

The Role of CT in the Diagnosis of Primary Lymphedema of the Lower Limb

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Twelve patients with primary lymphedema of the lower limb were examined with computed tomography (CT). A characteristic "honeycomb" pattern of the subcutaneous compartment was seen in 10 of these patients. CT scans in nine other patients with swollen leg secondary to chronic venous disease or lipedema did not show this characteristic pattern. CT may be helpful in the differential diagnosis of a swollen leg, thus obviating venography or lymphangiography.

Lymphedema, though less common than venous edema, is a well recognized cause of chronic swelling of the leg [1]. Secondary lymphedema may follow damage and obstruction to the lymphatic pathways. The causes of these include metastatic neoplastic disease involving the regional lymph nodes; surgical block dissections; radiotherapy; and filarial infestation (tropical elephantiasis) [2]. The diagnosis of secondary lymphedema is indicated by the primary pathology.

Although clinical examination and lymphangiography often establish the diagnosis of primary lymphedema, lymphangiography is time-consuming. It can be a difficult procedure and may involve complications, notably cellulitis [3]. In addition, the edema may be exacerbated by lymphangiography and may be dilatory in reverting to its former size. Moreover, venography may be required in some cases of primary lymphedema to differentiate it from swelling caused by chronic venous insufficiency [2]. The potential complications of this procedure have been described [4, 5]. In an effort to find a less invasive technique for the diagnosis of primary lymphedema, we investigated the role of computed tomography (CT) in 21 patients with swollen legs.

Materials and Methods

We reviewed the CT scans of 12 patients with primary lymphedema of the lower limb (10 women and two men; average duration of symptoms, 45 months) seen in the Swollen Leg Clinic at Hammersmith Hospital, London. Disease onset had occurred at less than 35 years of age (mean, 28) in 10 patients and at 42 and 47 years in the other two. The clinical diagnosis of lymphedema was confirmed by lymphangiography in the nine patients in whom this procedure was performed. All 12 patients were treated surgically after conservative management failed to improve their condition.

To examine the specificity of the changes in the subcutaneous tissue in patients with primary lymphedema, we also reviewed the CT scans of nine patients with swollen leg(s). Six of these (four men and two women; mean age, 55 years; average duration of swelling, 51 months) were shown by venography to have chronic venous disease, which was bilateral in four. The other three were women (mean age, 47 years; average duration of disease, 10 years) with bilateral lipedema. These CT scans were reviewed blind by one of us (D. H. C.), with the patient data and date of scan concealed.

All CT examinations were performed on a Siemens Somatom 2 body scanner at 125 kVp and 230 mAs. Slice thickness was 8 mm and scanning time was 5 sec. Four equidistant sections were obtained per patient, from 5 cm above the medial malleolus to 5 cm below the

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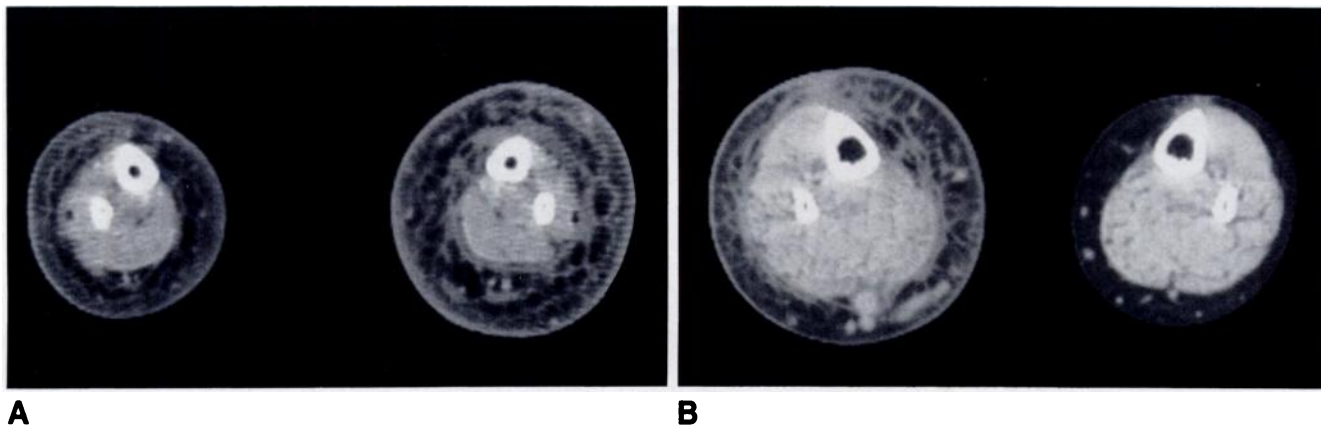


Fig. 1.—Two patients with bilateral lymphedema. Characteristic honeycomb appearance of subcutaneous compartment; bilateral (A) and unilateral (B) distribution. Normal organization of subfascial compartment.

