

Generalized Lymphangiomas: A Case Report

Jang-Gyu Cha¹, Jai-Soung Park¹, Sang-Hyun Paik¹, Hee-Kyung Kim²

Generalized lymphangiomas is a rare congenital malformation of the lymphatics. CT and MR scan have been used to evaluate lymphangiomas, which appear as large multicystic fluid-filled masses. CT and MR Imaging findings are often helpful in distinguishing lymphangiomas from various vascular disorders. We report the findings of CT, MRI and bone scan in a patient with generalized cystic lymphangiomas. Whole body 3.0-T MR scan using STIR sequence with a larger FOV could detect the additional lesions that were not seen at other imaging modalities. We believe that whole body 3.0 T MR imaging is a good modality to evaluate the extent of the disease and following up the patients with the generalized cystic lymphangiomas.

Index words : Congenital
Lymphangiomas
Magnetic resonance (MR)

Introduction

Generalized lymphangiomas is the term used for diffuse involvement of soft tissue, viscera (especially lungs and spleen) or bone by a lymphangiomas proliferation. The presentation, as in our case, is often with a chylothorax. It is categorized as a subtype of lymphangiectasia, which is owing to dilatation, and proliferation of lymphatics (1).

Lymphangiomas generally presents in the first two decades of life and is thought to be congenital in origin, although it does not appear to be inherited. The

characteristic lesions consist of a benign proliferation of lymphatic vessels within the soft tissues, bone, or parenchyma of organs. The majority of patients have both visceral and skeletal involvement, most commonly in the spleen, liver, lung, long bones, pelvis, skull, and vertebrae (2, 3).

Magnetic resonance (MR) imaging with its excellent tissue contrast, high spatial resolution, and detailed morphological information also appears promising for tumor screening (4, 5). Previously MRI has been employed for the assessment of focal pathologies in restricted anatomical regions or organ systems (5). With advent of whole body MR technique, patients could be

JKSMRM 13:190-194(2009)

¹Department of Radiology, Soonchunhyang University Bucheon Hospital

²Department of Pathology, Soonchunhyang University Bucheon Hospital

Received; April 8, 2009, revised; May 4, 2009, accepted; August 7, 2009

Corresponding author : Jang Gyu Cha, M.D., Department of Radiology, Soonchunhyang University Bucheon Hospital,
1174 Jung-dong, Wonmi-gu, Bucheon-si, Gyeonggi-do 420-767, Korea.
Tel. 82-32-621-5876 Fax. 82-32-621-5016 E-mail: mj4907@schbc.ac.kr

