Idiopathic inflammatory myositis: imaging

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Myositis: MRI

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Map

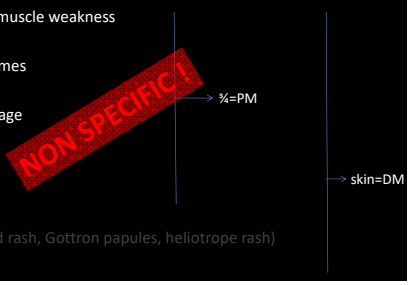
- Definition
- Diagnosis
- Imaging
  - When ?
  - Why ?
  - How ?
  - Where ?
- Take home

Definitions

- Heterogeneous group
- Rare: 10/100000, but high morbidity, mortality
- Systemic connective tissue diseases
- Hallmark: muscle inflammation
- Extramuscular manifestations
- Multiparameter based diagnosis

Diagnosis (Bohan & Peter diagnostic criteria, NEJM 1975)

- 1 Symmetrical proximal muscle weakness
  - Clinics
- 2 Increased muscle enzymes
  - Biology
- 3 Myogenic Muscle damage
  - EMG
- 4 Muscle biopsy
  - Histology
- 5 Skin
  - Dermatology (eyelid rash, Gottron papules, heliotrope rash)



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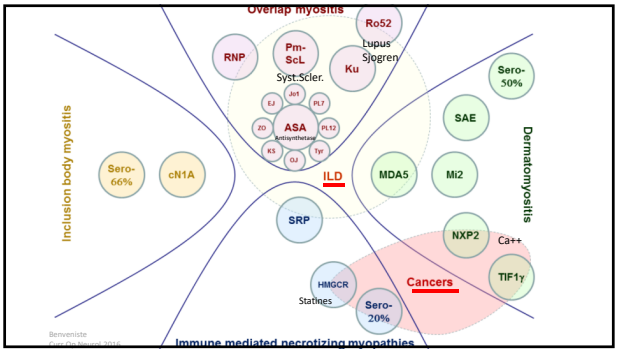
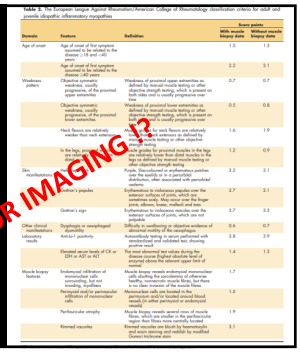
- **Myositis Specific or Associated Auto-antibodies : DM, IBM, IMNM, OVERLAP M,….**



Lunberg I, et al. 2017 **EULAR-ACR Classification Criteria** for Adult and Juvenile IIM and Their Major Subgroups. ARD 2016

NO PLACE FOR IMAGING !?



Aggarwal R, et al. 2016
ACR – EULAR Criteria for Minimal, Moderate and Major Clinical Response for Adult Dermatomyositis and Polymyositis: An International Myositis Assessment and Clinical Studies Group /Paediatric Rheumatology International Trials Organisation Collaborative Initiative.
ARD 2016

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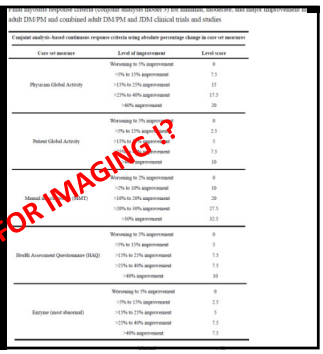
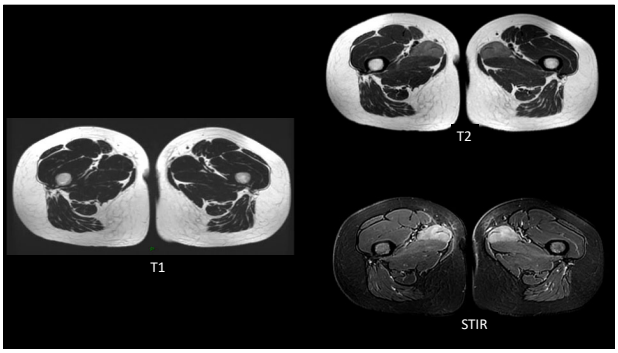


