■ Sun-247 ■

Lung Involvement in Neurofibromatosis

고려대학교 구로병원 호흡기내과

*박민정, 최주환, 오지연, 이영석, 민경훈, 허규영, 이승룡, 강경호, 심재정

Background and objective: Pulmonary manifestation of neurofibromatosis (NF) has not been highlighted compared to other phenotype of NF, such as skin and bone abnormality. Since respiratory problem manifested as abnormal chest image in NF patients affects their mortality and morbidity, characteristics of computed tomography (CT) scan in NF patients need to be researched. **Methods:** Patient data were collected retrospectively from 2005 to 2018 in Korea University medical center Guro- hospital. Among 113 patient diagnosed as NF, 26 patients who are older than 20 year old and underwent chest CT scan were selected. **Results:** The mean age of subjects were 44 years. Sixteen (61.0%) subjects out of twenty-six were male. Emphysema (30.8%) is the most dominant CT scan finding as well as no finding (30.8%). Bulla (26.9%) is the next most frequent characteristics followed by ground glass opacity (15.4%), and thoracic scoliosis (15.4%), ground glass density nodule (7.7%), small nodule (7.7%), non-small cell lung cancer (7.7%), thin walled cyst (7.7%), and pneumothorax (3.8%). **Conclusion:** Chest CT abnormality is found in over two-third NF patients in out data. Clinician should understand lung characteristics and concomitant physiology of NF patient in the clinical field.

Table 1. Radiologic findings of neurofibromatosis patient with lung involver

	No. of cases (%)
No finding	8 (30.8)
Emphysema	8 (30.8)
Bulla	7 (26.9)
GGO	4 (15.4)
Thoracic scoliosis	4 (15.4)
GGN	2 (7.7)
Small nodule	2 (7.7)
NSCLC (Adeno ca)	2 (7.7)
Thin walled cyst	2 (7.7)
Pneumothorax	1 (3.8)

■ Sun-248 ■

A case of direct hemoperfusion with polymyxin B-immobilized fiber for the Acute exacerbation of IPF

[1]일산 백병원, [2]호흡기내과

*정재우[1], 박소희[1,2], 이성순[1,2], 박혜경[1,2], 구현경[1,2], 강형구[1,2]

Acute exacerbation of idiopathic pulmonary fibrosis acute exacerbation (AE-IPF) is known to have very poor prognosis. AE-IPF with severe respiratory failure has very high mortality and, there is no proven treatment. Some studies have reported the effectiveness of direct hemoperfusion with polymyxin B-immobilized fiber column (PMX-DHP) for AE-IPF with respiratory failure. Here we reported the case of improving respiratory physiology of AE-IPF with PMX-DHP. An 82-year-old woman has been under observation for 2 years after being diagnosed with IPF in our hospital. In June 2018, the patient was admitted to our hospital due to exertional dyspnea and cough occurred 7 days ago. The patient's partial pressure of oxygen was 62.0 mmHg, although she had an oxygen supply at nasal prong 6L/min. Fine crackles were audible in both lungs. Laboratory findings upon admission revealed an increased WBC count (10580/uL), elevated CRP level (31.2mg/dL). Chest PA revealed progression of diffuse ground glass opacity (GGO) in both lungs. Comparing previous echocardiography, we excluded cardiac failure and pulmonary embolism. Also, there was no definite pneumonia evidence, so the patient was diagnosed with suspected AE-IPF. High flow nasal cannula and antibiotic therapy with ceftriaxone and azithromycin were initiated, but the patient's respiratory condition worsened, mechanical ventilation was initiated and, antibiosis was change to meropenem at 3rd day. We initiated methylprednisolone (1mg/kg) and, PMX-DHP because her respiratory condition was very quickly getting worse. PMX-DHP was performed for 12hours in two days. One day after PMX-DHP, the PaO2/FiO2 ratio improved from 113 to 400, we could decrease FiO2 from 75% to 35% . The lung compliance improved from 30.9(ml/cmH2O) to and 60.3. Chest PA revealed decreased GGO. Based on improving patient's lung physiology, we tried to wean for MV. But, massive bleeding occurred during removing dual lumen catheter on day 13. We could not control the bleeding because of bleeding focus. The patient status was getting worse after then and, died on day 25 PMX-DHP may be able to improve the prognosis of AE-IPF patients with severe respiratory failure