Case Report

rare tumors

Rare Tumors
Volume 16: 1–6

DOI: 10.1177/20363613241255668
journals.sagepub.com/home/rtu
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Enormous myxoid/round cell liposarcoma: A case report

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Abstract

Myxoid/round cell liposarcomas (MRCLPS) are a rare soft tissue sarcoma. We report the largest sarcoma in our institutional history. We discuss the patient's surgical management and treatment of the tumor and challenges given its dimensions. Several complications arose following primary resection that were managed by a multidisciplinary team. Although MRCLPS can vary in size, large MRCLPS must be treated cautiously given the potential for complications. Additionally, multidisciplinary treatment of MRCLPS is essential in diagnosing and treating these complex cases.

Keywords

Myxoid liposarcoma, soft tissue sarcoma, myxoid/round cell liposarcomas, large sarcoma, multidisciplinary

Date received: 31 March 2023; accepted: 29 April 2024

Introduction

Liposarcomas are a group of cancers that are derived from adipocytes or their precursor cells, lipoblasts and constitute 20% of soft tissue neoplasms.[1] There are several subtypes of liposarcomas outlined by the World Health Organization: atypical lipomatous tumor/well-differentiated liposarcoma, dedifferentiated liposarcoma, myxoid/round cell liposarcoma (MRCLPS), and pleomorphic liposarcoma.[2,3] MRCLS represent a morphological spectrum from low-grade myxoid liposarcoma, histologically characterized by round-to-oval mesenchymal cells and small signet ring lipoblasts, to higher-grade round cell liposarcomas, histologically characterized by progression to hypercellular and round cell components.[4] These tumors typically arise in the lower extremities, although they may rarely present in the retroperitoneum, subcutaneous tissues, and the abdominal wall.[5] In the vast majority of these cases, surgical resection with wide negative margins is the preferred treatment. However, some cases may demand additional measures, including radiation therapy, and more rarely, chemotherapy, to reduce the mass to an operable size, or to prevent local recurrence or metastasis.[6]

In this case report, we report a 51-year-old woman who was treated for the largest sarcoma in our institutional history which final diagnosis was consistent with myxoid/round cell liposarcoma (MRCLPS). We discuss the clinical course of the case, as well as the challenges associated with treatment due to its dimensions.

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Case Report

A 51-year-old female patient presented for evaluation of a mass in her left thigh that had been growing for the last 8 years. Although she experienced no pain, she did have paresthesias of her left lower extremity. On physical examination, a firm, mobile left thigh mass was identified extending from the medial left thigh, involving the skin, subcutaneous tissue, and down to the muscle. The mobile mass, approximately 40 cm, was found to be pedicled to the thigh by a 10–15 cms stalk. Magnetic resonance imaging (MRI) of the lower extremities revealed enlarged left inguinal lymph nodes, as well as a pedunculated, multi-lobulated, multiseptated T2 hyperintense heterogeneously enhancing mass arising from the left proximal thigh (Figure 1), measuring 26 × 23 × 36 cm in close proximity to the femoral vessels. These findings suggested the possibility of a large soft tissue sarcoma. A core needle biopsy was performed, returning a final diagnosis of MRCLPS with atypical features. Although neoadjuvant radiation therapy was considered, two factors led the multidisciplinary team to prefer postoperative treatment. First, the size and location of the mass would make positioning for radiation treatment difficult. Second, after a discussion with her medical oncologist, it was determined pretreatment, while beneficial in tumor reduction, would not substantially reduce the narrow communicating stalk (Figure 2(A)). After a multidisciplinary sarcoma conference, the patient underwent en bloc resection followed by delayed plastics reconstruction and adjuvant chemoradiation.

Intraoperatively, the entire femoral neurovascular bundle was discovered to be adherent to the tumor, just deep to the sartorius. Vascular surgery was consulted after resection of the superficial femoral artery and femoral vein was performed. Vascular surgery performed their reconstruction at that point (Figure 2(B)) followed by plastic surgical reconstruction of the soft tissue defect.

