



## Case Report

# Pseudosarcoma of the Thigh: A Rare Case of Massive Localized Lymphedema

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Massive localized lymphedema, also called pseudosarcoma, is a rare condition associated with morbid obesity. Accurate identification of this entity helps the physician make the distinction between this condition and other soft tissue tumors, especially with the increasing rate of obesity worldwide. Obesity and increased caloric intake lead to storage of the excess energy in the form of adipose tissue. The excess adipose tissue disturbs the lymphatic vessels, leading to massive edema localized mostly in the lower extremity, reaching a substantial size and weight that interferes with the quality of life of the individual in question. The mass can cause cosmetic but more importantly functional and structural defects, leading to altered biomechanics with increased risk of deep vein thromboembolism. Below is a presentation of a patient presenting with complaint of a massive medial thigh mass. After much investigation, consultation, and use of diagnostic radiologic modalities it was diagnosed as what is referred to in the literature as massive localized lymphedema, or pseudosarcoma. This mass was managed by surgical excision and the diagnosis was further confirmed by pathologic analysis. After the excision, our patient regained his ability to ambulate on his own, with the help of a rehabilitation program and physical therapy. Massive localized lymphedema needs to be on the differential diagnosis of any morbidly obese patient presenting with mass, especially in the lower extremities. Adequate knowledge about this condition enables the physician to make the distinction between pseudosarcoma and true soft tissue sarcomas.

*Key words:* Pseudosarcoma – Massive localized lymphedema – Lymphatics – Benign tumor – Obesity

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**Fig. 1** Preoperative image revealing the massive right medial thigh mass with the patient in supine position on the operating table.

The lymphatic system is designed to follow the vessels, and it transports the extravasated fluids and molecules back to the circulatory system; therefore, any derangement to and obstruction within this system can lead to collection of these fluids and molecules. Lymphedema occurs as the lymphatic system is disrupted, leading to the collection of protein-rich fluid in the interstitial space. *Pseudosarcoma* is the term used to describe massive localized lymphedema (MLL), a benign soft tissue mass resulting from advanced proliferation of lymphatic apparatuses, first discussed in series of cases and further discussed in other case studies distinguishing it from liposarcoma.<sup>1-3</sup> Farshid and Weiss<sup>1</sup> first shed light on this emerging problem with report of 14 cases, all presenting MLL of extremities in the proximal medial location. MLL has since been discussed in surgical, pathologic, and dermatologic journals and articles. This substantial and considerable lymphedema often presents with such a striking appearance and large size that it appears, pathologically and morphologically, to be a sarcoma.<sup>2-4</sup> This condition has been associated with morbid obesity, which itself is well associated with many comorbid conditions that occur in people with obesity. Some of these comorbidities are chronic conditions that physicians in all specialties encounter in the clinic on daily basis, such as hypertension, diabetes, joint pain, osteoarthritis, chronic back pain, lower extremity edema, Acanthosis nigricans, acrochorda, leg ulcers, skin excoriations, and hyperkeratosis of the plantar surface.<sup>2,3</sup> Surgical resection offers the best treatment, and the patients also need to be counseled about weight loss and diet modification to prevent recurrence.<sup>3,5</sup> This report presents

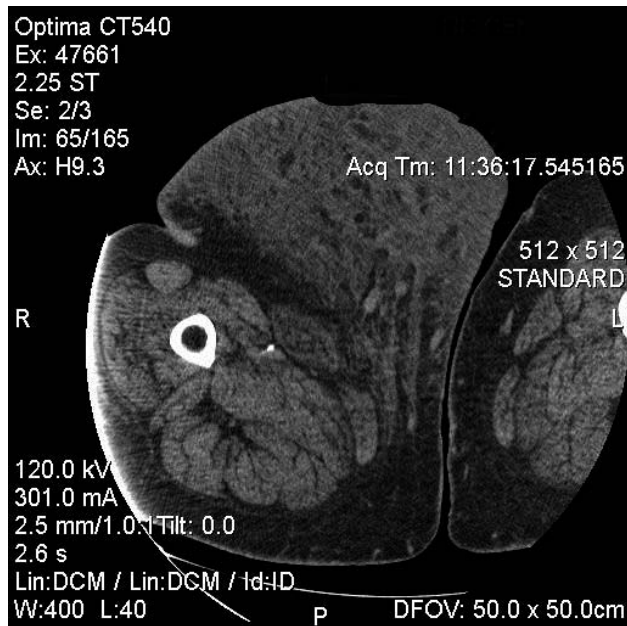
a patient complaining of a massive soft tissue mass of the medial thigh, diagnosed to be a pseudosarcoma.