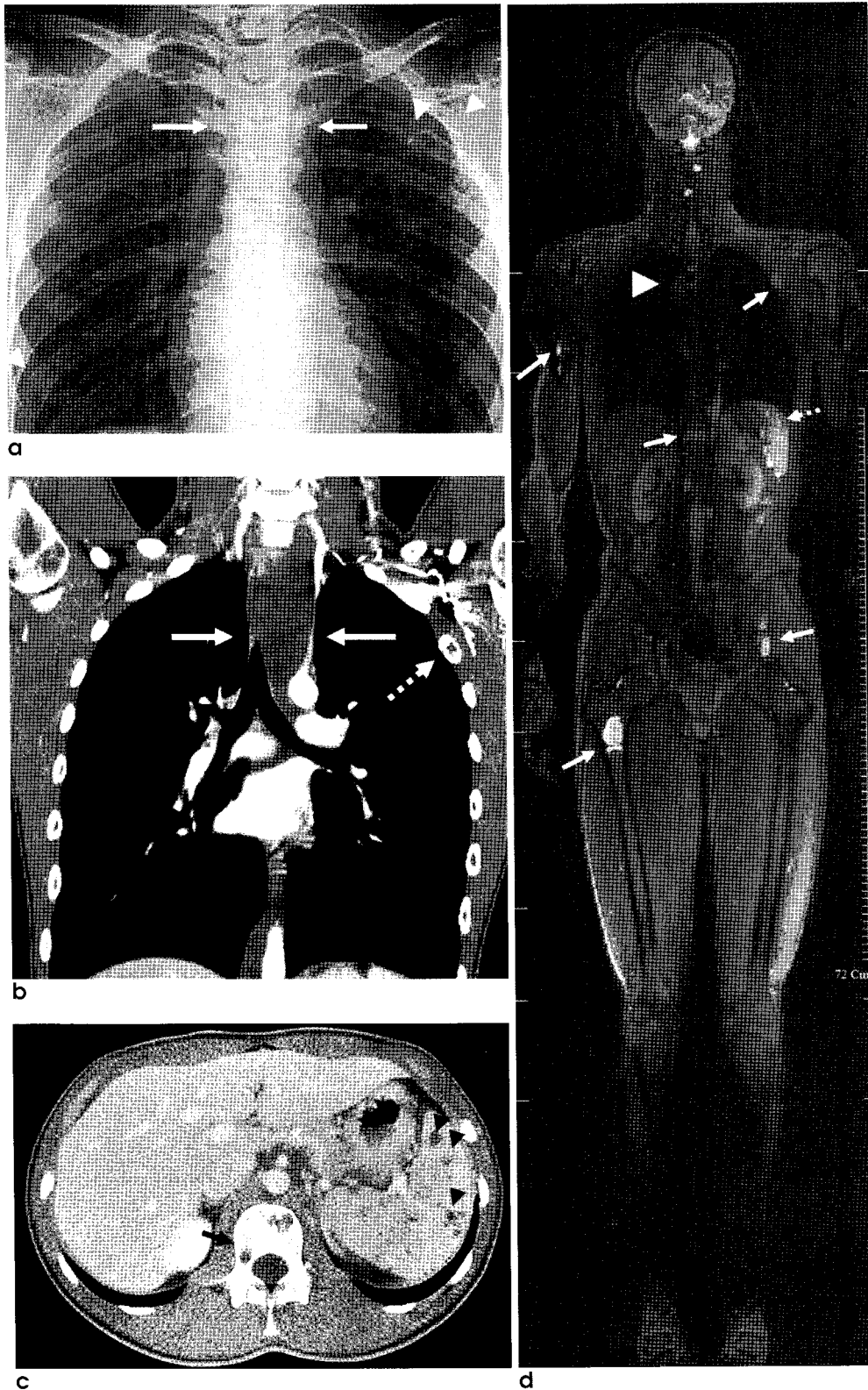
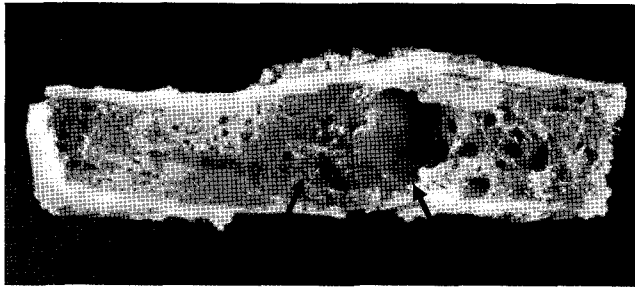


Fig. 1. **a.** Chest PA shows widening of mediastinum (arrows) and osteolytic lesions in right 8th rib, left 3rd and 4th ribs (arrowheads). **b.** Coronal reconstruction of contrast-enhanced chest CT shows non enhanced mediastinal mass (arrows) and osteolytic lesion in left third rib (dashed arrow). **c.** Chest CT at lower level of lungs than Fig. 1b shows multiple cystic lesions in spleen (arrowheads) and well defined osteolytic lesions in vertebral body (arrow). **d.** Whole body MR using STIR sequence demonstrates mediastinal mass (arrowhead) and cystic lesions in spleen (dashed arrow) and cystic lesions in left third rib, vertebra, right humerus, left iliac bone and right femur (arrows).



e
Fig. 1. e. Photograph of gross specimen obtained from right 8th rib shows multi-septated cystic lesions (arrows) in intramedullary cavity.

imaged from head to toe within one hour of scan time. There have been few reports on whole body MR imaging of the generalized cystic lymphangiomatosis (6-9). In this report, a case of generalized cystic lymphangiomatosis occurring in visceral organ, skeletal system is presented with whole body 3.0-T MR scan using short tau inversion recovery (STIR) sequence.

Case Report

A 16-year-old man was admitted to our hospital for a complete medical evaluation of abnormal mediastinal shadow and rib lesions found on a chest radiograph during his annual check-up.

