Lipomatous tumours of the extremity

Dr. Paul Clarkson

BC Cancer Agency

DISCLOSURES

- NIL

Overview

- Introduction to lipomatous tumours

Lipoma Types	ALT - Dediff	Myxoid-Round Cell	Pleomorphic
Lipoma	Atypical Lipomatous Tumours	Myxoid Liposarcoma	Pleomorphic Liposarcoma
Lipoblastoma	De-differentiated Liposarcomas	Round Cell Liposarcoma	
Angiolipoma			
Myolipoma ...			

Lipoma Types	ALT - Dediff	Myxoid-Round Cell	Pleomorphic
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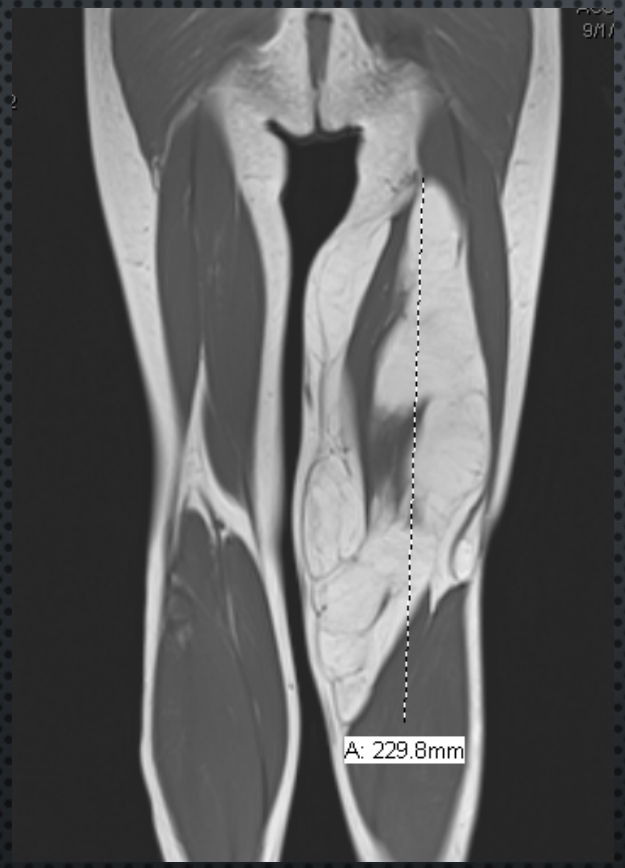
Benign lipomatous lesions

- Number of diagnoses.
- Simple Lipoma/Spindle cell lipoma
- Always benign
- Never under go malignant transformation

Lipoblastoma

- Infancy/early childhood (90% <3 years)
- Benign, no chance of transformation
- Infiltrative, high local recurrence rate
- If recurs – Re-excision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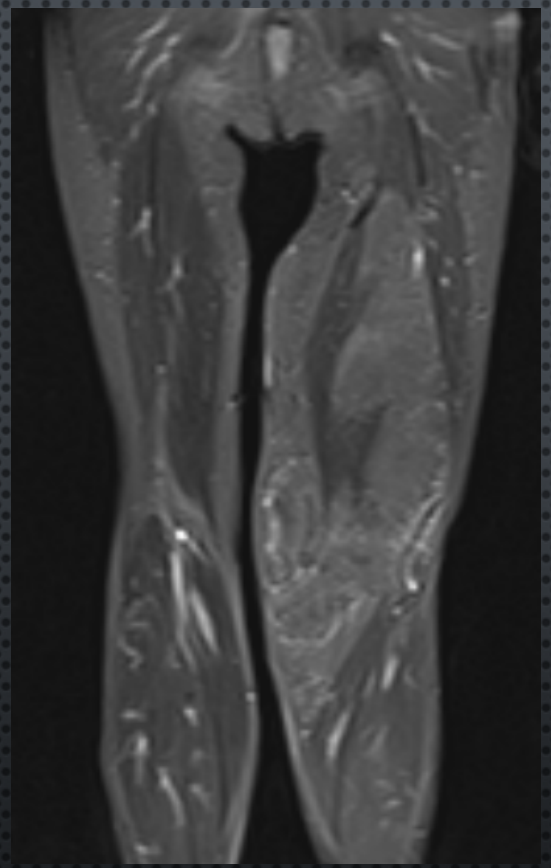

