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THE ROLE OF MAGNETIC RESONANCE IMAGING IN DIAGNOSIS OF PERIPHERAL LYMPHATIC DISORDERS

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ABSTRACT

Magnetic resonance imaging (MRI) was used in 13 patients with peripheral lymphedema and 2 patients with extensive cavernous lymphangioma of the limb for the purpose of evaluating its role in diagnosis of lymphatic disorders. In chronic lymphedema, MRI showed deformity of lymphatics at different tissue levels. In the subcutis, MRI characteristically displayed diffuse edema or a honeycombed pattern consistent with reticular lymphangiectasis and "lakes" with a marked increase in signal intensity with T2-weighted imaging. In lymphedema hyperplasia and chylous reflux, MRI depicted dilated retroperitoneal lymphatic collectors and lumbar trunks. In cavernous lymphangiomatosis, MRI demonstrated a prominent lattice-like pattern which had lower signal intensity on T1-weighted imaging and higher intensity on T2-weighted imaging. The findings of MRI are valuable not only for accurate assessment of lymphatic dysplasia syndromes but also provide a blueprint for treatment options.

Lymphedema is a common lymphatic disorder of the extremity, and a simple, easily repeatable diagnostic method is useful for accurate depiction of lymphatic anomalies. Conventional lymphography using oil contrast has long been the gold standard for displaying lymphatics and nodes (1). However, it is invasive, time-consuming, and may exacerbate lymphedema. Lymphangioscintigraphy, on the other hand, does not readily depict lymphatics and lymph nodes in remote anatomic areas especially when lymphatic pathways are blocked or absent.

Cavernous lymphangioma is another common lymphatic anomaly involving a limb. Evaluation of the extent of a cavernous lymphangioma (lymphangiomatosis) may also be difficult on clinical grounds alone. These lesions are often seen in children with diffuse involvement of an extremity (or extremities), which may be confused with primary lymphedema. Because the lesion typically has no direct communication with normal lymphatic vessels, lymphangioscintigraphy may not delineate the true nature of the disorder and biopsy and histology has been the usual way of substantiating the diagnosis.

MRI is a safe and noninvasive imaging modality and provides excellent information regarding soft tissue changes (1). However, its value for visualizing and depicting the lymphatic system has not been extensively studied. The aim of this study was accordingly to evaluate MR imaging characteristics of lymphatic disorders of a limb, i.e., lymphedema (both primary and secondary) and cavernous lymphangioma.

MATERIALS AND METHODS

Thirteen patients (6 female and 7 male, age 8-72 yrs) with chronic lymphedema of the lower extremities were examined. Ten

Fig. 1. A 42-year old woman with bilateral lower extremity lymphedema precox since 20 years of age. MRI shows diffuse thickening of the subcutaneous tissue and a honeycomb pattern of increased signal intensity. Above: coronal section, T1-weighted; Below: axial section, T1-weighted (left), T2-weighted (right).

patients had primary lymphedema. Four patients had lymphedema precox, and six patients had congenital lymphedema, and of these, three had chylous reflux. Two patients developed secondary lymphedema from repeated infection and one had lymphedema after trauma. The duration of symptoms ranged from 3 to 29 years.

Two infants with diffuse lymphangioma of the extremities were also studied. One child (5 months old) had extensive swelling of the left lower leg and was initially diagnosed as having Milroy's disease. The other (4 months old) had swelling of both left upper and lower extremities. Both the patients had findings since birth. The diagnosis of cavernous lymphangioma was established by biopsy and histologic examination.

