An Unusual Manifestation of Pulmonary Lymphangiomyomatosis: Airspace Consolidation Masking Cystic Lung Lesions

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We report a case of pulmonary LAM occurring in a 24-year-old pregnant woman in whom large areas of air-space consolidation partly masked interstitial lung disease on plain radiography. For correct diagnosis, high resolution CT was considered to still be a potent method for the disclosure of typical interstitial lung changes, including thin-walled cysts. Follow-up CT showed progressive worsening of these interstitial lung lesions.

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Pulmonary lymphangiomyomatosis (LAM), a rare idiopathic interstitial lung disease occurring almost exclusively in women of reproductive age, is pathologically characterized by irregular progressive proliferation of atypical smooth muscle in the pulmonary lymphatic vessels, blood vessels, and airways, resulting in the obstruction of pulmonary lymphatics, veins, and bronchioles (1, 2). The clinical outcome of this is chylos pleural effusion, pulmonary hemorrhage, and recurrent pneumothorax (1, 2). Although the hallmark of the radiological findings of pulmonary LAM is known to be multiple thin-walled cysts distributed diffusely throughout the lungs (3–8), air-space consolidation mainly as a result of pulmonary hemorrhage or lymphedema may accompany the cystic lesions (1, 2). To our knowledge, however, air-space consolidation if any-accompanying pulmonary LAM, has been rarely emphasized in the radiology literature (3–5). We report a case of pulmonary LAM occurring in a pregnant woman in whom, on plain radiography, large areas of air-space consolidation masked interstitial lung disease.

Case Report

A 24-year-old pregnant woman at the 35th week of gestation was referred to this hospital with a complaint of progressive dyspnea. This had developed two years earlier when she had had her first baby. She subjectively felt some relief of dyspnea after delivery, so no work-up was performed at that time. After the pregnancy during which she attended this hospital, her dyspnea worsened again. She denied any family or medical history of seizure or mental retardation, or previous episodes of pneumothorax or hemoptysis. Although there was a mild intermittent cough, it was not productive. No specific medication was given before or during pregnancy. Her temperature was 36.5°C, respiration rate was 24 per min, pulse was 100, and blood pressure was 140/80 mmHg. Physical examination revealed somewhat decreased breath sounds over both lung fields unaccompanied by rales or wheezing, as well as edema of the extremities, and multiple lymph nodes palpable at both inguinal regions. Heart sound was normal and there was no dermatologic abnormality. Neurologic examination was also negative. White blood cell count was 7,000/mm³ with a normal differential count and the hematocrit was 40.2 percent. Serum protein was 6.2g
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per 100ml with albumin of 3.2g per 100ml. Other blood chemistry and electrolytes were normal. Arterial blood gas analysis showed pH of 7.42, PCO₂ of 31 mmHg, PO₂ of 55 mmHg, and bicarbonate of 20 mmol per liter. Pulmonary function testing with spirometry revealed a mild restrictive pattern with a forced expiratory volume in one second (FEV₁) of 1.82 L (60% of predicted), a forced vital capacity (FVC) of 2.07 L (60% of predicted), and a FEV₁/FVC ratio of 104%. Electrocardiography was normal.

Chest radiograph showed large areas of air-space consolidation in both lower lung fields. In addition, widespread dissemination of fine reticular opacities was seen in both lung fields (Fig. 1A). On the fourth hospital day, she underwent cesarean section, and this revealed about 200 cc of chylous ascites. The operation

**Fig. 1.** 24-year-old pregnant woman with pulmonary lymphangiomyomatosis.
A. Supine anteroposterior chest radiograph before delivery shows large areas of air-space consolidation predominantly in both lower lung fields. There is a widespread dissemination of fine reticular opacities throughout the lungs. Note superior mediastinal fullness and borderline cardiomegaly indicating pregnancy-associated volume overload.
B and C. CT scans obtained three days after delivery. High-resolution CT (HRCT) at level of inferior pulmonary hilum (B) shows large areas of air-space consolidation associated with a marked irregular interstitial thickening in the right middle lobe and both lower lobes. Several small thin-walled cysts are clearly seen within or near the areas of air-space consolidation (arrows). Note bilateral pleural effusion. HRCT at level of apical lung (C) shows numerous thin-walled, air-filled cysts in both lungs.
D. Erect posteroanterior chest radiograph obtained 45 days later than in Fig. 1A shows significantly improved air-space consolidation. In contrast, small cysts and reticular opacities are more prominent in the areas of previously demonstrated air-space consolidation. Now, the heart and mediastinum appear normal.
E and F. CT scans obtained nine months after delivery at same level as in b and c show more localized air-space consolidation (E) and numerous cysts (E and F) which are now slightly more prominent in size and in number in comparison with those shown in b and c.
was uneventful and the baby was in good condition.

