

Massive localised lymphoedema: a new benign entity in the morbidly obese patient

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Massive Localised Lymphoedema (MLL) is a rare and benign entity that is rapidly gaining recognition due to its association with morbid obesity. It presents a real diagnostic challenge for clinicians, even with histopathological analysis. It is relatively unknown and its true incidence, pathophysiology and transformation into a malignant process will need to be further evaluated. This case report highlights the important features associated with MLL and management at our institution.

Case Report

A 65-year-old morbidly obese Caucasian man presented to our Emergency Department with an infected, large, right-

sided, inguinal, soft-tissue mass. The mass had gradually increased in size over 13 years, but he had only recently sought medical attention. The patient's medical history was significant for morbid obesity, with a body mass index of 71 kg/m². He was previously involved in a motor vehicle accident in 2003, where he suffered multiple traumatic injuries, including a fracture of his distal right femur, which required surgical intervention with an intra-medullary nail, and had a prolonged and complicated recovery secondary to hypertrophic non-union of his fracture. His other past medical history included diet-controlled type 2 diabetes, atrial fibrillation, hypertension and polyarthropathy. Family history and review of systems were

Figure 1



Figure 2: lesion shown with arrow.

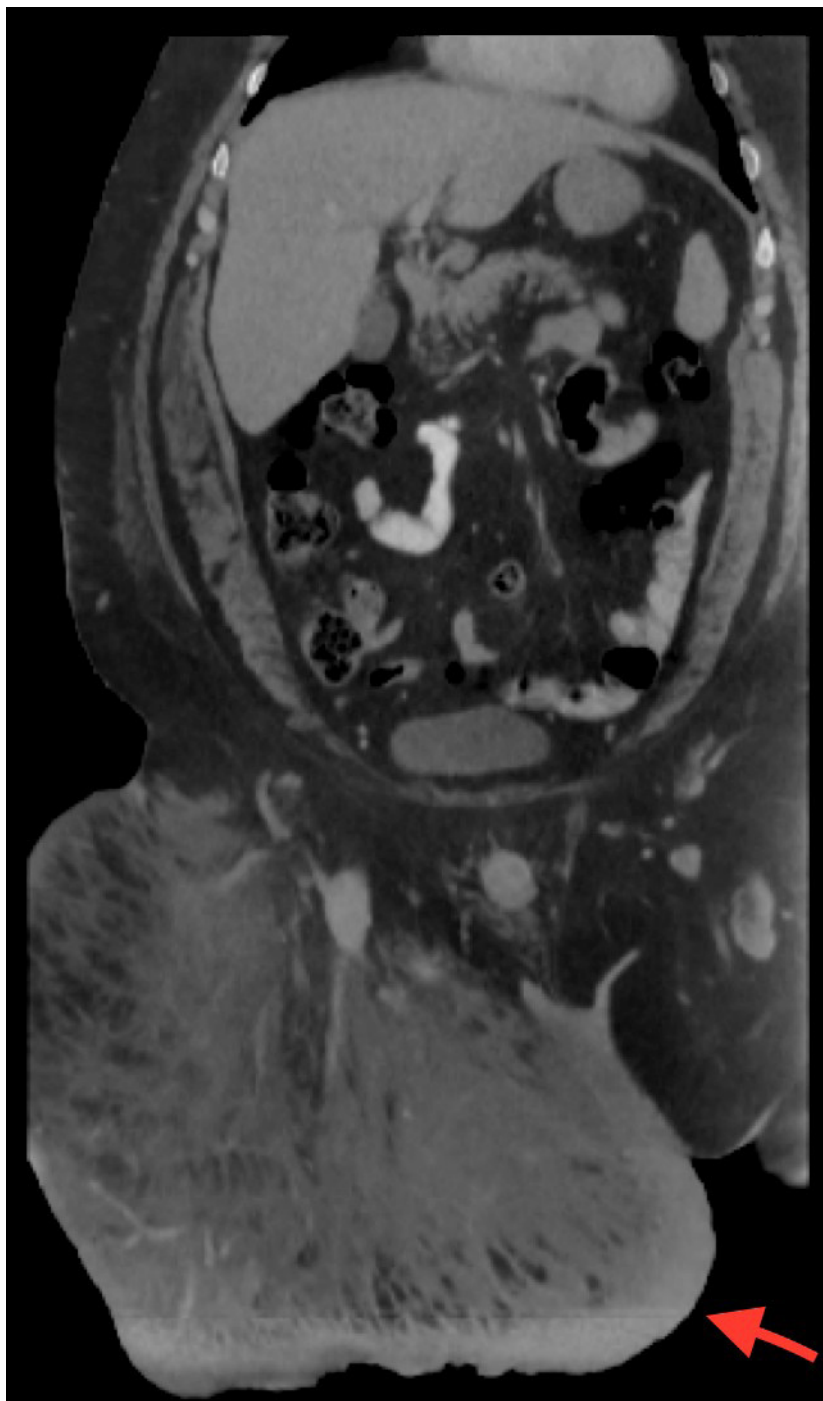


Figure 3: 10x magnification revealing fibroadipose tissue with intervening areas of oedema.

