American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2019; 20: 318-322
DOI: 10.12659/AJCR.914079

Received: 2018.11.11
Accepted: 2018.11.25
Published: 2019.03.11

Large Undifferentiated Pleomorphic Sarcoma of the Posterior Thigh

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF **Austin H. Allen**

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Conflict of interest: None declared

Patient: Female, 62
Final Diagnosis: Undifferentiated pleomorphic sarcoma
Symptoms: Pain in the leg
Medication: —
Clinical Procedure: Surgical excision
Specialty: Oncology

Objective: Rare disease

Background: Sarcomas account for less than 1% of all cancers. Undifferentiated Pleomorphic Sarcoma, formerly called Malignant Fibrous Histiocytoma, is a rare subtype identified by a lack specific immunohistochemical markers for a specific lineage of differentiation. These soft tissue tumors are aggressive and rapidly enlarge. Risk for metastasis increases almost linearly as the tumor increases in size, emphasizing the importance of early detection, treatment, and post-resection monitoring.

Case Report: This article reports a case of a large undifferentiated pleomorphic sarcoma of the posterior thigh in a 62-year-old female. Given the patient's history of thrombotic thrombocytopenic purpure, her initial mass was thought to be a hematoma following a hernia repair surgery. After diagnosis of undifferentiated pleomorphic sarcoma, she underwent radical excision revealing a 24×9.5×7cm lesion – one of the largest reported in the literature.

Conclusions: Sarcomas are very rare soft tissue neoplasms, but they should not be excluded in a physician's differentials when a patient presents with an enlarging soft tissue mass. Because sarcomas enlarge rapidly, delay in evaluation and management should be avoided and these patients should be quickly referred to a center specializing in sarcoma treatment. Magnetic Resonance Imaging (MRI) is the recommended initial imaging for all soft tissue masses of the extremities, trunk, and head and neck while Computed Tomography (CT) is the recommended imaging choice for retroperitoneal and visceral masses. After successful surgical excision with clean margins, patients should undergo serial monitoring by CT or MRI for surveillance of recurrence or late pulmonary metastases.

MeSH Keywords: Histiocytic Sarcoma • Histiocytoma, Malignant Fibrous • Neoplasms, Connective and Soft Tissue • Sarcoma • Soft Tissue Neoplasms

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/914079

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Background

Soft tissue sarcomas account for less than 1% of all cancers. Undifferentiated pleomorphic sarcoma (UPS), formerly called malignant fibrous histiocytoma and declassified by the World Health Organization in 2002, is a rare and malignant subtype [1]. These tumors are the fourth most common soft tissue sarcoma and have an incidence of about 0.08–1 per 100,000 [2]. Sarcomas typically present in the sixth and seventh decades of life and tend to occur in the extremities, retroperitoneum, viscera, and head and neck [3]. We present this case of a high-grade posterior thigh undifferentiated pleomorphic sarcoma due to the rarity of the disease, the complication of diagnosis due to a patient history of thrombotic thrombocytopenic purpura, and the abnormally large size of the tumor at the time of excision. To our knowledge, this is one of the largest reported UPS that was successfully removed by radical excision [4].

Case Report

A 62-year-old woman initially presented for an emergent hernia repair with suspected small-bowel obstruction. One month after her surgery, she felt pain in her posterior right thigh. Concerned, she presented to her primary care physician. Her medical history was significant for thrombotic thrombocytopenic purpura (TTP), hypertension, congestive heart failure, myocardial infarction, stroke, and pacemaker implantation, all of which were well-managed with outpatient treatment and surveillance. Physical exam was largely unremarkable, but did reveal a small palpable mass in her posterior right thigh.

Due to her permanent pacemaker, the patient was unable to have magnetic resonance imaging (MRI) therefore her primary care physician ordered an ultrasound and non-contrast computed tomography (CT) of her right leg to evaluate the mass. The first orthopedic oncologist to analyze her imaging suggested that the mass was consistent with a hematoma, given her recent hernia surgery and her history of TTP. The patient was not pleased with her first oncologist, so she sought a second opinion with another local orthopedic oncologist. This second physician ordered a CT with contrast of her right lower extremity which demonstrated a probable large, bulky, neoplastic mass. Following the concerning CT report, she then underwent her first fine needle biopsy. The biopsy revealed the lesion was consistent with sarcoma and she was referred to a university hospital where she could be treated in whole. One month later, her physical exam demonstrated a mass that was tender and palpable at 10cm. She underwent three more attempts at needle biopsy for further classification, but none of the samples were viable due to diffuse necrosis. One month after presentation to the university orthopedic oncologist, she underwent open biopsy with partial radical resection that revealed grade