The mass measured 39.0 cm in greatest dimension and was 10.2 kg. Sectioning through the mass showed a multilocular mass with heterogenous appearance (Figure 2(C)) ranging from smooth, flesh-tan to yellow-tan with focal areas of hemorrhage. The tumor was soft with focal, solid area. There was no necrosis identified on gross examination. Microscopic sections showed a heterogenous morphology reflecting the gross heterogenicity. The tumor showed a predominant growth of classic morphology consisting of stellate cells with scarce mitotic activity in branching vascular network (Figure 3(A)). There were areas containing mucin pools representing pulmonary edema-like pattern (Figure 3(B)) and some areas of multivacuolated lipoblasts (Figure 3(C)). There was an area of hypercellularity composed of small round blue cells consistent with high-grade myxoid liposarcoma comprising 5% of the tumor (Figure 3(D)). Overall, the tumor was grade 2 with a focal higher grade (5%) component and stage IIIB. The resection

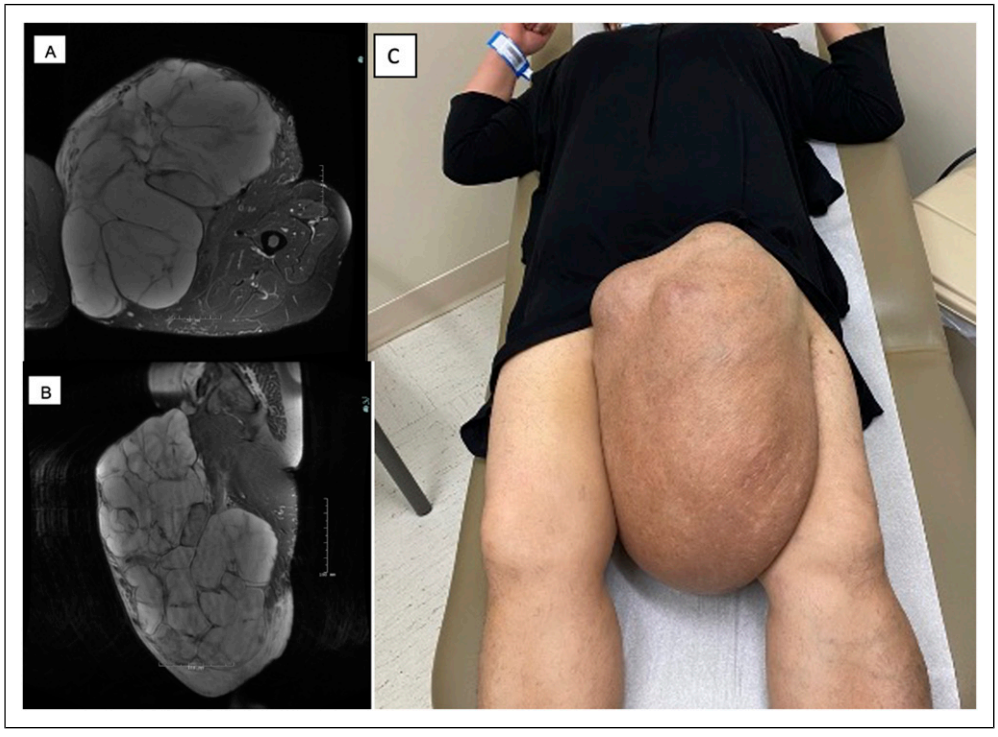

Figure 1. Axial (A) and sagittal (B) T2 MRI images demonstrating a pedunculated, multi-lobulated, multiseptated T2 hyperintense heterogeneously enhancing mass arising from the left proximal medial thigh. Clinical image of patient (C).

