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Case Report

Massive Localized Lymphedema in a Morbidly Obese Patient Associated with Renal Failure, Complete Tumor Resection and Normalization of the Kindly Function, Case Report and Review of the Literature

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Abstract

Massive localized lymphedema MLL is a rare disease of morbidly obese patients characteristically occur in the medial aspect of the thigh due to proliferation of lymphoproliferative tissue secondary to compression by the hypertrophied adipose tissue.

MLL is a peculiar soft tissue hypertrophy of the medial aspect of the thigh that can reach enormous size. The disease is seen in morbidly obese patients. We present a case of massive, localized lymphedema that reached enormous size and affected the patient mobility and function. There was associated renal insufficiency due to overwhelming sepsis from infected ulcers and skin defects. The initial Tru Cut biopsy was diagnosed as well differentiated liposarcoma. The tumor was resected without any cross-sectional imaging and the final diagnosis shows massive, localized lymphedema. Post-surgery the patient renal function returned back to normal after few days after surgery.

***Keywords**: Lymphedema; Massive Localized Lymphedema; Morbid Obesity*

Abbreviations

MML: Massive Localized Lymphedema; MRI: Magnetic Resonance Image

Introduction

Massive Localized Lymphedema (MLL) was first described as a distinct clinical entity by Farshid and Weiss1 in 1998.

The disease is considered a rare condition that appears only in morbidly obese patients and usually present as a painless mass in the lower extremity. The disease can present with large soft tissue swelling and can even reach an enormous size. The underlying cause of the disease is obstruction of the lymphatic vessels secondary to fatty deposition. The prevalence of obesity is increasing in the world and it is expected that the incidence of MLL will also rise. We present a case of massive, localized lymphedema that reached an enormous size and was associated with renal function impairment, soft tissue tumor resection was done and resulted in complete normalization of the kidney function.

Case Presentation

A 36 year old male patient referred to the orthopedic oncology clinic for evaluation regarding huge soft tissue mass in his right thigh. The patient is a known case of morbid obesity with current weight of 306 Kilogram and BMI of 104, he as well known to have obstructive sleep apnea, G6PD and sickle cell disease. The mass was progressively increasing in size during the past six years and reached an enormous size that made his ambulation, self-care and hygiene extremely impossible. Few weeks prior to his referral he developed multiple ulcers over the mass which got infected and went into septic shock. He eventually recovered from the sepsis and developed acute renal failure requiring Chronic renal replacement therapy for which a vascular access was planned.

Local examination showed huge mass in the medial aspect of the thigh reaching to knee. The skin overlying the mass showed a 'peau d'orange' appearance with multiple area of discharge and skin peeling Figure 1 (a). Ultrasound was done and showed echogenic soft tissue mass with venous and arterial blood flow, the peripheral aspect of the mass demonstrates soft tissue edema Figure 1 (b). MRI and CT scans were not done due to equipment weight limitations of the machines as per manufactures instruction. Laboratory investigations were done and all withing normal limits except a very high creating and urea levels.

A Tru-Cut biopsy from multiple sites was done and showed dense fibrosis and chronic inflammation with few adipose cells of variable shape and sizes in between the fibrous tissue raising the possibility of well differentiated liposarcoma. Preoperative optimization of his premorbid diseases was done, and patient was taken to the operating room for surgical resection. The tumor was excised through a circumferential incision around the tumor and the feeding vessels were ligated which were found to be huge in diameter, the wound was approximated and the remaining skin defect was covered initially by biological dressing and vacuum dressing Figure 1(c,d)

A week later the biological dressing was replaced by split thickness skin graft which eventually healed nicely Figure 1 (e,f).

