Pulmonary lymphangiectasia (PL) is a rare malformation characterized by dilatation of pulmonary lymphatics. This condition occurs mainly in neonatal and newborn patients. A few cases of PL occur in the young children and adult have been reported. We report another case of a 16-year-old female patient presenting with exertional dyspnea and nonproductive cough. Chest radiography shows bilateral chylous pleural effusions and perihilar interstitial infiltration. Contrast enhanced computed tomography (CT) shows bilateral pleural effusions, pericardial effusion, and lower lobes consolidation. High-resolution CT reveals interstitial changes and patchy areas of ground-glass opacity. Although no pathognomonic findings on HRCT, the interstitial process depicted this imaging tool provides the information to suggest clinical diagnosis when combined with appropriate clinical findings.

Key words: CT, high resolution; Lymphangiectasis, pulmonary; Radiography

CASE REPORT

A 16-year-old female patient was admitted to our hospital with exertional dyspnea, nonproductive cough, and sore throat for three days. There was no headache, chest pain, fever, neck stiffness, leg edema, diarrhea or conscious disturbance. She had a pneumonic episode one year ago and had no other medical disease. There was no significant family history. Physical examination revealed dullness to percussion and decreased breath sounds over both lower lung fields. Arterial blood gas obtained while the patient breathed room air revealed hypoxemia. Routine laboratory examinations were unremarkable. Mycoplasma IgG and IgM, cold agglutinin, rheumatoid factors, and antineutrophic cytoplasmic antibody levels were all within the normal range. Chest radiograph obtained during admission showed moderate bilateral pleural effusions and perihilar interstitial infiltration (Fig. 1). Pulmonary function tests showed mild restrictive lung function. Echocardiography showed moderate pericardial effusion and bilateral pleural effusions without abnormality of proximal pulmonary veins and cardiac...
chambers. Abdominal CT was negative. A right pleural thoracentesis was performed which revealed cloudy and milky fluid consistent with chylothorax. Cytologic analysis showed no evidence of malignant cells. Cultures for bacteria, tuberculosis, fungi, and virus were negative. The fiberoptic bronchoscopy examination showed no endobronchial lesion and bronchial washing cytology and bronchial alveolar lavage were negative. A lymphangiogram was suggested but was not performed for unknown reason. Contrast enhanced helical CT of the chest showed bilateral pleural effusions, pericardial effusion, and bilateral lower lobes consolidation (Fig. 2). HRCT of the lung was also performed which demonstrated patchy areas of ground-glass opacity, smooth interlobular septal thickening, and peribronchovascular bundle thickening (Fig. 3a, 3b). The diagnosis of pulmonary lymphatic disorder was arrived clinically with combination of HRCT findings. In order to confirmed the clinical diagnosis, lung biopsy obtained from the right middle lobe of the lung was performed by video-assisted thoracoscopic surgery. Histopathological examination of biopsy specimen demonstrated marked dilatation of the interlobular lymphatic vessels, chronic inflammatory cell infiltration in the interlobular septa, focal lung edema, and no significant fibrosis (Fig. 4). The final diagnosis was pulmonary lymphangiectasis.

The patient was treated conservatively by dietary control with low fat intake. After 10 hospital days, a repeated chest radiograph showed almost complete resolution of the chylos pleural effusions and mild cardiomegaly. On the 11th hospital days, she was discharged from the hospital free of symptoms. A follow-up chest radiograph nine months later showed only mild cardiomegaly (not shown). Follow-up HRCT about one year after initial CT examination revealed minimal residual pericardial effusion and mild smooth interlobular septal thickening (not shown). At the time of this writing the patient was free of symptoms and in good health.

**DISCUSSION**

Pulmonary lymphangiectasia is a rare malformation and thought to be caused by failure of normal regression of the pulmonary lymphatic during gestation, resulting in lymphatic dilatation [1, 7]. Pulmonary lymphangiectasis can be classified as follows: I, primary: (1) limited to the lungs, (2) with pulmonary and mediastinal involvement, (3) generalized lymphatic abnormality; II, secondary to obstruction of pulmonary venous outflow [5].

