MINIMAL WORKUP REQUIREMENTS

Introduction
Multidisciplinary patient management is absolutely crucial to optimize the therapy success. It is recommended that biopsy and surgery be performed by the same team/person.

When is a mass suspicious for malignancy (sarcoma)?
Soft tissue: - each subfascial mass (situated underneath the fascia)
- any rapidly growing mass > 3cm and/or symptomatic
- each superficial (epifascially located) mass >3-5cm (depending on localization).
- any superficial mass suspicious for sarcoma (except for "classic" lipoma)

Bone: - any aggressively looking lesion on conventional Xray.

What does an initial (local) imaging include to strengthen my suspicion?
- conventional radiographs of local tumor in 2 projections (mandatory for bone, optional for selected soft tissue lesions)
- MRI w/wo IV Gadolinium

When should a biopsy be planned / performed?
- any mass suspicious for sarcoma
- *after* completion of local imaging
- after presentation at regional Sarcoma Board (or at least contacting it; Addendum 4)

How is the biopsy organized?
- always in consultation with sarcoma surgeon to determine biopsy tract
- core biopsy (CT- or US-guided): whenever possible
- excisional biopsy only when tumor <2cm and superficial; or: after presentation at a sarcoma board
- fine needle biopsy: only recommended with experienced pathologist. Indications: confirmation of local recurrences and metastases or where a core biopsy risks significant morbidity
- incisional biopsy: usually not indicated as first line approach; after tru-cut failure
- histopathological diagnosis of the soft tissue mass has been confirmed/read by Sarcoma Center reference Pathologists (Addendum 5)

What do I do when sarcoma diagnosis is confirmed?
- send patient to regional Sarcoma Center (all bone sarcomas; Addendum 4); or:
- complete staging (see below)
- make sure that patient management strategy is discussed at regional Sarcoma Board.

How do I need to complete staging?
- chest CT (PET-CT usually not necessary)
- thoraco-abdominal CT for myxoid liposarcoma

Concluding remarks:
It is imperative that referring/family physician be informed about each therapy step/strategy. If there is any doubt regarding management, please always contact your nearest Sarcoma Center.

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ADDENDUM 1

What about "classic" lipomatous lesions?

* superficial (epifascial) lesion
    → proceed with surgery in case of "classic" lipoma <5cm
    → in case of doubt and/or >5cm:
        → proceed with MRI imaging;
          if still unclear:
        → proceed with biopsy
* subfascial (deep) lesion → proceed with biopsy as outlined above

MRI imaging of lipomatous lesions:

The following parameters increase the likelihood of atypical lipomatous tumor (ALT)
-deep (subfascial) location
-presence of non-fatty areas / septae / contrast enhancement
-age > 60 yrs
-size > 10cm
-lower limb location

Pathology of lipomatous lesions:

Ask for the expression of biomarkers: MDM2 and CDK4 proteins
MDM2 and CDK4 proteins are expressed in atypical lipomatous lesions (ALT), but not lipomas !
→ immunohistochemistry (IHC) has 80% sensitivity to distinguish between classic lipoma and ALT; in suspicious lesions proceed with IHC, when negative perform FISH.

Current Histopathological Classification of Lipomatous Tumors (incl. molecular features):

1. Lipoma (incl. variants) → benign fatty tumor
2. Atypical lipomatous tumor (ALT) – synonym well-differentiated liposarcoma (WDLS): lipoma like fatty tumor with risk of local recurrence and dedifferentiation, defined histopathologically by the demonstration of the overexpression of the products and/or amplification of mdm2/cdk4 (12q13-14) in lipomatous tissue. ALT is diagnosed in the extremities (most often resectable); WDLS is diagnosed in the retroperitoneum and mediastinum (R0 resection not possible in most instances; mortality risk)

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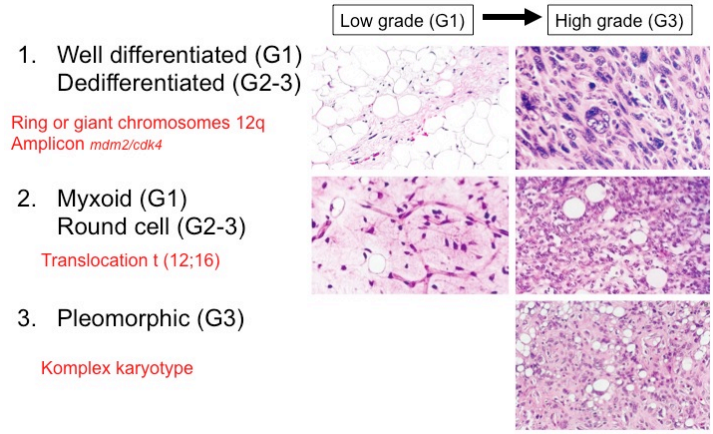

Therefore, performing a biopsy of lipomatous lesions has its importance; based on this information, the surgical approach can be adapted.

ADDENDUM 2

A Sarcoma Board includes the following 9 disciplines, which meet at a (bi-)weekly basis at the sarcoma center:

- Pathology
- Musculoskeletal Oncourgery (incl. Hand)
- Plastic & Reconstructive Surgery
- Medical Oncology
- Radiation Oncology
- Radiology
- Visceral Surgery
- Thoracic Surgery
- Pediatric Oncology

ADDENDUM 3

Each sarcoma diagnosis has to be established by one of the sarcoma center reference pathologists (confirmation by external consultation only in case of doubt). The sarcoma pathologists meet 4 times a year (the bone sarcoma group (under the lead of Daniel Baumhoer) and the soft tissue sarcoma group (under the lead of Beata Bode) each twice) to confirm the diagnosis of each sarcoma patient to enter the register.