Fun with Fat

General Rules

• Imaging: location (deep vs. superficial)
• Superficial lesions are seldom liposarcomas
• Deep lesions may be benign or malignant
• Myxoid stroma is common in benign and malignant lesions
• Lipoblasts can be seen in benign and malignant tumors
• Well diff. liposarcoma is a 4x diagnosis

Case

34 year old female with a 2cm mass lesion located on the chest, adjacent to the armpit. The lesion was excised and sent to pathology.
Diagnosis
Ruptured silicone breast implant

Case
40 year old male with a 2 cm neck mass. Lesion was excised and sent to pathology.
Diagnosis
Benign Lipoma with fat necrosis
Lipoma

- Benign tumors composed of cells resembling mature adipocytes
- Most common soft tissue tumors of adulthood (40-60 yrs)
- Simple excision. Rarely recur.
- Many subtypes; none with prognostic significance
- May be superficial or deep, intra- or inter-muscular
- Main differential: Well diff. liposarcoma

Lipoma with focal fat necrosis:
May mimic fibrous septa seen in WDLS.

Lipoma with focal fat necrosis:
May mimic variation in adipocyte size as in WDLS.
Lipoma with fat necrosis:
Sclerosis and variation in adipocyte size

“Lochkern” – intranuclear lipid invagination
Hyperchromatic “ugly cell” in a well differentiated liposarcoma

Good – 4X

Bad – 4X

Good - Oil

Bad - Oil
Intra-muscular Lipoma: Prone to recur [infiltrative]

Fibrolipomatous Hamartoma of Nerve

- Lipoma-like mass lesion attached to a large nerve with fibrosis
- Often median or ulnar nerves on hand, wrist, or fingers. Macrodactyly.
- Nerve is surrounded by ring of concentric fibrosis
- Benign

FLPH of nerve: Fat surrounds a nerve
FLPH of nerve: Fat surrounds a nerve with concentric fibrosis

FLPH of nerve: Fibrosis-rich lesion
Case

2 year old male with a slowly growing mass lesion on the upper left arm. The lesion was excised and sent to pathology.
Diagnosis

Lipoblastoma

• Benign tumor, typically arises during the **first three years of life**
• Slow growing painless mass most often on subcutaneous extremities
• Lipoblastomatosis: multiple poorly circumscribed masses, may be deep
• Local resection. May recur locally (9-22%). Does not metastasize
• Has features in common with lipoma (lobules of mature fat), WDLS (fibrous septa), myxoid liposarcoma (myxoid stroma with branching vessels, lipoblasts)

Lipoblastoma (mature areas): lobules, fibrous septa
Lipoblastoma (immature): Myxoid stroma with branching vessels

Lipoblastoma – Myxoid Stroma with prominent vessels

Lipoblastoma - Lipoblasts
Case

43 year old male with a 3 cm mass on the back of the neck. Lesion was excised and sent to pathology.
Diagnosis
Pleomorphic Lipoma

Lipoma Variants
• Many Variants, we will cover the most common entities
• All behave similar to regular lipomas
• Stromal collagen bands are very helpful to distinguish benign from malignant adipose tumors
• MDM2 immunostains/FISH

Angiolipoma
• Usually young adults
• Often on forearms
• Painful and multiple
• Numerous small capillaries with admixed fat in a lobular configuration
• Capillary supporting elements and fibrin thrombi
Angiolipoma: Lobular at low power. Fat, blood, and hypercellularity.

Angiolipoma: Lobular arrangements of small vessels with admixed adipocytes.

Angiolipoma: Stromal support elements surround and invest capillary proliferations.
Chondroid Lipoma

- Usually small, subcutaneous mass on extremity of an adult (female predominance)
- Well-circumscribed, lobulated, cellular
- Well-defined cells with intracytoplasmic vacuoles growing in “chondroid” matrix
- S100 protein, occasional focal Keratin
- Characteristic 11;16 translocation
Chondroid lipoma – lobulated, cellular

Chondroid lipoma – numerous vacuolated cells in an eosinophilic matrix that appears chondroid

Chondroid lipoma – Vacuolated cells resemble lipoblasts
Spindle Cell Lipoma

- Usually in males 45-70 years
- Solitary, painless mass on back of neck, shoulders, elsewhere
- Circumscribed and **subcutaneous**

Spindle Cell Lipoma

- Variable numbers of mature adipocytes, bland spindle cells, mast cells
- Wiry collagen fibers embedded in myxoid stroma, cleft-like spaces
- CD34+, S100-
Spindle Cell Lipoma: Well-circumscribed. Fibrotic and myxoid with fat at low power.

