CASE REPORT

Multicentric Myxoid Liposarcoma; a Case Report and Literature Review

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Abstract

Liposarcoma is a common malignant soft tissue tumor, accounting for 10% to 16% of all sarcomas. Multicentric myxoid liposarcoma is an uncommon condition. Differentiation between several primary tumors and metastasis of a single liposarcoma represents the main difficulty in diagnosis. A 47-years old woman presented with right thigh myxoid liposarcoma and underwent wide margin tumor resection. Other investigations like CXR, abdominal and pelvic CT-scans were negative. After 18 months another myxoid liposarma was found in her ipsi-lateral breast without any evidence of other organs metastasis.

Second lesion location, time between two presentation and cytogenetic differences are accepted criteria to site a sarcoma in multicentric category, but in myxoid liposarcoma these can be explained by the special features inherent to this tumor.

Key words: Liposarcoma, Multicentric, Myxoid

Introduction

Liposarcoma is a common malignant soft tissue tumor, accounting for 10% to 16% of all sarcomas. It typically affects patients between the fifth and seventh decade of life, and usually develops in the extremities or retroperitoneum (1).

Myxoid liposarcoma is a clinicopathologically and genetically distinct type characterized by its common occurrence in young adults, its location in the thigh, and the presence of at (12;16) translocation (2).

By definition multicentric liposarcoma is a lesion that develops in any of the typical locations of primary liposarcoma, such as the thigh, retroperitoneum, arm or pleura, without metastasis to conventional areas (lung, liver, bone), and having a differentiated histologic type (3)

Multicentric liposarcoma is an uncommon condition with a controversial diagnosis. Differentiation between several primary tumors and metastasis of a single liposarcoma represents the main difficulty in diagnosis.

Case report

A 46-years old woman presented with a history of right thigh pain of one month duration and a palpable mass at the posterior of right thigh from 6 months ago

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to our orthopedic tumor clinic at Imam Reza hospital, Mashhad, Iran. Conventional x-rays were negative. In her MR images we found encapsulated subcutaneous soft tissue tumor which was septated and noninfiltrating (Figure 1). On T1-weighted sequences there were lacy, amorphous foci of high signal within a low signal mass. These foci are believed to represent fat within the tumor, and its heterogeneous pattern suggested a malignant liposarcoma which was proved with core needle biopsy.

liposarcoma which was proved with core needle biopsy. Other investigations like CXR and double contrast abdominal and pelvic CT-scans were negative. She underwent wide margin tumor resection. The pathologic report was myxoid liposarcoma (Figure 2). The tumor margins were free of tumor.

There was not any local recurrence or metastases on her 17 months follow up. But on the eighteenth months she found a mass in her right breast. She referred to a breast surgeon. After evaluation by chest CT scan, she was scheduled for excisional biopsy (Figure 3).

Pathologic report was myxoid liposarcoma (Figure 4). The tumor margins were free of tumor (Figure 5).

Discussion

The first description of multicentric liposarcoma



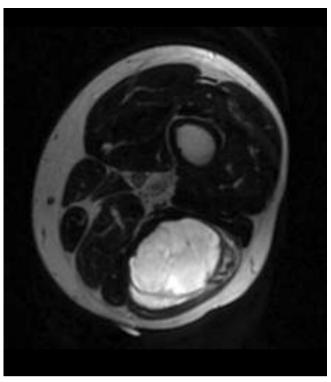


Figure 1. T1-weighted MRI of thigh, axial view.

was made in 1943; since that time, less than 50 cases have been described (4). The diagnosis of multicentric liposarcoma is not well established, the main problem being differentiation between multiple primary lesions and metastasis of a single tumor. One point we can use to define multicentric tumor is the presence of tumors in areas where metastasis does not usually occur. It is important to be familiar with the pattern of extension of liposarcoma. The most common site of metastasis is to the lung, but retroperitoneum, mesentery, bone and



Figure 3. Chest CT-scan. Axial image, Right sided breast mass.

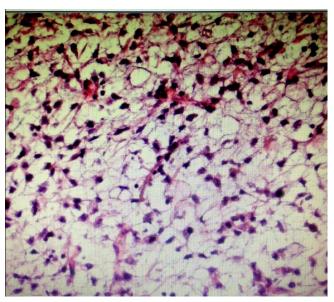


Figure 2. Myxoid liposarcoma of thigh, uni and multi-vacuolar lipoblasts. H/E 400X.

soft tissue of the trunk and glutei are other regions for myxoid type metastasis(1). Thus, when investigating extention of myxoid liposarcoma, the retroperitoneum and trunk should be evaluated in addition to the thorax, and the presence of sarcomatosis lesions in the extremities should be ruled out.

Advances in cytogenetic technique have enabled identification of the characteristic markers for liposarcoma, specially t(12;16)(q13;p11) translocation, which is seen in the myxoid and round cell subtypes(5). In our patient second lesion was found 18 months later

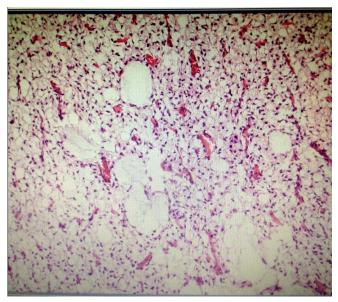


Figure 4. Myxoid liposarcoma of the right breast, many lipoblasts and curvilinear vessels with mucoid microcysts. H/E 100X.

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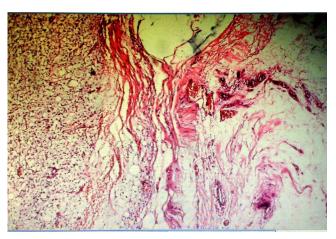


Figure 5. Sharp and free margin of breast tumor. H/E, 40X.

in the typical location of primary liposarcoma. Breast is not a conventional location for myxoid liposarcoma metastasis, especially when we couldn't find any other lesion in thorax, retroperitoneum or trunk. Moreover it's not logical to assume her breast as a primary location because detecting a lesion in the breast is easier and more accelerated than detecting its metastasis in the extremities. The third suggestive point is the time between first and second presentation, in contrast to the tendency of sarcomas to produce metastasis during the first 6 months after removal of primary tumor(6).

The cytogenetic techniques can be useful to recognize or rule out a colonal relationship between two lesions, but unfortunately it is not possible for us to do it in our centre. Furthermore the myxoid and round cell liposarcoma constitute a spectrum of the same condition and various histological forms of liposarcoma may be

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found in metastasis of a tumor (7).

Conclusion

To perform appropriate treatment we need to differentiate between several primary tumors and metastasis of single myxoid liposarcoma. Cytogenetic difference is not a characteristic clue for multicentric tumors because the myxoid and round cell liposarcoma constitute a spectrum of the same condition and various histological forms of liposarcoma may be found in metastasis.

Moreover in literatures few cases could be found with metastasis several years after primary myxoid liposarcoma in typical primary locations (1). So we need to define more criteria beside cytogenetic, time and location, to put a case in multicentric myxoid liposarcoma category.

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