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Atypical Imaging Characteristics of Metastatic Myxoid Liposarcoma

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ABSTRACT

Myxoid liposarcoma accounts for 30% to 50% of all liposarcomas, which is the second most common soft tissue tumor. It is usually painless slowly growing mass in the deep-seated soft tissue of the lower extremities or the peritoneum, with a strong metastatic predilection to the spine. Metastatic myxoid liposarcoma typically shows low signal intensity on T1 sequences with hyper-intensities on T2 sequences. In contrast to vertebral hemangioma, it shows increased signal intensity in both T1 and T2 sequences. We demonstrate 2 cases of metastatic myxoid liposarcoma with atypical MRI pattern. According to the histopathology and the clinical signs, we believe that increasing round cell cellularity and fat content is correlated to the increased T1 signal intensity and clinical aggressiveness. In conclusion, the atypical MRI pattern for vertebral metastatic lesions in patients with myxoid liposarcoma should be taken into consideration to prevent the delay in diagnosis and standard of care.

Keywords: Vertebral metastasis, Vertebral hemangioma, MRI, Atypical, High T1-signal

INTRODUCTION

Liposarcoma is the second most frequent type of soft tissue sarcoma, representing 9-18% of the total and exceeded only by pleomorphic undifferentiated sarcoma (PUS) (24%) [1,2]. According to WHO classification, myxoid liposarcoma is one of the four subtypes including "well differentiated, dedifferentiated and pleomorphic". Myxoid Liposarcoma accounts for 30%-50% of all liposarcomas [3].

Myxoid liposarcoma occurs predominantly in the lower limbs of young adults and it usually appears a decade earlier than other subtypes which are more commonly diagnosed in the 5th to 6th decade of life [4,5].

Myxoid liposarcoma presents mainly with an extra-pulmonary spread such as retroperitoneum, extremities, axilla and bone and this is what makes myxoid liposarcoma unique as compared to other subtypes [6]. Schwab, et al., reported skeletal metastasis as the most common site of metastatic with a predilection to the spine. In their study, 17% of patients developed bone metastasis accounting for more than half (56%) of all metastatic events [7]. This significant tendency to spread depends on the amount of hypercellularity/round cell component, as patients with >5% round cell component had shown an increased rate of metastasis than patient with <5% round cell component [8].

Previously, pathologists used to group myxoid and round liposarcoma into 2 distinct histopathological subtypes. However, the WHO classification of tumors of soft tissue has established now that both of them are representing one histopathological entity as both are associated with the exact same gene fusion FUS-DDIT3 or EWSR1-DDIT3 [9,10]. Now it is graded as low, intermediate or high-grade according to the degree of cellularity [9].

Microscopically, myxoid liposarcomas are composed of well-demarcated lobules of the myxoid matrix, characteristic plexiform capillary network of a stemmed vessel and a tree-like branching smaller ones 'arborizing', primitive

mesenchymal cells and signet ring lipoblasts in various stages of differentiation at the periphery of the lobule [11,12]. In addition to the round cell component which is arranged in solid sheets of round primitive cells with higher mitotic figures and necrosis [4].

The gross pathology of liposarcoma is variable and it is usually a reflection of the histological pattern [13]. Myxoid liposarcoma is a large well-circumscribed mass characterized by its nodularity and varies according to the percentage of the intermingled myxoid and round cell component. They appear as a gelatinous background matrix with interspersed focal areas of round cells containing white opaque nodules. This dual histological configuration is believed to be the main determinant of its grading as the upgrading from low to high-grade [14].

Clinical Features

The most common initial presentation of myxoid liposarcoma is a painless slow-growing soft tissue swelling, usually soft and non-tender, which gives it a misleading appearance of a benign lesion. Only 10% to 15% report pain and tenderness. It has a very strong predilection to the deep-seated soft tissue of extremities particularly the mid-thigh and popliteal region followed by retroperitoneum [15].

MRI Characteristics

Diagnosis of the specific cause for vertebral lesions can be challenging, especially in patients with established diagnosis of cancer. Description of a lesion as either benign or malignant, primary or secondary is crucial for accurate staging and optimal treatment [16,17].

