Case Report

Massive localized lymphedema of the thigh mimicking liposarcoma

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A B S T R A C T

Massive localized lymphedema represents a pseudosarcoma seen most commonly in middle-aged morbidly obese patients that radiologically can be easily confused for a soft-tissue sarcoma if one is not familiar with this entity. Although considered relatively rare, as the obesity epidemic continues to rise, the incidence of this entity will likely increase as well. We present a case of massive localized lymphedema occurring in the medial thigh, the most common location, with imaging and pathologic correlation.

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Introduction

Massive localized lymphedema (MLL) reflects a slow growing monstrous, pendulous appearing tumefactive pseudosarcoma seen most commonly in middle-aged morbidly obese patients, either as an isolated mass, or multiple masses [1–5]. Clinically, histologically, and radiologically this mass is easy to confuse for malignancy if one is not familiar with the entity. As the obesity epidemic continues to rise, the incidence of this relatively rare entity will likely increase as well. We present a case of MLL occurring in the medial thigh, the most common location, with imaging and pathologic correlation.

Case report

A 46-year-old morbidly obese woman was admitted through the emergency department with 2-3 months of painful redness and swelling of her medial right thigh. She denied any medical history but was found hypertensive on presentation. An ultrasound obtained to exclude deep vein thrombosis of the right lower extremity was negative. Contrast-enhanced computed tomography (CT; Fig. 1) ordered to exclude soft-tissue abscess demonstrated skin thickening and reticular soft-tissue attenuation compatible with edema throughout the subcutaneous fat at the site of redness.
The patient was given a presumptive diagnosis of right leg cellulitis. Diuretic therapy started while in the hospital for newly diagnosed hypertension appeared to minimally improve the right thigh swelling. She was also given intravenous antibiotics while being treated in the hospital, and eventually discharged with a 7-day course of oral antibiotics. In addition, a compressive stocking was recommended by the orthopedic surgery service to assist with reducing edema; however, the patient was without insurance and could not obtain this orthotic.

Over the course of the next 2 years, the patient was admitted to the same hospital 4 times for recurrent and/or persistent right thigh painful cellulitis and edema, managed with antibiotics. An ultrasound (Fig. 2) acquired during this time frame to exclude soft-tissue abscess demonstrated lobular contour of the subcutaneous fat with edema, but no organized abscess. Notably, during these admissions, she was afebrile with a normal white blood cell count.

Some 15 months later, the patient returned to the emergency department complaining of 3 weeks of right thigh pain.

Fig. 1 – A 46-year-old woman with redness and swelling of the medial right thigh—CT. Coronal contrast-enhanced CT images from anterior to posterior (A-C) demonstrate a mass-like pendulous fatty-appearing lesion with skin thickening and striated soft-tissue edema.
swelling with intermittent fevers and now with a larger and more well-defined medial right thigh mass when compared to initial presentation. Magnetic resonance imaging (MRI; Fig. 3) was obtained of the right thigh, demonstrating a large exo-phytic mass arising from the medial right thigh, involving only the subcutaneous fat plane, predominately isointense to muscle on T1-weighted imaging, hyperintense on the fat-suppressed fluid-sensitive sequences, and with minimal peripheral and/or dermal enhancement. Regions of interposed fat were noted, and a differential diagnosis including liposarcoma was provided by the radiologist.

An ultrasound-guided biopsy was performed, as a result of concern for malignancy, which was nondiagnostic. Thereafter, the mass was surgically excised in its entirety (Fig. 4). A 15 × 30-cm flap was created from the posterior thigh soft tissue and skin after the 20 × 40-cm tumor was fully excised. A 19-French drain was brought to lie through the area of dissection and through an inferior stab incision. Final pathology (Fig. 5), interpreted in conjunction with negative carboxypeptidase M amplification, was compatible with MLL.

Three-month follow-up MRI (Fig. 6) and CT (Fig. 7) were obtained demonstrating post surgical changes from recent resection, with a small postoperative fluid collection and surgical drain in situ.

Discussion

In a series of 14 cases all from morbidly obese patients, Farshid and Weiss [6] were the first to classify MLL as a distinct, reactive lesion. The investigators noted that the soft-tissue lesion mimics the histologic appearance of a well-differentiated liposarcoma (WDL), with lobules of mature fat interrupted by expanded connective tissue septa. The widened septa simulate the fibrous bands of sclerosing WDL, but MLL lacks the degree of nuclear atypia seen in the former. All lesions were extremely large (33.4 cm and 7408 g), and there was a nearly 2:1 female to male predominance [6].