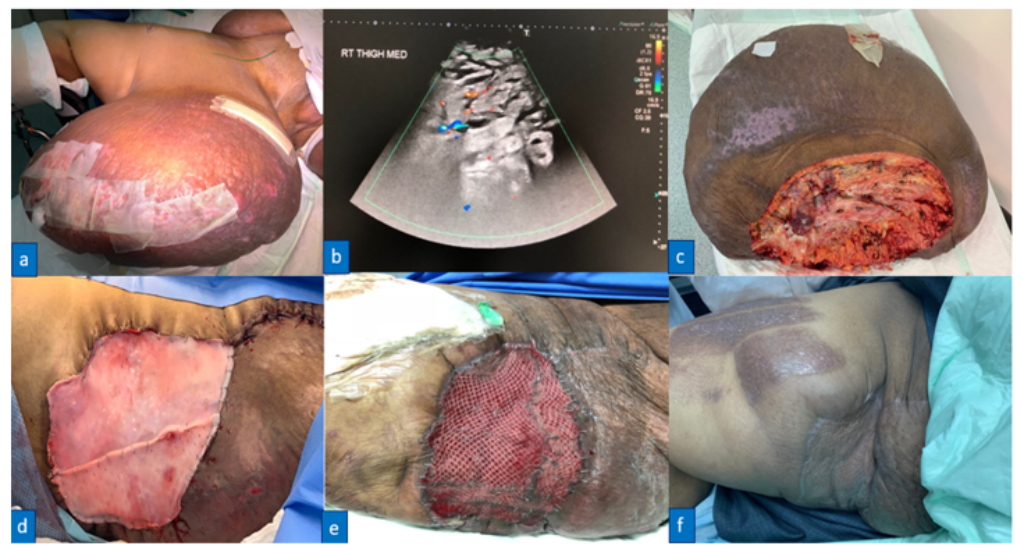

Figure 1: *A-Clinical Picture Showing The Mass Originating From The Medial Thigh. B- Ultrasound Image Shows The Mass With Marked Echogenicity And Abundant Vascularity And Oedema. C- The Resected Tumor. (D, E) The Defect Closed Using Biological Dressing And Later Split Thickness Skin Graft. F- The Final Clinical Picture Aftere Graft Healing.*

The resected specimen was sent to the pathology lab for examination. Grossly the mass weighs around 2800 gram and measuring 89x82x22 cm in dimensions Figure 1-c, and the final results showed the diagnosis of massive, localized lymphedema of morbid obesity figure 2.

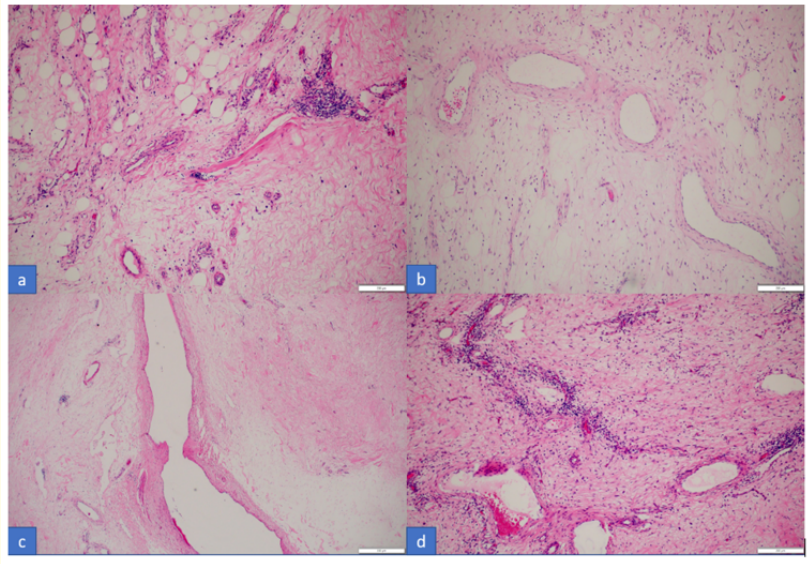

Figure 2: *H And E 20x (A,B,C,D,) Microscopic Picture Showing Mature Adipose Tissue With Expansion Of Fibrous Septae Between Asipocytes, Along With Marked Lymphovascular Proliferation And Dilatation, The Stroma Shows Lymphocytic Proliferation.*

Immediately post-surgery his renal function started to improve and in four days post-surgery his creating and urea were back to normal and remained such till his last follow up a year post surgery. Last follow up showed that the patient started to ambulate on walker and the surgical site showed excellent healing with no evidence of recurrence

Discussion

Massive Localized Lymphedema, MLL, is considered a secondary lymphedema occurring as a result of impaired lymphatic drainage due to morbid obesity. The disease usually occurs in the lower limb, hypogastrium, and mons pubis. The progressive growth can eventually result in an extremely large mass that affects the daily activities due to limitation of functional mobility as well it increases the risk of recurrent infections. The overlying skin often exhibits induration and a 'peau d'orange' appearance, consistent with chronic lymphedema and sometimes associated with skin pealing and ulcerations which can get infected. The treatment depends on the location and severity of the mass. Nonoperative treatment utilizing the conventional lymphedema treatment protocol of extremity elevation, compression stockings and observation is recommended to reduce the symptoms and to reduce the incidence of pressure ulcerations. In more severe cases of ulcerations, or infections, debridement and wound care with simple resection may be performed. Complex surgical resection and reconstruction is usually performed for larger soft tissue masses [1].