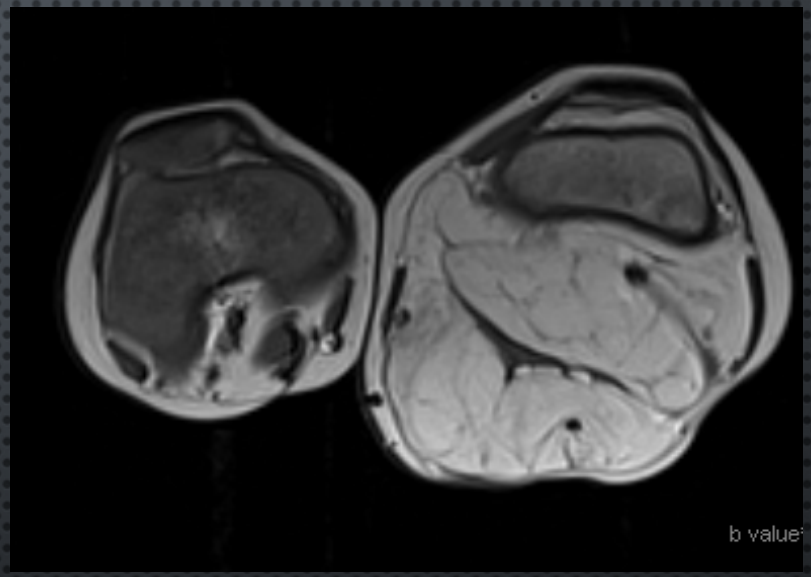
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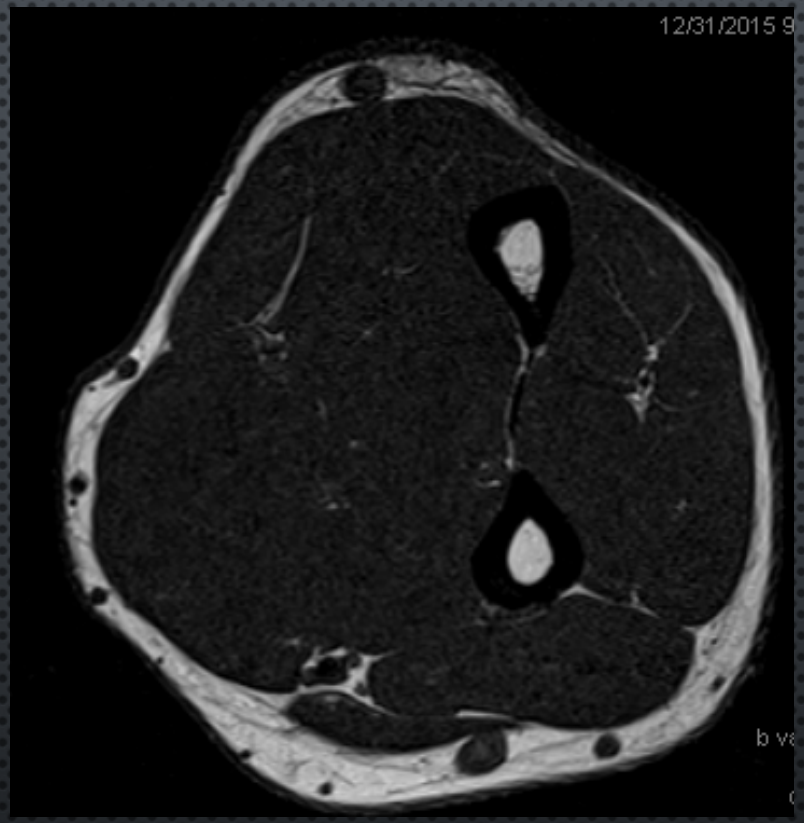
Angiolipomas

