

Case Report

Lipoedema: The Immitator “A case report with a review of literature”

**Uwais RUH, Dr.Khathija Hasan*

¹Department of General Surgery, Department of Anaesthesia, RIPAS Brunei

uwaisriazulhasan@gmail.com

Abstract

Lipoedema is a condition characterised by symmetrical lower limb enlargement. It was first described by Allen & Hines who coined the term lipoedema. The result of subcutaneous deposition of fat gives rise to fatty swelling in the buttocks, thighs and lower limbs sparing the feet. It is associated with obesity and hence upper limbs and rarely trunks are also affected. This Clinical condition affects females predominantly 1:4 and hence has a profound effect on the psychology therefore it is not uncommon to see these patients with a low self esteem, socially isolated and depressed. Due to its resemblance to lymphoedema it is not unusual to be misdiagnosed, thus leading to a spiral of inappropriate tests and improper treatment outcome.

Keywords: Lipoedema, Lymphoedema, Obesity

*Author for Correspondence.

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Introduction

The first description of lipedema was in 1940 by Allen & Hines when they noted that among the patients attending the Vascular clinic especially females had excess fat deposits in their Lower limbs. It was noted by Moncorps that the skin and joints of these patient were lax. Rapprich has describe lipodema as a condition characterised with abnormal fat depositions along the extremities. It is essentially a disorder of Adipocytes and a lot of studies have described it as Lipodema. We describe this case report with a review of literature of Lipodema along with the Spectra of lower limb disorders viz Obesity, venous disease, lymphatic insufficiency and lympho lipodema that Lipodema is easily labelled with.

Case Presentation

A 43 yr old Malay who was Obese was admitted to the Intensive care unit with a history of Painful swelling of her left buttock and lower legs for two months. A history of

low grade fever for a month and an exacerbation of her COPD for few weeks. A diagnosis of Morbid obesity with lymphoedema & Cellulitis was made and she was started on Antibiotics. The lower limb and buttocks were symmetrical bilaterally and noted to be out in proportion to the rest of her body. The fat distribution was noted to spare her feet bilaterally. An USG of the Gluteal region and Left lower limb revealed no evidence of an abscess collection. She did not have varicosities of lower limb, no pitting oedema and the stemmers sign was negative. She was noted to be depressed bed ridden for months and had OA of both her knees. No other member in her family had a similar condition. Her upper limbs were also noted to be enlarged symmetrically. (Figure 1).



Figure 1. This patient was non responsive to the initial line of treatment and hence a referral to a senior consultant was made. A Bed side USG showed a thickened subcutaneous tissue, our Diagnosis was revised to Cellulitis in Lipodema.

Discussion

Lipedema is a condition affecting women exclusively progressing gradually from puberty to middle age. Although the name implies oedema it is a disorder of adipocytes. The incidence is reported at 10% at lymphoedema clinics [18]. It was first reported in 1940 by Allen and Hines described a condition they called

“lipedema”. They recognised a subgroup of patients – all female – attending the vascular leg clinic at the Mayo Clinic with extensive lower limb swelling & sparing of the

feet. The etiology of lipedema is unknown. Many patients

with lipedema have a family history of similarly enlarged legs^[20] thereby implying a genetic link. In rare cases of lipedema males have been described. In 97% of the cases, lipedema is located in lower limbs and in 31% in upper extremities^[7,16]. Lipoedema is symmetrical in distribution from the hips to regions of thigh and lower limbs. Some patients report an increase in limb swellings towards the evening and an increased sensitivity to pain in their lower limbs this may be attributed to plantar fasciitis and altered gait alterations that comes with hip and knee joint damage as a result of the changing dynamics that accompany an increase in lower limb weight.

Clinically it is easily to be misdiagnosed as lymphoedema^[1]. Lymphoedema as we know occurs secondary to lymphatic dysfunction. The swelling developing as a result of seeping of protein rich interstitial fluid in the skin and subcutaneous tissue. Lymphoedema can either be unilateral or bilateral whereas lipoedema is bilateral. Lymphoedema is painless and commonly present a positive Stemmer sign this is because there's skin thickening with fibrosis making it impossible to pinch the skin of the second digit on the toe, whereas in pure lipoedema, Stemmer sign is negative, the feet are spared and the fat deposits begin abruptly above the ankles the so called cuffing sign. While lymphedema is usually painless, whereas lipedema patients report spontaneous or even minimal pressure induced tenderness. Lipoedematous skin gets easily bruised. Lipoedema is distinguished from obesity because it usually appears in lower limbs, more rarely in arms, whereas obesity may affect the whole body and is associated with an increased body mass index (BMI) while lipoedema can occur in patients with a normal BMI. This is important to recognize since lipoedema is frequently misdiagnosed as obesity as was in our patient. Varicosity are often seen among lipoedematous patients^[5] Can severe venous insufficiency be associated with lipoedema? it is reportedly albeit rare. The combination of lymphatic insufficiency and lipoedema is called lipolymphedema or lympho-lipedema. Morbus Dercum is a differential that needs to be considered especially with postmenopausal women. The limbs, trunk, the pelvic area and the buttocks have painful fatty depositions^[15,60]. Morbus Dercum usually has several typical accompanying symptoms like recurrent headache and depression that have not been described in lipoedema. Lipedema appears to be a distinct clinical entity but maybe confounded with lymphedema. The following descriptions pinpoint the most significant differences between lymphedema and lipedema^[16] Lymphedema can be either unilateral or bilateral while lipedema is always bilateral Lymphedema patients commonly present a positive Stemmer sign, whereas in pure lipedema, Stemmer sign is negative, the feet are spared and the fat deposits begin abruptly above the ankles (cuffing sign) in Lymphedema is usually painless and is associated with skin changes thickening of the skin, erythema of the affected area and thickening of the skin, which appears as peau d'orange skin and woody edema, Elephantiasis nostra verrucosa, an area of cobble-stoned, hyperkeratotic, papillomatous plaques most commonly seen on the shins with fissuring and ulcerations. In lipedema patients report spontaneous or even minimal pressure