Detailed history and clinical examination usually can point to the diagnosis if the disease is familiar to the surgical team. Imaging studies, such as ultrasound, CT, and MRI, are useful to further evaluate the mass as well as the associated vascularity, which is usually profound in these cases, however in morbidly obese patients they might be difficult or even impossible due to the size and weight limitations to CT and MRI studies as was in in our case. Although radiological investigations may be useful for diagnosis, the gold standard remains

tissue biopsy, but they can be easily misinterpreted as low grade liposarcoma as in our case. Routine blood works are usually necessary for preoperative evaluation and in our case, the patient was found to have renal impairment necessitating renal replacement therapy and once the tumor was resected the renal function started to return back to normal gradually possibly due to resolution of sepsis or to the reduction of the fluid loss secondary to continuous oozing from the skin overlying the mass.

Once the diagnosis is confirmed, surgical excision is usually the treatment of choice and the surgical incision has to be chosen cautiously preferably with the help of a plastic surgeon [2].

All reported MLL cases occurred in morbidly obese individuals, with an overall mean weight of 205 kg and the majority of patients were females in the middle age group, and the commonest site of presentation is the medial thigh (Table 1). There are few cases of tumor recurrence in the literature. As far as we know our case is the largest tumor mass reported in the literature both in weight and dimensions.

Author (Reference)	Year	Patients, N	Mean Age, Years	Mean Body Weight, Kg	Mean Mass Weight, G	Time To Presentation, Years	Most Common Site
Farshid and Weiss	1998	14	47	169	6727	1-10	Medial right thigh
Wu et al	2000	6	47	212	6764.5	2.9	Medial right thigh
Weston and Clay	2007	1	57	230	Not excised	5	Medial right thigh
Barr	2000	1	76	Not reported	Not reported	4	Pubic region
McCrystal and O'Loughlin	2007	1	41	210	3000	3	Medial right thigh
Robin James Evans and Chris Scilley	2011	2	44	260	2720	6	Medial right thigh
Furrukh Jabbar et al	2014	6	47	193	10886	1-8	Right Medial thigh – right calf – abdomen
Efstathios Kotidis	2015	1	61	140	1350	5	Lower abdominal wall
Jack Porrino and Josh Walsh	2016	1	46	Not reported	2000	3	Medial right thigh
Parviz Goshtasby and John Dawson	2016	1	49	182	8194	6	Medial right thigh
Dilek Ertoy Baydar	2018	1	53	85		17	Retroperitoneal
Aleksandra G and Rami Lutfi	2018	1	52	159	831	1	Medial left thigh

Table 1

Although the clinical presentation is understood, the pathogenesis of MLL is currently unknown. The commonest etiology is obliteration of the efferent lymphatic flow with secondary lymphedema. Occasionally, there is a decrease in the lymphatic drainage secondary to a large abdominal pannus [3].

Although it frequently mimics liposarcoma, given its appearance and massive size, pathology will reveal it to be a benign process with a lack of nuclear atypia. In our case the biopsy was read initially as low grade liposarcoma and the eventual resected specimen confirmed the diagnosis of MLL. This frequent resemblance leads us to use the term pseudosarcoma to describe this entity [2]. Both low grade liposarcoma and massive localized lymphedema share a similar architectural pattern seen under low power magnification, such as the expanded interlobular septa separating mature fat, however, the fibrous bands are considerably paler and there is a lack of a distinct mass as well as invasion of muscle or other structures as seen in liposarcoma. It also lacks the nuclear atypia hyperchromatic atypical stromal cells, atypical adipocytes, and lipoblasts. The disease characteristically shows mature adipose tissue with expansion of fibrous septae between adipocytes with associated lymphovascular proliferation.

These tumors are often longstanding, with the patients often delayed in seeking medical advice for up to 10 years and by that time the tumor would have reached enormous size that affects his ambulation, self-hygiene and care from recurrent attacks of skin ulcerations and infections [2].

The overall prognosis of MLL is good, with only two reported cases of transformation to malignancy. However, recurrence of the mass has been reported in up to 50% of patients within a 10-month to 10-year timespan, hence close follow-up is recommended [1]. Recurrence is managed with further excisions as necessary [2-8].

Conclusions

Massive, localized lymphedema MLL is a recognized clinical entity recently described in the literature. The tumor is characterized by a benign large soft tissue swelling that grows slowly and commonly found in the lower extremity of morbidly obese patients, however, unusual sites such as retroperitoneum in non-obese patients were also reported. The disease is usually treated by simple surgical resection.

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