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Diagnosis and management of lipoedema – a review paper

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Abstract

Lipoedema is a chronic progressive disorder of adipose tissue leading to an enlargement of lower extremities. It is considered to be rare; however, the prevalence of the disease is underestimated because it is commonly misdiagnosed as obesity or lymphedema and the general awareness is poor. The etiology of the disorder is considered to be multifarious, including genetic inheritance, hormonal imbalance and microcirculation alterations. Diagnosis is mainly based on medical history and physical examination. Management of lipoedema is focused on reducing the symptoms, improving the quality of life and preventing further progression of the disease. The aim of this paper is to raise the awareness of the disease and provide appropriate clinical guidance for the assessment of lipoedema. We searched through the PubMed/MEDLINE database and took into consideration all of the results available as of 6 September, 2020 and outlined the current evidence regarding lipoedema epidemiology, etiology, clinical presentation, differential diagnosis, and management.

Better understanding of lipoedema is crucial for establishing an early diagnosis and a proper treatment, which in turn will reduce the psychological and physical implications associated with the disease.

Key words: lipoedema, lymphoedema, lipolymphoedema

Introduction

Lipoedema is a chronic progressive disorder of adipose tissue. It affects almost exclusively women and often develops around the age of puberty. Lipoedema is considered to be a rare disorder, however the prevalence of the disease is underestimated because of a poor awareness and common misdiagnoses. Recent findings estimate the prevalence of lipoedema to be 1:72.000 in the general population.¹ While the etiology of lipoedema is unknown, multiple mechanisms have been suggested, including genetic inheritance, hormonal imbalance, and microcirculation alterations. Abnormally disproportionate distribution of fat leads to a bilateral enlargement of lower extremities and, in some cases, of arms. Lipoedema is often misdiagnosed as obesity or lymphoedema, leading to an improper management of the disorder. However, a thorough assessment would reveal the disproportion of shape, sparing of the feet and resistance to dieting in patients suffering from lipoedema. Treatment consists of conservative and surgical options. Increased awareness and a better understanding of the disease would enable clinicians to distinguish lipoedema from other similarly presenting diseases and to treat patients at the earlier stages.

Etiology

The etiology of lipoedema remains unknown, although it is likely that multiple factors are involved in the development of the disease. Genetic predisposition, hormonal imbalance and histopathological changes have been suggested.

The exact genes responsible for the development of lipoedema have not been fully identified; however, a genetic predisposition has been reported in 60% of patients suffering from the disease.² Furthermore, a study including 330 family members has suggested a possible autosomal dominant inheritance of the disease. Positive family history has been reported in affected patients in values ranging between 16% and 64%.³ Even so, given the lack of sufficient research, lipoedema is currently excluded from the list of hereditary diseases.

Lipoedema affects almost exclusively women, with the age of onset at the start or following puberty in 78% of cases.⁴ There have also been reports of the disease developing after pregnancy or even after menopause,⁵ as well as in male patients as a result of hormonal imbalance.⁶ Estrogen is known to modulate lipid metabolism through estrogen receptors, but the exact role of estrogen in the pathophysiology of lipoedema is unclear. Defects of estrogen receptors and altered signaling pathway are suggested.⁷

Microangiopathy is a histopathological feature reported in patients with lipoedema. This finding is supported by elevated VEGF plasma levels, a known stimulative factor for angiogenesis, found in patients with lipoedema.⁸

The hypertrophy of adipocytes, found in immunohistochemical analysis, may lead to their necrosis and subsequent macrophage infiltration, activating the inflammation response in the affected area. Both angiogenesis and inflammation are thought to be independent factors altering the microcirculation in patients with lipoedema. A certain study points out an increased lymph flow at the early stages of the disease and a decrease in the lymphatic flow in advanced stages.⁹ It should be mentioned that this finding is inconsistent with another study using the same method of lymphoscintigraphy to assess the flow of lymph and lymphatic drainage in the affected areas.¹⁰ These contradicting results suggest lipoedema may be responsible for the damage of microlymphatic vessels through adipocytes hypertrophy, rather than a lymphangiopathy being the primary cause. Further research is needed to clarify these findings.

Clinical presentation

While lipoedema affects almost exclusively women and usually starts during or after puberty, its onset has been reported up to the third decade of life.¹¹ Family history may be commonly positive. The disorder is characterized by a bilateral and symmetrical enlargement of legs, hips and buttocks, and in some cases affected areas may also include arms (gynoid type of fat distribution). Hands, feet and trunk are usually spared, resulting in the disproportionate shape of the body. The sharp distinction between enlarged legs and spared feet is referred to as a “cuff sign”, and disproportion between hips and waist is called a “riding breeches” sign.⁷ Pain is a commonly reported complaint, either elicited by touch or occurring spontaneously. The affected area is tender to touch and may easily bruise, possibly as a result of microcirculation alterations.¹² Orthostatic edema, which also may result from microangiopathy, is often present in patients suffering from lipoedema and shows no improvement with limb elevation. Another distinctive feature of the disease is resistance to dieting. Many patients are diagnosed with obesity and recommended to lose weight, however losing weight shows little to no effect on lipoedema.

