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ENDOSCOPIC TREATMENT OF PULMONARY ALVEOLAR PROTEINOSIS WITH WHOLE LUNG LAVAGE. THE FORLANINI HOSPITAL EXPERIENCE

Top Author: Gabriele Lucantoni

Servizio Endoscopia Toracica, Azienda Ospedaliera San Camillo- Forlanini, Roma Italy

Area and Category(at submission):

[WCBIP] Interventional pulmonology

Presentation Preference:

Poster

Case Report: NO

NO

BACKGROUND:Pulmonary Alveolar Proteinosis (PAP) is a rare lung disease characterised by abnormal intra-alveolar accumulation of surfactant-like lipoproteinaceous material. Clinical presentation is usually with non specific respiratory symptoms such as dyspnoea or minimally productive cough. The clinical course can be variable, ranging from spontaneous resolution to respiratory failure and death. The appearance on HRCT is characterised by the typical crazy paving pattern (FIGURE 1A). Recent advances now allow diagnosis by examination of the milky fluid obtained by Bronchoalveolar Lavage (BAL) fluid. Although there are no randomised controlled studies Whole Lung Lavage (WLL) is actually considered the most effective form of treatment. Our expericed would contribute to confirm the efficacy and safety of this therapeutical approach.

METHODS:From 2001 to 2011 n° 10 subjects (6 male and 4 female; median age 35±5) were referred

to our Endoscopy Center for the diagnosis and tretment of PAP. All subjects had the typical HRCT bilateral pattern, and showed slight-moderate dyspnea (Borg Scale 3 ±1). Patiens were subjected to serial monthly WLL alternating the lung. Under general anesthesia they were intubated by Harrel-Dumon rigid bronchoscope and serial aliquots (50 ml each) of warm (36-37°C) neutral sterile saline solution were instilled into the whole bronchial segments, using a flexible instrument, and draining the milky fluid from the selected lung. The number of WLL for each patients was decided on the basis of clinical-radiological improvement and progressive clarification of BAL.

RESULTS: A median of 19±5 WLL were performed for each patients. All subjects had improvement

of dyspnea, a progressive pulmonary clarification was showed at HRCT (FIGURE 1B). No significant post-operative complications occurred during these procedures.

CONCLUSIONS:Our experience confirms the WLL could be considered the treatment of choice for PAP.

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Figure 1A



Figure 1B