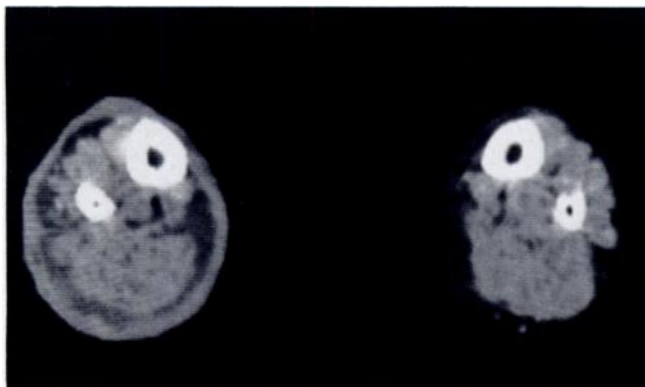


Fig. 2.—Bilateral chronic venous insufficiency. Thickening of subcutaneous compartment with disorganization of subfascial compartment.

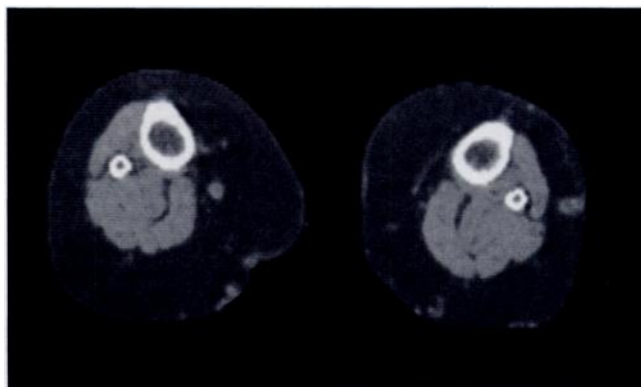


Fig. 3.—Bilateral lipedema. Enlargement of subcutaneous compartment. No specific pattern.

knee joint. If the thigh was affected also, two additional slices were taken 15 and 25 cm above the knee joint.

Results

Characteristic changes of thick, fibrous subcutaneous tissue or lakes of encapsulated fluid were encountered at surgery in all patients with lymphedema. Microscopic examination of specimen sections and tissue fluid protein estimation were compatible with the clinical diagnosis [6].

The CT findings in the lymphedema patients were as follows: (1) There was an increase in the thickness of the subcutaneous tissue compartment in all cases. (2) The skin appeared thickened in 11 of the 12 patients, but we were unable to distinguish skin thickness from any underlying fluid collection. (3) In 10 of the 12 cases, a characteristic "loofah sponge" or "honeycomb" appearance of the subcutaneous tissue was noted (fig. 1). The CT attenuation numbers suggested that these were fat pockets surrounded by fluid or fibrous tissue or both. These structures showed no enhance-

ment after intravenous administration of contrast material in two patients. (4) The appearance of the subfascial compartment was normal in all 12 patients.

CT in the patients with chronic venous disease showed enlargement of the subcutaneous compartment in all six patients and skin thickening in four. The subfascial compartment was obviously abnormal in four, with minor changes in the other two (fig. 2). The characteristic honeycomb appearance was not seen in any of these patients.

CT features in lipedema comprised enlargement of the subcutaneous compartment in all three patients, normal skin thickness, and a normal subfascial compartment (fig. 3). No specific pattern was present. CT numbers of the subcutaneous compartment were not notably different from those in patients with lymphedema.