This may lead to reduced self-esteem, negative effects on mental health and depression.⁴ High prevalence of misdiagnosis, poor understanding of the condition and inappropriate dietary recommendations are the challenges that both medical professionals and patients face when assessing the condition.

Although the development of lipoedema is unpredictable and differs among individuals, the disease may gradually progress over time. Most researches describe three stages of lipoedema when assessing the severity of the disease. In stage I, skin is usually soft and smooth, and the underlying subcutaneous tissue is thickened; in stage II, the skin surface has an irregular texture and small nodules are present in the subcutaneous tissue; in stage III, deformed fat deposits occur, causing impairments of function. Further progress of the disease leads to secondary lymphoedema and is identified as a stage IV (lipo-lymphoedema).¹³

Differential diagnosis

A proper assessment and an accurate diagnosis are an important part of management of lipoedema. However, due to the poor awareness of the condition amongst health professionals and the lack of sufficient research, the disease is often underdiagnosed or misdiagnosed. The differential diagnosis includes the disorders with similarly presenting symptoms, such as obesity or lymphoedema. Another challenge faced by clinicians when assessing the disorder is the absence of specific diagnostic markers or pathognomonic diagnostic measures for lipoedema. Warranted the clinician is familiar with the disease, the diagnosis is mainly based on medical history and physical examination.

Most distinctive features in patient history include gender, as lipoedema affect almost exclusively women, and positive family history. In lipoedema, progression of the disease is excluded to the affected areas, mostly legs, whereas obesity affects the whole body and lymphoedema spreads proximally in most cases.³ There is no response to dieting and minimal effect of elevation on oedema reduction in lipoedema. A physical examination reveals disproportion in body shape and a symmetrical bilateral enlargement of legs, hips and buttocks. Lymphoedema may be unilateral and asymmetric, while there is no shape disproportion in obesity. Further distinctive features of lipoedema are the typical sparing of the feet and the presence of a retromalleolar fat pad, which may cause the lack of Achilles tendon definition. Pain on affected limbs and easy bruising is present in lipoedema, but lacking in obesity. Contrary to lymphoedema, Stemmer sign, which is the ability to pinch the skin at the base of the second toe, is negative in lipoedema.¹⁴

Management

Early diagnosis and treatment are essential for managing the symptoms of the disease and preventing its progression. The multidisciplinary therapeutic approach should take patient's needs, expectations and abilities into consideration. Main goals of the management of lipoedema include reducing the symptoms, improving the quality of life and preventing further progression of the disease. There are some limitations to the treatment, stemming from complex and not fully identified etiology, as well as limited research regarding the subject.

Currently, options of management include education and psychosocial support, enhancing self-care approach, weight management through diet and physical exercise and skin care. Conservative treatment involves compression therapy, manual lymphatic drainage and mobilization. Surgical options include liposuction and lipectomy.

A personalized approach based on a collaboration between the patient and the medical professional is recommended.¹⁵ Psycho-education implemented at the early stages of diagnosis is essential in facilitating the self-care approach. Psychological support is an important component of the management of lipoedema and should not be overlooked by clinicians. Although the effects of dieting on lipoedema are little to none, lifestyle changes, including weight management and physical exercise, prevent obesity and reduce the risk of obesity-related comorbidities, enhance wellness and improve mobility. Skin care helps to maintain good skin health and avoid developing cellulitis. Conservative treatment may be prescribed by a clinician to reduce oedema. Compression therapy involves the use of compression bandaging, compression sleeves or other types of compression garments. Manual lymphatic drainage is a type of a light massage technique, which stimulates the lymph flow in the affected area, following the anatomic lymphatic pathways. Conservative therapy aims to reduce the pain and discomfort by improving the lymph flow from the affected limbs.

Simultaneously, because of the tenderness and discomfort in the lower extremities, the conservative therapy is challenging for these patients and requires a careful, personalized assessment.¹⁴ Surgical management is recommended when the conservative measures have resulted in minimal to no improvement of the condition.⁷ Surgical options aim to permanently reduce the amount of subcutaneous fatty tissue. Liposuction proves to be the most effective treatment for preserving long-term effects, improving the quality of life and reducing pain.¹⁴ Lipectomy consist of surgical excision of fat deposits, but since it is a more invasive approach, the complications including relapse, developing lymphoedema, or scaring, should be taken into consideration.

Conclusion

Lipoedema is an uncommon disorder of adipose tissue that affects almost exclusively women at the age around puberty. It is often underdiagnosed or misdiagnosed; therefore, it is crucial for clinicians to be able to differentiate lipoedema from obesity or lymphoedema. The diagnosis is based on medical history, such as the age of onset or positive family history, and physical examination. A proper clinical assessment should include areas affected, shape disproportion, color and texture of the skin, pain and bruising, effects of dieting, and an impact on daily living. The therapeutic approach should take patient's needs and abilities into consideration. Currently, options of management of lipoedema include education and psychosocial support, enhancing the self-care approach, weight management, and skin care. Compression therapy and manual lymphatic drainage are parts of a conservative treatment, and surgical options, such as liposuction or lipectomy, should be considered when conservative treatment is ineffective.

We have provided this article to raise awareness of lipoedema and outline the current evidence regarding the subject. Better understanding of the disease is warranted to establish a proper diagnosis and an early treatment, which has a tremendous effect on patients.

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