The clinical pictures and prognosis of PL vary according to the presenting type. Primary PL is generally considered to occur exclusively in young children. Primary form has a dismal prognosis when occur in neonatal period [3,5,6,8]. Patients with generalized lymphangiectasia usually have a milder form of pulmonary involvement and a better prognosis. White et al [3] reported a 25-year-old male with a mild generalized form of PL without intestinal involvement. The authors suggested that the lack of gastrointestinal features might partially be explain the late presentation of the patient. Our patient presented at aged of sixteen may be of same reason because she also had no gastrointestinal disorder.
Chest radiographic findings are nonspecific in this entity which include interstitial changes, hyperinflation, cystic changes, and pleural effusion [2, 3, 5, 6, 8, 9]. Conventional chest CT findings in a newborn patient of cystic lesions and perihilar interstitial infiltrate have been reported [9]. White et al [3] demonstrated the CT appearance of a generalized PL as massive mediastinal lymphatic dilatation surrounding which encase the mediastinal structures and appearing to extend out into the lungs. In the present case, CT revealed bilateral pleural effusions, pericardial effusions, and bilateral lower lobes consolidation, but there were no cystic lesions and no mediastinal involvement. Faul JL et al [10] reported the magnetic resonance findings in an infant patient presented as widespread cystic formation and abnormal lymphatic vessel. Scott C. et al [6] reported the first case of primary PL in an infant who was examined by HRCT, which showed scattered diffuse reticular changes in all lung zones. In our patient, in addition to reticular opacities, areas of ground-glass attenuation are also demonstrated on HRCT. To our knowledge, the present case may be the first adolescent and second case of PL that was studied with HRCT. In present case, the interlobular septal thickening and peribronchovascular bundle thickening are due to dilated lymphatic vessels filled with fluid, whereas the ground-glass attenuation may be due to reversible inflammatory process. In this report, although no pathognomonic features was identified on HRCT, interstitial changes caused by dilated lymphatic vessels depicted on HRCT combined with clinical features (large volume of chylous pleural effusion) had provided important information for clinical impression.

Histologically, the disease is characterized by marked dilatation of lymphatic vessels in the pleura, subpleural interstitium, interlobular septa, and peribronchovascular bundle [5]. The size of dilated lymphatic vessels varies from those that are only microscopically recognizable to those that can be seen grossly as small cysts [5].

The HRCT findings of PL, which is dilated lymphatic channels seen on imaging studies, also can be seen in other pulmonary diseases, like diffuse pulmonary lymphangiomatosis, pulmonary lymphangitic carcinomatosis, pulmonary edema, and pulmonary lymphangioleiomyomatosis. The typical HRCT features of diffuse pulmonary lymphangiectasis.

Figure 3. (a) High resolution CT at the lower lung shows interlobular septal thickening (arrows, right middle and inferior lingular segment) and patchy areas of ground-glass opacity (white arrowheads, right middle lobe). (b) High resolution CT at the level of carina shows bronchovascular bundle thickening (arrows, left lung).

Figure 4. High-power magnification of lung tissue shows marked dilatation of the lymphatic vessels (1) in the interlobular septa and mild interstitial chronic inflammatory cell infiltration.
giomatosis include diffuse, smooth thickening of interlobular septa and bronchovascular bundles with extensive infiltration of the mediastinal fat and associated perihilar infiltration [11]. Histologically, the non-proliferative dilatation of normal lymphatic vessels in PL is distinct from the increased number and complexity of lymphatic spaces in diffuse pulmonary lymphangiomatosis. Pulmonary lymphangitic carcinomatosis can also cause peribronchovascular bundle and interlobular septal thickening, but the interstitial thickening is usually nodular or beaded rather than smooth in appearance [12]. Hilar and/or mediastinal lymphadenopathy is often seen in lymphangitic carcinomatosis but not in the patients with PL. Although the lesion of pulmonary edema and PL both show basilar distribution, differentiating these two entities can be made on clinical ground. Lymphangioleiomyomatosis is a rare idiopathic disease characterized by progressive smooth muscle proliferation in the pulmonary lymphatic vessels, blood vessels, and airways. The typical HRCT findings of lymphangioleiomyomatosis are numerous thin-walled cysts distributed throughout both lungs whereas cystic spaces were not presented on CT in patients with PL.

Pulmonary lymphangiectasis may leads to respiratory failure. Treatment of PL and chylothorax is difficult. Treatment with dietary control, includes a low-fat and high protein diet, in addition to repeated aspiration of lymph accumulation, have been shown to be of some benefit [3, 13]. Although not often surgical procedures have been used and success have been reported [5, 13].

In conclusion, we reported the radiographic, CT, and HRCT findings of a generalized PL presented at a later age. Although there is no pathognomonic feature of this entity on imaging studies, the diagnosis can still be suggested with combined clinical features and HRCT findings, which is helpful especially when lung biopsy cannot be performed.

**REFERENCES**

肺部淋巴血管擴張症的X光片，電腦斷層及高解析度電腦斷層影像發現：病例報告

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肺部淋巴血管擴張症是一罕見異常，其特性為淋巴血管擴張。這類疾病大部分發生於新生兒或嬰兒，有極少發生於青少年或成人。本文報告一十六歲的女性病人，主訴為運動性呼吸困難及乾咳。胸部X光素片顯示雙側肋膜積水及肺門旁間質浸潤。電腦斷層攝影影像顯示雙側肋膜積水，心包膜積水及雙側下肺葉堅實變化。高解析度電腦斷層影像顯示肺間質變化及多處斑樣毛玻璃狀陰影。雖然淋巴血管擴張症在高解析度電腦斷層攝影並沒有示病徵象，但組織間隙的病變可在高解析度電腦斷層攝影顯現，這可提供有用的資料，再合併臨床所見，便可做出正確診斷。

關鍵詞：高解析力電腦斷層，淋巴血管擴張症，X光攝影