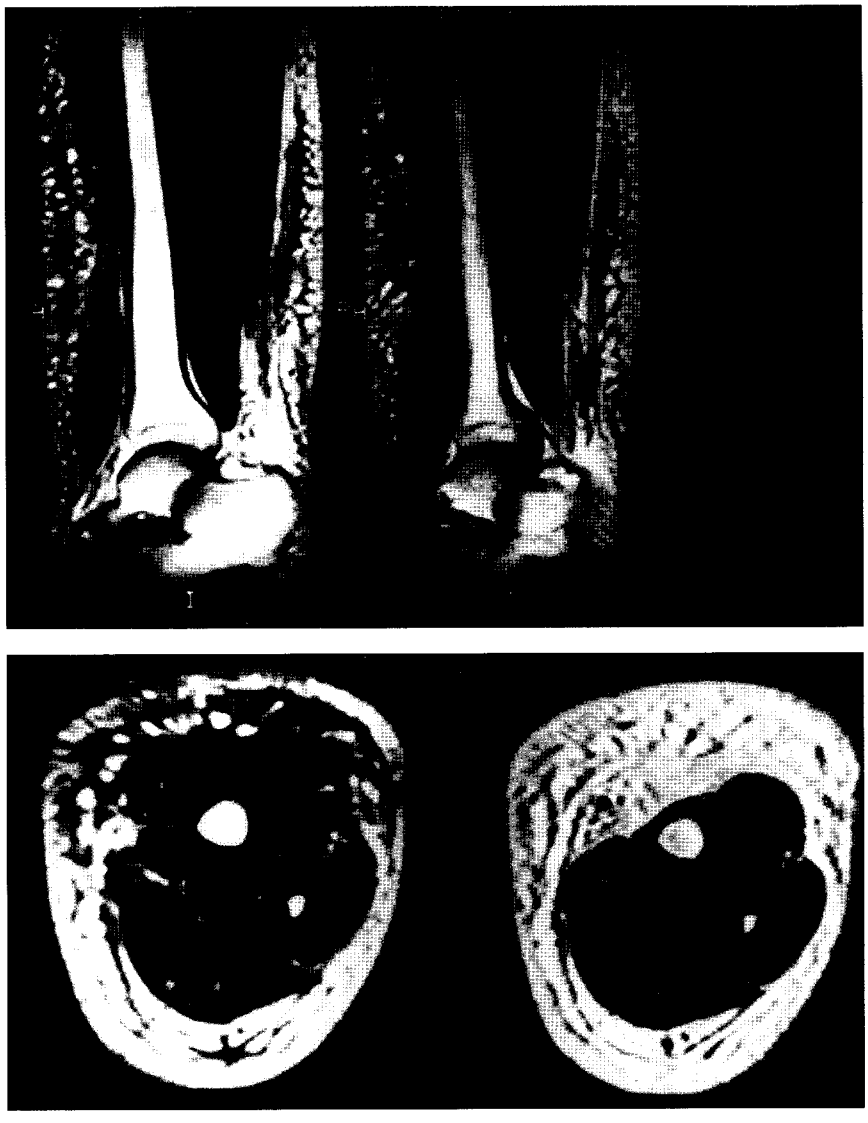


Fig. 2. A 38-year old man with bilateral secondary lymphedema of lower legs since 18 years of age. MRI shows marked subcutaneous edema with prominent dermal lymphatics and a honeycomb pattern of changes. Above: coronal section, T1-weighted; Below: axial section (left), T2-weighted (right).

MRI was performed with an 0.35-T scanner (Diasonic, USA). The imaging matrix was 256x100-256 (body) and 160x256 (extremity). T1-, T2- and protein density weighted transaxial and coronal magnetic resonance images with 5-10 mm slice thickness were obtained on each patient. T1-weighted images: TR=500 msec, TE=30 msec; T2-weighted images: TR=1800, 2000 msec; TE=40, 60, 80, 120 msec.

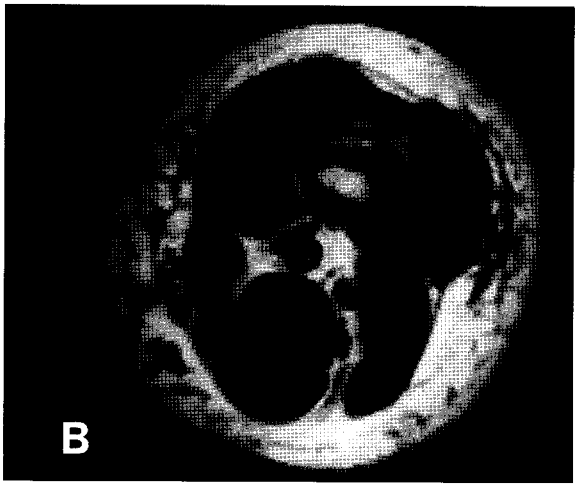
RESULTS

The MRI characteristic images of chronic lymphedema included thickening of the subcutaneous layer with extensive edema. In most patients (primary or secondary edema), there were dilated dermal lymphatics or

Fig. 3. An 8-year old boy with primary lymphedema of the left leg (hyperplasia). MRI shows lymphangiectasis in the subcutaneous tissue. Above: T1-weighted coronal image showing dilated lymphatics (arrow); Below: axial section, T2-weighted.

