Pediatr Radiol (2000) 30: 35–37

Orhan Konez
Pranav K. Vyas
Manish Goyal

Disseminated lymphangiomatosis presenting with massive chylothorax

Received: 22 June 1999
Accepted: 7 October 1999

O. Konez (✉) · M. Goyal
Department of Radiology,
NEOUCOM Affiliated Hospitals,
Aultman Hospital, 2600 6th Street SW,
Canton, OH 44710, USA

P. K. Vyas
Children's Hospital Medical Center
of Akron, Akron, Ohio, USA

Abstract *Background.* Lymphangiomatosis is a disease characterized by involvement of various body constituents and can involve the skeletal system, connective tissues, and visceral organs.
Materials and methods. We present a case of a 9-year-old girl where this entity presented with extensive right-sided chylothorax. Conventional imaging, including skeletal scintigraphy and contrast enhanced CT of the chest and abdomen, may have underestimated the extent of the disease, as seen on follow-up T2-weighted MR images of the chest and abdomen in our case.
Results. MRI easily demonstrated additional bone lesions as well as multiple small splenic lesions, which were difficult to appreciate on prior CT examinations.
Conclusion. We suggest that MRI may be helpful to assess the extent of this disease more accurately.

Introduction

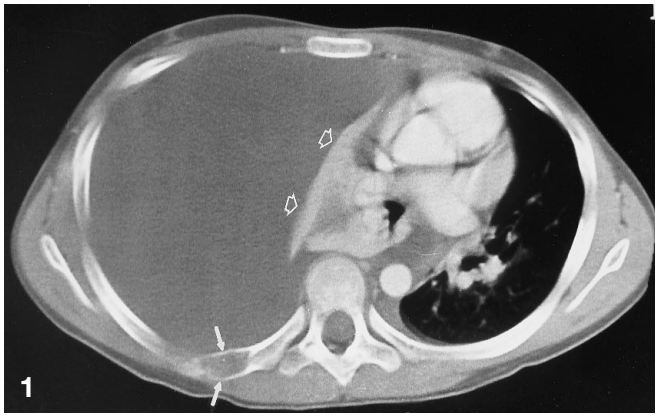
Lymphangiomatosis is a disease characterized by maldevelopment of lymphatic drainage and can result in chylothorax, as well as a multitude of other manifestations. We report a case involving an adolescent female that was investigated by multiple modalities, including radiography, skeletal scintigraphy, CT, and MRI.

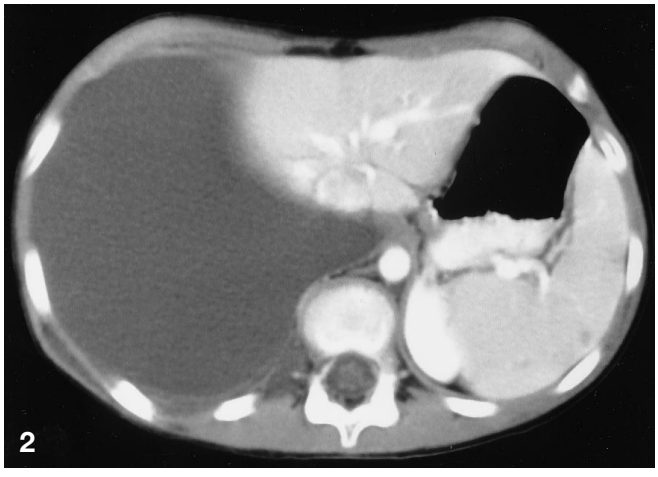
Case report

A 9-year-old girl presented with severe dyspnea that had progressed over the last few weeks. Chest radiographs demonstrated a large right pleural effusion resulting in mediastinal shift and lytic rib lesions on the same side. She was admitted for further work-up of her condition. A massive right pleural effusion as well as lytic ill-defined rib lesions involving the posterior aspects of the sixth and seventh ribs were better demonstrated on a subsequent CT of the chest (Fig. 1). Additionally, there were suspicious tiny peripheral splenic hypodensities on contrast-enhanced CT of the abdomen (Fig. 2). These findings were consistent with lymphangiomatosis involving the right sixth and seventh ribs, resulting in massive chylothorax with questionable involvement of the spleen. ^{99m}Tc-methylene diphosphonate scintigraphy was then performed to evaluate the metabolic activity of the observed rib lesions and to screen the entire bony skeleton for other lesions. The bone scan was essentially normal except for minimally increased uptake in the abnormal right ribs seen at radiography (Fig. 3).