MRI is a very sensitive modality for localizing skeletal lesions and metastasis with a 95% sensitivity and a 90% specificity [18].

Myxoid liposarcoma imaging findings on MRI depend on several factors; the most important one is the fat content of the tumor. Other variables include cellularity, vascularity, myxoid material content, and the presence or absence of necrosis [12].

Metastatic myxoid liposarcoma typically shows low signal intensity on T1 weighted sequences due to high water content, accompanied by lacy, globular, amorphous or linear hyperintense foci that are believed to represent the fat content of the tumor. In addition to marked T2 hyperintensity of the myxoid component, it also reveals long homogenous T1 and T2 relaxation time similar to water due to the presence of the myxoid matrix [4,19] (Figure 1).

Figure 1 A: A 53-year-old man with metastatic myxoid liposarcoma. Sagittal T1W MRI demonstrated a typical imaging characteristic with low T1 signal (Red arrow). B: A 53-year-old man with metastatic myxoid liposarcoma. Sagittal T2W MRI demonstrated a typical imaging characteristic with increased T2 signal (Red arrow)

Fat-containing lesions like vertebral hemangioma have classical MRI imaging features are characterized by the increased signal intensity in both T1 and T2 weighted images in correlation to increased fatty content [20,21] (Figure 2).

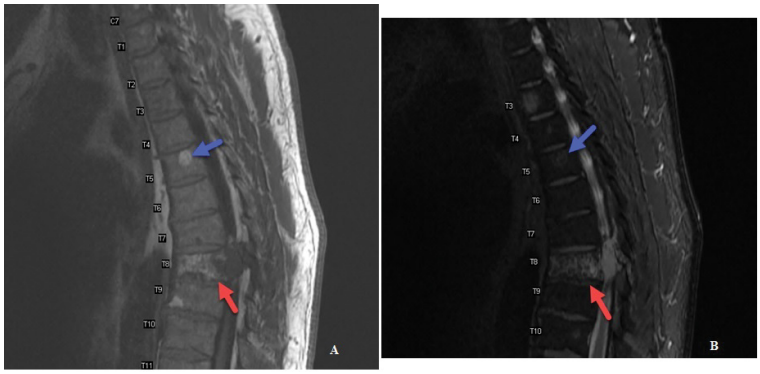

Figure 2 A: A 61-year-old male with hemangioma of the thoracic spine. Sagittal T1W MRI demonstrated two hemangiomata at fifth and eighth thoracic vertebrae. Benign hemangioma at T5 shows increased T1 signal (Blue arrow). Aggressive epithelioid hemangioma at T8 with epidural extension and mass effect on the cord also shows increased T1 signal (Red arrow). B: A 61-year-old male with hemangioma of the thoracic spine. Sagittal T2W Fat Saturation MRI demonstrated two hemangiomata at fifth and eighth thoracic vertebrae. Benign hemangioma at T5 shows increased T2 signal (Blue arrow). Aggressive epithelioid hemangioma at T8 with epidural extension and mass effect on the cord also shows increased T2 signal (Red arrow)

DISCUSSION

While reviewing 50 cases with metastatic myxoid liposarcoma during a retrospective analysis, working on an update about the incidence of spinal metastasis in relation to other sites, we found that 48 patients demonstrated the classic MRI characteristics. After reviewing the pathology reports, we found that this is corresponded to tumors having no more than 15% hypercellularity. Osseous metastasis in 2 patients showed a unique pattern demonstrating increased signal on both T1 and T2 weighted images. One case had additional metastatic lesions, which showed typical MRI characteristics of low T1 signal, increased T2 but with interval increase in size and T1 signal intensity on follow up examination (Figures 3 and 4).