Physical examination revealed no evidence of tenderness and palpable mass around chest wall. Breathing sound was normal in both lungs on auscultation exam. There was no splenomegaly. Laboratory exam was normal. Chest radiographs showed osteolytic expansile lesion with Irregular internal linear densities in the right 8th, left 3 and 4th ribs (Fig. 1A). Contrast enhanced- chest CT revealed multi-locular cystic mediastinal lesions containing fluid-attenuated material without invasion of adjacent organ and splenic involvement, which lesions showed no enhancement (Fig. 1B-C). Other multiple geographic osteolytic lesions with sclerotic borders were seen in ribs and thoracolumbar vertebrae (Fig. 1C). Whole-body 3.0-T MR imaging was performed to evaluate the nature of masses and extent of the disease. Whole body MR demonstrated additional bony lesions including humerus, pelvis and femur as well as mediastinal mass, and cystic lesions in spleen, rib, and vertebra. All of the bony lesions showed low signal intensity on T1

weighted images and high signal intensity on T2 weighted images.

Coronal STIR images were obtained with the following parameters: TR/TE = 4000-6000/27, inversion time = 150 msec, slice thickness = 8 mm, bandwidth = 62.5, number of excitation = 2, matrix = 320×192. Above thoracic, splenic and skeletal lesions on T1 and T2 weighted images were clearly depicted on STIR images (Fig. 1D). Bone scan showed no abnormal increased uptake. Gross specimen (Fig. 1E) and histopathology after rib resection (not shown) demonstrated dilated cystic spaces lined by flattened endothelial cells without intervening stroma, which revealed a diagnosis of lymphangioma.

Discussion

Lymphangiomatosis is thought to be originated from a persistence of dilated lymphatics in the 14th to 20th week of intrauterine development (8). Lymphangiomatosis is a condition characterized by diffuse or multicentric proliferation of lymphatic vessels resembling lymphangioma, which most frequently involves the liver, spleen, mediastinum, lungs, or soft tissues (10). The gross-pathologic substratum of generalized cystic lymphangiomatosis consists of dilated chyle-filled lymphatic spaces along the lymphatic pathways (2).

On the basis of histologic analysis, three types of lymphangiomas have been described: simple lymphangiomas, cavernous lymphangiomas, and cystic hygromas. Histologic classification, however, may eventually be confusing because of the wide morphologic variation of potential lesions. Also, histologic classification may be hampered by the potential overlap of generalized cystic lymphangiomatosis with other generalized mostly vascular disorders, such as generalized fibromatosis, lymphangioliomyomatosis, and diffuse hemangiomas. Therefore, imaging has been advocated as a first-line diagnostic option to assess patients with generalized cystic lymphangiomatosis (6). There has been no reported correlation with race or gender. In previous report of thoracic lymphangiomatosis, it presented as chylothorax, mass, pulmonary infiltrates, bone, spleen and cervical involvement (8). The skeletal changes frequently consist of multiple, rounded, cyst-

like lesions, found particularly in the pelvis and upper femur. However, lesions in the spine are not uncommon and MRI can be helpful in suggesting the correct diagnosis. The diagnosis is sometimes made by bone biopsy that shows that these lytic lesions are lymphangiomas containing lymph fluid.

Radiographic findings are nonspecific but included pleural effusions, mediastinal widening, or soft-tissue masses, sometimes with coarse calcifications (9). Lymphangiography shows multiple lesions of the thoracic duct, dilated lymphatic channels, and lymphangiomas throughout the bones and lungs (2). CT and MRI are the current techniques used to evaluate lymphangiomas, which appear as large multicystic fluid-filled masses. Imaging is often helpful in distinguishing lymphangiomas from various vascular disorders (8) and is useful in the preoperative planning of surgery (9). Furthermore, whole-body MR imaging has recently been proposed for tumor screening of asymptomatic individuals, and potentially life-changing diagnoses, such as formerly unknown malignancy, have been reported (4). The diagnostic value and characteristics of MR imaging for evaluating the generalized lymphangiomatosis have been previously reported on, including two reports using whole body MR imaging (3, 6–8).

