Fat saturation sections on magnetic resonance imaging (MRI) (Figures 1 and 2) reveal multiple diffuse cutaneous fat globules of the legs consistent with lipedema.

Lipedema is not an unusual cause of lower-extremity enlargement. However, it infrequently is diagnosed because of its clinical similarity to lymphedema. Whereas lymphedema results from an abnormal accumulation of protein-rich interstitial fluid within the skin and subcutaneous tissue due to lymphatic dysfunction, lipedema is a primary lipodystrophy that occurs in the absence of lymphatic or venous insufficiency. In longstanding disease, secondary lymphedema may complicate the clinical picture. The precise pathogenesis of lipedema remains to be elucidated.

Lipedema and lymphedema can be distinguished clinically and radiologically. In lipedema lower-extremity involvement is always bilateral and symmetric, whereas lymphedema tends to involve one leg more severely than the other. Pitting edema typically is marked in lymphedema but is mild or absent in lipedema. Furthermore, the skin overlying lipedematous legs typically is soft and pliable in contrast to the firm, indurated, thickened skin found with lymphedema. Prominent malleolar fat pads are characteristic of lipedema, but the feet and ankles will be normal in size and the skin over the fold of the second toe mobile (negative Stemmer sign). In contrast, lower-extremity lymphedema usually involves the feet and toes, and the skin over the toes will be tightly bound down, giving a positive Stemmer sign.

Although many cases are sporadic there often is a family history of similar findings. Patients may report pain with pressure (eg, from compression stockings) or with prolonged standing. Whereas cellulitis and other lower-extremity infections are common occurrences in lymphedematous legs, infection is unusual in lipedema. Although our patient had a history of recurrent lower-extremity cellulitis, these infections were likely secondary to trauma to her insensitive feet.

Although MRI is probably the best tool to evaluate the soft tissue in these patients, both MRI and computed tomography (CT) scans can be used to distinguish lipedema from lymphedema. MRI examination demonstrates thickening of the subcutaneous tissue in both disorders but lipedema shows diffuse homogenous bilateral increase of subcutaneous fat with normal peripheral lymphatics and no subcutaneous edema. By contrast, chronic lymphedema shows circumferential edema, a honeycomb pattern in the subcutaneous compartment representing pockets of fat surrounded by fluid or fibrous tissue, and marked dermal thickening.

Unfortunately, treatment options for lipedema are limited. Patients with lipedema show little to no improvement with compression stockings, a mainstay of lymphedema therapy. Dieting and diuretics have little impact. Attempts at surgical debulking of fatty tissue have been met with limited success. Liposuction, in conjunction with limited skin and subcutaneous tissue excision, may produce significant improvements in the contour and size of the extremities.

References