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Myxoid-round cell liposarcoma: MRI appearance after radiation therapy and relationship to response

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ABSTRACT

Liposarcomas are classified into 4 different subtypes, with the myxoid-round cell variant demonstrating increased morbidity and metastatic potential dependent on cell composition. Unique to sarcomas, the myxoid-round cell liposarcoma is remarkably sensitive to radiation therapy in the pretreatment setting, owing to the tumor morphology and vascular distribution.

Herein we report a case of myxoid-round cell liposarcoma within the deep soft tissues of the thigh of an 81-year-old male with excellent neoadjuvant response to radiation. We briefly review treatment options.

Introduction

Liposarcomas are currently classified into 4 different subtypes based on histologic or genetic analysis according to the World Health Organization (WHO), including well-differentiated, dedifferentiated, myxoid, and pleomorphic [1]. The round cell subtype is now representative of a higher grade of the myxoid variant.

The myxoid variant accounts for one-third to one-half of all liposarcomas, commonly occurring in a younger demographic with peak incidence in the fifth decade, in comparison to the dedifferentiated and well-differentiated subtypes which demonstrate peak incidence in the seventh decade. The most common sites of tumor involvement include the lower extremities with a predilection for the medial thigh and popliteal area [2,3]. The myxoid variant encompasses a continuum of lesions, ranging from predominantly well-differentiated myxoid

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areas to predominantly round cell type with poorly differentiated lipoblasts. The higher percentage of round cell areas within the tumor results in increased metastatic potential and mortality [3–5].

Magnetic resonance imaging (MRI) features of myxoid-round cell liposarcomas are diverse, owing to the continuum of differing myxoid and small cell components. Imaging appearance of typical myxoid-round cell tumors include low signal intensity on T1 given high water content and high signal intensity on T2-weighted sequences of the myxoid element [3–5]. Additional imaging characteristics include encapsulation, lobulated fat, and diffuse, globular or nodular intense enhancement. T1-hyperintense fatty septa can help differentiate these tumors from cystic lesions [5,6].

With regard to treatment, sarcomas are generally considered radiation resistant [7]. Local control and preservation of functional outcome is the goal for preoperative radiation therapy [7]. Currently, use of radiotherapy (RT) is accepted in both the pre- and postoperative settings for local control after a National Cancer Institute of Canada clinical trial demonstrated comparable results [7]. Myxoid liposarcoma is unusual in that it is shown to be highly sensitive to radiation therapy and can be treated successfully with both adjuvant and neoadjuvant radiation therapy. The mechanism of tumor response to RT is unknown; however, it is thought to be related to the distinctive vasculature of the tumor [7].

We present a case of a myxoid round cell tumor with excellent neoadjuvant radiation therapy response on MRI with histopathologic correlation.

Case report

An 81-year-old male presented with an enlarging left thigh mass and pain. Initial MRI demonstrated a $7 \times 7 \times 12$-cm encapsulated intermuscular mass within the left thigh deep to the sartorius muscle and superficial femoral neurovascular bundle (Fig. 1). Typical imaging characteristics of myxoid liposarcoma were seen, including T1-hyperintense fatty lobular regions comprising approximately 50% of the mass, marked T2 hyperintensity of nonfatty components, and postcontrast avid nodular enhancement predominantly of the nonfatty components. Ultrasound-guided biopsy yielded a tissue diagnosis of liposarcoma, myxoid type with round cell components, grade 2 (stage IIb). Patient subsequently underwent neoadjuvant radiation therapy with 50 Gy in 2 Gy/fraction using an intensity-modulated radiotherapy plan.