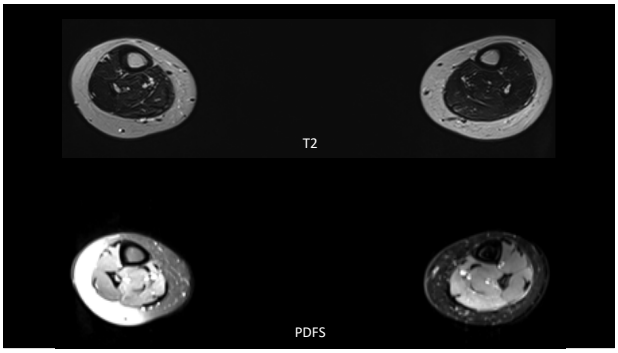
When and why : indications & roles

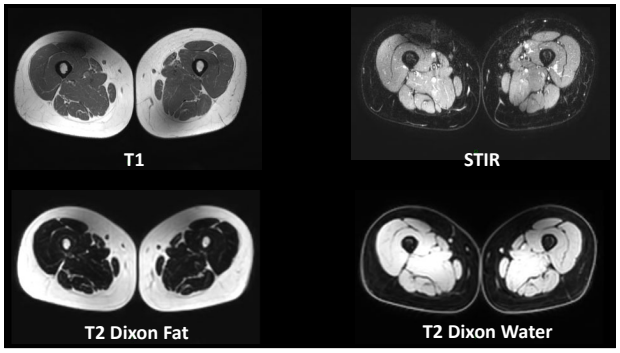
- Suspected (especially if AB-serology is negative)
  - Rule in (Limited specificity !)
  - Rule out (Limited sensitivity !)
  - Phenotype
  - Guide biopsy
- Once diagnosed
  - Severity
  - Related disorders
  - Complications
  - Response evaluation

How : sequences (basic)

- T1 and T2
  - Historical, lack sensitivity
  - Evolution
- … « super » T2 : mandatory
  - STIR
  - T2 FS
  - Dixon T2

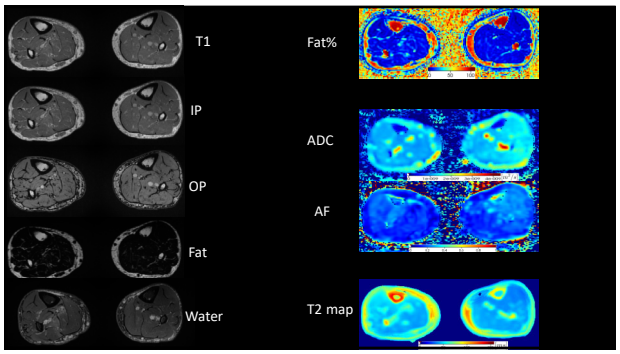






How : sequences (advanced)

- DWI ?
  - Morpho
  - ADC
- DTI ?
- Fat fraction?...

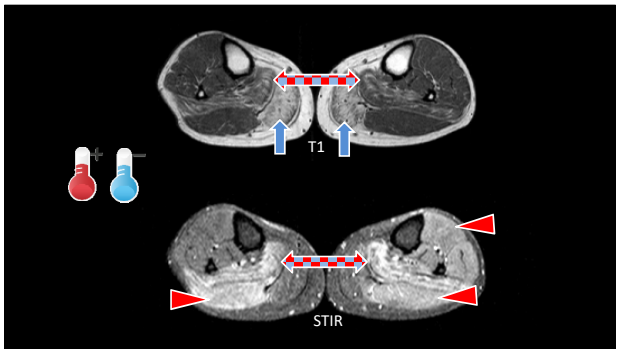


Elementary lesions

- Muscle signal
  - Edema = inflammation
  - Fat = chronic, quiescent, endstage
- Muscle trophicity
  - Swelling
  - Atrophy

