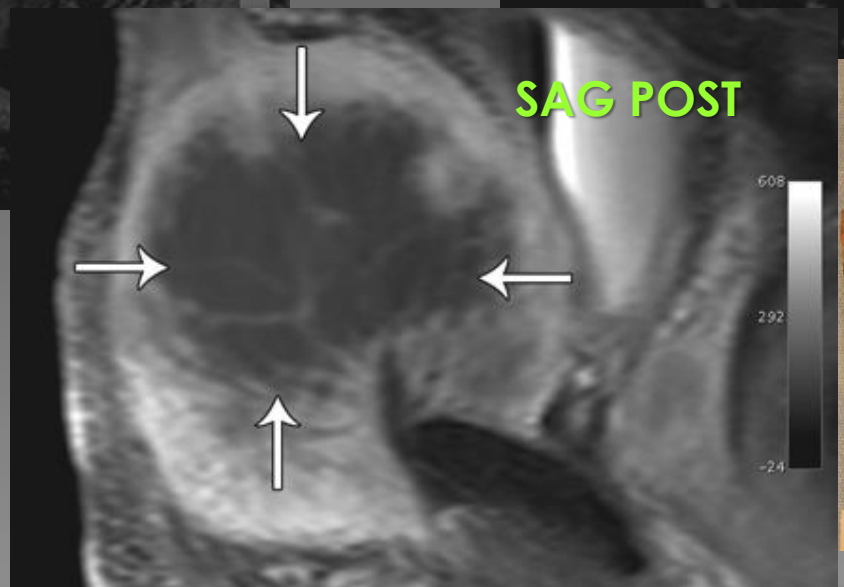

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Abdominal Wall Fibromatosis

- **Treatment & Prognosis**
- Local management of extraabdominal DTF and abdominal wall desmoid tumors **similar**, with attempted **wide-margin resection** being initial treatment of choice
- *Propensity to recur locally* (within first 2 yrs after excision or in connection with subsequent pregnancies or deliveries)
- Local recurrence rate 15%–30% (lower than extraabdominal lesions)
- Reports of regression with menopause

Abdominal Wall Fibromatosis

- **Treatment & Prognosis**
- Preoperative adjuvant radiation therapy may be required
  - Radiation may cause central necrosis
- Regimens of high-dose antiestrogen and progesterone agents (such as tamoxifen and raloxifene), luteinizing-releasing hormone, and testosterone have also been used with some success

Summary

- **Superficial MSK Fibromatoses**
  - Palmar Fibromatosis
  - Knuckle Pad Fibromatosis
  - Plantar Fibromatosis
- **Deep MSK Fibromatoses**
  - Desmoid Type Fibromatosis
  - Abdominal Wall Fibromatosis

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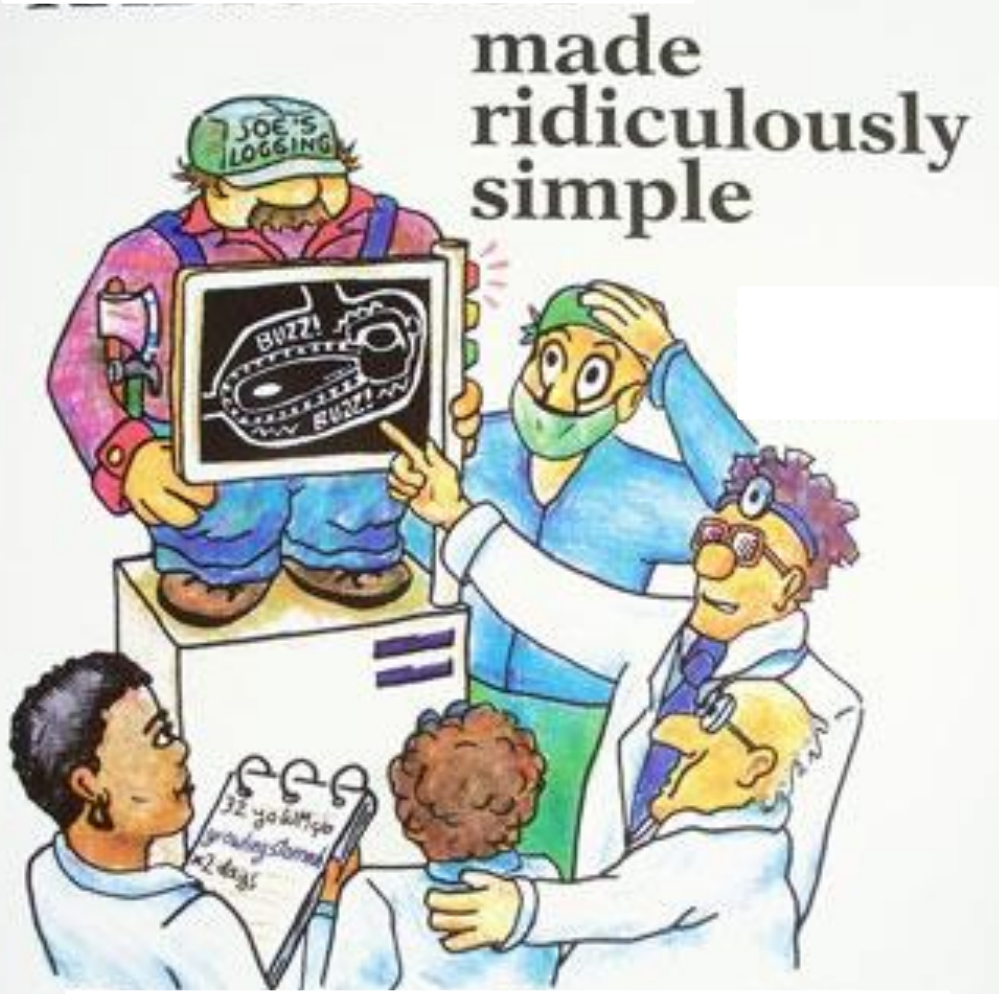