The patient was thought to be suffering from LAM, and CT of the chest and abdomen was performed on the third postpartum day. In both lungs, and especially in the right middle lobe and both lower lobes, high-resolution CT (HRCT) of the chest demonstrated large areas of multifocal air-space consolidation associated with irregular thickening of interlobular and intralobular interstitial lines (Fig. 1B). Numerous air-filled cysts, 2–10 mm in diameter, were also visualized in both lungs; they were more conspicuous in both upper lobes and the right costophrenic sinus (Fig. 1C). A small amount of bilateral pleural effusion was also seen. CT of the abdomen after the intravenous administration of contrast material showed multiple enlarged lymph nodes in the retrocrural and retroperitoneal areas. Except for the physiologically enlarged and hypertrophied uterus, no visceral abnormality was found.

The diagnosis of LAM was confirmed by biopsy from the right inguinal lymph node. The patient was injected intramuscularly with a monthly dose of a 500 mg medroxyprogesterone acetate; a series of follow-up chest radiographs after delivery showed the gradual disappearance of areas of air-space consolidation, while reticular and cystic opacities were seen more conspicuously in those areas (Fig. 1D). HRCT of the chest obtained nine months after delivery demonstrated more localized air-space consolidation (Fig. 1E). The cystic lesions had become slightly more prominent in size and in number (Fig. 1E and F), indicating radiologically the downhill course of the disease. As at the time of this report, the patient is still being treated with progesterone with dyspnea waxing and waning.

Discussion

The radiological hallmark of pulmonary LAM is multiple thin-walled cysts distributed diffusely throughout the lungs. CT, particularly HRCT, has been reported to be superior to conventional radiography in showing the presence and morphology of lung abnormalities accompanying LAM (3–8). Smooth muscle proliferation can occur anywhere along the walls of lymphatic channels and venules throughout the lung, resulting in a varying spectrum of clinical and radiographic manifestations (1, 2). This can sometimes cause accumulation of lymph or blood within the alveoli, manifested on radiography as air-space consolidation (1, 2). To our knowledge, however, such alveolar opacities associated with LAM have seldom been emphasized in the radiology literature (3–5).

In their study of eight patients with pulmonary LAM, Sherrier et al (3) found patchy, ill-defined areas of increased attenuation in two patients. Lenoir et al (4) demonstrated dependent alveolar areas of attenuation in one of nine patients with LAM. Müller et al (5) found small, localized areas of air-space consolidation in three of 14 patients with LAM. The present case shows that areas of air-space consolidation can be large enough to mask small cysts and reticular opacities on conventional radiography. Although there was evidence of interstitial disease in other areas of the lung, this was not sufficient to reach a correct diagnosis. In contrast, HRCT clearly demonstrated small thin-walled cysts as well as air-space consolidation, making the diagnosis of LAM straightforwardly. As shown in the present study, CT was also useful for follow-up monitoring of the disease.

The relationship between the severity of dyspnea and pregnancy or delivery in the present case support sex hormone theory as the pathogenesis of LAM (9, 10). With regard to the pathogenesis of smooth muscle proliferation in LAM, there have been different opinions; these include a traumatic rupture of the thoracic duct with secondary smooth muscle proliferation, a neoplastic proliferation of lymphangiopericytes, a hamartomatous malformation, a hormonally stimulated hamartoma, and a forme fruste of tuberous sclerosis (9). The almost exclusive occurrence of LAM in women of child-bearing age as well as its reported association with oral contraceptives, human chorionic gonadotropins, menses, pregnancy, and hormonal receptors has suggested, however, that sex hormones play a role (9, 10). Despite these observations, attempts at hormonal manipulation or organ ablation surgery have not been invariably successful. It might be inferred that treatment failed partly because therapy was initiated late in the course of the disease (9, 10).

Although the exact nature of air-space consolidation in the present case was not pathologically proven, the presence of pleural effusion and chyloous ascites, and absence of a history of hemosiysis favor lymphedema rather than hemorrhage resulting from venous obstruction, if the latter cannot be entirely excluded. Air-space consolidation is unlikely to be caused by muscle growth per se because of its rather rapid clearance after delivery. Pregnancy-induced volume overload is another consideration; since on follow-up some alveolar opacity still remained, this overload might not entirely explain such opacity, though it might have aggravated the situation by elevating the venous pressure, thus further obviating lymphatic drainage. Cardiac-related problems, infections, and other medical reasons could be clinically excluded; based on clinical
and radiologic findings, we speculate that pregnancy accompanied by volume overload significantly contributed to lymphedema by inducing or accelerating smooth muscle proliferation along lymphatic vessels.

We conclude that areas of air-space consolidation due to either lymphedema or hemorrhage can be large enough to mask the characteristic findings of pulmonary LAM on conventional radiography. HRCT discloses typical interstitial changes in thin-walled cysts, and so is extremely valuable for correct diagnosis.

References

7. 이경수, 최은완, 이병호 등. 폐 림프관 평활근증증 증례보고. 대한방사선의학회지 1991; 27: 240-244
8. 문우경, 임정기, 한만청. 폐 포진 증·흡기 폐포성 폐경결: 고해상 CT소견, 대한방사선의학회지 1991; 27: 543-546