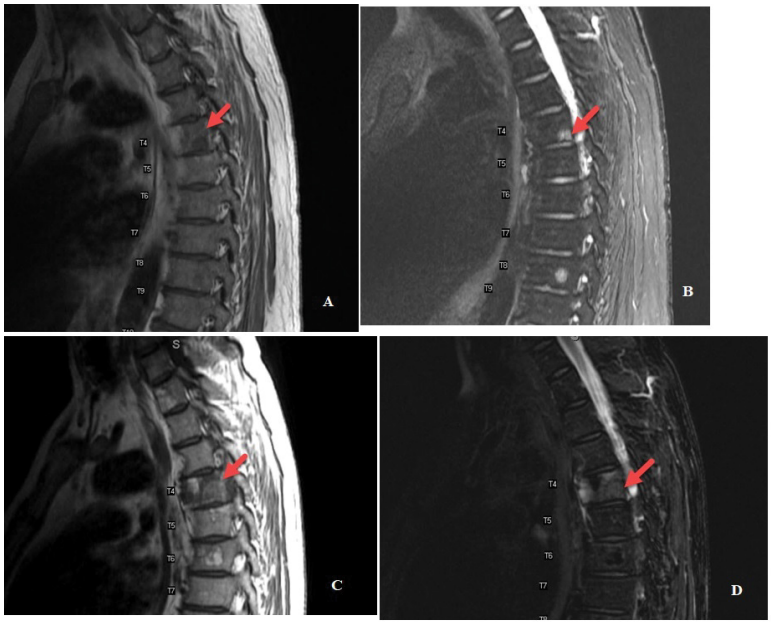

Figure 3 A: A 49-year-old man with metastatic myxoid liposarcoma. Sagittal T1W MRI demonstrated a low T1 signal lesion (Red arrow. B: A 49-year-old man with metastatic myxoid liposarcoma. Sagittal T2W MRI demonstrated an increased T2 signal lesion (Red arrow). C: A 49-year-old man with metastatic myxoid liposarcoma. 9 months later, Sagittal T1W MRI without contrast demonstrating an increase in T1 signal intensity with interval increase in size (Red arrow). This is an atypical pattern for a metastatic myxoid liposarcoma. D: A 49-year-old man with metastatic myxoid liposarcoma. 9 months later, Sagittal T2W MRI without contrast demonstrating an increase in T2 signal intensity with interval increase in size (Red arrow)