1. Mature fat
2. Small hyperchromatic spindle cells
3. Myxoid matrix
4. Wiry Collagen
5. Mast cells

Spindle Cell Lipoma: Mast Cells
Spindle cell lipoma – myxoid with cleft-like spaces

Spindle cell lipoma – clefts, myxoid, wiry collagen
Spindle cell lipoma – clefts, myxoid, wiry collagen

Pleomorphic Lipoma
- Basically a spindle cell lipoma with atypia, thicker collagen bundles
- Hyperchromatic multinucleated cells in a “floret” arrangement
- Lipoblasts may also be present
- How to distinguish from WDLS - collagen pattern, MACROSCOPIC appearance, MDM2(-)
Pleomorphic lipoma – circumscribed, fat, fibrotic and myxoid stroma with large cells at low power

Pleomorphic Lipoma: Atypical Hyperchromatic cells

Pleomorphic Lipoma: Bizarre Atypia
Pleomorphic Lipoma: Lipoblasts

Pleomorphic Lipoma: Mast Cells and Collagen

Pleomorphic Lipoma: dense eosinophilic collagen bands and cellular myxoid areas
Hibernoma

- Uncommon benign tumor with features of brown fat; younger patients than lipoma (late 30s).
- Arise in several locations, including abdominal cavity and retroperitoneum. Usually 9-10cm. Benign.
- Three cell types: mature adipose, finely vacuolated, granular eosinophilic. Small branching capillaries.
Case

41-year-old male with a large ileoinguinal mass lesion. The lesion was resected and sent to pathology.
Diagnosis
Dedifferentiated liposarcoma

Liposarcomas – Three general types
• Well-Differentiated /Dedifferentiated
• Myxoid/High grade myxoid (Round Cell Liposarcoma)
• Pleomorphic Liposarcoma

Well-Differentiated Liposarcoma
• 40-45% of liposarcomas
• Middle aged adults – 6th decade, M=F
• Deep soft tissues of limbs [esp thighs – where the fat is] followed by retroperitoneum and groin
• Metastasis is a risk in lesions with dedifferentiation (28% of retroperitoneal lesions)
• Wide Local excision. 40-50% recurrence rate for extremities. Retroperitoneal and groin lesions nearly always recur (80-90%)
Atypical Lipomatous Tumor

- ALT terminology is applied to superficial/ extremity lesions
- Same histology and genetic alterations as deep lesions
- Different natural history: much lower likelihood of dedifferentiation (5% vs. 28%) and metastasis

Well-Differentiated Liposarcoma: Lipoma-like

- Thick fibrous bands
- Variation in mature adipocyte size
- Pleomorphic hyperchromatic cells visible within fibrous bands at 4x scanning magnification
- +/- Lipoblasts: *Not necessary* for diagnosis.

Retroperitoneal well-diff liposarcoma
Paratesticular well-diff liposarcoma

Thick fibrous bands with hyperchromatic cells

WDLS: Hyperchromatic cells at low power
Hyperchromatic cells with no lipoblasts

Pleomorphc cells in thick collagen bands
- The eosinophilic fibrillar collagen bands seen in pleomorphic lipoma are absent.

WDLS: Needle Biopsy at 4x mag.
Fat, hyperchromatic cells, fibrous septa
Well-Differentiated Liposarcoma: Inflammatory type

- Extensive infiltration of tumor with lymphoid cells with follicles and germinal centers
- May also have eosinophils and neutrophils
- Atypical cells of WDLS may cause confusion with lymphoma
- Focal inflammatory-type areas may be seen in other subtypes
Inflammatory WDLS: Fat, fibrous bands, myxoid areas, inflammation, lymphoid follicles.

Well-Differentiated Liposarcoma: Sclerosing Type
- May not appear adipocytic on imaging
- Mostly fibrotic or hyalinized stroma with hyperchromatic cells in sclerotic areas
- Infrequent mitoses, moderate to low cellularity, no fascicles

Sclerosing WDLS: Hyperchromatic and pleomorphic cells in sclerotic stroma with few adipocytes
Dedifferentiated Liposarcoma

- A confluent focus of high grade sarcoma arising within or adjacent to a WDLS. No size criteria.
- Bestows metastatic capability (15-30% metastatic rate).
- May occur at any site and also increases risk of local recurrence.