"lakes" with sequestered "lymph" in the subcutis, which displayed a typical honeycombed or reticular pattern. On T1-weighted images, tissue resembling fat was visualized by an intermediate signal intensity, whereas the lymphatics and "lakes" displayed a lower signal intensity. On T2-weighted images, edema fluid gave a higher signal intensity and was clearly distinguished by its brightness *(Fig. 1,2)*.

In primary lymphedema, several patients displayed diffuse edema without visible lymphatics or "lakes" on T2-weighted images; the entire subcutis simply showed a higher signal intensity. In those patients, lymphangioscintigraphy documented



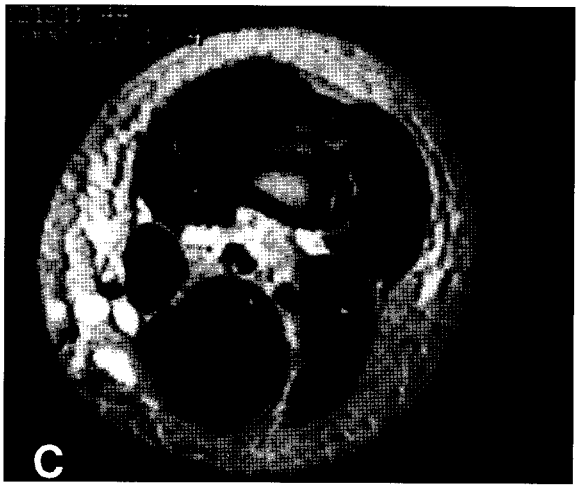
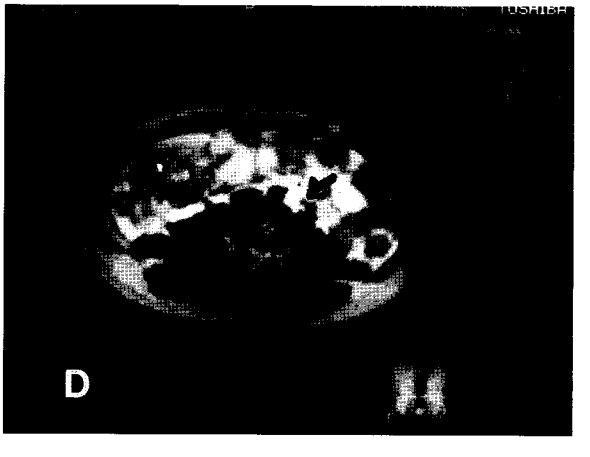
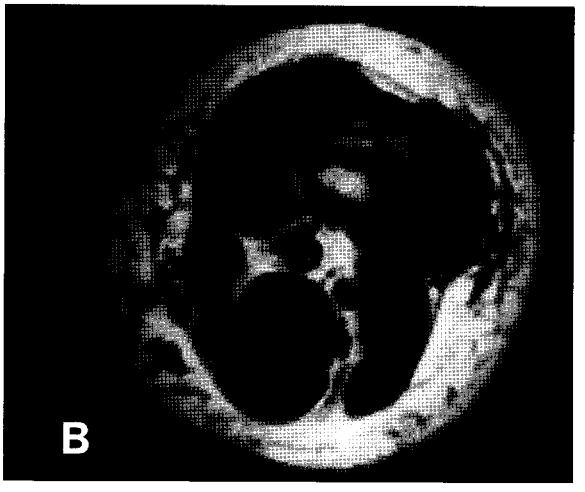
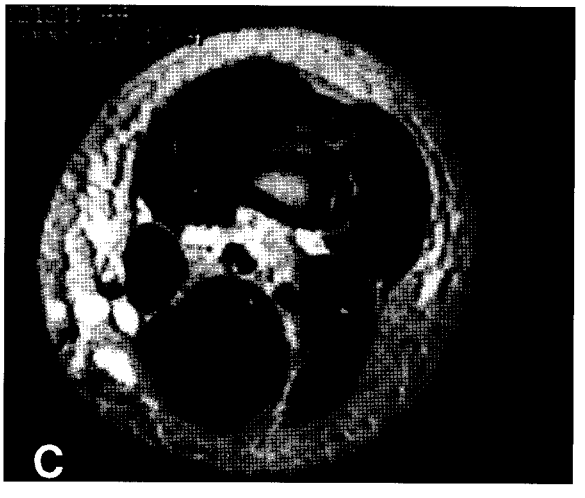