- Benign
- Often multiple
- Never transform
- Often have some increased signal on MRI
- Usually small and excisable on clinical grounds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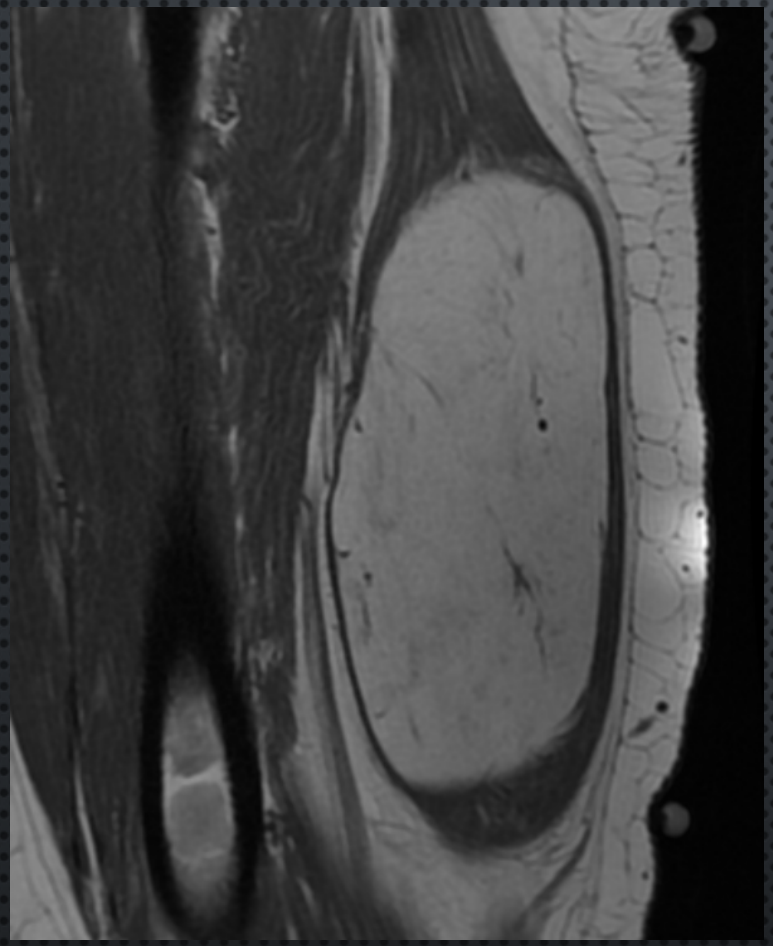
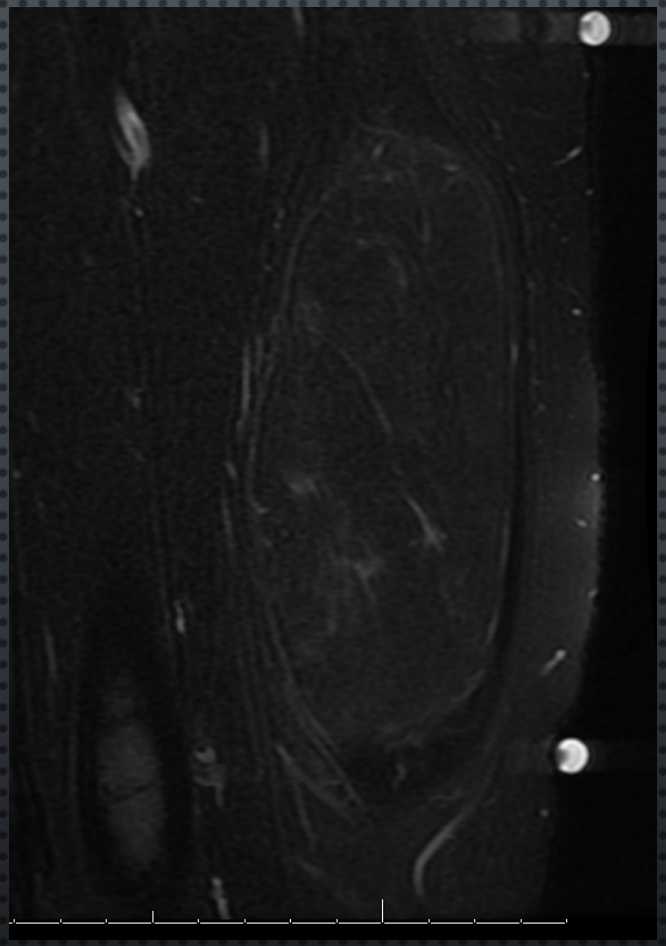
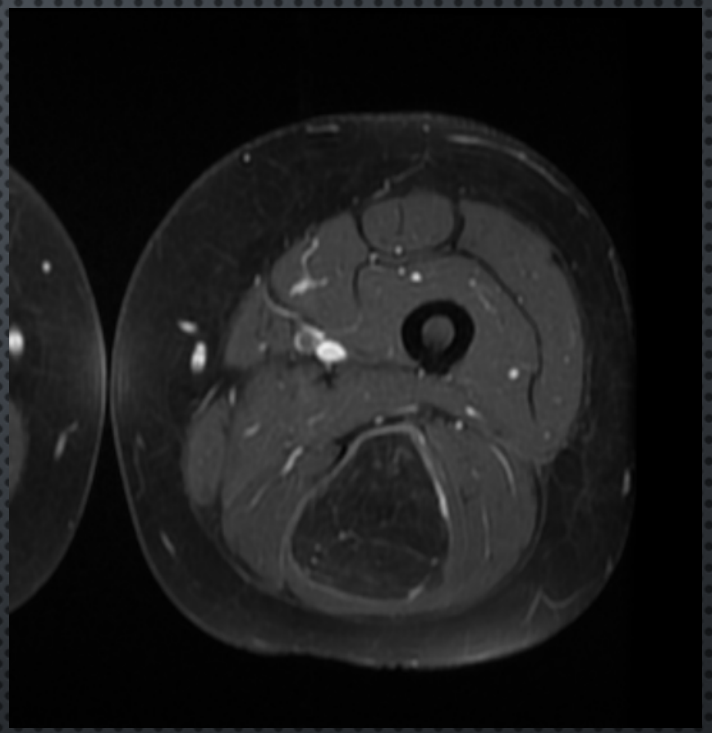
HIBERNOMA

- Occurs in 30-50 years
- Sites of brown fat, and essentially a “lipoma of brown fat”
- Benign, no chance of transformation
- High local recurrence rate
- Treatment is excision







Myxoid - round cell liposarcoma

- The same tumour – "round cell" is now obsolete
- Specific translocation (FUS-DDIT3 or EWSR1-DDIT3)
- 20% of all liposarcomas
- Children/young adults – peak 20s/30s
- >60% in thigh, rare in RP/subcutaneous