Reimers CD,et al. MRI of skeletal muscles in IIM. Neurol. 1994 Mar;241(5):306-14.
In acute myositis, oedema-like abnormalities were visible on T2
In chronic diseases, muscle lipomatosis is seen on T1

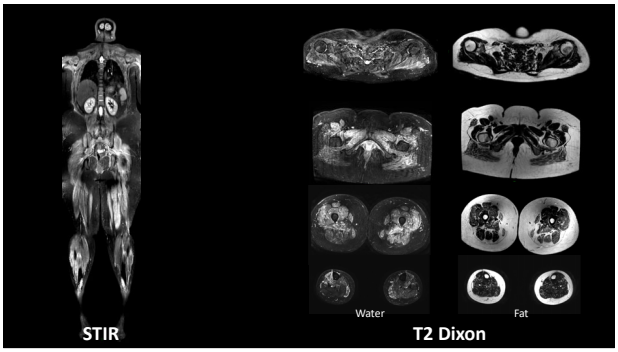
Schedel H, Reimers CD, et al. Muscle edema in MRI of neuromuscular diseases. ActaRadiol. 1995
Contrast-enhanced T1-weighted images were not more sensitive than T2 images.

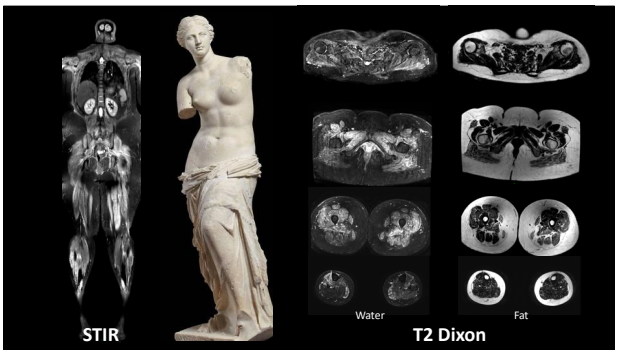


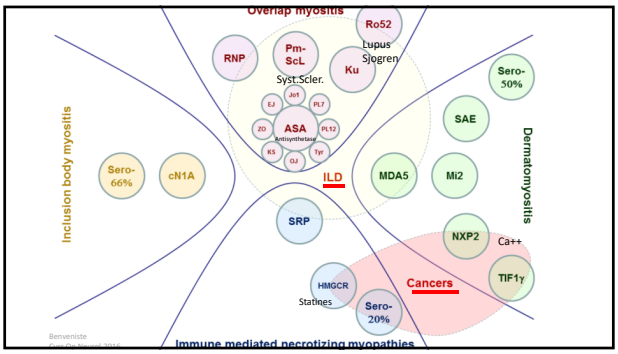
Where ? (anatomy and planes)

- Regional (limited) : girdles (scapular, pelvic), thighs, calves
  - Transverse
  - Large field of view
- Global (Total body)
  - Usefulness: trunk ?
  - Coronal or transverse (minimum)

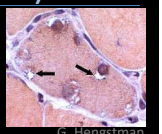
Filli L, et al. WB-MRI in adult inflammatory myopathies: Do we need imaging of the trunk? Eur Radiol 2015
Huang ZG, et al. An efficacy analysis of WB-MRI in the diagnosis and follow-up of PM and DM. PLoS One 2017.



