

Case

P R E S E N T A T I O N

LIPOEDEMA A DISTINCT CLINICAL PRESENTATION

Case report of swollen legs - An uncommon cause for a common problem - swollen legs

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Introduction

Allen and Hines first described lipoedema under this name in the medical literature in 1940¹. It is known by a number of different names including lipodystrophy, adipocyanosis², painful fat syndrome³ and a subgroup called rusticanus monocorps lipoedema.

It is a distinct entity from classical primary or secondary lymphoedema (Table 1). The aim of this article is to present clinical characteristics of lipoedema to assist in diagnosis of this interesting condition and to help distinguish from lymphoedema.

Case Presentation

History

A 59-year-old female presented with painful “swollen legs” to a phlebology clinic .

The enlargement of her legs first appeared in her late teens. She stated that she had always had thick ankles and her sister was of a similar build. Her previous diagnosis had been lymphoedema for which she had been prescribed class one and two compressive calf stockings. The patient found stockings unhelpful and painful.

There was no history of cellulitis, eczema or ulceration. Her mother suffered from varicose veins but there was no family history of thrombophilia.

The patient had no visible varicose veins.

The patient stated that her legs were tender to touch and there was no relief from her “swelling” on rest or elevation. Diuretics were ineffective and while she had no problem wearing shoes she would get occasional swelling of her feet on hot days and after prolonged standing.

ABSTRACT

Case presentation of 59-year-old female with symptomatic swollen legs is presented.

A diagnosis of lymphoedema as cause for symptoms and appearance had previously been given.

This patient’s history and physical examination with colour compression ultrasound is consistent with diagnostic criteria for a condition called lipoedema which is a non-pitting oedema with abnormalities in lipocytes and abnormal prelymphatic pathways.

This case will discuss relevant history, examination findings and the role of imaging in diagnosis.

Table 1.

Deficiency in Lymphatic Drainage (High Protein Oedemas)

a. Primary Lymphoedema

- i. Congenital Lymphoedema (agenesis or hypogenesis)
 - Genetic and hereditary (Milroy’s)
- ii. Lymphoedema praecox (hypogenesis)
 - Begins at puberty and may be triggered of by trauma, wounds, infections or allergies
 - Initially mainly unilateral but 50 % develop bilateral disease
- iii. Lymphoedema Tarda (late onset)
 - After age of 30

b. Secondary Lymphoedema

- i. Obstruction to the lymphatic tracts
 - Extrinsic
 - Tumour
 - Fibrosis of lymphatic walls
 - Intrinsic
 - Infection (Lymphangitis)
 - Parasites (Filariasis)
- ii. Damage to lymphatic ducts or lymph nodes
 - Surgery
 - Radiotherapy
 - Trauma

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Over the last several months she had increasing pain in both feet and intermittent burning sensation at the end of the day. This had been the reason for her presentation.

The patient's past history was unremarkable, in particular there was no history of DVT.

On examination the patient was found to be obese generally with a normal cardiovascular examination including normal blood pressure and JVP readings.

Examination of the legs revealed the following findings:

- Pulses, reflexes and sensation normal
- Allodynia over lower anterior shins bilaterally
- Left calf and right calf were equal in diameter
- No visible varicose veins, minor telangiectasia only
- No signs of chronic venous dysfunction
- No swelling of foot or toes
- Stemmer sign negative*
- Swelling non-pitting
- Skin soft, no induration

* Stemmer test for lymphoedema (positive = inability to pinch the skin of the second toe between first two fingers)



Figure 1: Lipoedema post view



Figure 2: Lipoedema "Egyptian Column" shape.

- Note
- fat pad anterior to lateral malleolus
 - Ring of fatty tissue just above ankle



Figure 3: Lipoedema distal view

- Note:
- Fat pad and distinctive "ring" at ankle
 - Foot and toes normal, negative Stemmers sign
 - No oedema noted distal to the ankle

Investigations

Pathology

- FBC/ESR, electrolytes, albumin normal
- CPK normal
- TSH normal range
- Total cholesterol and triglycerides mildly elevated

Colour Duplex Ultrasound Left leg

- Normal Deep system
- Dilated, incompetent GSV from groin to ankle
- 6.7 cm x 3cm x 2.3 cm Baker's cyst (see figure 4)
- Normal skin thickness at ankle level
- Minimal or trace oedema, absence of "lymphatic lakes"
- Marked increase in subcutaneous fat at the ankle level with homogeneous echogenic appearance (see figure 5)



