Nine-Year Experience of Pulmonary Alveolar Proteinosis In Children

Tıp Fakültesi

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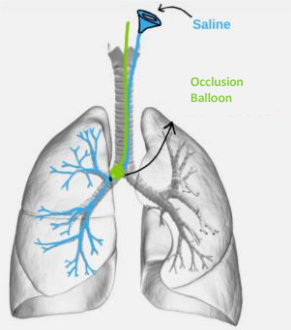
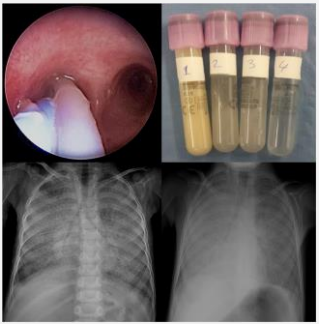
Aim and Background: Pulmonary alveolar proteinosis (PAP) is a rare interstitial lung disease that occurs with surfactant accumulation in the lung parenchyma in childhood. The gold standard treatment is whole lung lavage (WLL). The aim of the study is to evaluate the outcomes of patients followed in our clinic due to PAP.



Material and Methods:

- May 2015 - July 2024, retrospective
- Demographics and associating pathologies
- Genetic examination
- Histopathological evaluation
- Intervention technique, number and volume
- Blood gas levels before and after bronchoscopy
- Postoperative mechanical ventilation (MV)
- Complications
- Follow-up data



Results:

- 4 patients, all male
- Median age 32,5 (28-88) months
- One patient with genetic mutation, unknown relevance
- One patient with mitral insufficiency
- One patient with wandering atrial pacemaker
- Diagnosis:
  - 3 bronchoalveolar lavage
  - 1 thoracoscopic wedge biopsy
- Immunhistopathology reports:
  - Periodic acid-Shiff stain (PAS +)
- MV support for 24 hours after interventions
- No complications due to therapeutic bronchoscopy
- Mean follow-up time 20 (6-89) months

p	Procedure	Lavage	Mean WLL volume (ml/kg)	Mean preop pH	Mean postop pH	Preop pCO2	Postop pCO2	Continuous respiratory support	Follow-up
1	Rigid bronchoscopy	2	136 (100-172)	7,41	7,29	31	36	Nasal O_2	Ongoing WLL
2	Rigid bronchoscopy	20	136 (110-180)	7,33	7,24	47	60	None	Ongoing WLL
3	Rigid bronchoscopy	4	131,5 (104-180)	7,41	7,30	34	51,5	None	Cure
4	Rigid bronchoscopy	29	100 (60-180)	7,38	7,34	44,5	44	None	Cure

Table 1: Whole lung lavage data of the patients

Conclusion: PAP is a lung disease in childhood regarding an unknown aetiology most of time. The management of PAP requires repeated bronchoscopic whole lung lavage and multidisciplinary approach. Increasing lavage volume may lengthen the intervention intervals in resistant cases.