MLPS - CLINICAL

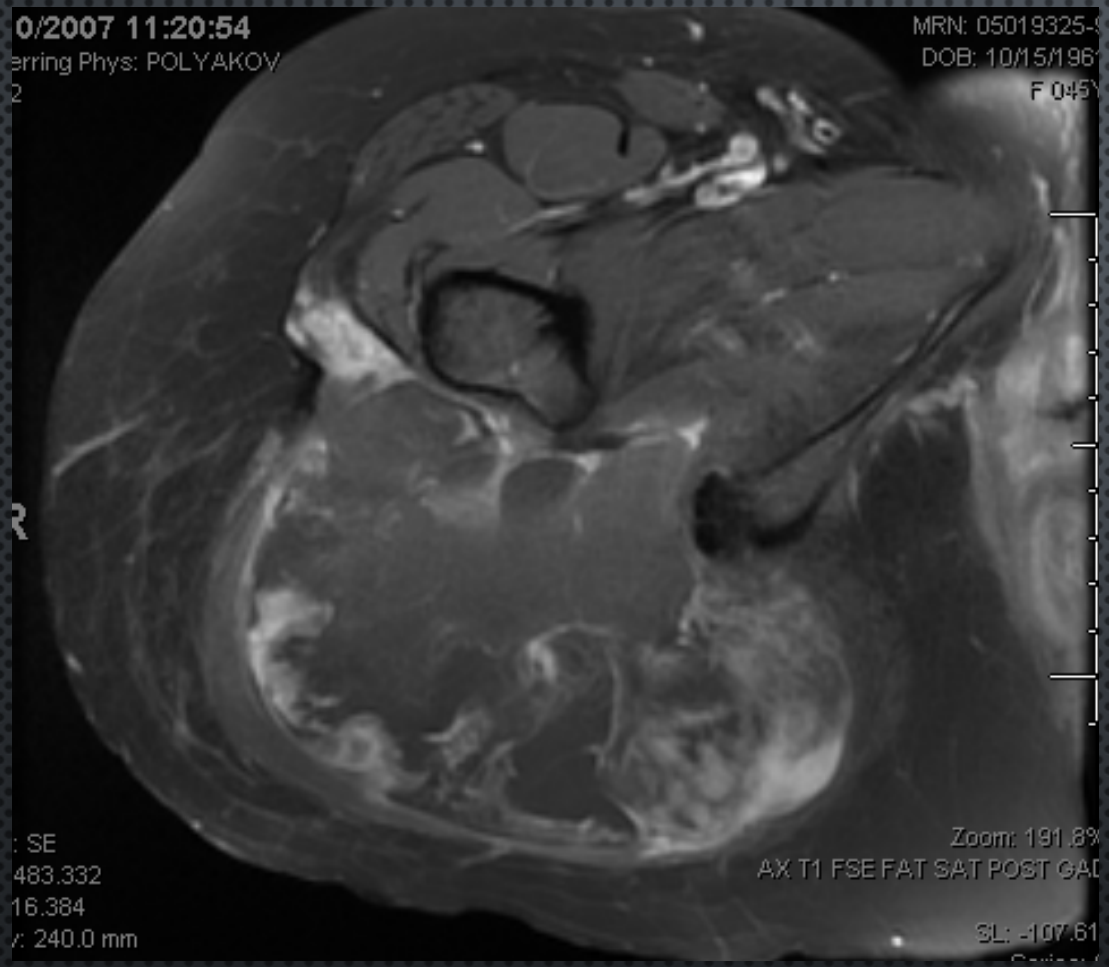
- Presents as large painless mass
- 1/3 develop metastases
- Mets go to odd places = ?role for total body MRI
- Can have systemic mets without lung mets
- Low grade have >90% 10 year survival

MLPS - MANAGEMENT

- Clinical suspicion – large, deep mass
- MRI
- Wide surgical resection of the primary
- Very radiation sensitive
- ?Role for Sentinel Node Biopsy?