## Patient and Methods

The patient presented here is a 54-year-old, morbidly obese African American with an extensive past medical history of sleep-disordered breathing managed on BiPAP at home, poorly controlled diabetes, anemia, lower extremity cellulitis with lymphedema, gastroesophageal reflux disease, chronic pain syndrome, deep vein thrombosis in the right lower extremity, peripheral neuropathy, benign prostatic hypertrophy (BPH), depression, hypertension, chronic back pain, and open wounds; he has also tested positive for human immunodeficiency virus, which has been managed and treated with highly active antiretroviral therapy. Patient presented for the consultation of a huge mass of the right thigh (Fig. 1). He complained of an increase in size and weight of the mass, which thus affected his mobility and quality of life. Family history was also significant for a father who died of colon cancer at age 85 years and a mother who died at age 80 years of breast cancer. Patient's problems with the right leg dated back to 1992. At the time of presentation, he had developed a massive amount of swelling in the proximal medial thigh. He had stated that he had significant complications with activities of daily living, walking, gait, and balance, and back pain that had chronically worsened with the increasing size of the mass.

The physical exam showed a morbidly obese man with a pedunculated mass of the medial proximal right thigh and lower extremity edema. Magnetic resonance imaging scan of the right lower extremity showed a mass of 33 × 27 cm, an enhancing soft tissue mass arising from the subcutaneous tissues of the medial right thigh (Fig. 2).

The patient was counseled about surgical excision, which was presented as the only option in the treatment of the mass. The patient was transported to the operating room and positioned in the supine position with general anesthesia administered. Medial and lateral flaps were outlined and marked, and after the surgical incision dissection was made down to the subcutaneous tissue with electrocautery, cauterizing the subcutaneous vessels. Large bridging veins and feeder veins were then clipped, ligated, and transected. We worked down the fascia to identify the plane of the tissue and finally transected it off of the quadriceps fascia. Area was



**Fig. 2** Preoperative magnetic resonance image of the mass, illustrating the mass obscuring the right medial thigh anatomy.

thoroughly irrigated and suctioned clear. A large Blake drain was placed and attached to a Hemovac, secured in place with silk ligature. Using deeper subcutaneous layers the skin was reapproximated by 3-0 interrupted sutures, and the flaps were stapled together. The patient tolerated the procedure well and was transported to the recovery area with cardiovascular anesthesia.

The pathology report of the resected mass exposed the specimen to be 18 lb. The gross description indicated the specimen to be a 39.6-kg, 49.0 × 30.0 × 13.0 cm, yellow, lobulated soft mass, with multiple large-caliber, thin-walled vessels throughout. The microscopic examination revealed large bands of fibrous septa showing minimal cellularity, evidence of benign vascular proliferation between the areas on adipose tissue, a focally more solid fibrotic area which is again hypocellular containing benign multiple benign appearing lymphoid aggregates. The pathology report further noted that there was no evidence of necrosis, hypercellularity, lipoblasts, or other atypical cells. Of further note, there was no evidence of malignancy in the mass, and the overall features were those of MLL associated with morbid obesity. The patient was discharged to rehabilitation on postoperative day 4 and has been seen in follow-up at 4 and 6 weeks, with complete healing of the flap (Fig. 3). Over time the patient regained the ability to



**Fig. 3** Postoperative image of the right medial thigh with the patient in sitting position.

ambulate on his own without the use of a walker or a wheelchair. At 18 lb, or 8.16 kg, this mass stands as one of the largest pseudosarcomas reported.

## Results

Morbid obesity is defined as a body mass index >40 kg/m<sup>2</sup>. MLL is typically a chronic lesion and is seen in middle-aged adults with a varying high mean weight of close to 400 lb, with reports of women outnumbering men from 2:1 up to 10:1.<sup>5,6</sup> It has been observed that patients commonly delay treatment up to a decade from the onset of the problem, when the mass starts interfering with ambulation and most common daily activities.<sup>1-3,7</sup> In these morbidly obese individuals, excess adipose tissue in the body is deposited haphazardly, disturbing the lymphatic channels, constricting and hindering them, leading to retention of lymphatic content and subsequent distension of the tissue.<sup>3,6</sup> Along with the disruption of the lymphatic system, there is lymphoproliferative growth that is benign in nature.<sup>3</sup> The blockade

of protein-rich lymphatic fluid leads to its seepage in the breakage of hyperkeratotic skin, creating a sanctuary for bacterial proliferation and infection, further exacerbating the condition by formation of cellulitis, which has been reported to be chronic and recurrent in these patients and also seen in the patient presented here.<sup>2,5,6</sup>

