Myxoid Liposarcoma. Radiological findings.

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Authors: G. M. Santandreu¹, J. Llauger Rosselló², J. Palmer Sancho², S. Valverde Lavinien³, L. F. Granados Palacio²; ¹08025, Ba/ES, ²Barcelona/ES, ³Barcelona, barc elona/ES
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Learning objectives

1. To describe the radiological spectrum of myxoid liposarcoma with special emphasis on MRI findings.
2. Describe the main epidemiological, clinical, and pathological aspects of this tumor.
3. To highlight the most characteristic findings of this tumor.
4. To document unusual forms of metastatic spread.
Background

I - INTRODUCTION and EPIDEMIOLOGY:

Liposarcoma is a malignant tumor of mesenchymal origin, rather than mature fat cells.

The first description of liposarcoma is attributed to Virchow in 1857, as tumor arising from adipose tissue which he called myxoma lipomatoides indicative of a lesion comprised of myxoid and fatty components.

Liposarcoma is the second most common of the adult soft-tissue sarcomas, accounting up to 18% of all soft-tissue sarcomas.

Although LPS can be classified into five subtypes, recent developments in molecular biology and genetic mapping now support designation of three major categories:

- Atypical Lipomatous Tumor, Well-Differentiated Liposarcoma, and Dedifferentiated Liposarcoma: the commonest (50%).
- Myxoid Liposarcoma and Round Cell Liposarcoma: is the second most common subtype.
- Pleomorphic Liposarcoma: is the least common subtype.

Myxoid LPS and round cell LPS were formerly classified as distinct entities but are now recognized as a continuum of the same entity on the basis of a common chromosomal translocation found in up to 90% of cases.

Myxoid liposarcoma represent an intermediate grade of malignancy (if predominantly myxoid, but high-grade if they contain a substantial round cell component) whereas the well-differentiated is a low grade and the pleomorphic are regarded as highly malignant tumors with a tendency toward local recurrence and metastasis.

This is a typical tumor of adulthood that occurs most often between the fourth and fifth decades of life, ten years before the other histologic subtypes of liposarcoma.
They are mainly detected in extremities (representing the 30%-40% of all liposarcomas in the extremities), buttocks, retroperitoneum, trunk, ankle, proximal limb girdle, head and neck and wrist.

The clinical presentation includes a large, slow-growing, painless mass. The mass is relatively soft and nontender at palpation and may be quite large at presentation.

II - PATHOLOGIC FEATURES:

Myxoid liposarcomas are well-circumscribed and multinodular masses whose gross pathologic appearance varies depending on the degree of myxoid and round cell components.

Predominantly myxoid lesions are gelatinous, whereas lesions with focal areas of round cells contain opaque white nodules. Lesions with a predominant round cell component have a nonspecific, white fleshy appearance similar to that of other soft-tissue sarcomas.

At microscopic analysis, myxoid liposarcoma consists of a myxoid matrix, delicate arborizing vascular networks, and lipoblasts. The radiological features will depend of the predominant histological elements found in the tumor.

III - RADIOLOGICAL FEATURES:

A - PLAIN FILMS:

Radiographs of patients with liposarcoma may identify a soft tissue mass but are rarely specific.

Calcification is uncommon but has been reported in up to 10 per cent of cases. The well-defined ossification are rarely seen. Fig. 1 on page 9

B- CT, PET-CT and US:

At CT, these lesions typically have attenuation lower than that of adjacent muscle, but they are not isoattenuated relative to pure fluid; indeed, they may be confused with cystic lesions.
They can be distinguished from them by the enhancement of the mass with intravenous contrast medium which is obviously absent in the cysts.

Also US can be useful to distinguish for depicting the solid nature of these tumors: the finding of a complex hypoechoic lesion that fails to demonstrate through transmission easily rules out the presence of simple fluid.

The assessment of this type of tumor using functional imaging tests such as PET, can make one, although not exact, diagnostic orientation of the histological type by metabolic tumor characteristics and quantifying the maximum tracer uptake (SUV max) reflecting the most metabolic areas of the tumor.

The assessment of maximum SUV by FDG-PET can be considered a useful parameter in guiding the histologic subtype of liposarcoma; it has been described that maximum SUV values of 3.5 g/ml orientates to the mixoid variant of liposarcoma.

C - NONENHANCED MR IMAGING:

Myxoid liposarcomas has a characteristic appearance on MRI: usually seen as a multilobulated intramuscular mass, the high water content of these tumors produces low signal intensity on nonenhanced T1- and high signal intensity on T2-weighted sequences.

These features are due to the fact these tumors consist of a myxoid matrix as the predominant component and small amounts of mature fat.

On T1-weighted images, most myxoid liposarcomas exhibit high-signal-intensity foci within a predominantly homogeneous low-signal-intensity mass; these foci represent fat within the tumor.

Some myxoid liposarcomas may instead appear as cystic masses on nonenhanced images; the absence of high-signal-intensity foci on T1-weighted images makes a myxoid liposarcoma indistinguishable from most other soft-tissue masses (as well as the heterogeneous aspect of the pure round cell variant).
In these cases US can be useful to differentiating solid masses from cystic ones as well as contrast enhanced MR.