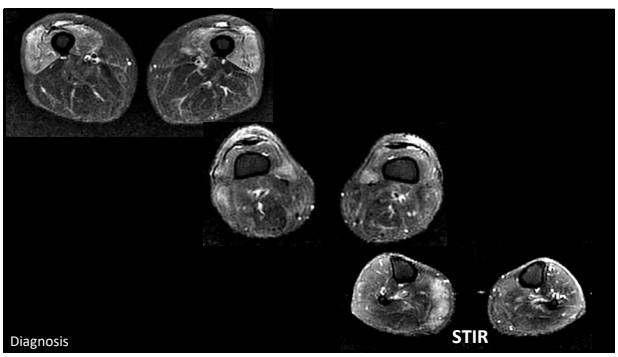


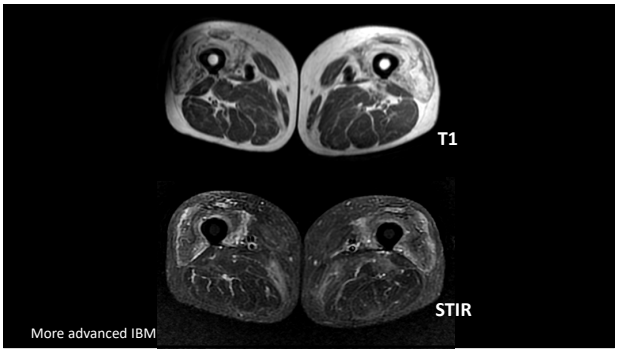


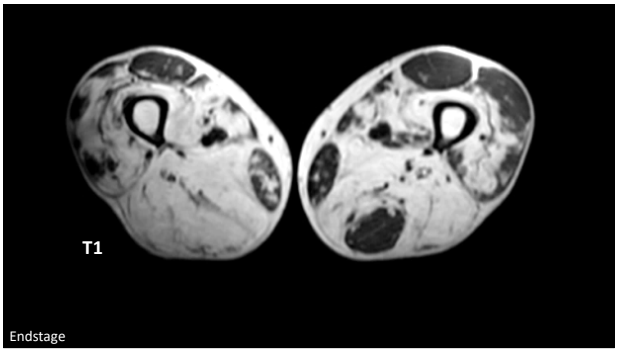
Inclusion Body Myositis (IBM)


G. Hengstman

- > 50 y
- M > F
- 15-30 % IIM
- Insidious progressive proximal and distal weakness
- Marked atrophy quadriceps and forearm muscles (wrist & fingers flexors)
- Poor response to therapy and prognosis
- Dysphagia frequent (30-50%)
- No sytemic sign of inflammation (CRP)
- No association with lung, cardiac disorders or cancer

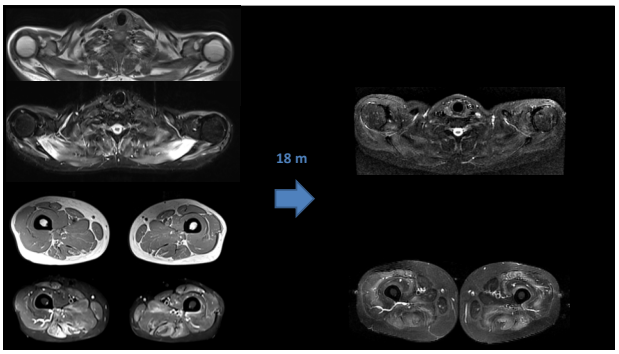






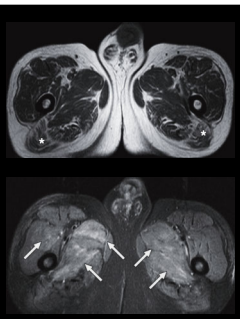
Immune mediated necrotizing myositis (IMNM)

- 15 % IIM
- Profound proximal weakness in weeks or months
- Very high CK
- More widespread edema, atrophy, fatty transfo >> PM & DM
- Statin exposure with Anti-HMGCR
- Other Anti-SRP (more severe muscle lesions)



Statins

- self-limited myopathy, up to 5 % of patients
- HMGCR AB-associated IMNM : proximal muscle weakness, higher CK, severe diffuse edema, muscle necrosis. Even years after statin exposure. Treatment : aggressive immunotherapy (including IVIG)

