

**Case Report.**

## **Lipedema: An underdiagnosed Condition Predisposing to Lymphedema.**

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### **INTRODUCSION:**

Lipedema of the lower limbs is a common but rarely diagnosed disease or frequently confused with obesity. It is a chronic disease of lipid metabolism that results in the symmetrical impairment of fatty tissue distribution and storage combined with the hyperplasia of individual fat cells. Lipedema occurs almost exclusively in women and is usually associated with a family history and characteristic features. It can be diagnosed based on clinical history and physical examination. Lipedema is usually symmetrical, extends from hip to ankle but spares the feet and it is often painful to palpation <sup>(1,2)</sup>.

Disease onset is usually at, or soon after puberty or during periods of hormonal changes such as pregnancy. Lipedema results in considerable frustration and distress resulting from the cosmetic appearance <sup>(3)</sup>. This condition bears some clinical resemblance to lymphedema and is frequently misdiagnosed as such.

However, in contrast to lipedema, the swelling of lymphedema is due to accumulation of protein-rich interstitial fluid within the skin and subcutaneous tissue caused by lymphatic dysfunction. Lipedema is usually diagnosed after exclusion of other cause of lower limb edema such as venous or lymphatic obstruction. It is believed that lipedema may predispose to lymphedema by virtue of extrinsic pressure of the fat cells on the tiny lymphatic vessels <sup>(4)</sup>.

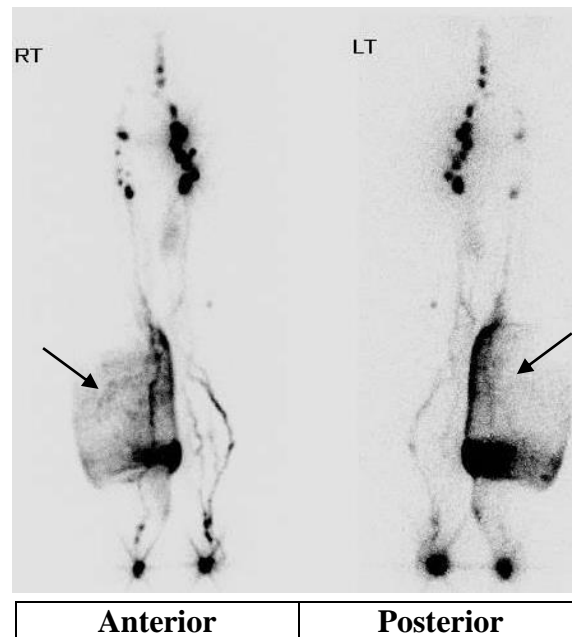
This report describes three female patients presented to Nuclear Medicine Section, Department of Clinical Imaging at Hamad General Hospital, for evaluation of massively enlarged lower extremities. Three cases finally diagnosed with lipedema, based on clinical picture of massive lower extremities swelling sparing the feet and unremarkable ultrasound Doppler of lower limbs deep veins.

The first case was a 59 years old obese lady with history of bilateral lower limb surgeries (lipectomy/liposuction) in France. She complained of left thigh localized swelling, which has increased in size significantly and became painful. She had also intermittent fever. Physical examination revealed massive bilateral lower extremity swelling with minimal pitting sparing the feet. The patient had a negative duplex venous imaging, thus ruling out any vascular causes of the edema. The lymphoscintigrams revealed significant lymph-stasis and dermal backflow at the right leg without significant obstruction as the proximal thigh lymphatics and draining inguinofemoral nodes were visualized. This limb expresses lipolymphedema. The left lower limb revealed normal lymphoscintigraphy but for localized lymph