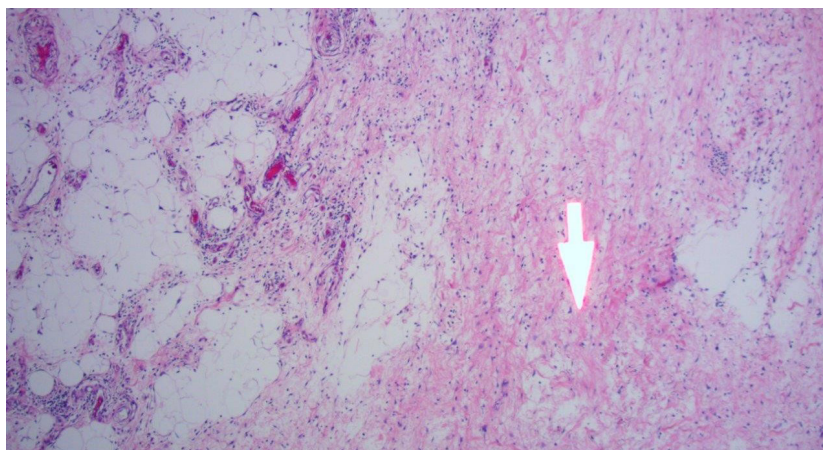
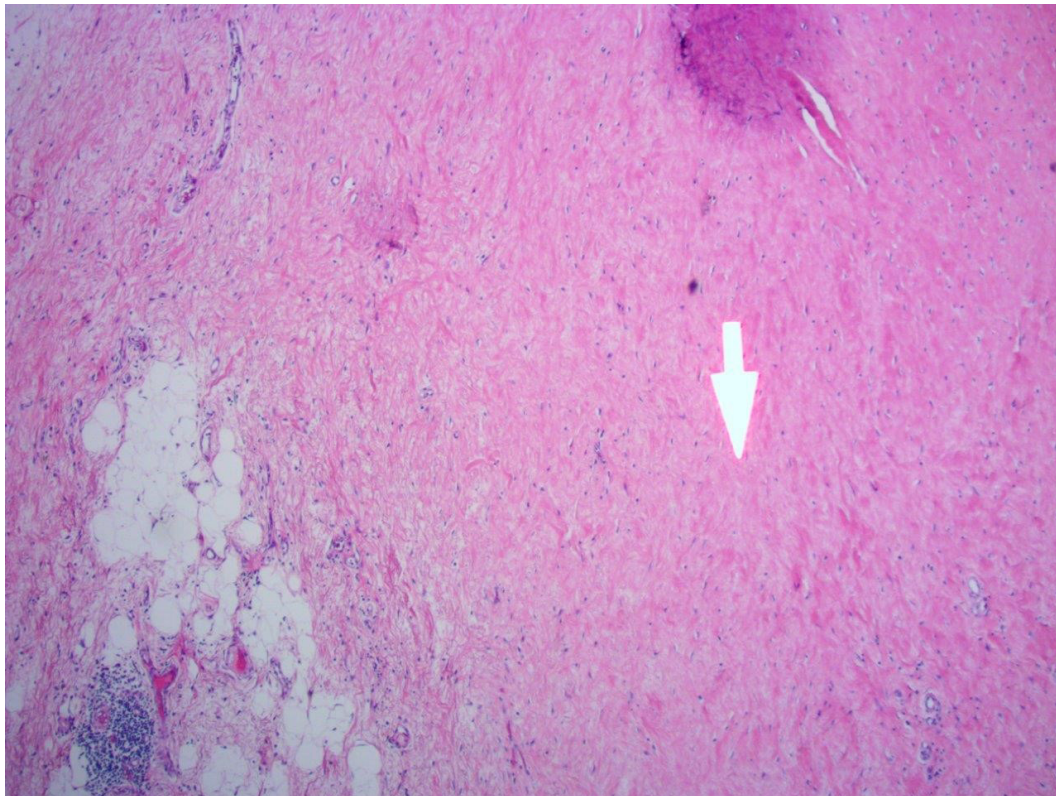


Figure 4: 10x magnification revealing dense fibro-collagen tissue with adipocytes.



noncontributory. He was mostly confined to bed and dependent on carers for most of his activities of daily living.

On physical examination, he had a temperature of 39 degree Celsius, and was tachycardic at 115 beats per minute. The rest of his vitals were within limits. A large pedunculated inguinal mass was obvious from the end of the bed. It was erythematous and interspersed with areas of patchy ulceration on closer inspection. It was leathery and nodular and was separate from the testis which was obscured. (Figure 1)

He had a raised WCC of 21 (normal: 4–11 x 10E9/L), and a low albumin level of 27 (normal: 34–48 g/L) with the rest of his blood panel including thyroid function test within normal limits. Further imaging with CT of abdomen and pelvis with contrast demonstrated a right inguinal mass measuring 42 cm x 41 cm x 23 cm (Figure 2), with extensive thickening and oedema of skin and soft tissue stranding throughout with signs of chronic venous congestion and multiple enlarged lymph nodes in the femoral triangle, but no intra-abdominal or testicular involvement.