48 female, buttock mass

- Presented 2007
- Proven MLPS on biopsy, low grade
- Pelvic nodes positive on biopsy
- Received pre-operative RT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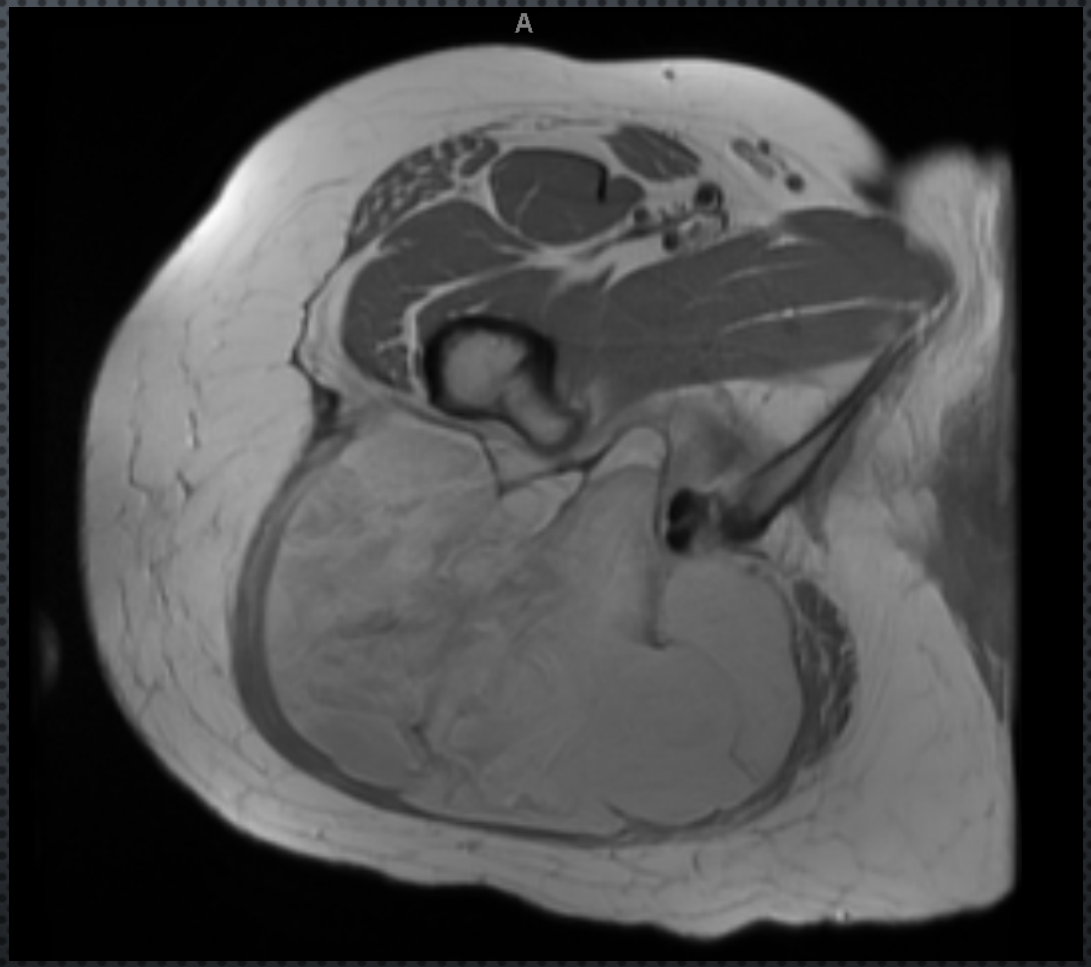
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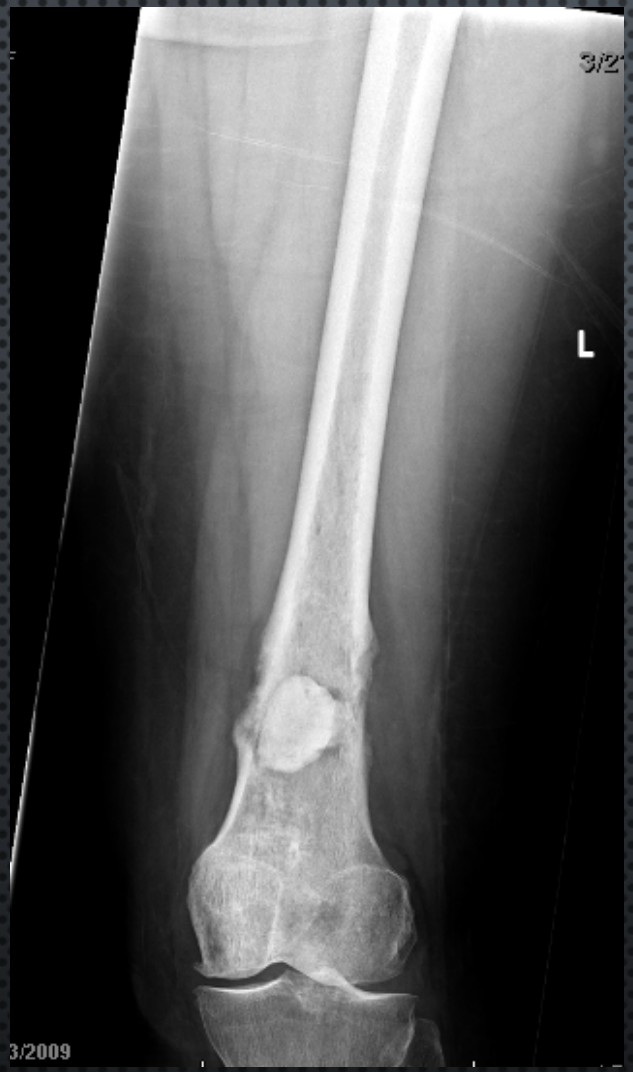
- Wide resection of primary
- No positive nodes identified
- Postop chemo
- 2009 – leg pain = met, resected
- Now 12 years out from primary, clear of disease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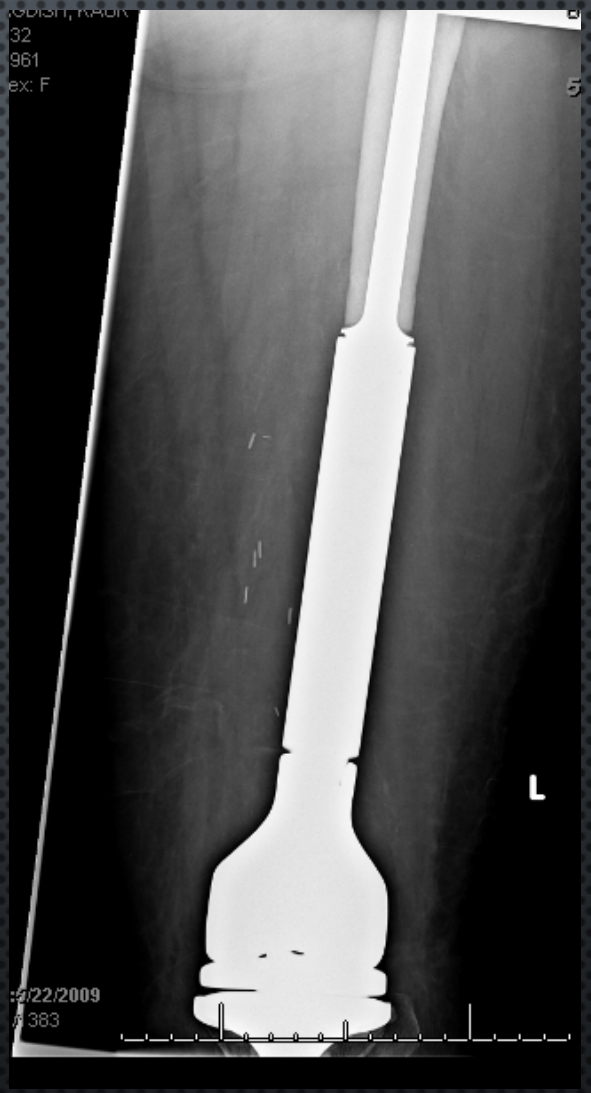
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LESSONS

- 40 years old
- MRI clearly not alt
- Idiosyncratic distribution of mets
- long disease free intervals even with mets
- SNLB - ?