Discussion

In 1934, Allen [7] introduced a classification of lymphedema into primary and secondary. Primary lymphedema is some-

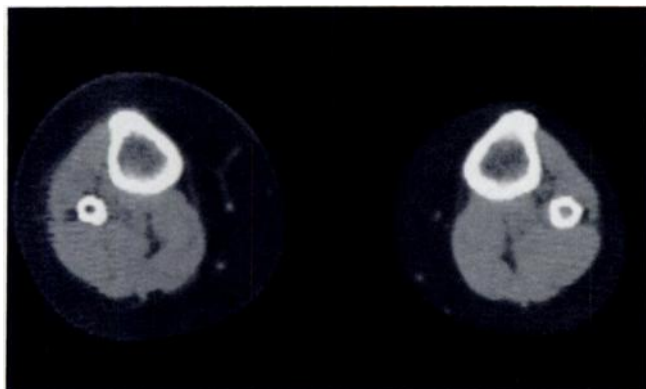


Fig. 4.—Bilateral lymphedema. Thickening of subcutaneous compartment but without honeycomb pattern.

times congenital, but in most cases it appears early in life with a predilection for the female: lymphedema precox [2]. Late onset of the disease (after age 35), known as lymphedema tarda, is uncommon. Rarely, the edema is both congenital and familial; this condition is known as Milroy disease. Lymphangiography has revealed a variety of deformities of the lymphatic pathways in primary lymphedema. There is strong evidence that these are due to inborn errors [8]. By far the most common deformity is hypoplasia: lymphatic vessels that are deficient in size and number [2]. Hyperplasia is seen in about 15% of cases and is characterized by large, dilated lymphatic vessels. However, the classical concept that lymphedema is always precipitated by abnormal lymphatics has been questioned by some authorities [9].

The differential diagnosis of primary lymphedema includes chronic venous disease, lipedema, and cyclical edema, the latter seen almost exclusively in premenopausal women [10]. The more common causes of leg edema (i.e., cardiac failure and renal failure) should be easily excluded. Similarly, a thorough medical history should elicit the cause of edema secondary to trauma and/or infection. Early venous edema may be difficult to distinguish from lymphatic edema, particularly when the thigh is affected; phlebography may be necessary in such cases [2]. Not all patients with chronic venous disease manifest diagnostic skin changes; conversely, patients with lymphedema may also have varices [2]. This may explain why many patients with lymphedema are inappropriately evaluated with venography.

Although the use of CT to demonstrate lesions of the musculoskeletal system is well documented [11], we are aware of no published work on the potential role of CT in the differential diagnosis of the swollen leg. A recent report on the CT appearance of primary lymphedema in the lower limb [12], based on two patients, did not examine the specificity of the detected changes in the subcutaneous compartment. Even though we investigated only patients with primary lymphedema, CT also helps to differentiate primary from secondary lymphedemas by demonstrating the presence or absence of a pelvic mass that may be causing obstruction.

In our series, a characteristic honeycomb appearance of

the subcutaneous compartment was present in 10 (83%) of 12 patients with lymphedema. The CT numbers suggest that the pattern represents pockets of fat surrounded by fluid or fibrous tissue. Findings at surgery and lymphangiography militate against the hypothesis that the rimlike "halos" represented lymph within dilated lymphatics. Although this question could be answered by lymphangiography preceding the CT, the previous demonstration of lymphatic hypoplasia in all nine patients in whom the procedure was done made it unlikely that the halos represented dilated lymphatics; thus, we thought repeat lymphangiography was unwarranted. It is interesting to note that of the 10 patients with the characteristic honeycomb pattern, six had bilateral disease—but the honeycomb pattern was present in both legs in only four of these. The CT features of the two limbs without the pattern were indistinguishable from those of the two patients who did not exhibit the pattern in either limb.

Since clinically unilateral disease often progresses to involve the other leg, which may then become worse than the initially involved leg, we looked at the CT scans of the clinically uninvolved legs of the four patients with the characteristic pattern who had unilateral disease. There were no features distinguishing these limbs from the normal limbs of patients with unilateral chronic venous disease.

The absence of the honeycomb pattern in two patients with lymphedema (fig. 4), both with bilateral disease, could not be explained on the basis of chronicity, since the age of onset and duration of swelling in these two patients were not different from those in patients who exhibited the pattern. The severity of symptoms, including episodes of streptococcal infection, was also not different in the two groups.

Although we examined the specificity of the CT findings in patients with primary lymphedema by comparing them only with patients with swollen legs secondary to chronic venous disease or lipedema, it is important to note that the honeycomb pattern was not seen in any of the latter patients. Moreover, other studies reporting the CT appearance of lesions of the lower limb, including tumors, abscess, and hemorrhage, have failed to demonstrate a specific pattern [13]. If this pattern specificity were to be confirmed by CT findings in a larger series, it could form the basis for noninvasive diagnosis of primary lymphedema.

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