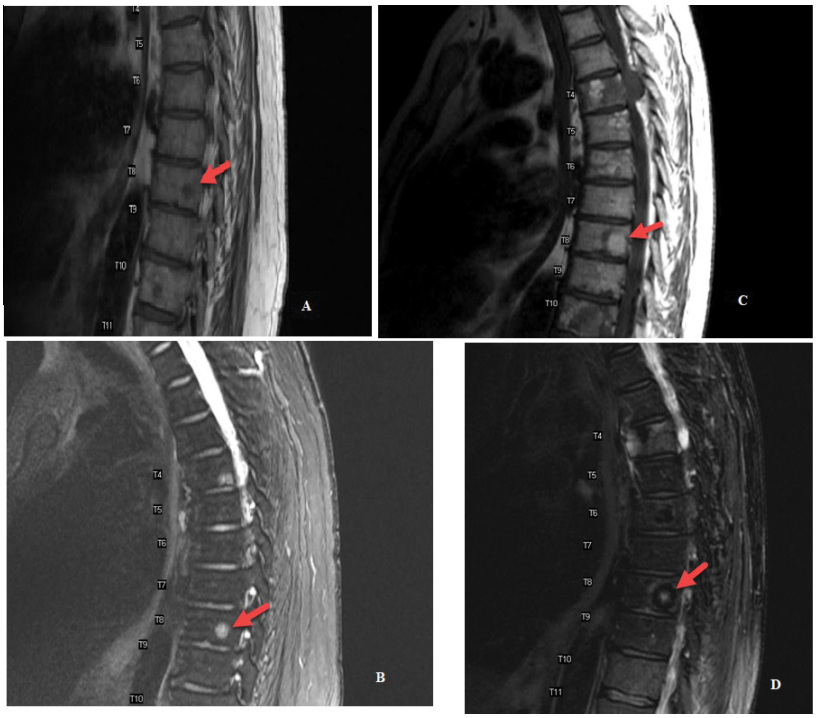

Figure 4 A: A 49-year-old man with metastatic myxoid liposarcoma. Sagittal T1W MRI without contrast demonstrating a classic decrease in signal intensity correlates to metastatic myxoid liposarcoma lesion at T8 (Red arrow). B: A 49-year-old man with metastatic myxoid liposarcoma. Sagittal T2W MRI without contrast demonstrating a classic increase in signal intensity correlates to metastatic myxoid liposarcoma lesion at T8 (Red arrow). C: A 49-year-old man with metastatic myxoid liposarcoma. 9 months later, Sagittal T1W MRI without contrast demonstrating an increase in T1 signal intensity with interval increase in size (Red arrow). This is an atypical pattern for a metastatic myxoid liposarcoma. D: A 49-year-old man with metastatic myxoid liposarcoma. 9 months later, Sagittal T2W MRI without contrast demonstrating an increase in T2 signal intensity with interval increase in size (Red arrow)

Other metastases also showed an atypical appearance from the start and then showed even more increased signal and size consistent with increased clinical aggressiveness (Figure 5).

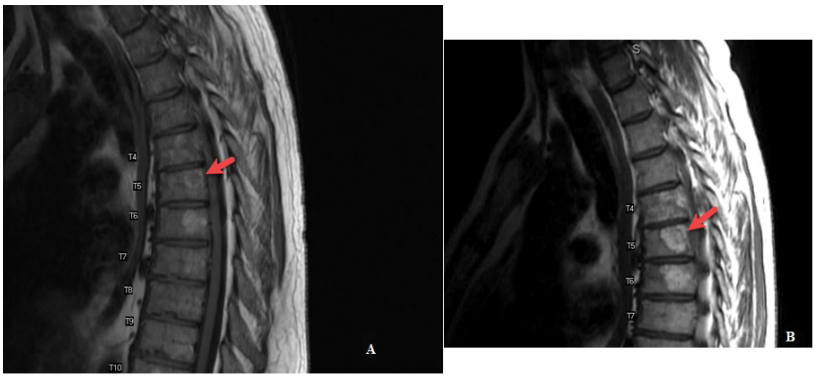

Figure 5 A: A 49-year-old man with metastatic myxoid liposarcoma. Sagittal T1W MRI without contrast demonstrating a lesion with increased signal intensity at T5 (Red arrow).This is an atypical pattern for a metastatic myxoid liposarcoma. B: A 49-year-old man with metastatic myxoid liposarcoma. 9 months later, Sagittal T1W MRI without contrast demonstrating an increase in T1 signal intensity with interval increase in size (Red arrow)

Follow up scans shows new metastatic lesions, which have dedifferentiated, to a more clinically aggressive state (Figure 6).

Figure 6 A 49-year-old men with metastatic myxoid liposarcoma. Sagittal T1W MRI without contrast demonstrating a new lesion at T6 with an increase in T1 signals intensity (Red arrow). This is an atypical pattern for a metastatic myxoid liposarcoma

The other case had a free interval with no metastasis for 2 years, with MRI, follow up we discovered multiple vertebral metastatic lesions some of them demonstrating the atypical pattern while others follow the typical MRI characteristics (Figure 7).