He underwent complete excision of this mass, as there was a suspicion of this being a malignant lesion. A resection pattern was first marked around the base of this lesion and then excised.

The specimen weighed 4,520 grams. It was well vascularised and had a lobulated fibro-fatty appearance. Microscopically, it had a mature pale fibro-collagen tissue, adipocytes, intervening areas of oedema with no evidence of cytological atypia, mitotic activity, necrosis or haemorrhage (Figures 3 and 4).

MDM2 and D12Z3 was not amplified on Fluorescence InSitu Hybridisation (FISH) analysis. This was consistent with Massive Localised Lymphoedema.

He was discharged to a peripheral hospital one week after his initial surgery for rehabilitation.

Discussion

MLL is a new clinical entity that we as clinicians need to be aware of. There are a handful of papers in the English literature identified on PubMed on MLL since it was first described by Farshid and Weiss¹ in 1998. However, the nature of MLL and similarity to soft tissue sarcomas still presents a diagnostic dilemma.

It is 1.5 times more common in females than males, with the average BMI reported with MLL to be 60.9 kg/m², and age of patients with MLL range from 19 to 81 years, making age a less relevant factor.² It characteristically occurs as a single lesion at the lower

extremities, at the medial aspect of lower limbs, lower abdomen, scrotum or supra-pubic regions in morbidly obese patients. There is a single case report in the literature of concurrent lesions presenting in a single patient.³ Nevertheless, they are all asymmetrical and tend to run a slow growing process over many years, with recurrent episodes of cellulitis and ulceration to the lesion, as seen with our patient. The asymmetry and sheer size of MLL lesions restricts ambulation and mobility, thus limiting our patients to the bed or their home.

Gross evaluation of the MLL lesions tend to resemble thickened and indurated skin, similar to chronic lymphoedema and what other authors have described as a *peau d'orange* appearance.³ Subcutaneous tissue seen intraoperatively resemble a network of white fibrous streaks intersecting with lobulated pale adipose tissue, giving it a striking marbled appearance. This diffuse unencapsulated lesion seem to extend to the deep margins with no discrete solid component identified.

The microscopic findings of widened fibrous bands or septa separating lobules of mature fat seen with MLL are similar to well differentiated liposarcoma (WDL) or atypical lipomatous tumour (ALT).^{1,3}

However, MLL is different histologically when compared to a neoplastic process, as there is an absence of atypical stromal cells, lipoblasts and atypical adipocytes.^{1,3} In addition, the collagen in MLL is finer and paler in contrast to the dense eosinophilic collagen type seen with WDL.¹ Finally, the presence of reactive vessels at the junction of adipose and fibrous tissue is typical for MLL.¹

The natural progression and propensity of MLL lesions to transform from a benign to a malignant lesion is still unknown. There was a reported series of 5 patients where angiosarcoma was identified within a MLL lesion by Shon et al.⁴ Cutaneous angiosarcoma, in the setting of chronic lymphoedema, is also known as Stewart-Treves syndrome, where it was first

reported in 1948 in a cohort of post-mastectomy patients who subsequently developed chronic lymphoedema of the upper limb.⁵ A recent literature review, by Chopra et al, reported a 14% incidence of angiosarcoma within a MLL lesion.² Su et al described a case of widely differentiated squamous cell carcinoma arising from a scrotal MLL in a patient with a previous history of radiotherapy and surgery for rectal adenocarcinoma many years prior.⁶

The exact pathogenesis of MLL is uncertain and is likely multifactorial. It is a form of secondary lymphoedema that is strongly associated with morbid obesity and lymphatic stasis, as it is the common denominator seen in all cases reported.

Farshid and Weiss¹ have suggested that the pathogenesis of MLL can be secondary to an enlarged abdominal pannus causing obstruction of regional lymphatics. Local trauma to regional lymphatics can also lead to impairment of efferent lymphatic flow thus contributing to lymphatic stasis.

Other factors associated with MLL include hypothyroidism³ and immobility, which understandably causes sluggish lymphatic contractility, chronic lymphatic engorgement and lymphatic valvular insufficiency which eventually leads to lymphatic stasis.⁷

Conclusions

MLL is still a relatively rare and under diagnosed condition that when resected can bring upon a successful and better outcome for the patient. MLL is typically described as an asymmetrical, chronic, diffuse soft tissue lesion occurring in the lower extremity with an oedematous appearance akin to chronic lymphoedema in a morbidly obese patient. It is crucial that the main clinical features of MLL are highlighted to the histopathologist to facilitate the diagnosis of MLL.

As MLL is a recently described entity, a longer surveillance period after primary excision is needed to establish incidence of recurrence and malignant transformation.

Competing interests: Nil**Author information:**

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