More recent studies highlight that this pseudosarcomatous tumefactive benign entity occurs mostly in the obese population, with a female predilection. The lesions range in size from 19.5 to 61.5 cm [1,2,5]. There is controversy in the literature as to the association of pain with MLL [1,7].

The disease remains a diagnostic challenge to separate clinically and histologically from a variety of other tumors or tumor-like processes (lipoma, lipomatosis, WDL, desmoid-type fibromatosis, lipomatosis, angiomatosis, lymphangiomatosis, angiomyoma, angiosarcoma, low-grade fibroblastic neoplasm, and myxoid liposarcoma) if one is not familiar with this process. The history of morbid obesity and prominent skin changes, including induration and “peau d’orange” of congested dermal lymphatics, assist with making the correct diagnosis. In challenging cases, immunohistochemical stains or fluorescent in situ hybridization can be performed [2–5,7,8].

There is a predilection for the inner thigh, but other locations, such as the lower abdominal wall, suprapubic region, mons pubis, vulva, inguinoscrotal, penis, perianal, popliteal fossa, lower leg, and arm have been described [1–5,9]. The pathogenesis is indeterminate, with associations including lymphatic obstruction related to massive adipose tissue, trauma, surgery, and/or hypothyroidism [2–5,8]. The lymphatic system follows the vessels, transporting extravasated fluids and molecules back to the circulatory system. With dysfunction of the lymphatic system, these fluids and molecules can collect as lymphedema [8].
Sectioning of the mass yields abundant mature adipose tissue separated by thick edematous fibrous septae, and with substantial edema. On occasion, dystrophic calcification may be present [3]. Recent reports describe the histologic appearance of dermal fibrosis, expansion of fibrous septa between fat lobules with increased numbers of stromal fibroblasts, lymphatic proliferation and lymphangietasia, multinucleated fibroblastic cells, and marked vascular proliferation [7].

The CT and MRI appearance of the lesion is that of a large pendulous mass of adipose tissue without fascial or muscle involvement, and directly corresponds to the histopathologic findings of dermal fibrosis, edema, and fat partitioned by fibrous septae. Specifically, edema tracks along the fibrous septa in a lace-like fashion with associated dermal thickening. After contrast administration, there is only mild enhancement in the dermis, as is seen in lymphedema [2,3,7].

Fig. 3 – A 50-year-old woman with a large medial right thigh mass—MRI. Coronal short-tau inversion recovery (A), axial T2 fat-suppressed (B), T1 (C), T1 fat-suppressed (D), and T1 fat-suppressed postintravenous gadolinium-based contrast-enhanced (E) MR images demonstrate a large fat-containing mass arising from the medial right thigh with increased signal on fluid-sensitive sequences and faint peripheral and/or dermal enhancement.
Fig. 4 – A 50-year-old woman with MLL—resection specimen. Large resected specimen with pathologic diagnosis of MLL.

Fig. 5 – Photomicrographs of the resected MLL. Adipose tissue is present with focal nuclear atypia associated with fibrosis, patchy chronic inflammation, and increased vascularity.
Surgery is the standard of treatment for MLL to restore mobility (large lesions of the inner thigh may preclude ambulation), prevent recurrent infections that presumably result from mechanical dermal abrasion, and for diagnosis confirmation [1–4,8]. Conservative treatment alone with antibiotics for associated cellulitis and compression dressings is prone to failure [1]. Since the underlying cause of MLL is believed to be related to lymphatic obstruction, persistence or recurrence of the lesion is expected after resection [4,5]. Notably, cases of squamous cell carcinoma and cutaneous angiosarcoma arising from long-standing MLL have been reported [1,4,5].

Fig. 6 – A 50-year-old woman with large medial right thigh MLL status post resection—MRI. Coronal short-tau inversion recovery (A), axial T2 fat-suppressed (B), T1 (C), T1 fat-suppressed (D), and T1 fat-suppressed postintravenous gadolinium-based contrast-enhanced (E) MR images demonstrate postsurgical changes, including a small soft-tissue fluid collection.
Acknowledgment

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Fig. 7 – A 50-year-old woman with large medial right thigh MLL status post resection—CT. Axial contrast-enhanced CT images from proximal to distal (A–D) demonstrate postsurgical changes, including a small soft-tissue fluid collection with gas tracking to the skin surface (a drain was present but poorly seen on the provided figure).