Surgical Management of Massive Localized Lymphedema: A Case Report

Joseph Daodu MD, Alexys N. Ramos DO, Rainna Coelho MD, Natalia Cavagnaro DO, Mike K. Liang MD

HCA Houston Healthcare

UNIVERSITY of HOUSTON COLLEGE OF MEDICINE

Background

- Lymphedema is a pathologic accumulation of interstitial fluid and fibroadipose tissue
  - Unknown etiology; Possibly abnormal lymphatic vessel development or reduction in efferent lymphatic flow due to excess adipose tissue
  - Secondary lymphedema results from another identifiable source (trauma, surgery, hypothyroidism, cancer, medication induced, etc.)
- Massive Localized Lymphedema (MLL) is a large, deep, nonneoplastic benign pedunculated lymphedematous mass
  - Considered a form of secondary lymphedema, where excessive weight interrupts lymphatic channels, leading to a localized soft tissue mass.
  - Most commonly in morbidly obese individuals on the lower limbs
  - Typically present with heaviness, tightness, discomfort, swelling of the impacted limb. Two thirds of cases are unilateral.
- Severity can be assessed through measurements of limb circumference or limb volume (using water displacement, optoelectronic volumetry or calculation of limb volume).
- No gold standard system for clinical staging of lymphedema, though the International Society of Lymphology (ILS) staging system is widely accepted and used by many for initial assessment

Clinical Presentation

- 37 year old male with PMHx of obesity and hypertension, presenting with MLL involving bilateral thighs, associated with ulcerations and recurrent episodes of cellulitis
- Massive pedunculated lesions, thickened skin with induration and peau d'orange, draining wounds, chronic bacterial and fungal infections despite antibiotics
- Symptoms began as localized edema on the right thigh, and progressed for the next three years into bilateral growing masses
- Weight of 503 lbs with a BMI of 69. Patient motivated to lose weight, but efforts towards lifestyle changes limited due to grossly restricted mobility
- Patient was medically stable, chronic ulcerations without active infection, and patient was scheduled for elective surgery

Treatment

- Conservative management is first line tx, includes good skin hygiene, compression garments, daily exercise and decongestive therapy
- Indications for surgical intervention: Recurrent cellulitis, limitation of function, persistent pain, diminished quality of life, and leakage of lymph into a body cavity or organ, and disfigurement
  - Physiologic interventions are aimed towards creating new lymphatic channels to increase drainage.
  - Reductive interventions involve removal of fibroadipose tissue, and are preferred for cases in advanced stages (ILS stage II and III)
- Patient classified as stage III , taken to OR for resection of bilateral MLL masses
- Skin was loosely approximated, incisional negative pressure wound therapy (iNPWT) and compression wraps were applied
- Weight on POD 1: 438lbs. Largest specimen 38x33x16cm, weighing 34.2lbs

Intraoperative Findings

Fig. 1: Pre-operative physical assessment in clinic

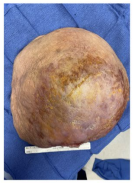

Fig. 2: Surgical specimen, L medial thigh mass

Fig. 3: Surgical specimen, R medial thigh mass

Fig. 4: Immediate post-operative period

Discussion

- MLL mimics malignant conditions, and is often misdiagnosed as neoplasm such as sarcoma
- Benign lesion but is associated with increased risk of angiosarcoma or Stewart-Treves syndrome
- Postoperative care involves regular dry dressing changes and early, frequent ambulation
- Patients should be warned to expect excess drainage for 3-8 weeks following surgery
- Unlike classic lymphedema, MLL is localized and irreversible. Conservative treatment is rarely successful
- Physiologic procedures are best utilized in patients with early stage disease, prior to the deposition of excess fat and tissue necrosis, suggesting early diagnosis and management may improve patient outcomes

Conclusion

- Incidence of MLL is rising with the increased prevalence of morbid obesity. Due to the associated morbidity of this condition, it is important for surgeons to acquaint themselves with the technical challenges of resecting these lesions.
- Reductive intervention is the recommended treatment for MLL for individuals with impaired ambulation, as immobility promotes complications including worsening obesity.
- Further research is required to identify pathogenesis, pathophysiology and further treatments
- A multidisciplinary approach with meticulous post operative care is required for best patient outcomes

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