**D - CONTRAST ENHANCED MR:**

As we said above myxoid liposarcomas can be mistaken for cystic lesions cause its very high signal and clear demarcation of the tumor with homogeneous signal intensity.

The enhancement patterns are: homogeneous (total enhancement), heterogeneous (partial enhancement), and no enhancement and these depends on the degree of cellularity, the vascularity and presence of necrosis.

Tumors with a homogeneous enhancement pattern have a prominent plexiform capillary pattern, increased cellularity, and abundant myxoid substance.

In tumors with heterogeneous enhancement, the enhancement pattern is variable. These tumors consist of two distinct zones: one has compact cellularity representing the enhancing areas and the other contains necrosis and mucinous material that represents the nonenhancing areas.

Some liposarcomas consist of multiple histologic subtypes within the same lesion. In this setting, the signal intensity throughout the lesion can vary depending on the combination of histologic subtypes.

In case of recurrent or multicentric tumors, the MR imaging findings are similar to those of the primary tumors.

Postoperative hygromas and seromas usually appear as homogeneous cystic lesions on T1 and T2 weighted images, and recurrent myxoid liposarcomas may appear as cystic lesions on nonenhanced images.

**IV - MULTICENTRIC TUMORS:**
Myxoid liposarcomas may be multicentric, with involvement of two or more anatomic sites.

In 10% of patients with primary liposarcomas of the thigh, a second liposarcoma occurs in the retroperitoneum two or more years after removal of the primary tumor.

V - PATTERN OF METASTATIC SPREAD:

Myxoid liposarcomas differs from other subtypes in its unusual pattern of metastatic spread specially in extrapulmonar sites (unlike most of the other soft-tissue sarcomas).

Fuglo et al analysed 45 patients with myxoid liposarcoma, and founded distant metastases in 16% of them with an unusual pattern of spread compared to what is seen in other liposarcomas and soft-tissue tumors.

The most common metastatic sites include the paraspinal regions, bone, retroperitoneum, and opposite extremity followed by the lungs and liver.

Fig. 9 on page 16 Fig. 10 on page 17 Fig. 11 on page 18

MRI is the most sensitive modality in the detection of osseous and soft tissue metastases, and is the recommended modality for the diagnosis and follow-up of bone and soft tissue involvement.

It is recommend that in patients treated for myxoid liposarcoma, with no or few round cells, diagnostic imaging including both the lungs and abdomen / retroperitoneum (e.g., CT of thorax and abdomen) should be performed as a part of the postoperative control.

Bone metastases from myxoid liposarcoma are common unlike other types of liposarcomas.

They may not be detected in PET-CT studies due to its hypometabolic condition: thus, the study of metastatic spread of myxoid liposarcoma should include a whole body MR study or, at least, an spine MR study.

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It should be considered to extend the followup beyond five years postoperatively.

VI - DIFFERENTIAL DIAGNOSIS

Many benign and malignant soft-tissue tumors contain myxoid tissue.

Such tumors include:

1. **Extraskeletal myxoid chondrosarcoma**: contrast-enhanced images may show rings and arcs, which reflect the typical lobulated growth pattern of cartilaginous tumors of bone.

2. **Intramuscular myxoma**: at MR imaging, myxoma appears as a well-defined mass with prolonged T1 and T2 that mimics a cyst on nonenhanced images.

   Differentiation of myxoma from myxoid liposarcoma may be difficult because of the variable and often complex components. However myxoid liposarcomas shows intense enhancement of the tumor volume in most of the cases including those cases with a cystic appearance on nonenhanced images and, second, the high signal intensity from fatty foci on T1-weighted images is quite characteristic.

3. **Ganglion**: they occur uncommonly in muscle bundles. The MR imaging appearance of ganglion is similar to that of other cysts. No enhancement or peripheral enhancement is observed on gadolinium-enhanced images.

4. **Epidermoid Cyst**: epidermoid cysts are usually found in the subcutaneous tissue. These cysts most often show the signal intensity characteristics of a simple cyst. No enhancement is observed at gadolinium-enhanced imaging.
Fig. 1: Nodular-morphology soft-tissue tumor in the distal third of the left thigh, without bone involvement. No calcifications are seen. Diagnosis: myxoid liposarcoma.

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**Fig. 2:** Axial T1 weighted image (same patient of figure 1). Well defined mass in the distal third of the left thigh with high-signal-intensity foci. These foci represent fat within the tumor. Diagnosis: myxoid liposarcoma.

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Fig. 3: Axial T2 weighted image (same patient of figure 1). It shows a high signal well defined mass in the distal third of the left thigh. Diagnosis: myxoid liposarcoma.

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Fig. 4: Axial post contrast MR image (same patient of figure 1). It shows a well defined mass in the distal third of the left thigh with diffuse enhancement. Diagnosis: myxoid liposarcoma.