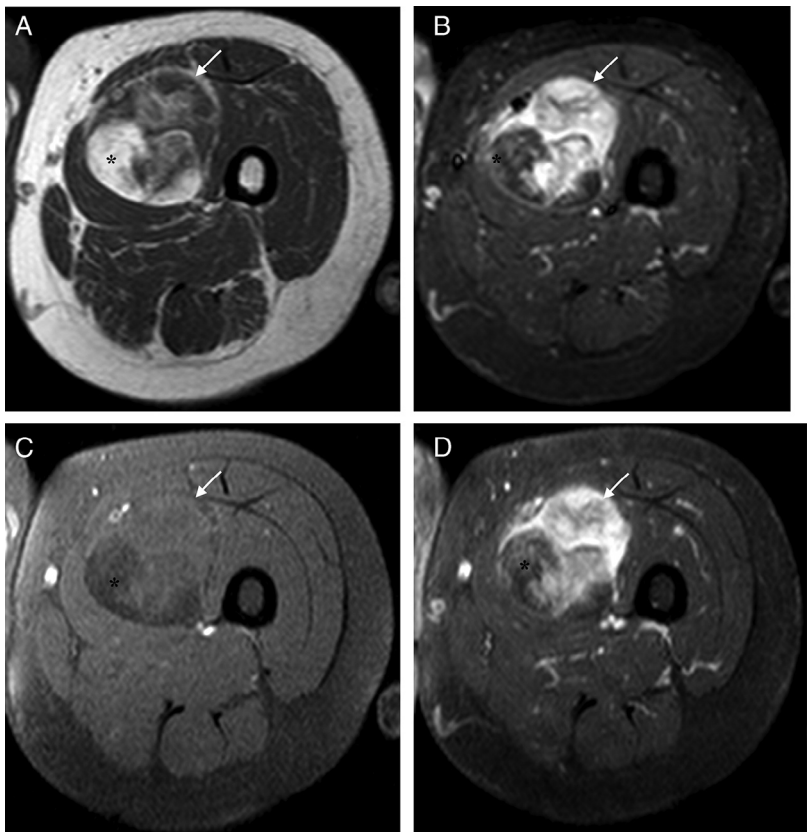

Fig. 1 – An 81-year-old male with myxoid-round cell liposarcoma. Initial magnetic resonance imaging (MRI) demonstrated a $7 \times 7 \times 12$-cm well-circumscribed intermuscular mass within the left thigh deep to the sartorius muscle and superficial femoral neurovascular bundle with resultant mass effect upon the adductor and quadriceps musculature. (A) Axial T1, (B) Short Tau Inversion Recovery, and (C) T1-weighted fat saturated images show a T1-hyperintense fatty component (*) with fat saturation and a nonfatty, T2-hyperintense, T1-hypointense nodular component (arrows). (D) Axial T1-weighted fat saturated postcontrast image demonstrates avid enhancement primarily of the nonfatty components.

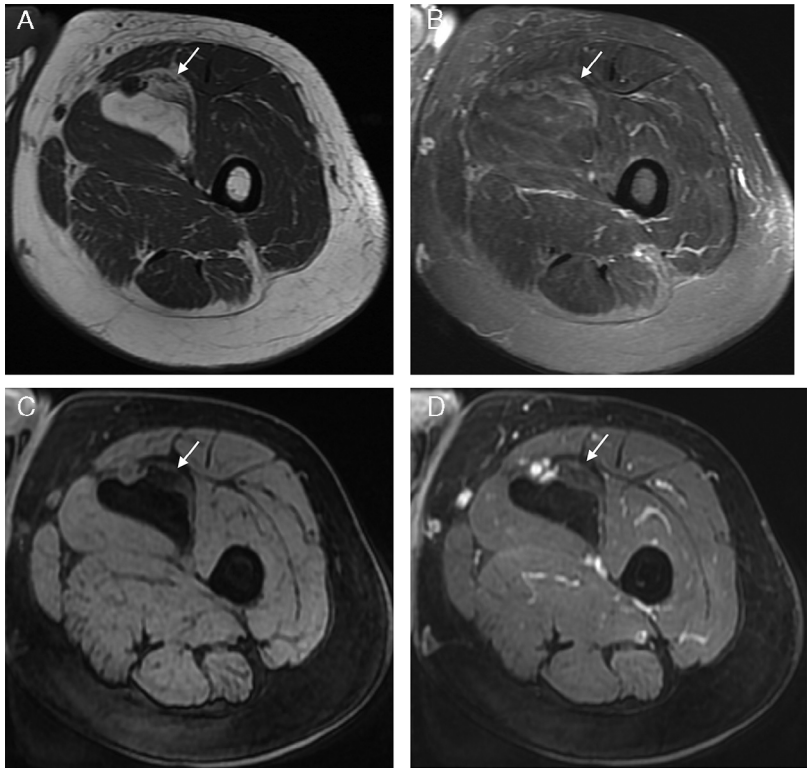

Fig. 2 – Postradiotherapy magnetic resonance imaging (MRI) of myxoid-round cell liposarcoma. (A) T1-weighted image demonstrates decreased size of the tumor with residual mass composed of mainly fatty tissue. (B) T2-weighted fat saturated (FS) image reveals minimal T2-hyperintensity within the mass anterolaterally (arrows) likely representing residual myxoid tissue. (C) T1 fs gradient echo precontrast and (D) T1 fs gradient echo postcontrast image demonstrates minimal residual enhancement.