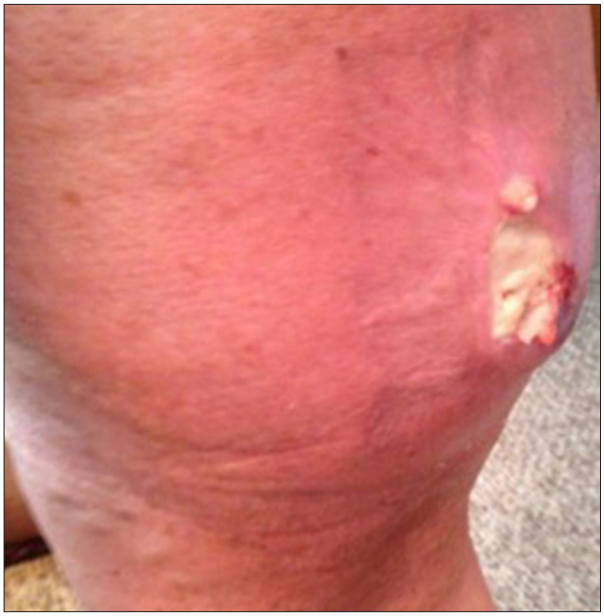

Figure 1. Superficial fungating lesion post-radiation pretreatment.

3 UPS arising from the adductor magnus muscle. Chromosomal investigations revealed 47, XX, +X, t(1;1)(p36.3;p34), t(4;19)(q35;q13.1)[cp17]/47, i(6)(p10)[xp3]. Immunohistochemical investigations revealed vimentin was strongly positive while desmin, HMB-45, smooth muscle actin, Cam 5.2, S100, smooth muscle myosin, CD34, CD45, TLE1, CD99, Keratin AE1/AE3, and SOX10 were all negative. Immunohistochemical evaluation allowed for the pathological diagnosis of UPS.

Due to the large size of the lesion, the patient was advised to undergo preoperative radiation therapy in an attempt to reduce the tumor. She completed the radiation therapy in 3 months and developed a 3cm lesion with serosanguinous drainage that allowed the tumor to fungate superficially (Figure 1). After labs and a CT scan of her lungs to evaluate for metastasis, she underwent operative radical tumor resection. The tumor had not invaded any osseous structures and had not compromised any neurovasculature in the leg making removal straightforward (Figures 2, 3). The fungating portion of the tumor was closed and sealed and a 20 cm ellipse-style incision was made so as not to contaminate the fungating area along the entire posterior thigh. The tumor was dissected medial to lateral, proximal to distal, and superficial to deep down to the level of the sciatic nerve. The pes anserine tendon was taken with the tumor so as to expose the popliteal fossa where the popliteal vessels were identified and protected. The dissection was then carried distal to proximal and a neuroplasty was performed on the sciatic nerve through its division into the peroneal and tibial trunks. The dissection was then carried distal to proximal protecting the nerve and releasing sequential vascular branches to the tumor all the way up into the ischium. The portion of the

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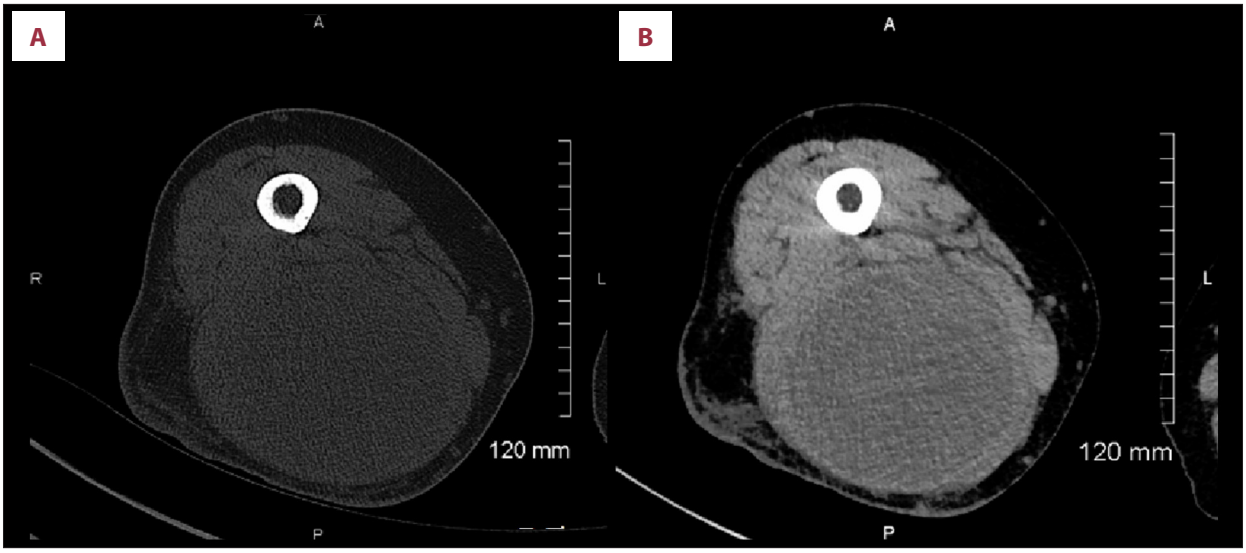

