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Pulmonary Metastasis In Recurrent Myxoid Liposarcoma Despite Histological Grade -A CASE REPORT

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ABSTRACT

Background: Myxoid liposarcoma is an uncommon malignancy of adipocyte origin in soft tissue. Despite being characteristically low-grade, it has an uncharacteristic tendency for recurrence and distant metastasis, most frequently to the lung. A case of recurrent myxoid liposarcoma with swelling of the right leg and metastasis to the lung in a 60-year-old male is described here.

Case Report: A 60-year-old male patient presented with a history of recurrent calf swelling of the right leg and multiple previous excisions (2005, 2010, 2014) presented with a new calf swelling. Imaging and biopsy revealed recurrent myxoid liposarcoma. PET-CT revealed local invasion and pulmonary metastasis. Chemotherapy with gemcitabine and docetaxel was initiated. Lifestyle counseling and a follow-up visit were advised.

Conclusion: This case highlights the importance of Myxoid liposarcoma, though a low-grade sarcoma, is characterized by extensive local invasion and distant metastasis. Early treatment and appropriate systemic management are essential to manage recurrence and metastasis.

Keywords: *Myxoid Liposarcoma, Soft Tissue Sarcoma, Pulmonary Metastasis, FNCLCC Grade 1, Chemotherapy, Recurrence*

1. INTRODUCTION

Myxoid liposarcoma is an adipocytic sarcoma of primitive cell origin and is one of the most common subtypes of liposarcoma. It accounts for approximately 30–40% of liposarcomas and is primarily seen in adult patients in the third to sixth decades of life. The most common site of presentation is in the upper and lower deep soft tissues of the lower extremities (ground), including the thighs and calves, but it is not limited to these areas.

Histologically, the myxoid liposarcoma is characterized by myxoid matrix with a capillary network, round to oval, uniform and regular non-lipogenic cells, lipoblasts, and the characteristic chromosomal translocation t(12;16)(q13;p11)[1]. The

presence of the FUS-DDIT3 fusion gene resulting from this translocation and the histopathological and molecular features differentiate myxoid liposarcoma from other soft tissue sarcomas.

Clinically, myxoid liposarcoma is characterized by a slow-growing, painless, deeply seated mass. In its early phase, it can be asymptomatic and is diagnosed incidentally or when it has attained significant size. Despite having low to intermediate histologic grading, this tumor shows clinically aggressive behavior in an unusual manner with very high local recurrence and distant metastasis. The metastatic pattern is also distinctive with the lungs. The pattern of metastasis is consistent with the lungs, retroperitoneum, and soft with extrapulmonary sites being the predilection sites. Although pulmonary metastasis may not be the most common, it is clinically significant and usually indicates advanced disease.

A diagnosis of myxoid liposarcoma is made based on a combination of histopathological findings and imaging studies. The preferred imaging study for local staging is a magnetic resonance imaging (MRI) scan due to its ability to delineate the extent of soft tissue involvement. Computed tomography (CT) and positron emission tomography-computed tomography (PET-CT[2]) are also helpful in assessing distant sites of metastasis. Immunohistochemistry is always helpful, while S-100 and vimentin are helpful as markers to identify the tumor.

Recurrence or metastasis may be difficult to identify in myxoid liposarcoma due to its indolent presentation and lack of specific symptoms. Furthermore, an initial surgery may obfuscate the clinician's ability to notice a recurrence. The disease may initially show benign lesions which leads to a potential delay in required oncologic treatment. As is typical with recurrences, they occur after several years of disease-free status, where appropriate long-term follow-up is an important recommendation.

The standard treatment is wide local excision with negative margins. In cases of non-resectable disease or metastatic disease, systemic chemotherapy is the first line of treatment. Chemotherapeutic agents such as gemcitabine, docetaxel, and others have been reported to be effective, especially in those with metastatic disease who are not amenable to curative resection. Radiation therapy may be used as an adjunctive treatment, specifically for non-resectable malignancies, or to potentially reduce the risk of local recurrence after resection.

2. CASE REPORT

A 60-year-old male patient was admitted to the department of surgical oncology with swelling in the right calf area that had been present for three days. The swelling had a recurrent pattern. The patient was not experiencing any related symptoms like pain, paresthesia, color change, or loss of motor function in the involved limb. Interestingly, he had previously had surgical removal of such swellings from the same location three times in 2005, 2010, and 2014, although no thorough oncological workup was done during those instances.

On general examination, the patient was afebrile, and his blood pressure was 130/90 mmHg, pulse rate 92 beats per minute, respiratory rate 20 breaths per minute, temperature 98.1°F, and oxygen saturation (SpO2) 98%. Cardiovascular and neurological exams were normal. He is a chronic diabetic and was on oral anti diabetic treatment in the form of Metformin 1000 mg, Acarbose 50 mg, and Glimepiride 2 mg, all twice daily.

Diagnostic assessment

Investigation	Result	Interpretation
MRI Right Leg (03/06/2024)	Multiple well-defined soft tissue masses are present in the posterior and posteromedial areas of the right knee, involving the tibial marrow.	This suggests a myxoid tumor.
Biopsy (04/06/2024)	Pleomorphic spindle cells appear in a myxoid background with a branching vascular pattern.	Indicative of malignant neoplasm, no necrosis or vascular/perineural invasion
CECT Chest (05/06/2024)	Multiple hypodense, poorly enhancing soft tissue nodules are found in both lungs.	This indicates pulmonary metastases, with the largest nodule located in the left upper lobe.
Immunohistochemistry	S-100 and Vimentin positive; MDM2, CD99, and CD34 are negative; Ki-67 index is 9%.	This confirms the diagnosis of low-grade myxoid liposarcoma.
PET CT (10/06/2024)	Metabolically active nodular lesions are seen in the posterior and posteromedial areas of the right knee and leg, with invasion of subcutaneous, muscular, and cortical tibial regions.	This confirms recurrence and metastatic progression.