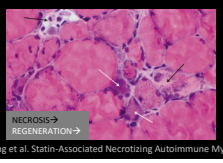


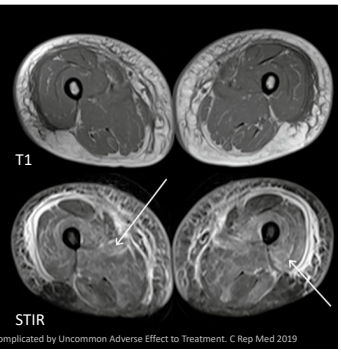
Peters SA, et al. MRI in Lipid-Lowering Agent–Associated Myopathy: A Retrospective Review of 21 Cases. AJR 2010

53-y old woman
Type 2 diabetic
Hyperlipemia
High dose statins

Weakness lower and later upper limbs

CK 28,000 U/L
HMGCR AB +

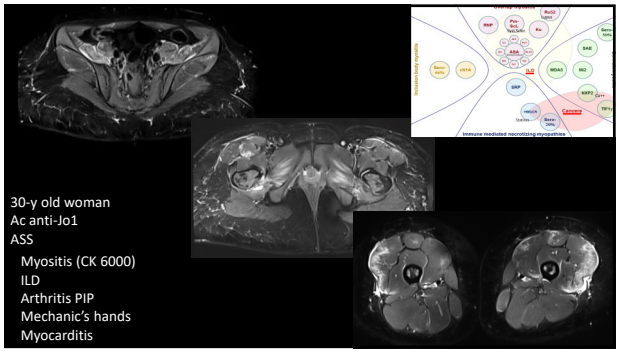




Fung et al. Statin-Associated Necrotizing Autoimmune Myositis Complicated by Uncommon Adverse Effect to Treatment. C Rep Med 2019

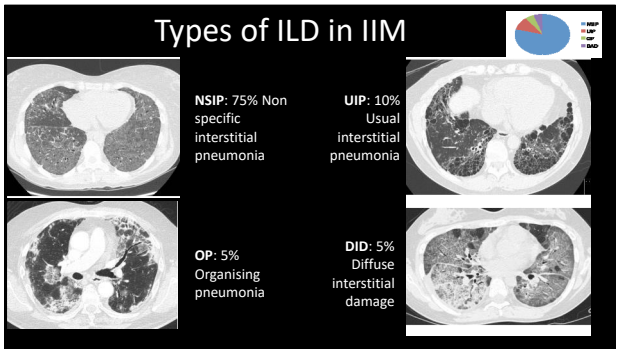
Antisynthetase Syndromes (ASS)

- Constellation of clinical features:
  - Myositis
  - ILD
  - Inflammatory arthritis
  - Fever
  - Raynaud
  - Mechanic's hands
- Anti-Jo1 (most common ASS), higher risk of myositis
- Other than Jo1: often predominant ILD



30-y old woman
Ac anti-Jo1
ASS
Myositis (CK 6000)
ILD
Arthritis PIP
Mechanic's hands
Myocarditis

Types of ILD in IIM



NSIP: 75% Non specific interstitial pneumonia

UIP: 10% Usual interstitial pneumonia

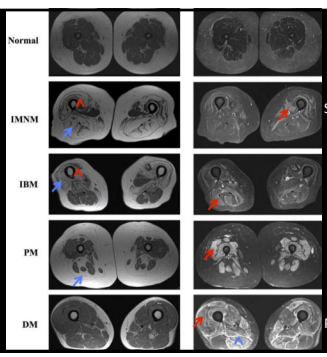
OP: 5% Organising pneumonia

DID: 5% Diffuse interstitial damage

Roles? (reporting)

- Positive or not ?
- Active or not ?
  - Edema, swelling
  - Fat, atrophy
- Cartography (phenotyping)
- Guide a biopsy
  - Active
  - Accessible