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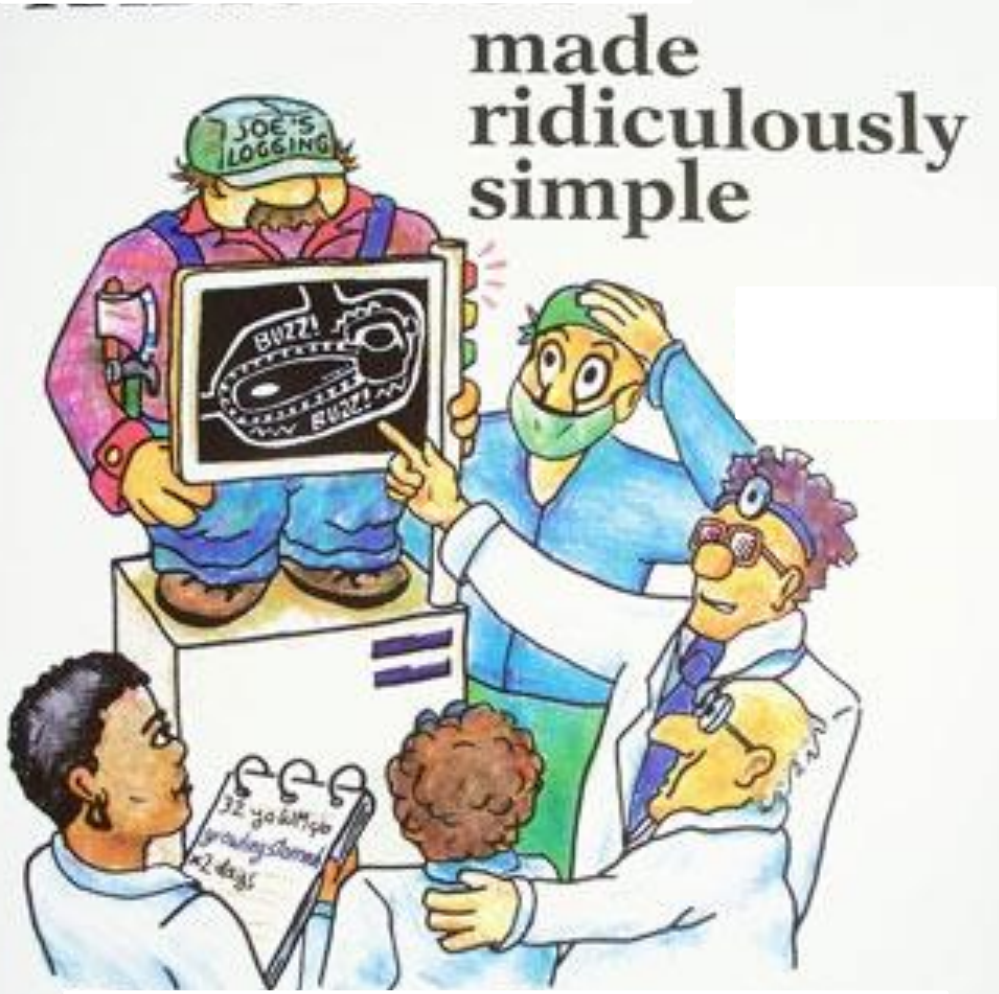

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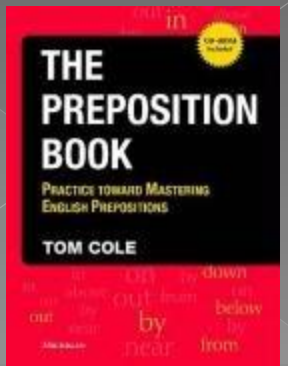


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