Dedifferentiated Liposarcoma

- May present as any non-adipocytic sarcoma, MFH most common
- Most retroperitoneal MFHs represent dediff. liposarcoma
- Classically abrupt transition from WDLS to dediff. component
- Hypercellularity without adipocytes, nuclear pleomorphism, high mitotic rate

Dediff. liposarcoma  Lipoma-like WDLS
Lipoma-like WDLS

Sclerosing WDLS

Dediff. liposarcoma

Dedifferentiated Liposarcoma

MFH-like

Myxofibrosarcoma-like

Dediff. Liposarcoma: MDM2 immunostain
**Low-grade Dedifferentiation**

- >1 low power field with no fat
- More cellular and fascicular than sclerosing WDLS
- Basically a low grade fibrosarcoma component
- Does not improve prognosis
- Chief differential: Sclerosing WDLS

**Fooler for Atypical Lipomatous Tumor/Well Diff LPS**

- Massive localized lymphedema


Massive localized lymphedema in the morbidly obese – Reactive fibroblasts in septa appear similar to the cells in atypical lipomatous tumor – CONTEXT of superficial mass with ectatic lymphatic vessels is key
Massive localized lymphedema in the morbidly obese – Reactive fibroblasts in septa appear similar to the cells in atypical lipomatous tumor – note the hemosiderin

Case

43 year old male with a 7cm soft tissue mass deep in the thigh. The lesion was biopsied and sent to pathology.
Diagnosis
Myxoid Liposarcoma

Case
An incidental plaque-like nodule on the abdominal wall of a 78 year old female was noticed during an exploratory laparotomy. It was excised and sent to pathology.
Diagnosis
Fat Atrophy
Cytomegalovirus infection with fat atrophy in an HIV/AIDS patient

Myxoid/Round Cell (high grade form) Liposarcoma

- About a third of liposarcs, deep extremities (esp. thigh), younger patients (4th decade)
- ~30% metastasize. Peculiar metastatic tropism to other soft tissue sites, lung, and bone
- A translocation sarcoma: bland, uniform oval to spindled tumor cells in a myxoid matrix with finely branching vasculature
- Cells often cluster at periphery of lobules or around vessels. Lipoblasts present
- “Round” cell is high grade variant of myxoid
Myxoid Liposarcoma: Lobules with clustering of tumor cells at periphery and around vessels in a myxoid background.
Myxoid Liposarcoma: Uniform oval cells

Myxoid Liposarcoma – Uniform spindled cells. Children may present with purely spindled cells with little to no fat or lipoblasts.

Round Cell Liposarcoma – t(12;22) – TLS(FUS) – CHOP (DDIT3) T(12;22) – EWSR1-CHOP (DDIT3)
Transitional Myxoid/Round Cell Liposarcoma
Partial preservation of background stroma

Pleomorphic Liposarcoma
- Rare and aggressive. M=F, >50yrs
- Deep soft tissues of the thigh, trunk or retroperitoneum
- Typically exceed 10.0 cm
- High incidence of metastases and tumor related mortality (5yr survival of 50-63%)
- Variable areas of pleomorphic lipoblasts and MFH-like morphology, some resemble myxofibrosarcoma

Pleomorphic Liposarcoma: Numerous atypical Lipoblasts.
Pleomorphic Liposarcoma

- MFH-like areas
- Lipoblast areas

Pleomorphic liposarcoma: Epithelioid lipoblasts

Pleomorphic liposarcoma: Myxoid stroma
Case

63 year old female with a history of myelofibrosis with a 3cm perirenal mass lesion. The lesion was biopsied and sent to pathology.
Diagnosis
Sclerosing extramedullary hematopoietic tumor

Case 5
• 76 year old female with a 5cm subcutaneous mass on the foot. The lesion was excised and sent to pathology.
Diagnosis

• Myxofibrosarcoma

Myxofibrosarcoma: Fake Lipoblast
Pleo. Liposarcoma: Real Lipoblast
Myxofibrosarcoma (Myxoid MFH)

- One of the most common sarcomas during 6th-8th decade; typically limbs, ~50:50 superficial:deep
- Multilobulated with incomplete fibrous septa; multiple infiltrative nodules in superficial locations vs. a solitary mass deep
- Spindled and pleomorphic cells in myxoid matrix with curvilinear vessels, pseudolipoblasts
- Graded primarily on cellularity: low grade lesions do not metastasize, 21-38% of intermediate and high grade lesions do
- All grades have high recurrence rates, and low grade lesions may recur as a higher grade
Myxofibrosarcoma: pleomorphism, myxoid stroma, prominent vascularity

Myxofibrosarcoma: curvilinear vessels

Myxoid LPS: Uniform cells
Who needs DDIT3 with good H&E????

Thank you!!!!