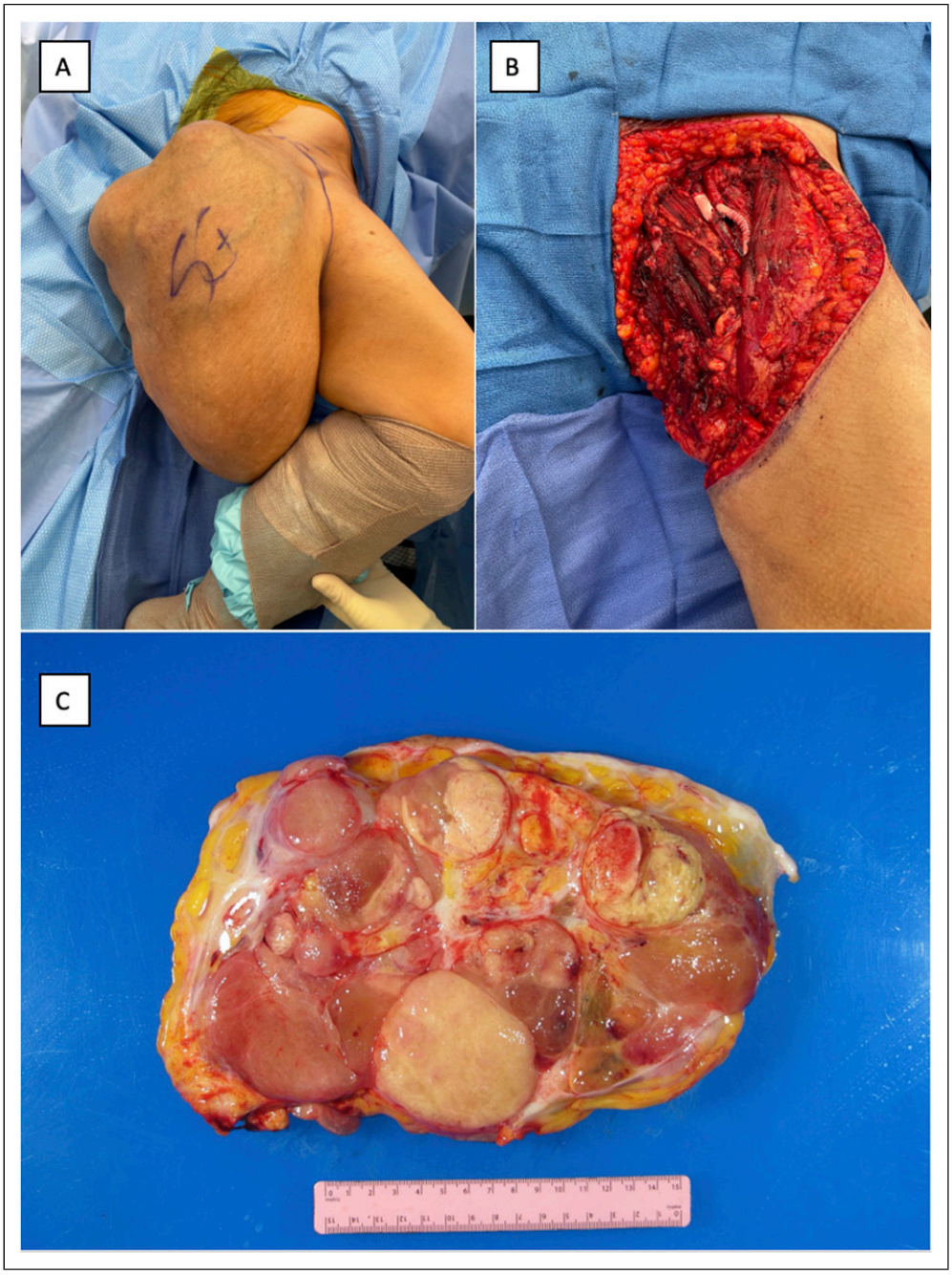

Figure 2. Preoperative image demonstrating pedicled nature of sarcoma (A). Intraoperative image demonstrating soft tissue defect with vascular repair (B). The cross section shows multilobulated pink tan to yellow, fleshy cut surface (C).

margins were negative, and the 3 lymph nodes that were examined due to likelihood of metastasis were negative.

Following resection, a 25 × 27 cm soft tissue defect with exposed vasculature remained. Plastic surgery utilized an ipsilateral pedicled sartorius flap to cover the vasculature and a wound vacuum assisted closure (VAC) was placed to cover the remaining wound. Five days after the index procedure and after confirmation of negative margins, the wound VAC was removed, and an ipsilateral vertical rectus abdominis myocutaneous (VRAM) flap and bone temporizing matrix (BTM) were placed for wound coverage. A new wound VAC was placed as a bolster.