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Fig. 5: Coronal T1 weighted image. Soft tissue tumor with low signal in the distal third of the left thigh, located between biceps femoris and adductors. Diagnosis: myxoid liposarcoma (recurrence after two years of surgery).

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**Fig. 6:** Coronal MR STIR image (same patient of figure 5). Soft tissue tumor with high signal in the distal third of the left thigh, located between biceps femoris and adductors. Diagnosis: myxoid liposarcoma (recurrence after two years of surgery).

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Fig. 7: Contrast-enhanced CT scan of the pelvis (same patient of figure 5). Large heterogeneous mass (of predominantly hypodense) with higher density areas inside and fat component. Diagnosis: myxoid liposarcoma (metastatic lesion after three years of surgery of the thigh primary lesion).

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Fig. 8: CT scan of the chest. Patient with history of fast growing right axillary mass: nodular tumor, located in the soft tissues of the right axillary region (surrounding the subclavian vein) with heterogeneous solid content and lipomatous component. Diagnosis: myxoid liposarcoma.

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Fig. 9: CT of the chest. Soft-tissue mass located on top of the right hemithorax with pulmonary apex involvement (arrow). Diagnosis: myxoid liposarcoma metastatic lesion after four years of surgery of the primary tumor (located in the right popliteal fossa).

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Fig. 10: CT scan of spine (same patient of figure 9). Soft tissue density mass in the apex of the right hemithorax. This lesion produces an expansion of the contiguous T1-T2 intervertebral foramen (arrow). Diagnosis: myxoid liposarcoma metastatic lesion after four years of surgery of the primary tumor (located in the right popliteal fossa).

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**Fig. 11:** PET-CT image of the chest: acquisition 60 min after iv injection of 18-FDG(same patient of figure 9). Pulmonary nodule with abnormal uptake in contact with T1 transverse apophysis(SUV 3,6). Diagnosis: myxoid liposarcoma metastatic lesion after four years of surgery of the primary tumor (located in the right popliteal fossa).

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Fig. 12: Coronal T-1 weighted image with fat saturation obtained in a 24-year-old man. High signal large mass affecting the posterior compartment of thigh. Diagnosis: myxoid liposarcoma recurrence one year after surgery.

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Fig. 13: Whole body PET-CT image acquisition 60 min after iv injection of 18-FDG (same patient of figure 12). It shows no pathological deposits of the FDG in a patient with extensive metastatic spread of a myxoid liposarcoma (see figures 14 to 18).

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**Fig. 14:** Whole body MR: coronal T1-weighted image of the pelvis (same patient of figure 12). Soft-tissue mass located along the lower and medial rim of the left gluteus maximus (arrow). Lesion not detected on PET-CT study. Diagnosis: myxoid liposarcoma (metastatic lesion after a year of recurrent tumor surgery).

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**Fig. 15:** Whole body MR: coronal T1 weighted image of the pelvis (same patient of figure 12). Bone metastatic lesions detected in left femur and supraacetabular region (arrows). Lesions not detected on PET-CT study. Diagnosis: myxoid liposarcoma (metastatic lesion after a year of recurrent tumor surgery).

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Fig. 16: CT of the spine (same patient of figure 12). Blastic lesion in the body of second lumbar vertebra. Lesion not detected on PET-CT study. Diagnosis: myxoid liposarcoma (metastatic lesion after a year of recurrent tumor surgery).

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**Fig. 17:** Contrast CT scan of the abdomen (same patient of figure 12). Involvement of adjacent left psoas muscle (arrow). Involvement of adjacent left psoas muscle of the lumbar blastic metastatic lesion showed in figure 16 (arrow). Lesions not detected on PET-CT study. Diagnosis: myxoid liposarcoma (metastatic lesion after a year of recurrent tumor surgery).

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Fig. 18: Contrast CT scan of the chest (same patient of figure 12). Hypodense solid mass in the upper mediastinum, located in the right paratracheal space, with extensive contact with the esophagus, trachea and brachiocephalic vessels. Lesion not detected on PET-CT study. Diagnosis: myxoid liposarcoma (metastatic lesion after a year of recurrent tumor surgery).

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Findings and procedure details

We performed a retrospective review of all myxoid liposarcomas studied in our hospital from January 2005 to June 2013.

All patients were evaluated by magnetic resonance imaging and CT.

With MRI, we studied:

Tumor size and location of the tumor, margins of the lesion, MR signal, contrast enhancement, bone invasion and articular and/or vascular involvement.

With CT we studied characteristics and location of metastatic lesions.
Conclusion

1- A wide spectrum of MR imaging features may be noted in myxoid liposarcomas of the soft tissue that are due to several factors.

2- Gadolinium-enhanced imaging can be helpful in differentiating myxoid liposarcomas from benign cystic tumors and in directing biopsy.

3- Thoraco-abdominal CT is required in extension studies in these patients due to unusual metastatic spread.

4- Bone metastasis of myxoid liposarcoma may not be detected in PET-CT studies due to its hypometabolic condition; thus, the study of metastatic spread should include a whole body MR study or, at least, an spine MR study.
References