In addition, state-of-the-art imaging techniques such as T1-weighted and STIR imaging have proven highly efficient for the assessment of soft tissue and bone structures (5). Previous reports described that CT imaging showed that the generalized lymphangiomatosis was showed as sharply defined (2), non-enhanced cystic lesion and MR imaging showed that the lesions showed hypointense signal intensity on T1-weighted images and hyperintense on T2-weighted images (3, 6, 7). In our case, the radiologic findings was consistent with those in other studies of the generalized lymphangiomatosis and three additional bony lesions including humerus, pelvis and femur could be clearly depicted with whole body 3.0-T MR scan using STIR sequence (7). Especially, coronal STIR imaging could find thoracic lesion, visceral lesions such as spleen and skeletal lesions in rib, spines, humerus, pelvis and femur through the whole body within 45 minutes. Therefore, we believe that the whole body 3.0-T MR

imaging is very useful for evaluating the characteristic nature and the extent of generalized lymphangiomatosis.

No treatment for the disease has been found. Although surgical excision can be recommended for localized lymphangioma, complete excision of generalized lymphangiomatosis can be difficult or impossible. Palliative procedures often include medical therapy with interferon-alpha, draining the pleural effusions or performing pleurodesis for recurrent pleural effusions (7).

In conclusion, whole body 3.0-T MR scan using STIR sequence with a larger field of view (FOV) could detect the additional lesions that were not seen at other imaging modalities. We believe that whole body 3.0T MR imaging is a good modality for evaluating the extent of the disease and following up the patients with the generalized cystic lymphangiomatosis.

References

- Noonan JA, Walters LR, Reeves JT. Congenital pulmonary lymphangiectasis. *Am J Dis Child* 1970;120:314-319
- Faul JL, Berry GJ, Colby TV, et al. Thoracic lymphangiomas, lymphangiectasis, lymphangiomatosis, and lymphatic dysplasia syndrome. *Am J Respir Crit Care Med* 2000; 161:1037-1046
- Konez O, Vyas PK, Goyal M. Disseminated lymphangiomatosis presenting with massive chylothorax. *Pediatr Radiol* 2000;30:35-37
- Schmidt GP, Kramer H, Reiser MF, Glaser C. Whole-body magnetic resonance imaging and positron emission tomography-computed tomography in oncology. *Top Magn Reson Imaging* 2007;18:193-202
- Schmidt GP, Haug AR, Schoenberg SO, Reiser MF. Whole-body MRI and PET-CT in the management of cancer patients. *Eur Radiol* 2006;16:1216-1225
- Wunderbaldinger P, Paya K, Partik B, et al. CT and MR imaging of generalized cystic lymphangiomatosis in pediatric patients. *AJR Am J Roentgenol* 2000;174:827-832
- Yang DH, Goo HW. Generalized lymphangiomatosis: radiologic findings in three pediatric patients. *Korean J Radiol* 2006;7:287-291
- Maki DD, Nesbit ME, Griffiths HJ. Diffuse lymphangiomatosis of bone. *Australas Radiol* 1999;43:535-538
- Marom EM, Moran CA, Munden RF. Generalized lymphangiomatosis. *AJR Am J Roentgenol* 2004;182:1068
- Alvarez OA, Kjellin I, Zuppan CW. Thoracic lymphangiomatosis in a child. *J Pediatr Hematol Oncol* 2004;26:136-141

전신성 림프관종증: 증례 보고

¹순천향대학교 부천병원 영상의학과

²순천향대학교 부천병원 병리학과

차장규¹ · 박재성¹ · 백상현¹ · 김희경²

전신성 림프관종증은 림프관의 드문 선천성 기형이다. CT와 MR 영상은 림프관종 진단에 이용되고 있고 액체로 가득찬 다낭성의 큰 종괴로 보이고 이러한 영상 소견으로 림프관종과 다양한 혈관 질환을 감별하는 데 도움이 된다. 저자는 전신성 림프관종증 환자의 CT, MR영상과 방사선 동위원소 영상소견을 보고한다. STIR 영상 연쇄 기법을 이용한 전신 3.0-T MR 영상은 고해상도 다른 영상 장비에서 진단하지 못한 부가적인 병변을 발견할 수 있었다. 저자는 전신 3.0T MR 영상이 전신성 림프관종증 환자의 침범정도 평가나 추적관찰에 유용한 진단 장비라고 생각한다.

통신저자 : 차장규, (420-767) 경기도 부천시 원미구 중동 1174, 순천향대학교 부천병원 영상의학과
Tel. 82-32-621-5876 Fax. 82-32-621-5016 E-mail: mj4907@schbc.ac.kr