Laboratory Investigations:

Test Category	Parameter	Value	Normal Range
Electrolytes	Sodium	136 mmol/L	135–145 mmol/L
	Potassium	4.0 mmol/L	3.5–5.0 mmol/L
	Magnesium	1.4 mg/dL	1.5–2.5 mg/dL
Renal Function Test	Blood Urea	28 mg/dL	10–40 mg/dL
	Serum Creatinine	0.9 mg/dL	0.6–1.3 mg/dL
Liver Function Test	Total Bilirubin	0.8 mg/dL	0.2–1.2 mg/dL
	SGOT (AST)	32 IU/L	5–40 IU/L
	SGPT (ALT)	30 IU/L	7–56 IU/L
Hematology	Hemoglobin	13.6 g/dL	13.5–17.5 g/dL (male)
	Total Leukocyte Count	7900 cells/cumm	4000–11000 cells/cumm
	Platelet Count	2.1 lakh/cumm	1.5–4.5 lakh/cumm
Inflammatory Markers	ESR	14 mm/hr	<20 mm/hr (male)
Others	Blood Glucose (Fasting)	112 mg/dL	70–100 mg/dL

3. DISCUSSION

Myxoid liposarcoma (MLS) is a rare variant of liposarcoma, accounting for approximately 30–40% of all liposarcomas and typically occurring in adults between the third and sixth decades of life. It exhibits a characteristic histopathological pattern, featuring a myxoid matrix, an arborizing capillary arrangement, and monotonous non-lipogenic mesenchymal cells, typically associated with the FUS-DDIT3 fusion gene resulting from the t(12;16)(q13;p11) translocation is considered a diagnostic hallmark of MLS (Antonescu et al.)[1].

Although histologically classified as low to intermediate grade (according to the FNCLCC Grade 1 in this instance), MLS is characterized by its unpredictable nature, including a high incidence of local recurrence and distant metastasis, particularly to extrapulmonary locations such as the lung, retroperitoneum, and soft tissues. Recurrent disease was noted in our patient following prior surgeries in 2005, 2010, and 2014, without formal oncologic follow-up, highlighting the insidious, progressive behavior of the disease and the danger of underdiagnosis.

In our case, recurrent disease was diagnosed with the (MRI, PET-CT), biopsy, and immunohistochemical staining. The tumor was S-100 and vimentin positive and MDM2, CD99, and CD34 negative—markers usually positive in other soft tissue tumors. A Ki-67 index of 9% also indicated a low proliferative activity, but the finding of pulmonary metastases on CECT and PET-CT established progression at the systemic level.

In contrast to most other soft tissue sarcomas metastasizing to the lungs in a hematogenous fashion, MLS has a periphasic preference for fat-containing tissues, though the lungs are also an important metastatic location. Literatures indicate that such metastasis can be possible even years after initial therapy, and this is a reflection of the utmost significance of lifetime surveillance and interval imaging, especially in recurrence.

The patient was started on chemotherapy with gemcitabine and docetaxel, a regimen proven to be clinically effective in metastatic soft tissue sarcomas that are refractory to initial therapy (Chung et al.[2]). The multidisciplinary strategy—combining oncology, pathology, radiology, and pharmacologic support—was essential to the treatment of this advanced case. Like SCORTEN scoring in Stevens–Johnson syndrome in order to predict prognosis, prompt imaging and grading in MLS are essential to therapeutic planning and prediction of outcome.

This case highlights the paradoxically aggressive nature of myxoid liposarcoma, even in its histologic low-grade form. Silent recurrence and late risk for metastasis necessitate increased clinical suspicion and vigorous systemic treatment when surgical options are not favorable. Early detection, oncologic assessment, and proper systemic treatment are mandatory to minimize morbidity and enhance survival.

4. FOLLOW-UP AND OUTCOME

Following the start of chemotherapy with gemcitabine and docetaxel, the patient responded well to the first cycle and had no immediate side effects. He was discharged in stable condition and received instructions for outpatient follow-up and ongoing chemotherapy sessions under the care of an oncologist. The patient and caregivers learned about dietary needs, preventing infections, taking medications as prescribed, and the importance of regular check-ups to monitor treatment response and detect complications early.

During the last follow-up, the patient remained clinically stable. Further evaluations, including repeat imaging and clinical assessments, will be done after the first few cycles of chemotherapy to check how well the treatment is working and to track disease progression. Early signs of a positive response are encouraging and offer a chance for continued management focused on controlling the disease and maintaining quality of life.

5. CONCLUSION

This case shows the aggressive nature of myxoid liposarcoma, even though it is classified as low-grade histologically. Recurrent disease and distant metastases, especially to the lungs, can happen years after the initial diagnosis and surgery. Because of this, long-term follow-up with proper imaging and tissue evaluation is crucial. Using various diagnostic tools and a team-based treatment approach, including chemotherapy with drugs like gemcitabine and docetaxel, is important for improving clinical results. Identifying recurrence and systemic involvement early is essential for starting timely treatment and maintaining the patient's quality of life. This case underscores the need for heightened clinical awareness and the significance of management strategies for patients with soft tissue sarcomas.

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