Fig. 4. A 25-year old woman with right leg congenital lymphedema (chylous reflux). MRI show lymphangiectasis and edema in subcutis. Lymphatic varices in the medial aspect of thigh are clearly seen (arrow). A: coronal T1-weighted; B: axial section T1-weighted section; C: axial section T2-weighted section; D: axial section of abdomen T2-weighted show dilated lumbar lymphatics (arrow).

lymphatic aplasia or marked hypoplasia. In patients with lymphatic hyperplasia, MR images demonstrated dilated lymphatic trunks paralleling the long axis of the extremity *(Fig. 3)*. In chylous reflux syndromes, MRI of the legs displayed dilated lymphatic collectors. On T1-weighted images, these lymphatic vessels displayed a decreased signal intensity while on T2-weighted images the lymphatics gave a higher signal intensity which made it easier to distinguish from blood vessels *(Fig. 4)*. MR of the abdomen showed dilated lumbar lymphatics with extensive retroperitoneal lymphatic dysplasia *(Figs. 4D,5)*.







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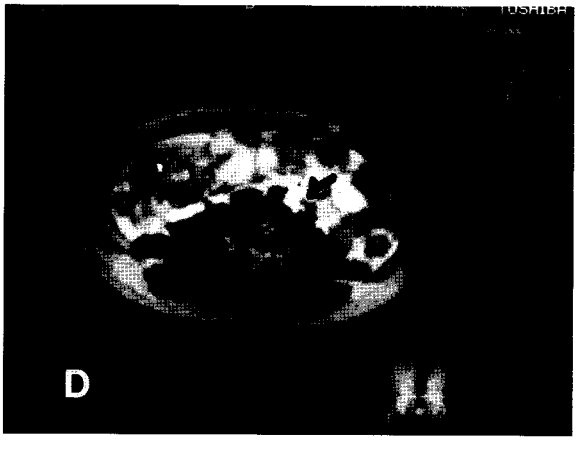

Fig. 5. A 15-year old woman with left lower extremity lymphedema since age 3 years (chylous reflux). MRI shows extremely thick subcutaneous tissue with massive lymphatic "lakes". A: coronal section T1-weighted; B: axial section T1-weighted (left), T2-weighted (right); C: axial T2-weighted MR imaging of abdomen (c) shows hugely dilated lumbar lymphatics around the inferior vena cava and aorta (arrow).