Dermatologically, the mass causes excoriation and cosmetic defect, along with the obvious gigantic pedunculated mass. The edema begins as pitting in early stages, with the disappearance of the pitting and the development of induration in later stages.<sup>8</sup> Radiologic studies are used to confirm the diagnosis of the MLL and to show skin and subcutaneous edema with a lack of muscular tissue involvement lying beneath the edema.<sup>2,5,6</sup>

Histopathologically, there is no evidence of atypical nuclei, fibroblasts, or lipoblasts, and therefore a lack of evidence of malignancy, distinguishing it from liposarcoma.<sup>2,5</sup> Microscopically there is a thickening of the epidermis; dermal papillary lymphatic channels that are dilated and corresponding tissue that is fibrotic and edematous, with dermal expansion and fibrosis; and peripheral infiltration of lymphocytes, generally perivascular ones.<sup>3,5-8</sup> Deeper tissue examinations at the dermal layer reveal an increase in the number of normal fibroblasts in enlarged fibrous septa with split fat lobules that are mature and contain normal lipoblasts.<sup>3,5,7</sup> Fibrosis is the result of the chronic inflammation, which is itself a consequence of malfunctioning macrophages and a fall in oxygen pressure.<sup>6</sup> One of the grave consequences of untreated lymphedema, albeit a rare one, is a disposition toward developing cutaneous angiosarcoma, hence the necessity of close follow-up and monitoring for the patients.<sup>3,5</sup>

The etiology of MLL has been widely speculated to be multifactorial, with alteration of local anatomy and physiologic changes on the cellular and molecular levels.<sup>2,8</sup> Stasis of the lymphatic fluid acts as stimulus for tumorous growth and proliferation of multiple lines of cells, adipocytes, fibroblasts, myofibroblasts, keratinocytes, and adipose tissue associated with the lymphatic system.<sup>2,3</sup> As the lymphatic fluid extravasates the vessels, growth factors in the fluid are thought to spark the differentiation of preadipocytes and inflammatory mediators, with local ischemia further exacerbating the process.<sup>2,3,8</sup> Other factors producing lymphedema are massive blunt local trauma, local surgery, history of vein stripping, myxedema of hypothyroidism, and lymphadenectomy.<sup>2,6,8</sup> Diagnosis of

MLL has proven to be a challenge to the pathologist because of its very close appearance to sarcoma and its rarity.<sup>7</sup> In the diagnosis of MLL there are a few differentials that repeatedly make the list. Of these, atypical lipomatous tumor/well-differentiated liposarcoma is most often submitted, along with benign processes encompassing multiple fat and lymphatic conditions.<sup>7</sup> Surgical resection, diet counseling, and weight loss offer the best steps in management of the patient to allow relief from the symptoms. It needs to be mentioned that a competent physician can benefit from having adequate knowledge about obesity and the lengthy list of comorbidities accompanied with it. MLL needs to be on this lengthy list and needs to be a part of a differential diagnosis with any obese patient presenting with an evolving mass, especially on the medial proximal lower extremities. Because many of the patients with MLL wait until the mass interferes with their daily activities, it is important for the physician to perform a thorough history and physical exam to identify any masses obscuring the normal anatomy and to document the growth over time. Proper management and education regarding decreasing caloric intake and stepwise increases in the level of activity and exercise should be part of both preoperative and postoperative care provided to the patients. Patients also need to be instructed to adhere to regularly scheduled follow-ups to track the evolution of the mass and/or recurrence of a previously removed mass.

## Conclusion

With morbid obesity becoming an epidemic not just in the United States but also in other parts of the world, in both industrialized and developing countries, it is imperative for physicians to have adequate knowledge about MLL, or pseudosarcoma. This condition is caused by disordered deposition of extra adipose tissue acquired by increased caloric intake, which leads to derangement in the lymphatic system. It is important to distinguish pseudosarcoma from true lipomatous sarcomas in order to have proper assessment and to plan the treatment and management of the mass. Progressively, with poor diet and lack of physical activity the pseudosarcoma can grow to the point that it can impose restrictions on an individual's mobility and daily living activities. At this point surgical resection would offer the best treatment, and the patient also needs to be counseled about weight loss and diet modification to prevent recurrence. It

is also vital to schedule follow-up appointments with the patient and to keep an eye on the recurrence of the mass.

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