Pleomorphic liposarcoma

- Aggressive tumours - 5 year survival 60%
- Older adults – 70s
- Most common centrally (I rarely see them in the limbs)

Atypical lipomatous tumours

- Locally aggressive benign tumour
- Most common in limbss (esp. Thigh) > Retroperitoneum > other
- Can be subcutaneous, but rare
- Can become very large

Pathology

- Morphologic types: Lipoma-like / sclerosing / inflammatory
- MDM2 amplification with nuclear MDM2 staining
- Supernumerary and ring chromosomes

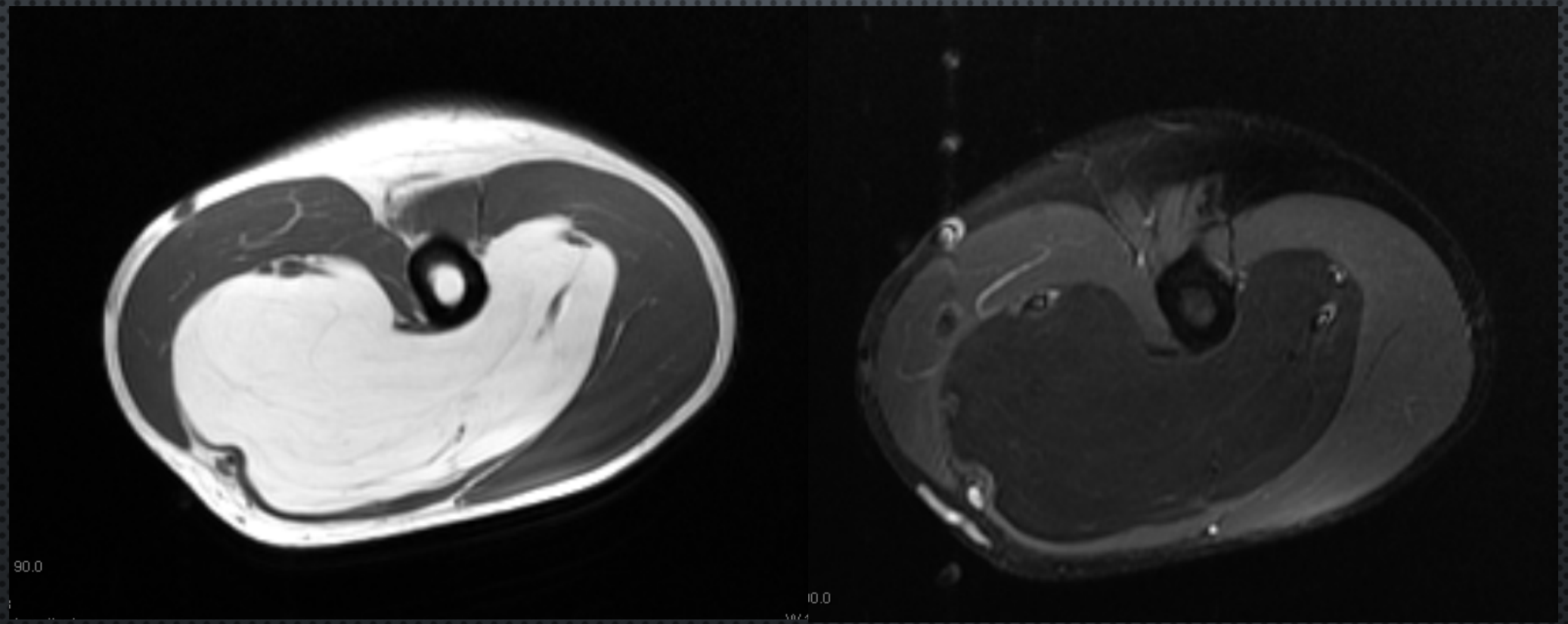
	Extremity	Retroperitoneum
Local Control	Local Recurrence 10-20 %, usually treated with re-excision	Local Recurrence not treatable and often leads to death
Dedifferentiation	<2%	>20%
Overall mortality at 10-20 years	0%	>80%

Distinguishing from a lipoma/other

- Can generally regard all superficial lipomatous tumours as benign
- Deep tumours are more likely to be ALT if –
  - Large >5 cm
  - INTER-muscular, not INTRA-muscular
  - History of growth over last 12-24 months
  - Do not strictly follow signal of fat in all sequences
- Extensive overlap