The patient underwent a right thoracotomy and pleural drainage, with evacuation of all chylous fluid present at that time. No parenchymal lesion was found at operation, and there was no isolated intrathoracic lymphatic leak. However, throughout the operation, there was a continuous leakage of chylous fluid from the chest wall into the pleural space. The abnormal portions of the involved ribs were resected, with biopsies taken of the ribs, pleura, and lung. Despite resection, the chylous drainage persisted in the postoperative period. A second thoracotomy was performed 3 days later, at which time a right parietal pleurectomy was also performed. The right chylothorax reaccumulated rapidly thereafter.

CT of the chest was repeated approximately 5 months after the initial diagnostic work-up, which showed post-surgical changes in the right hemithorax. No additional bony abnormalities were noted in the thoracic skeleton at that time. The patient was then referred to another institution for treatment, where an MRI of the chest and abdomen was obtained to evaluate the skeleton and intra-abdominal organ involvement (Fig. 4) (approximately 6 months after initial diagnostic work-up). Several other lesions in the vertebrae and a single lesion in the sternum were noted, as well as splenic lesions that were not well seen on a prior contrast-enhanced CT examination. All bone and splenic lesions consistently showed high-signal intensity on T2-weighted spin-echo images that were easily differentiated from normal bone marrow. Retro-





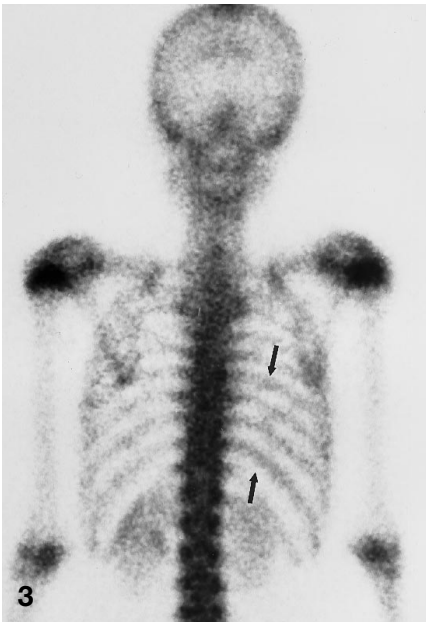


Fig. 1 A CT scan of the chest demonstrates massive fluid accumulation in the right hemithorax, resulting in collapse of the right lung (*open arrows*) as well as an ill-defined lytic lesion involving the posterior aspect of the right seventh rib (*solid arrows*). The mediastinum is shifted to the left

Fig. 2 A CT scan of the abdomen was performed with spiral technique and rapid bolus contrast infusion (power injector, 20-s delay, 2 cc/kg). There are multiple, small, peripheral splenic lesions showing less contrast enhancement than normal splenic parenchyma

Fig. 3 ^{99m}Tc-methylene disphosphonate scintigraphy (posterior view) demonstrates minimally increased uptake in the abnormal right ribs (*arrows*). No other bone abnormalities are seen

spective evaluation of the bone-window images of the post-operative CT of the chest revealed very subtle lytic changes corresponding to the additional bone lesions noted on MRI (Fig. 5).

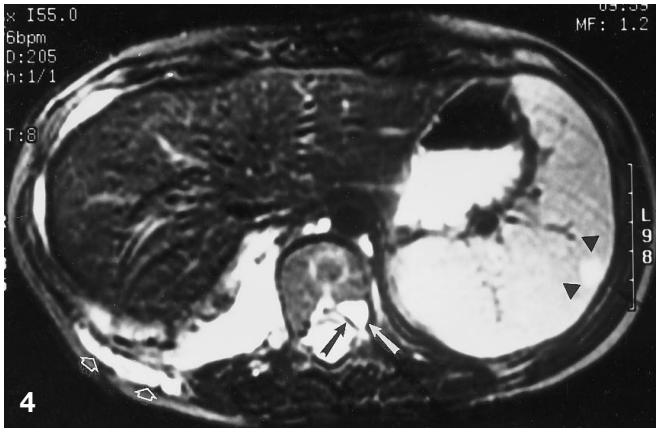
Discussion

"Lymphangiomatosis" is the term used for diffuse or multifocal involvement of bones, parenchymal organs, or soft tissues by lymphangiomas. This very rare entity occurs mainly in children or adolescents and is considered to be a malformation of the lymphatic system, rather than a neoplastic process. Its etiology is unknown, but it is assumed to arise from a sequestration of peripheral lymphatic vessels that fail to connect with the main lymphatic channels [1].

Replacement of a single or several contiguous bones by lymphangiomatous tissue is known as Gorham's disease [2]. Widespread bony involvement associated with diffuse soft tissue disease occurs in younger age groups and is known as generalized lymphangiomatosis [3]. Thoracic involvement with this process can result in chylothorax.