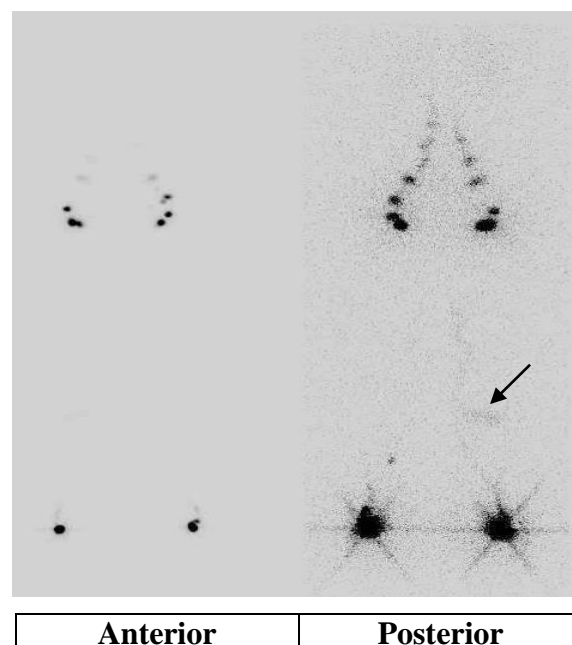
collection at the thigh (lymphocele), (*Figure 1*).

The second case was 30 years old obese female with long standing bilateral symmetrical lower limb edema. In addition she complained of localized swelling of the posteromedial aspect of the right leg. The Duplex ultrasound was normal and so was the lymphoscintigrams, except for a localized lymph collection, corresponding to the right leg localized swelling (lymphocele) (*Figure 2*).

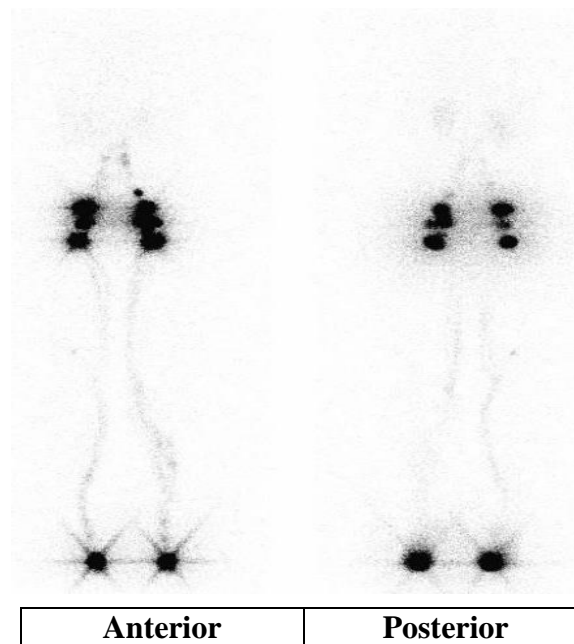
The third case is 33 years old average-built female with 6 years history of bilateral massive symmetrical edema of the lower limbs sparing the feet, the onset of which is related to the last month of pregnancy. Both venous Duplex ultrasound and lympho-scintigraphy was unremarkable, (*Figure 3*).



**Fig. 1:** Lower extremity Tc-99m Nano colloid lympho-scintigraphy revealing significant interstitial lymph-stasis in the right leg with dermal back flow, more at the medial side of the leg, the proximal lymphatic channels of the right thigh and the right inguino-femoral nodes were visualized thus excluding significant obstruction. The left leg showed patent normal lymphatic drainage except for a localized area of lymph stasis seen at medial side of upper left thigh representing lymphocele (arrow).



**Fig. 2:** Lower extremity Tc-99m Nano colloid lympho scintigraphy reveals normal lymph drainage; with localized interstitial lymph-stasis at the right leg posteriorly representing a lymphocele (arrow).



**Fig. 3:** Normal lymphoscintigrams of both lower limbs are displayed with normal visualization of the main medial lymphatic channels of the legs and thighs and fairly symmetrical appearance of the draining inguinofemoral and external iliac nodes.