Figure 2. CT scan of the lower extremity without (**A**) and with (**B**) contrast demonstrating the bulk of the tumor.

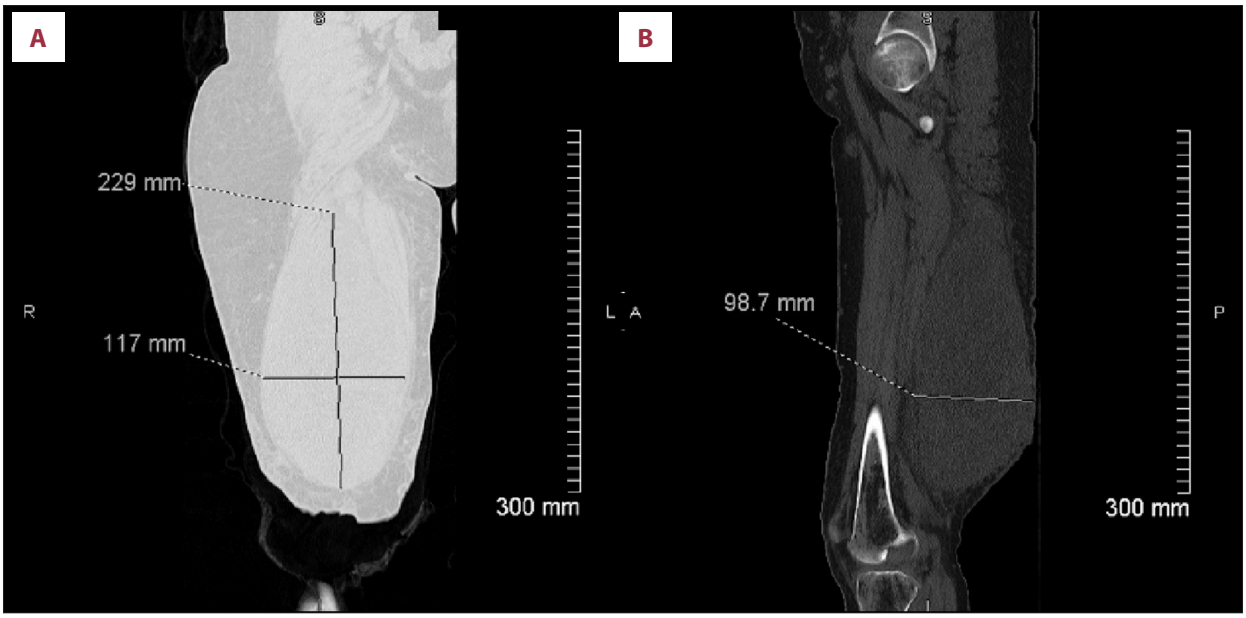

Figure 3. Anterior (**A**) and lateral (**B**) CT scans of the tumor demonstrating its large size prior to excision.

hamstrings musculature was then released from the ischium and the tumor was removed. Soft tissue repair of the adductor musculature to the remaining hamstrings was performed to cover the sciatic nerve and the wound was then closed in surgical layers. The 20 cm ellipse that could not be closed was covered with a negative pressure wound vacuum and sterile dressing for later closure by plastic surgery. The tumor was sent fresh to the pathologist for evaluation. The lesion was a well-defined and entirely circumscribed tan/white friable mass with diffuse necrosis that measured 24×9.5×7 cm.

The patient was extubated the same day of surgery in the post-anesthesia care unit and had no postoperative complications. Her wound vacuum was left in place for 7 days. She then had surgical placement of an Integra dermal regeneration template, sterile dressings, and a knee immobilizer by plastic surgery. She was discharged home after the surgery with a plan of serial wound care follow up and a referral to physical therapy for movement rehabilitation. The surgical advisory board also developed a plan for serial CT monitoring every 3 months for 2 years post-operative and every 6 months for 2 years afterwards to evaluate for recurrence or late pulmonary metastases.

