112 年奇美醫院胸腔內科臨床病例討論會

日期：中華民國 112 年 09 月 26 日(星期二)

時間及地點：16:00-17:00

課程活動題目：**Lymphangioleiomyomatosis in CXR**

主講人：謝俊民主任

主辦單位：奇美醫院胸腔內科

課程地點：10 樓討論室

教育積分：台灣胸腔暨重症加護醫學會

參加對象：主辦單位所屬院內醫師

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摘要:

Lymphangioleiomyomatosis (LAM)

Female predominance-Occurs exclusively in females, natural history variable. Symptoms-Dyspnea (third of fourth decade of life); pneumothorax (50-80%), chylothorax (7-39%), hemoptysis (28-40%).

Radiology – The chest radiographic findings are variable

Possible appearances include:

- Normal (early in the course of the disease).
- Interstitial opacities (reticular, reticulonodular, miliary). These opacities result from the compression of smooth muscle-rich interstitial tissue by more dilated, cystic airspaces.
- Honeycomb changes.
- Hyperinflation (33 to 62 percent of patients) with severe emphysematous-like changes in advanced disease.

Chest CT – High resolution chest CT (HRCT) scanning is very useful and is much more sensitive than routine chest radiography in demonstrating the cystic nature of the disease. The findings of diffuse, homogeneous, small (less than one cm diameter) thin-walled cysts can be highly suggestive of the diagnosis in an appropriate clinical context. There is a close correlation between the extent of the cystic parenchymal changes (as measured by quantitative HRCT) and disease severity (as determined by spirometry, diffusing capacity, lung volumes, or exercise performance). Thus, HRCT scanning may have both a diagnostic and a prognostic role.

Pulmonary Function Tests- Pulmonary function testing usually reveals an "obstructive" or "mixed" pattern. The lungs are often hyperinflated, with an increased total lung capacity (TLC) and increased thoracic gas volume. Gas trapping, as evident by an increase in residual volume (RV) and RV/TLC ratio, is commonly present. Airflow limitation and reductions in diffusing capacity occur in the majority of patients. Studies of pulmonary mechanics show that mean elastic recoil is reduced and that upstream resistance is increased. Both a loss of elastic recoil and an increase in airway resistance contribute to the observed airflow limitation.

Gas exchange is often abnormal, with a markedly reduced DLCO and an increase in the alveolar-arterial oxygen difference. Diminished exercise performance is found in most patients, generally characterized by:

- Reduced oxygen consumption.
- Low anaerobic threshold.
- An abnormal and excessive ventilatory response, with a high respiratory rate, excessive minute ventilation, and reduced breathing reserve.
- Abnormal baseline or exercise dead space to tidal volume (VD/VT) ratio.

The primary determinants of the exercise limitation are related to airflow limitation and to mechanical factors (eg, decreased breathing reserve, increased work of breathing). In addition, pulmonary vascular dysfunction/destruction also leads to severe impairment in exercise performance in many patients.

Therapy-

- oophorectomy and antiestrogen regimens tried but disappointing results.
- Transplantation.