## DISCUSSION:

Lipedema is a disorder characterized by symmetric enlargement of the legs due to deposits of fat beneath the skin. It is a common condition that is underdiagnosed, occurring almost exclusively in women and affecting up to 11% of women <sup>(4,5)</sup>.

The diagnosis of lipedema is frequently missed because it clinically resembles lymphedema. However, sparing of the feet is a clinical characteristic of lipedema. Also, after exclusion of systemic cardiac, renal or hepatic causes of limb oedema, a normal lympho-scintigraphy and a normal venous duplex scan would then support the diagnosis of lipedema and exclude the

diagnosis of other familiar causes of lower limb oedema <sup>(6)</sup>.

The etiology of lipedema is unknown, but there are evidences of hereditary and hormonal influences <sup>(4,6)</sup> *Allen and Hines* first described it in 1940 as having a female predilection and a family history of similar problems <sup>(3)</sup>.

The body's hormonal milieu also appears to play a role given that lipedema occurs almost exclusively in women and onset occurs typically during puberty or other periods of hormonal change, including pregnancy and menopause <sup>(4)</sup>.

This applies to third case, where the onset of lower limb oedema was related to pregnancy and to our first case where the onset was related to menopause. The second case is a 30-year-old unmarried female who could not provide a definite answer about the onset of her lower limbs swelling but empathized that the oedema was long standing. We presume that the onset might date back to puberty.

Unlike lipedema, patients with lymphedema will have a positive Stemmer sign. They may also have a history of renal, hepatic, or vascular abnormalities. The diagnosis can become complicated in patients with longstanding lipedema who may develop lipolymphedema. In lipolymphedema, the accumulation of adipose in subcutaneous tissues causes lymphatic dysfunction and subsequent lymphedema as seen in the right limb of our first case<sup>(3)</sup>.

Localized contained lymph stasis (lymphocele) was seen at the left thigh of our first case and at the right leg of our second case. It is not known whether such a lymphocele seen in 2 out of our 3 cases is a chance finding or a tendency in lipedema. The fact about this query necessitates further extended studies on large samples of patients with lipedema.

To date we did not find in the literature review information about possible association between lipedema and lymphocele.

Lipedema, lipolymphedema and lymphoceles may be inter-related, as hyperplasia of the fat cells causes pressure on the normal lymphatic channels, which in turn results in interstitial leak of lymphatic fluid causing lymphedema. In one of three cases lipolymphedema was diagnosed (one of six limbs), where lymphedema was extensive. In two of the three cases (two out of six limbs) localized lymphoceles were seen<sup>(2,6)</sup>.

The diagnosis of lipedema may involve ultrasound, MRI, lymphangiogram and/or lympho-scintigraphy. However, test result may be normal in the early stages of the disorder<sup>(7, 8, and 9)</sup>. Whereas normal lympho-scintigraphy was encountered in our third case. There is no one effective treatment for lipedema. Exercise, diet and nutrition and emotional support are important factors.

The main conservative treatment is complete decongestive therapy (CDT), which involves several approaches, such as manual lymph drainage by a massage technique, compression therapy by long elastic leg socks and physical mobilization by exercise<sup>(10,11)</sup>.

Follow up of third case revealed that she is under CDT with regular lymph drainage massage sessions, exercise and compression therapy.

Surgery may be considered if conservative therapy is not effective. Surgical option may include liposuction using specialized technique for lipedema, such as water jet-assisted liposuction and excision removal of large fat deposits <sup>(7, 11)</sup>.

In fact, the first patient underwent liposuction in France some 6 months before she came for lympho-scintigraphy

which revealed extensive lymphedema at the right leg and a painful lymphocele at the left thigh.

## **CONCLUSIONS:**

The current knowledge about lipedema is scarce, but the scientific interest is increasing. More studies are required to know the real prevalence and to reach an earlier diagnosis of this disorder. Lymphedema is a likely complication of lipedema, whether diffuse or localized in the form of lymphocele.

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