Bone scintigraphic findings in this entity have been reported to be decreased [4] and increased [4–6] in affected areas. In a case report by Chu et al. [5], in a 12-year-old patient, skeletal lesions that were not apparent radiographically displayed increased ^{99m}Tc-MDP uptake. In our case, the bone scan showed very minimal increased radiotracer uptake in areas corresponding to the rib abnormalities that were much more conspicuous on chest radiography and CT. Additionally, the bone scan was negative for other skeletal lesions, whereas the MRI showed multiple other skeletal lesions. This suggests that bone scintigraphy may not always be a reliable modality to evaluate bony lesions in this condition fully, especially considering the fact that lack of new bone formation is a characteristic of the disease process.

MRI can provide reliable information regarding the extent of disease, as seen in this case. On T2-weighted spin-echo images, the lesions involving the ribs, vertebrae, and sternum demonstrated high-signal intensity, which gave lesions high conspicuity. The characteristic homogeneous high T1 and relatively low T2 signal of the normal bone marrow can be easily differentiated from pathological tissue [6]. Additionally, in our case



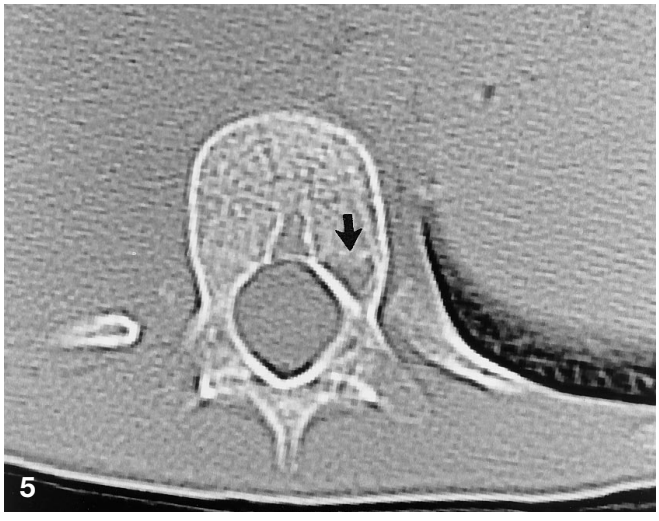


Fig. 4 A T2-weighted axial spin-echo image (TR/TE: 2307/102) demonstrates high-signal intensity corresponding to sites of lytic rib lesions (*open arrows*). Focal high signal intensity is also noted in the left posterior portion of the vertebral body (*solid arrows*). Additionally, similar focal high signal is present in the spleen, consistent with splenic involvement (*arrowheads*) (courtesy of Sara M. O'Hara, M. D., Duke University)

Fig. 5 A magnified bone-window CT image (post-operative) showing a very subtle bony lesion involving the left vertebral body (*arrow*) that was noted only in retrospect. The corresponding MR image demonstrates a conspicuous lesion within the same location (Fig. 4)

the MRI showed parenchymal splenic involvement, which was not easily seen on the initial CT scan. A standard abdominal CT technique consisting of rapid bolus contrast infusion and spiral scanning is essential to help identify visceral parenchymal lesions in most organs, but this technique can cause inhomogeneous enhancement of the spleen. Therefore, splenic lesions can be misinterpreted as normal inhomogeneity when spiral scanning with bolus contrast infusion was used. The other consideration could be that the increased conspicuity of lesions seen on the MRI but not seen on the initial CT examination may reflect interim progression of the disease process in the 6-month interval between the two studies rather than a false-negative CT result. This is probably not the case in our example based on the fact that demonstrated splenic lesions have similar size, location, and configuration in both examinations. In conclusion, MRI has greater contrast resolution and consequently lesion conspicuity than other modalities for many soft tissue lesions. Therefore, it may be helpful to evaluate patients with lymphangiomatosis more definitely than CT or bone scintigraphy alone.

References

1. Moerman P, Geet CV, Devlieger H (1997) Lymphangiomatosis of the body wall: a report of two cases associated with chylothorax and fatal outcome. Pediatr Pathol Lab Med 17: 617–624
2. Gorham LW, Wright AW, Shultz HH, et al (1954) Disappearing bones: a form of massive osteolysis. Am J Med 17: 674–682
3. Levine C (1989) Primary disorders of the lymhatic vessels – a uniform concept. J Pediatr Surg 24: 233–240
4. Taybi H, Lachman RS (1996) Radiology of syndromes, metabolic disorders, and skeletal dysplasias. Mosby Year Book, St Louis
5. Chu JY, Graviss ER, Danis RK, et al (1977) Lymphangiography and bone scan in the study of lymphangiomatosis. Pediatr Radiol 6: 46–48
6. Damron TA, Brodke DS, Heiner JP, et al (1993) Case report 803: Gorham's disease. Skeletal Radiol 22: 464–467