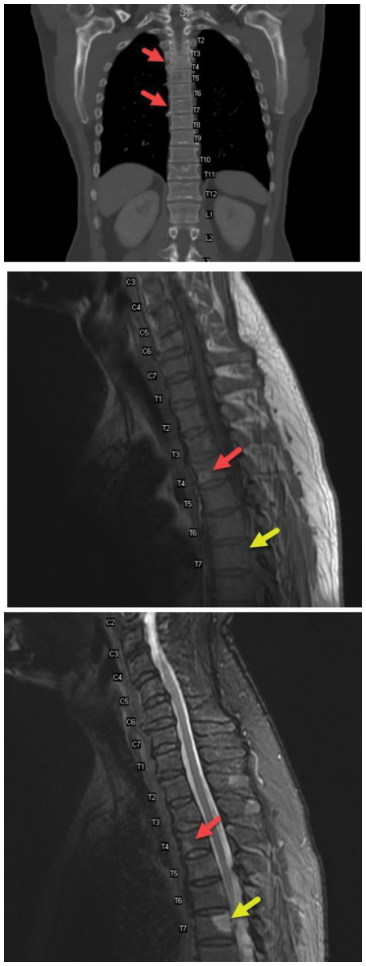

Figure 7 A: A 48-year-old man with myxoid liposarcoma with no vertebral metastasis. Coronal Chest-Abdomen-Pelvis CT scan shows no vertebral metastasis at T4 or T7 (2 red arrows). B: A 48-year-old man with metastatic myxoid liposarcoma. Sagittal T1W MRI without contrast demonstrating a lesion at T4 with increased T1 signal intensity (Red arrow) which shows an atypical pattern. Another lesion at T7 with intermediate to hypointense T1 signal (Yellow arrow) follows the typical pattern. C: A 48-year-old man with metastatic myxoid liposarcoma. Sagittal T1W MRI without contrast demonstrating two lesions at T4 (Red arrow) and T7 (Yellow arrow) both showing increased T2 signal intensity

According to the pathology slides, we believe that the increased signal on T1 weight images is secondary to increased cellularity and fat content in the metastasis (Figure 8). These characteristics are important to recognize as they reveal the evolving histopathology (increased cellularity) associated with tumor progression (60% increase in cellularity compared to 15% or less in the other 48 cases). In addition, distinguishing these from benign fat containing spinal osseous lesions such as vertebral hemangioma is critical to accurate staging and treatment planning.

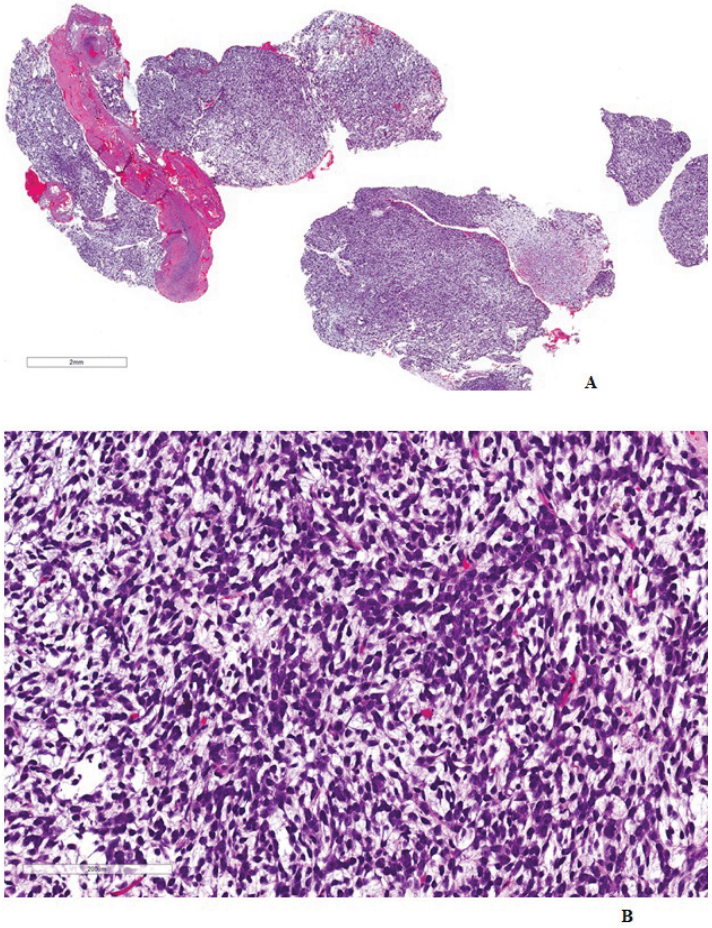

Figure 8 A: A 49-year-old man with metastatic myxoid liposarcoma. Histopathology slide (Low power) for an excisional biopsy from T4 lamina. B: A 49-year-old man with metastatic myxoid liposarcoma. H and E stained slide (200X) for an excisional biopsy from T4 lamina demonstrates increased cellularity and less myxoid stroma than often encountered

CONCLUSION

In conclusion, the atypical MRI pattern for vertebral metastatic lesions in patients with myxoid liposarcoma should be taken into consideration to prevent the delay in diagnosis and standard of care.

DECLARATIONS

Conflict of Interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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