Postradiation therapy MRI showed a decreased size of the mass, measuring $5 \times 5 \times 7$ cm, with predominantly T1-hyperintense fatty regions and only minimal residual enhancement (Fig. 2). There were minimal T2-hyperintense areas corresponding to likely residual myxoid elements. Tumor resection was subsequently performed, with pathologic examination yielding a 6-cm mesenchymal mass (Fig. 3) with rare foci of residual myxoid liposarcoma (<10% viable tumor exhibiting maturation of tumor cells) in a background of extensive treatment effect. Round cell morphology characterizing the original biopsy was not identified. Post-treatment tumor was classified as grade I (Fig. 4).

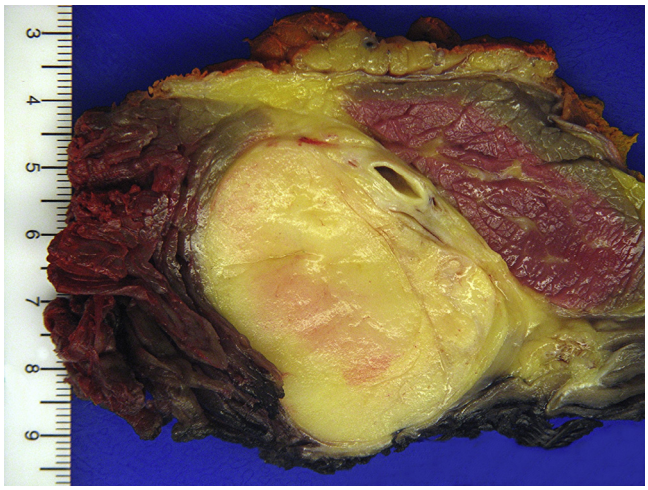

Fig. 3 – Gross photo after surgical resection demonstrates a well-circumscribed yellow mass abutting the sartorius muscle and the superficial femoral artery.

Discussion

Myxoid liposarcoma and round cell liposarcoma have a common chromosomal translocation t(12;16)(q13;p11) resulting in the fusion transcript of the *TLS* and *CHOP* genes and are now recognized as a continuum of the same entity [5,6,8]. MRI features of myxoid-round cell liposarcoma have prognostic significance [6]. According to Tateishi et al., imaging characteristics associated with improved overall survival and lower grade tumors include thin septa or tumor capsule. Pronounced enhancement was the most significant prognostic factor with regard to adverse outcomes [6].

Previous studies have illustrated that areas with greater than 5% nonfatty, nonmyxoid, enhancing components are associated with higher grade tumors [4]. According to Tateishi et al., patterns of enhancement including pronounced enhancement mainly at the periphery of lesions and globular or nodular enhancement within the lesion center were characteristic of intermediate to high-grade tumors [6]. Previously reported

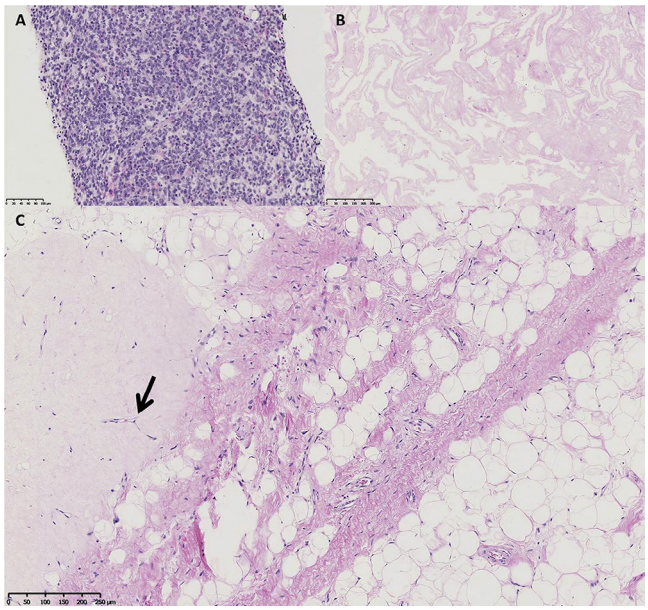

Fig. 4 – Histologic sections of the tumor. (A) Preoperative, pretreatment biopsy reveals a predominance of small round blue cells, with hyperchromatic nuclei and sparse granular cytoplasm, consistent with the round cell liposarcoma phenotype (hematoxylin-eosin stain, original magnification ×200). (B and C) Postoperative post-therapeutic excision (hematoxylin-eosin stain original magnification ×100). In (B) large areas of sclerotic fibroconnective tissue in the tumor bed are shown. In (C) note residual myxoid stroma with the characteristic delicate thin-walled curved "chicken wire" (arrow) shown on the left consistent with residual myxoid liposarcoma phenotype in approximately 10% of the residual mass, juxtaposed with mature adipose tissue shown on the right.