Over the next month, the patient remained admitted in the hospital for several complications, including a hematoma under the VRAM flap that was evacuated surgically, a BTM graft infection that was removed, several wound

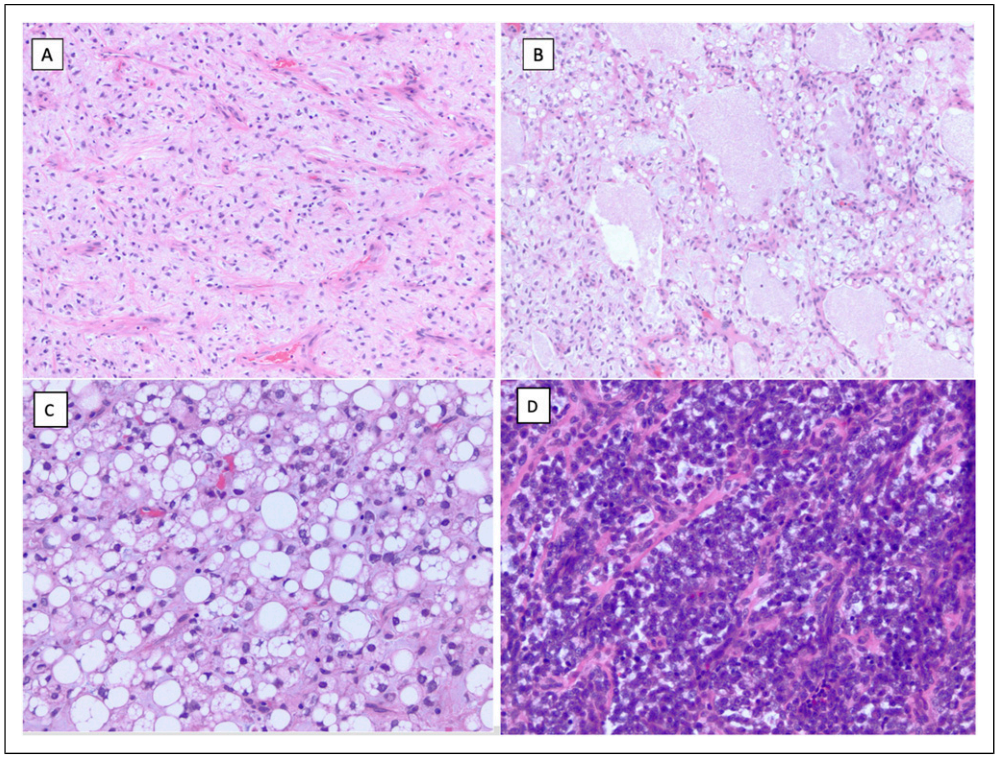

Figure 3. Low cellular areas with spindled to stellate cells with scarce mitotic activity (A) and areas with pools of mucin are appreciated, resembling pulmonary edema-like pattern (B). Multivacuolated lipoblasts are present. Mild nuclear pleomorphism is appreciated (C). Focal hypercellular area composed of small round blue cells comprising 5% of the tumor (D).

washouts, VAC changes, and inferior vena cava (IVC) filter placement. The patient was discharged to a subacute rehabilitation facility with a wound VAC in place. One month following discharge, she returned to the operating room for coverage of her wound with split thickness skin graft (STSG). Upon removal of her wound VAC, adjuvant chemotherapy of six cycles of doxorubicin and ifosfamide and 4005 cGy in 15 fractions of radiation were initiated. Three months postoperatively her wound appeared to be healing well with minimal swelling.

The patient tolerated the remainder of her chemotherapy treatment with no complications. However, 11 months after her index procedure she developed an area of wound necrosis in the setting of radiation. This large wound was treated by plastic surgery through outpatient tissue debridement. The operation was successful, but as anticipated, necessitated a wound VAC placement for a few months, and ultimately a STSG was placed over the wound. At most recent follow up which is 16 months after the index procedure, the patient's STSG has fully healed, and the patient is on her way to a full recovery.