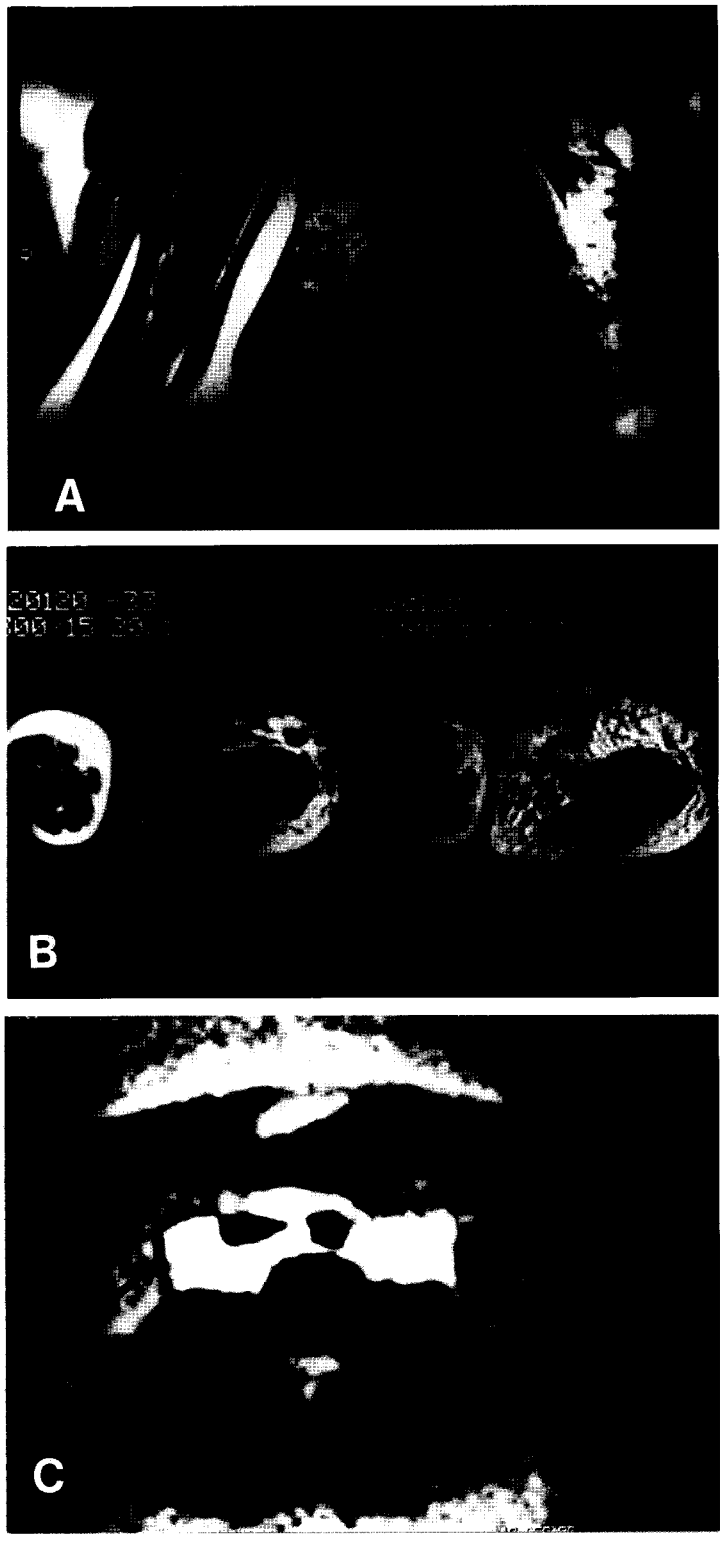

Fig. 6. A 4-month old boy with cavernous lymphangioma of the left leg. T1-weighted coronal sequence shows a marked lattice-like pattern in the subcutaneous tissue of low-signal intensity.

In cavernous lymphangioma, an increased volume of the leg was demonstrated by MR images with thickness of the subcutis and prominent lattice-like abnormal signals. The lesions were characterized by hyperintensity in T2-weighted images and decreased intensity in T1-weighted images *(Figs. 6,7)*. The muscular (subfascial) compartments were structurally intact.

DISCUSSION

MR images provide excellent detail of soft tissues (1). Tissue fluid and lymph yields high intensity signals on T2-weighted sequences and, accordingly, in lymphedema, thickening of the subcutis is easily differentiated from obesity (so-called lipedema). MRI is also useful to distinguish "phlebedema" from lymphedema. In the former condition, there typically is an increase in size of the subfascial muscular compartment with an increase in signal intensity in both the subcutis and muscle layers (2).

In lymphedema, MR imaging has the advantage of displaying lymphatic abnormalities at different levels. The typical picture of lymphedema by MRI is a honeycombed or reticular subcutaneous tissue pattern. According to our observation during operation on these limbs, the honeycomb pattern and "lakes" are likely accumulation of fluid between fat surrounded by fibrotic tissue. The reticular structure may represent dilated dermal lymphatics. These changes are not usually detected on lymphangioscintigraphy because the tracer diffuses in the subcutaneous tissue (dermal backflow) or stagnates at the injection site when lymph pathways are completely blocked or absent. MR imaging also displays more central (retroperitoneal) lymphatics, lumbar lymphangiectasia, which is poorly defined by lymphangioscintigraphy. In a patient with retroperitoneal lymphatic dysplasia, MR scanning may yield detailed information regarding the site of obstruction, the width and extent of the lymphatic anomaly and its relationship to adjacent structures and organs.

In patients with peripheral cavernous lymphangiomatosis, the diagnosis is often delayed or mistaken for congenital lymphedema. Lymphangioscintigraphy may show normal lymphatics if the lymph circulation itself is intact and, accordingly, diagnosis usually requires biopsy and histologic confirmation. MRI, as shown here, depicts characteristic features of cavernous lymphangiomatous lesions, namely prominent lattice-like signals in the fatty tissue with notable thickness of the subcutis. MRI also provides information on the size and depth of the lesion and its relationship with deeper structures (3).