pathologic examinations have demonstrated round cell components to be located at the periphery of lobules, adjacent to fibrous septa extending through the tumor and surrounding large vessels [6]. Given these concordant findings, MRI enhancement pattern can potentially be used to stratify tumor grade based on round cell component.

Myxoid liposarcoma is unique with regard to its unusual pattern of metastatic spread to extra pulmonary sites including bone, retroperitoneum, paraspinal regions, and opposite extremity, with skeletal metastases reported as the most common [4,5,8]. According to Jagannathan et al., osseous metastases that are detected with ease on MRI are difficult to identify on conventional computed tomography and positron emission tomography-computed tomography secondary to lack of significant osseous destruction and decreased fluorodeoxyglucose (F18-FDG) avidity owing to the myxoid matrix. Therefore, several investigators recommend whole-body MRI for screening, especially in the setting of higher grade tumors [4].

In our case, pre-RT imaging findings demonstrated features of an intermediate to high-grade tumor, including pronounced enhancement. Post-RT findings demonstrated significant response to treatment with only minimal residual enhancement. Postsurgical pathologic findings of <10% residual tumor and absent round cell component correlate with the post-treatment enhancement pattern. This case report further illustrates the use of MRI in imaging myxoid-round cell liposarcomas before and after RT, which can provide valuable information regarding tumor grade and treatment response based on enhancement patterns.

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REFERENCES

[1] Crago AM, Dickson MA. Liposarcoma: multimodality management and future targeted therapies. Surg Oncol Clin N Am 2016;25(4):761–73. doi:10.1016/j.soc.2016.05.007. PubMed PMID: 27591497. PubMed Central PMCID: PMCPMC5010855.

[2] Goldblum JR. Liposarcoma. In: Enzinger and Weiss's soft tissue tumors. Sixth ed. Saunders; 2014, p. 484–523.

[3] Murphey MD, Arcara LK, Fanburg-Smith J. From the archives of the AFIP: imaging of musculoskeletal liposarcoma with radiologic-pathologic correlation. Radiographics 2005;25(5):1371–95. doi:10.1148/rg.255055106. PubMed PMID: 16160117.

[4] Jagannathan JP, Tirumani SH, Ramaiya NH. Imaging in soft tissue sarcomas: current updates. Surg Oncol Clin N Am 2016;25(4):645–75. doi:10.1016/j.soc.2016.05.002. PubMed PMID: 27591491.

[5] O'Regan KN, Jagannathan J, Krajewski K, Zukotynski K, Souza F, Wagner AJ, et al. Imaging of liposarcoma: classification, patterns of tumor recurrence, and response to treatment. AJR Am J Roentgenol 2011;197(1):W37–43. doi:10.2214/ajr.10.5824. PubMed PMID: 21700993.

[6] Tateishi U, Hasegawa T, Beppu Y, Kawai A, Satake M, Moriyama N. Prognostic significance of MRI findings in patients with myxoid-round cell liposarcoma. AJR Am J Roentgenol 2004;182(3):725–31. doi:10.2214/ajr.182.3.1820725. PubMed PMID: 14975976.

[7] de Vreeze RS, de Jong D, Haas RL, Stewart F, van Coevorden F. Effectiveness of radiotherapy in myxoid sarcomas is associated with a dense vascular pattern. Int J Radiat Oncol Biol Phys 2008;72(5):1480–7. doi:10.1016/j.ijrobp.2008.03.008. PubMed PMID: 18448271.

[8] Asano N, Susa M, Hosaka S, Nakayama R, Kobayashi E, Takeuchi K, et al. Metastatic patterns of myxoid/round cell liposarcoma: a review of a 25-year experience. Sarcoma 2012;2012:345161. doi:10.1155/2012/345161. PubMed PMID: 22550416. PubMed Central PMCID: PMCPMC3329861.