Discussion

Myxoid/round cell liposarcoma is the second most prevalent of five WHO-defined subtypes, encompassing approximately one-third of all liposarcomas.[5] The most common location of myxoid tumors, excluding myxoid pleomorphic liposarcomas, is the lower extremities. The average dimensions reported for this tumor type are between 8 and 12 cm, however, they can vary from 1.5 to 33.0 cm.[7] Although there is a range in sizes of MRCLPS, the tumor in our case report is 39 cm.

Historically, surgical treatment for tumors of this size could result in drastic losses of limb functionality.[8] As a tumor increases in size, there may be an increased risk of enveloping vital structures such as the limb neurovasculature, which could further jeopardize function in the limb.[9] The concern for damage to vital structures and permanent impairment is heightened when neoadjuvant radiation, an important aspect of the treatment plan, is logistically unfeasible.[10] Additionally, while pure myxoid liposarcoma generally conveys a favorable prognosis with a 5-years survival rate of approximately 90%, tumor diameters greater than 10 cm and higher grade round cell composition of greater than 5% of the tumor drastically worsen this prognosis, with increased risk for metastasis and death.[11] In our case, the patient developed some focal areas of higher grade round cell tissue, comprising about 5% of the tumor and was diagnosed with grade 2 MRCLPS, but did not develop metastases. However, our patient did

experience several complications following treatments, including the development of a hematoma deep to the VRAM flap and wound necrosis secondary to radiation. As the patient had been on anticoagulation since her index procedure, she developed a hematoma deep to the VRAM flap. The wound necrosis and clinical challenges in healing the skin graft are expected complications in the setting of radiation treatment.[12,13] While radiation therapy has not been widely proven to increase overall survival, local control outcomes have certainly been improved with its utilization.[14,15] In this case, adjuvant radiation was performed due to the size and constraints of this specific tumor.

Another important aspect of this case is the necessity of a multidisciplinary surgical team for this patient. Following wide local excision performed by the orthopedic oncology team, the plastic surgery team encountered a soft tissue defect of 27 × 25 cm. They provided immediate coverage with a pedicle sartorius flap followed by delayed VRAM coverage and BTM skin substitute. Although there were initial postoperative complications, the multispecialty surgical team was able to preserve the limb. After preoperative consultation, radiation oncology and medical oncology determined adjuvant treatment to be appropriate. Once again, in the adjuvant course she developed complications in her STSG related to her radiation treatment. However, the plastics team was able to debride the wound and place a new STSG that has since well healed with no issue at the wound or donor site. The role of chemotherapy and radiation in this patient's treatment course were also extremely important in preventing local recurrence of this disease, given that the patient was unable to receive neoadjuvant radiation.

Conclusion

This report summarizes the presentation, workup, treatment, and postoperative conditions of a 51-year-old female who developed a 39 cm MRCLPS of the left medial thigh. Although MRCLPS is often an indolent growth, very few cases of this magnitude have been published or even reported. We emphasize the importance of a multidisciplinary effort in work up and treatment in complex soft tissue sarcoma cases.

Authors' note

Authorship has been granted only to those individuals who have contributed substantially to the research or manuscript.

Author contributions

JRF, AY, and GV researched literature. ATB, DW, AA, IM, GK, and SG conceived the study. GV, AY, AA, AB, IM, and JRF were involved in obtaining images and writing and editing. All authors reviewed and edited the manuscript and approved the final version of the manuscript.

Declaration of conflicting interests

The author(s) declared the following potential conflicts of interest with respect to the research, authorship, and/or publication of this article: ATB is on the Rare Tumors editorial/governing board and DW is the Editor in Chief. All other authors have no pertinent financial disclosures or pertinent conflicts of interest.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethical statement

Ethical approval

Rush IRB approval was waived as a non-Human Subject Research (NHSR) form was completed for this case series.

Informed consent

The authors confirm that informed consent of the patient was taken for publication of this case report.

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Data availability statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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