Phenotyping



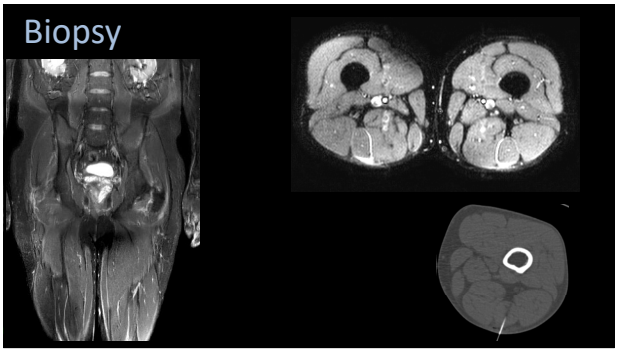
Pinal-Fernandez, et al.
Thigh muscle MRI in immune-mediated necrotising myopathy: extensive oedema, early muscle damage and role of anti-SRP autoantibodies as a marker of severity
ARD 2017
(666 subjects (101 IMNM, 176 PM, 219 DM, 17 CADM, 153 IBM) (John sHopkins)

Table 1. MRI patterns in myositis subtypes.

Specific MRI patterns	Oedema	Atrophy, fatty replacement	Fascial oedema
PM	· Symmetrical · Diffuse, homogeneous oedema · Both anterior and posterior muscle groups (18) (58)	· More prevalent in anterior compartment (58)	· Fascial oedema of rectus femoris uncommon (14)
ASS	· Predominantly anterior compartment (13)	· Predominantly posterior compartment (13)	· Equal in all compartments (13)
Overlap myositis	· Predilection in the gluteal and thigh muscles (59)	· Not known	· Not known
IMNM	· Mostly in the lateral rotators, glutei, medial and posterior compartment; frequent oedema of adductor brevis (14) · Anti-SRP: most affected vastus lateralis with relative sparing of vastus intermedius, rectus femoris, biceps femoris, adductor magnus (60) · Anti-HMGCR: more symmetrical involvement than anti-SRP (14)	· Frequent atrophy of obturator externus (14) · Anti-SRP: predominant affection of adductor magnus, gluteus maximus, biceps femoris long head, semimembranosus and semitendinosus; the least affected m. quadriceps femoris (60)	· Infrequent (14)
DM	· Symmetrical · Focal and patchy oedema · Frequent involvement of quadriceps femoris (6)	· Milder than PM (58)	· More prevalent in the anterior (esp. rectus femoris), medial compartment and surrounding m. semimembranosus (14)
JDM	· Symmetrical · No differences between proximal and distal muscles (19)	· Atrophy of quadriceps, especially vastus medialis (61)	· On WB-MRI fascial oedema limited to the limbs (19)
IBM	· Distal predominance · Anterior group involvement (14), esp. quadriceps muscle (18) and sartorius (25)	· Predominantly forearm and anterior compartment of the thigh · Relative sparing of rectus femoris compared to other quadriceps muscles (22, 23) · Medial gastrocnemius more affected in the lower leg (22-25)	· No oedema, but "undulating fascia" sign – fascia drawing a line between atrophic vastus intermedius and vastus lateralis (18)

Kubínová et al. The Role of Imaging in Evaluating Patients With Idiopathic Inflammatory Myopathies. Clin Exp Rheumatol 2018

Biopsy



Imaging beyond muscles

Guided by Auto-Antibody Profile

- Lung (CT)
- GI tract (dysphagia…)
- Cancer screening
  - WB-MRI
  - PET/CT

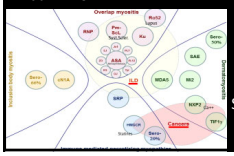
Huang ZG. An efficacy analysis of WB-MRI in the diagnosis and follow-up of PM and DM. PLoS One 2017

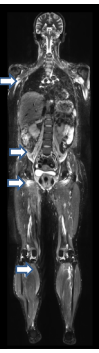
39 y-old female
Proximal weakness
Typical skin lesions