Practical differentiation

- 1. Biopsy – not helpful as no good MDM2 test
  - But if differential is something else, then helpful
- 2. Observation
  - Serial MRI for 2 or 3 years. If no growth, probably a lipoma
- 3. Take it out
  - If symptomatic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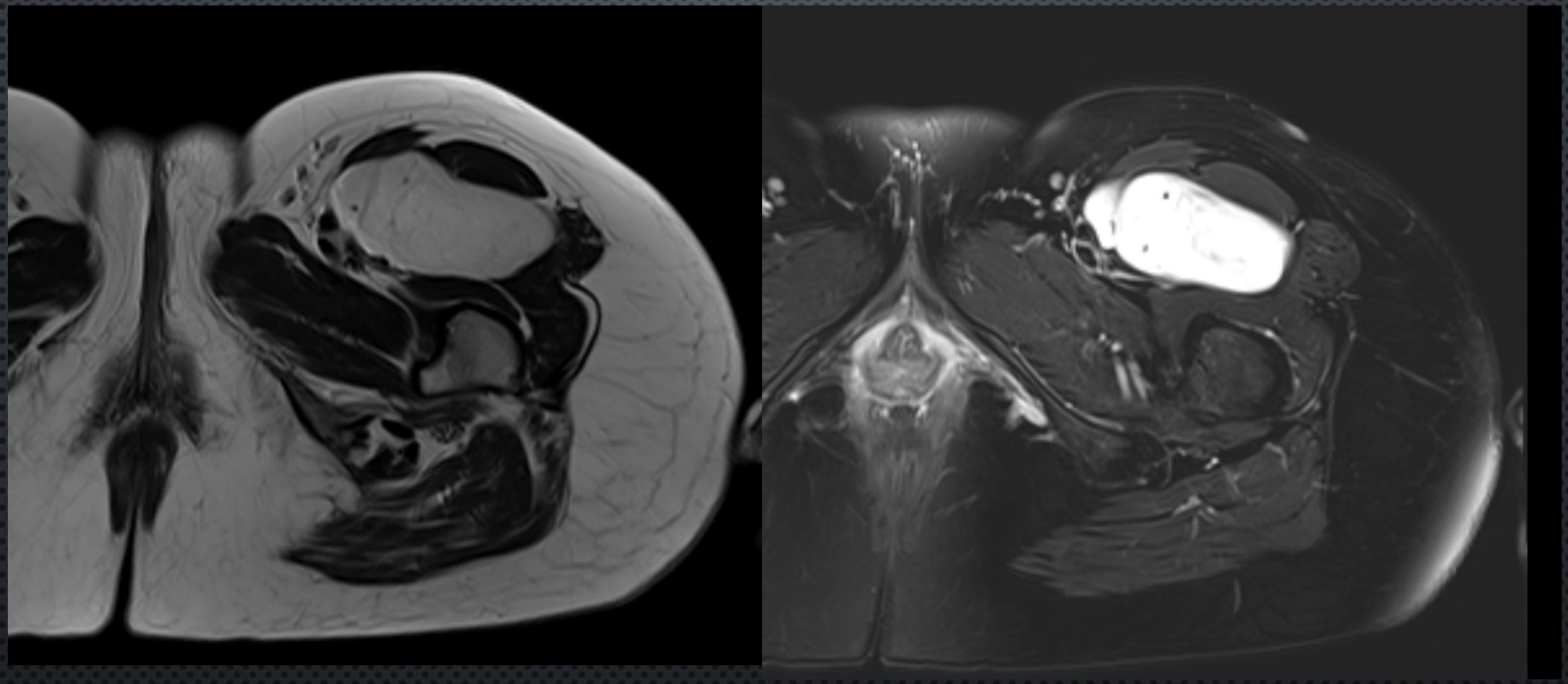
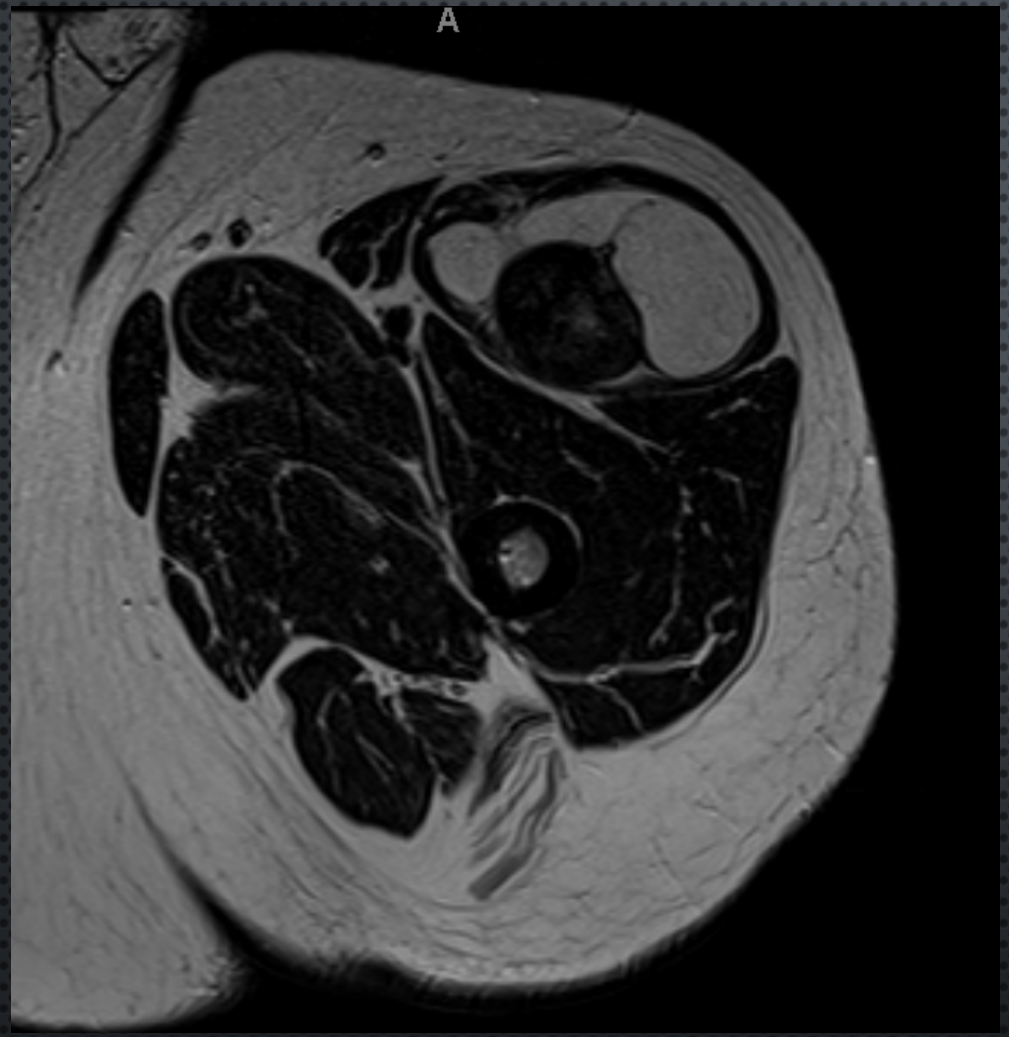
Surgical considerations