Figure 4: Cross sectional ultrasound left popliteal fossa

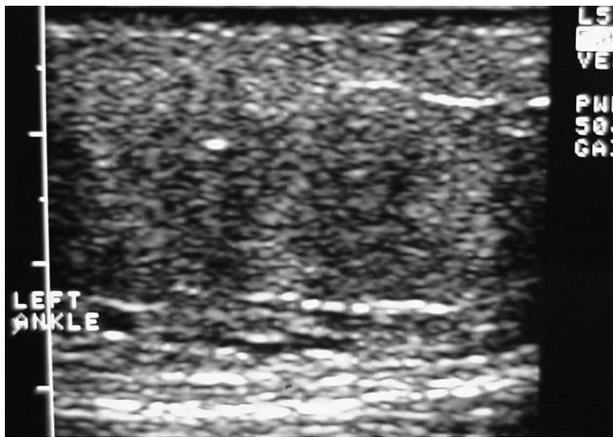


Figure 5

Discussion

Lipoedema is a diagnostic consideration in female patients who present with bilateral non-pitting swelling of the legs. It needs to be distinguished from low protein oedemas and bilateral lymphoedema.

Allen and Hines described lipoedema in 1951⁴ as a condition in females with symmetrically enlarged legs and

buttocks with sparing of the feet. Prominent malleolar fat pads were noted in some patients. The lower limb tissue was described as soft and pliable, unlike that seen in the later stages of lymphoedema.

Elevation and compression stockings were noted to be of little assistance, with stockings frequently causing considerable discomfort. Unlike lymphoedema, a history of cellulitis was never or rarely obtained, whereas a family history of the condition was frequently obtained.

Plastic surgeons, Rudkin and Miller⁵ summarised the history and physical signs of lipoedema and lymphoedema as Table 2.

The most striking clinical features include prominent malleolar fat pads and the absence of any swelling distal to the ankles.

The character of the subcutaneous tissue is dramatically different from that seen in patients with lymphoedema. There is absence of the dermal and perivascular fibrosis consistently seen in patients with lymphoedema.

Skin and subcutaneous biopsy of patients with lymphoedema show increased fibrous tissue in perivascular and dermal areas.

Biopsies of patients with lipoedema show absence of skin thickening and fibrosis in the subcutaneous fat.⁶

Table 2.

Signs and Symptoms	Lymphoedema	Lipoedema
Sex	Male or Female	Female
Generalised Obesity	+/-	+/-
Unilateral /bilateral	Usually unilateral	Bilateral
Nature of swelling	Firm	Soft
Pitting	Often Marked	Minimal
Pain on Pressure	Rare	Common
Foot involvement	Yes	No
Effect of Elevation	Initially beneficial	Ineffective
Family history	Rare	Often
History of Cellulitis	Common	Rare
Stemmer sign	Positive	Negative

Pathogenesis

Until recently, lipoedema was considered solely as genetically determined obese legs.⁸

However, Partsch⁹ found abnormalities in the prelymphatic channels by indirect lymphangiography.

The question arises as to whether the lymphatic alterations are primary or secondary to the fat deposition.

The fact that lipoedema is exclusively located in the legs, in the majority of cases in the lower third, would suggest that hydraulic factors play a main role in the genesis of increase of subcutaneous fat.¹⁰

In summary, there certainly exists alteration in the lymphatic system. This can be primary or secondary to the excess of subcutaneous fat. The role of the venous circulation in the pathogenesis of the disease is unknown.

Table 3.

Ultrasound findings	Lymphoedema	Lipoedema
Skin Thickness	Increased	Normal
Subcutaneous tissue	Increased	Increased
Ultrasound	Subcutaneous tissue shows increased areas of echogenicity of irregular pattern demarcation by areas more or less without echo ("lymphatic lakes")	Subcutaneous fat with homogeneous echogenic appearance

Treatment

Very little appears in the literature regarding effective treatment.

There is mention of complex decongesting therapy by Foldi¹¹

Surgical treatments that have been tried include liposuction and minimal skin and subcutaneous excision, extensive skin and subcutaneous excision. The results for extensive surgery could be described as poor as all patients in one trial suffered long term foot oedema. Liposuction provided better results.¹²

Conclusion

Lipoedema is a distinct entity from primary or secondary lymphoedema and should be adequately differentiated on the basis of history and examination.

Lipoedema should be distinguished clinically from lymphoedema on the basis that lipoedema has fatty deposits (anterior fat pads to lateral malleolus and a ring of fat just above the ankle) and particularly shows no sign of induration or pitting oedema as seen in lymphoedema.

It can be characterised as a symmetrical enlargement of the lower limbs of females with soft fatty tissue and minimal oedema. Prominent malleolar fat pads are present and the feet are normal. Skin changes that occur in lymphoedema are completely lacking.

Investigations including Ultrasound may be of benefit to confirm diagnosis and exclude other pathologies. In this case presentation, multiple pathologies were present. They included lipoedema, concealed superficial venous insufficiency and a Baker's cyst.

The art of medicine is to determine the main causes for this patients symptoms so that accurate considerations can be given to proposed therapeutic options.

Further research needs to be undertaken on the pathogenesis of lipoedema.

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