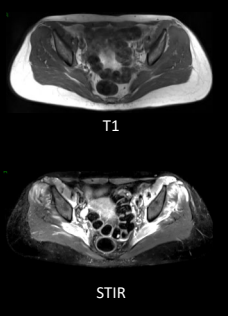
Anti TIF1 gamma
No biopsy

→ DM

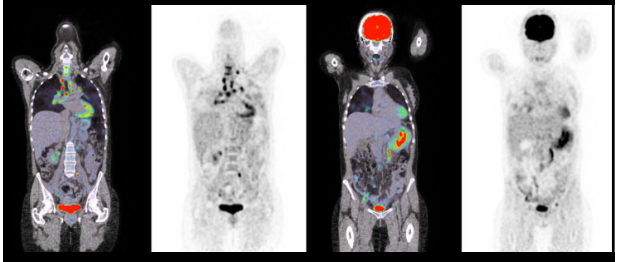
Initial PET CT -

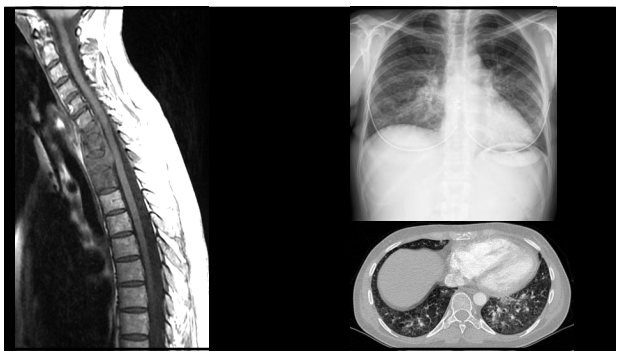






1 year follow-up PET/CT

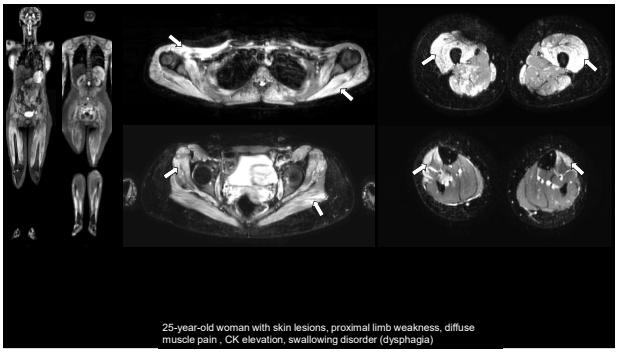


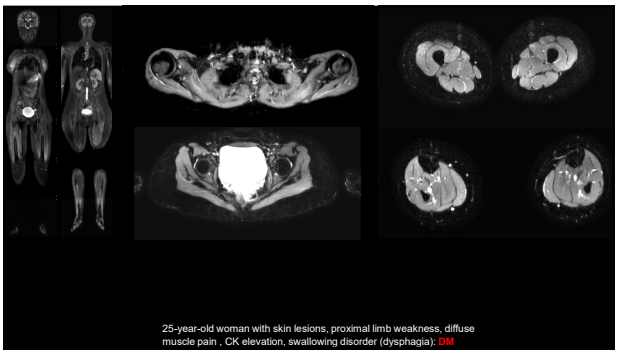


Imaging beyond diagnosis

- Response ?
  - Limitations of biology
  - Qualitative, Semi-quantitative (edema, fat), Quantitative …
- Complications ?
  - Vertebral compression fractures
  - Epiphyseal osteonecrosis (10 %)

Huang ZG. An efficacy analysis of WB-MRI in the diagnosis and follow-up of PM and DM. PLoS One 2017.





Huang ZG, et al. An efficacy analysis of whole-body magnetic resonance imaging in the diagnosis and follow-up of polymyositis and dermatomyositis. PLoS One 2017.

Idiopathic Inflammatory Myositis

- Complex spectrum of diseases
- Clinico-serological diagnosis; autoimmunity biomarkers are key
- MRI may help
  - Positive / negative
  - Phenotype
  - Biopsy
  - Response, complications
- Think associated diseases (WB-MRI?)
- Stay humble and be careful !