In conclusion, MR imaging displays abnormalities of lymphatics and lymph nodes

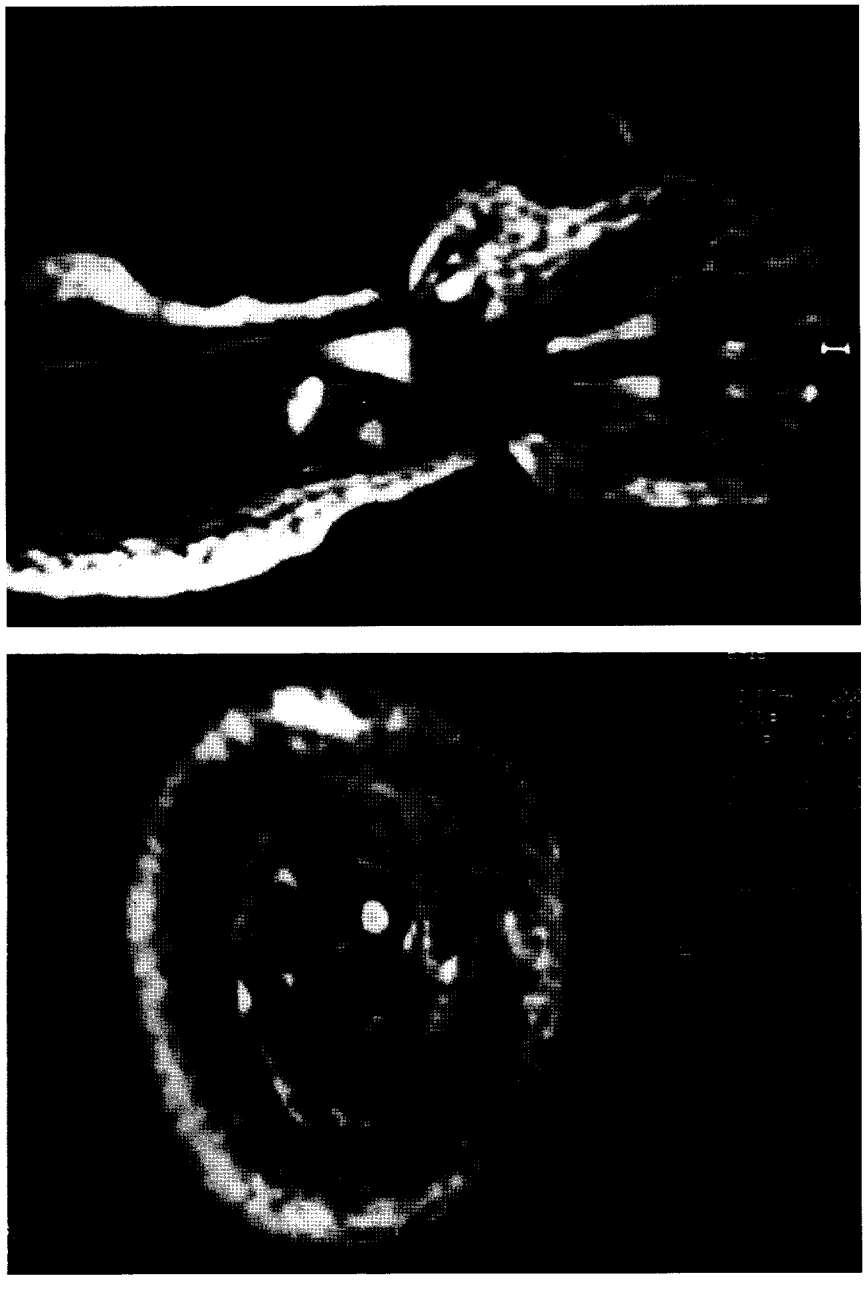

Fig. 7. A 5-month old girl with extensive cavernous lymphangiomatosis of both the upper and lower extremity. T1-weighted coronal (above) and axial (below) scan of arm and hand negatively displays a massive low-signal reticular pattern in the subcutaneous tissue.

at different levels, as well as depicts soft tissue changes even when the peripheral lymphatic circulation is intact as with lymphangiomatosis. It is especially valuable for evaluation of primary lymphedema with lymphatic hyperplasia and chylous reflux. When MRI is used in conjunction with lymphangioscintigraphy, lymphatic morphology and function is well outlined. For cavernous lymphangiomatosis, MR imaging is especially useful and renders diagnosis without need for biopsy and histologic examination.

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