induced tenderness. Lipoedematous skin gets easily bruised. lipedema results from the subcutaneous deposition of fat and occurs independently of lymphatic or venous insufficiency^[19]. Fifty percent of women attending a lymphoedema services with lipoedema were also found to have generalised obesity^[18], Lipoedema is distinguished from obesity because it usually appears in lower limbs, more rarely in arms, whereas obesity may affect the whole body presents with an increased body mass index (BMI), Lipoedema on the other hand can occur in patients with a normal BMI. It has been reported that Lipedema, unlike obesity, hardly ever responds to low-calorie diet^[9]. A restricted dietary intake is advised as a preventive measure against further weight gain in patients with lipoedema^[15]. As a differential lipohypertrophy should also be considered. As Lipoedema is often preceded by symmetrical lipohypertrophy, it becomes difficult to diagnose one from the other^[16,13].

Pathology and pathophysiology

Microangiopathy has been proposed by Földi and Földi with area of the affected adipose tissue seeping in higher permeability proteins and increased capillary fragility leading to spontaneous and minor traumatic injury^[9]. Hypoxia is reported a potent inducer of angiogenesis and this factor has a significant role in lipedema^[6]. There are fourfold higher levels of plasma vascular endothelial growth factor (VEGF) levels at baseline than normal values of VEGF^[6].

The European Lymphology Society describes lipoedema with five subtypes.

- Type I—Involves buttock and hips/ Saddle bag phenomenon
- Type II—Involves Buttock to knees
- Type III—Buttock to ankles
- Type IV—Arms & Legs
- Type V Lipo-lymphoedema

Diagnosis

Lipoedema is usually made on a Clinical basis^[16]. Usually, the medical history and clinical examination are enough to suspect the diagnosis. Bed side investigations like Duplex ultrasound examination of lipoedema shows a thickened subcutaneous issue with increased echogenicity, whereas in lymphedema, the typical echo-less gaps are not compressible. Bed side sonography is superior to clinical examination and is increasingly being used as an accessory tool in the diagnosis of lipoedema^[12,18]. It is reported computed tomography scan has a sensitivity of 95% and specificity of 100%^[13] for the diagnosis of lipoedema. The subcutaneous fluid accumulation, the honeycomb pattern and muscle enlargement are not seen in lipoedema, in contrast to lymphedema^[13]. Magnetic resonance imaging and its modified form, magnetic resonance lymphangiography have an important role when the lymphatic involvement is unclear^[9].

Treatment

Lipoedema is a chronic disorder and hence accurate diagnosis with Multi Disciplinary approach is the Step forward. A dedicated team of Specialists offering a

Psychological Assessment, The Role of the Dietician in a Multidisciplinary weight management programme with the Aim for weight Stabilization needs to be emphasised. Physicians for management of Co Morbidities. It is reported that Dieting, diuretics, leg elevation, and compression appear to be of minimal benefit ^[11,9] and attempts to treat invasively via lipectomy or liposuction ^[13] are not recommended because they risk causing mechanical damage to the lymphatics ^[16].

There are some German authors however who recommend tumescent liposuction results of which would benefit for lipodematous limbs without serious side effects ^[13]. Liposuction around the knees decongests and is reported to drastically improves pain perception, mobility and range of motion, especially at knee joints and results appear to be sustained ^[20,9]. There are other authors ^[24] state that liposuction is the treatment of choice for patients with an acceptable waist measurement and has stressed on weight management in the post op period. The role of Compression is to reducing ankle cuffing. Lipoedema with venous oedema can be managed with low levels of compression.

Conclusions

It is imperative that an accurate Diagnosis is made and early effective treatment is initiated to avoid pit falls in the Chronic Management of these disorders. Multidisciplinary treatment is the step forward.

Conflict of Interest Statement

No conflict of interest to be declared.

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