- Morbidity
- Incisions should be extensile and well-planned for future surgery
- Do not disrupt capsule
- Take loose tissue or even muscle surrounding it
- drain



Distinguishing a de-diff component

- Does still happen in the extremity
- Usually a clear focus of change on MRI
- If biopsy –ve, need to excise in its entirety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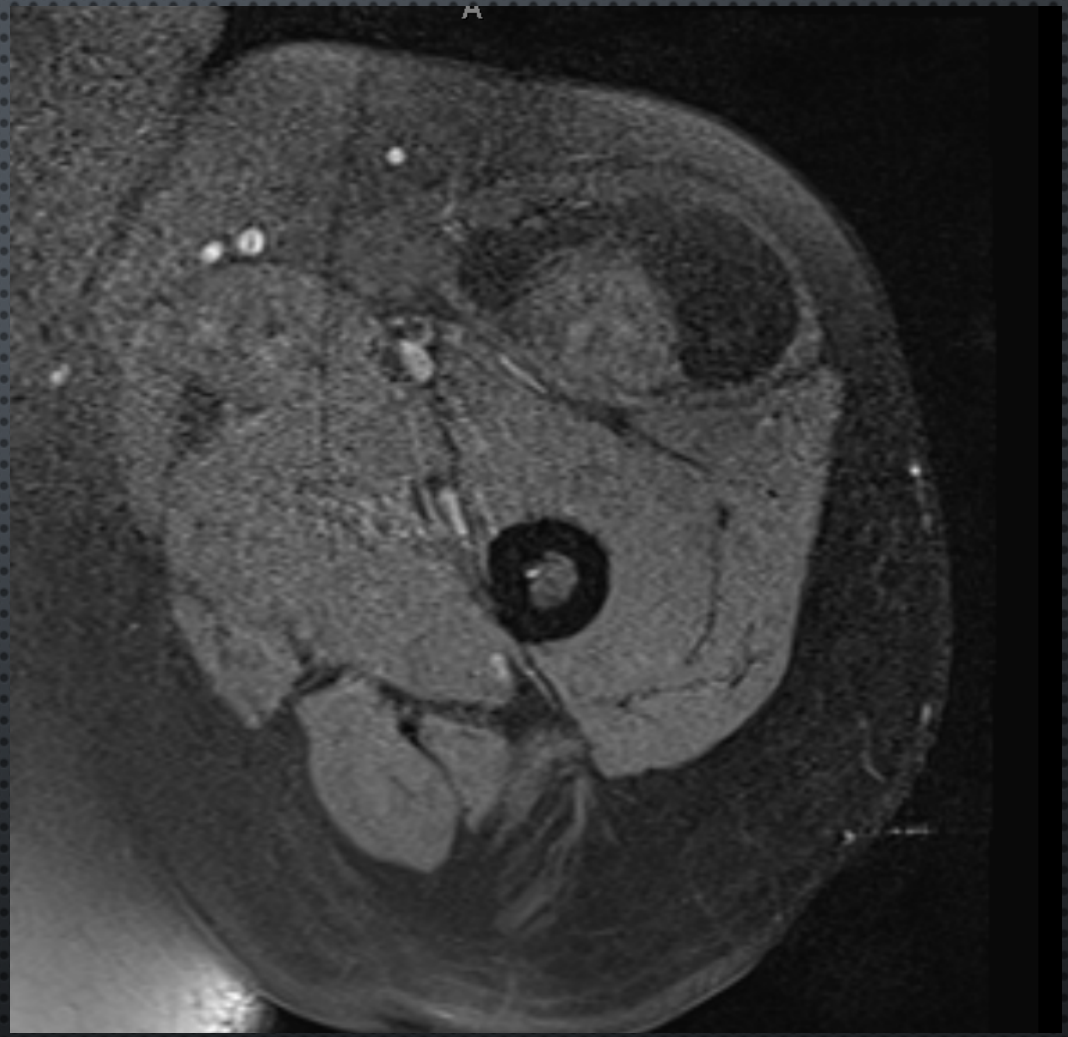
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DE-DIFFERENTIATED LIPOSARCOMA

- Arise in a pre-existing ALT
- Usually retroperitoneal
- Exceptionally rare in subcutaneous tissue
- IN extremity Metastases uncommon (10%)
- Management RT and surgery

Practical approach

- Superficial:
  - 5-8 cm, just take it out
  - Fungating, fast-growing, then need a biopsy/referral
  - >8 cm probably mandates an MRI

Practical approach

- Deep
  - Intramuscular, small – probably lipoma. Reassure.
  - Other
    - Asymptomatic: monitor for a few years.
      - If stable – discharge
      - If grows - remove
    - Symptomatic
      - Remove but respect the high LR
      - Need a good Clearance of all tissue

Practical approach

- Beware of traps
  - Look at the MRI yourself
  - Growing/very large lesions
- Don't hesitate to send us the images to review.

THANK YOU