The patient is now post-operative status 16 months at the time of reporting. Serial CT monitoring has not revealed any evidence or concern for recurrence or pulmonary metastases. Her

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wound has healed well and she has full strength and sensation in her right lower extremity with no obvious neurological, musculoskeletal, or vascular deficits. She will continue her serial CT monitoring and follow up appointments with oncology and her primary care provider.

Discussion

High-grade pleomorphic malignant tumors that lack specific immunohistochemical markers for a specific lineage of differentiation are classified as "undifferentiated pleomorphic sarcomas/malignant fibrous histiocytomas" [5]. These tumors are rare and account for <1% of all malignant cancers. They most commonly present in the 6th and 7th decades of life and tend to occur in the extremities, retroperitoneum, and head and neck [3,6].

Retrospective studies have determined that adequate resection margins during treatment is the single most important prognostic factor for long-term survival as local recurrence and distant metastases are significantly associated with higher mortality [4]. Of note, 5- and 10-year survival calculations have been conducted and demonstrate that tumors that are deep, high grade, and >5cm have the worst overall 10-year survivals, only 38%, indicating the importance of early diagnosis and management [7]. Tumor grade, size, and depth have not, however, been statistically significant prognostic factors for local recurrence. Instead, microscopically positive surgical margins, tumor location in the upper extremities, or distal tumors have been shown to be strong predictive factors for local recurrence of sarcomas [7]. It should also be noted that sarcomas have a propensity to metastasize frequently to the lungs [3,8,9]. According to Grimer [9], the risk of metastasis in sarcomas increases almost linearly as the tumor enlarges.

Delay in diagnosis of sarcomas is extremely common. Because they are rare, they frequently bypass a physician's differentials. In addition, patients frequently do not seek prompt care due to the often-painless nature of the mass. The United Kingdom Department of Health has published criteria for the urgent referral of any patient with a soft tissue lesion: mass >5 cm (golf ball size), painful lump, lump that is increasing in size, lump of any size that is deep to the muscle fascia, or any lump that recurs after excision [10]. It is also noted in the literature that masses are often excised inappropriately before biopsy can confirm the diagnosis because physicians assume benignity. Data underscore the importance of transferring any patient with an unidentified soft tissue mass to a center that specializes in treating sarcomas so they may undergo appropriate initial resection [11].

MRI is the recommended initial imaging for all soft tissue masses of the extremities, trunk, and head and neck while CT is the recommended imaging choice for retroperitoneal and visceral masses [12]. It is also recommended that all patients diagnosed with sarcoma undergo chest imaging with CT for evaluation of pulmonary metastasis. CT of the abdomen may also be indicated to evaluate for extrapulmonary metastasis to the abdomen and retroperitoneum [12]. Positron Emission Tomography (PET) is not routinely recommended as a component of the initial staging workup of soft tissue sarcoma for evaluation of either pulmonary or extrapulmonary metastatic disease. Regarding biopsy, core needle biopsy is considered the preferred initial method in most cases due to its low incidence of complications and high diagnostic accuracy. Fine needle biopsy, as was done in this case, is not recommended due to lower diagnostic accuracy [13].

Because sarcomas have a substantially better prognosis with early diagnosis and smaller size, it is crucial to recognize and evaluate any enlarging mass promptly. Although rare, sarcomas should not be overlooked in the differential of any soft tissue mass. It is extremely important to refer any suspected cases to appropriate management centers early and treat them aggressively to prevent complications.

Conclusions

Sarcomas are very rare soft tissue neoplasms, but they should not be excluded as a differential when a patient presents with an enlarging mass, especially if the mass is located in the extremities, retroperitoneum, or head and neck. Sarcomas enlarge rapidly, in this case growing 24×9.5×7 cm in only 9 months. Delay in evaluation and management should be avoided in aggressive sarcomas as the risk of metastasis increases almost linearly as the tumor enlarges [9]. MRI is the recommended initial imaging for all soft tissue masses of the extremities, trunk, and head and neck while CT is the recommended imaging choice for retroperitoneal and visceral masses [12]. The recurrence rate and incidence of pulmonary metastasis in sarcomas is high [3,8,9]. Serial monitoring with CT or MRI should be conducted on a regular basis for several years after surgical excision of a sarcoma to evaluate for recurrence or late pulmonary metastases.

Acknowledgements

L. Daniel Hall, DO: Dr. Hall provided medical records for this patient as well as general support in the preparation of this manuscript. He read and edited the original manuscript prior to submission.

Laura C. Reese, DO: Dr. Reese provided medical records for this patient and read and edited the original manuscript prior to publication.

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