IMAGES IN THORAX

Wandering pulmonary shadows coinciding with pericardial and pleural effusions

Yusaku Nishikawa,[1] Osamu Nishiyama,[1] Shigeki Shimizu,[2] Akiko Sano,[1] Yuji Tohda[1]

[1]Department of Respiratory Medicine and Allergology, Kindai University Faculty of Medicine, Osakasayama, Osaka, Japan
[2]Department of Pathology, Kindai University Faculty of Medicine, Osakasayama, Osaka, Japan

Correspondence to
Dr Osamu Nishiyama, Department of Respiratory Medicine and Allergology, Kindai University Faculty of Medicine, Osakasayama 589-8511, Japan; nishiyama_o@yahoo.co.jp

Received 31 March 2020
Revised 3 May 2020
Accepted 18 May 2020
Published Online First 10 June 2020

CASE PRESENTATION

A 43-year-old man was referred to a nearby hospital because of exertional dyspnoea. Chest radiography revealed an enlarged heart and bilateral lower lung infiltrates (figure 1A). Chest CT revealed bilateral basal pulmonary consolidations, pericardial effusion and small amounts of pleural effusion bilaterally (figure 1B). He was referred to our hospital and underwent pericardiocentesis. Because of a high triglyceride concentration in the pericardial effusate (1287 mg/dL), the patient was diagnosed with chylopericardium. He was prescribed a low-fat diet with subsequent resolution of the pericardial effusion and exertional dyspnoea. Although the bilateral consolidations on chest CT also resolved (figure 1C), on the following year, ground-glass opacities appeared, mainly in the right upper lobe and right S6 (figure 1D). The patient's physical examination and blood test results were unremarkable. His pulmonary function tests were normal. The findings of bronchoalveolar lavage fluid obtained from the right S3 were as follows: total cell count 1.6×10^7/mL, alveolar macrophages 68.0%, lymphocytes 26.8%, neutrophils 0.8%, eosinophils 4.4%, CD4/CD8 ratio 1.73. A transbronchial lung biopsy was non-diagnostic. A surgical lung biopsy was then performed to obtain a definitive diagnosis. Specimens from the patient's right S2, S3 and S8 lobes showed marked proliferation of anastomosing lymphatic channels in the visceral and parietal pleura, interlobular septa and bronchovascular bundles (figure 2A,B). The bland appearance (figure 2C) and low MIB-1 index of the lymphatic endothelium suggested a benign process. The lymphatic endothelium was found to be D2-40 positive by immunostaining (figure 2D). The lesion did not contain HMB45-positive cells. Focal pulmonary oedema with proliferation of lymphatic channels was also seen in the S2 and S3 specimens. These findings were diagnostic of diffuse pulmonary lymphangiomatosis (DPL). One month after the biopsy, pleurodesis was performed to control right chylothorax, which had appeared. The patient remains alive without signs/symptoms 1 year after the diagnosis, except for worsening ground-glass opacities on chest CT.

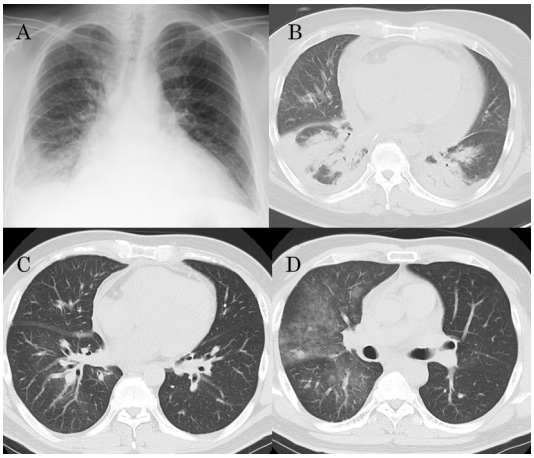

Figure 1 Chest X-ray image shows enlarged heart and bilateral lower lung infiltrates (A). Chest CT image shows bilateral basal pulmonary consolidations, pericardial effusion and slight bilateral pleural effusion (B). Athough the bilateral basal pulmonary consolidations on chest CT resolved, on the following year, ground-glass opacities appeared in the right upper lobe and right S6 (C,D).

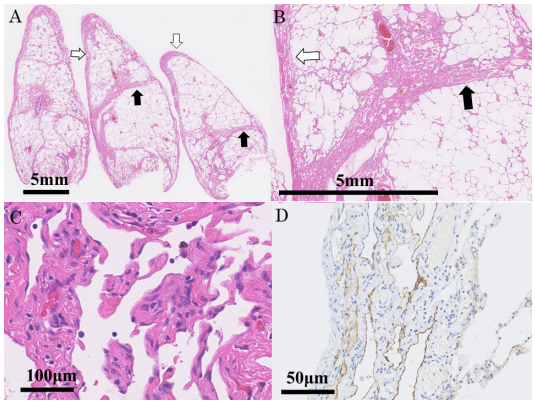

Figure 2 Surgical lung biopsy specimen of the right S8 shows proliferation of lymphatic channels in the pleura (white arrows) and interlobular septa (black arrows) (A,B; H&E). The lymphatic channels are anastomosing and are shaped irregularly with a bland-appearing endothelium (C, H&E). The lymphatic endothelium was D2-40 positive (D, D2-40 immunostaining).

DISCUSSION

Lymphangiomatosis is a very rare disease that is characterised by diffuse infiltration of lymphangiomas, which can occur in any part of the body that contains lymphatic vessels.[1] The differential diagnosis includes lymphangioleiomyomatosis and lymphangiectasis. Lymphangioleiomyomatosis contains HMB45-positive cells, and lymphangiectasis does not show an increased number of anastomosing lymphatic vessels. Because the disease usually occurs in children and adolescents, an adult

To cite: Nishikawa Y, Nishiyama O, Shimizu S, *et al*. *Thorax* 2020;**75**:817–818.

patient with this condition is even more rare. If the abnormality predominantly involves the thorax, it is called DPL.[2] In addition to mediastinal soft-tissue infiltration and enlargement, abnormalities of the peribronchovascular bundles, interlobular septal thickening and diffuse ground-glass opacities on chest CT have been reported in patients with DPL.[3] Chylothorax and chylopericardium are possible complications. Both the bilateral basal pulmonary consolidations and subsequent ground-glass opacities in the right upper lobe and S6 in our patient are consistent with the previously reported findings. However, to our knowledge, this is the first reported case in which the features of the pulmonary shadows changed with time. The reasons for the changes are unknown. However, perhaps the pressure in the abnormal lymphatic vessels changed after drainage of the pericardial effusion.

Contributors YN contributed to patient management. ON conceived the idea for the case report. YN and ON drafted the initial manuscript. SS are responsible for pathological findings. ON and AS critically reviewed the manuscript. YT approved the final version of the manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement There are no data in this work.

REFERENCES

1 Blei F. Lymphangiomatosis: clinical overview. *Lymphat Res Biol* 2011;9:185–90.
2 Faul JL, Berry GJ, Colby TV, *et al*. Thoracic lymphangiomas, lymphangiectasis, lymphangiomatosis, and lymphatic dysplasia syndrome. *Am J Respir Crit Care Med* 2000;161:1037–46.
3 Sun X, Shen W, Xia S, *et al*. Diffuse pulmonary lymphangiomatosis: MDCT findings after direct lymphangiography. *AJR Am J Roentgenol* 2017;208:300–5.