4.06 Presentation, mode of diagnosis and management of pulmonary alveolar proteinosis syndrome in an Irish population.

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Rationale: Pulmonary alveolar proteinosis (PAP) is a rare syndrome characterized by abnormal accumulation of pulmonary surfactant. Autoimmune PAP results from presence of anti-GM-CSF antibodies and accounts for 90% of cases. Positive serum GM-CSF autoantibodies are sensitive and specific for PAP. Whole-lung lavage (WLL) is current gold standard treatment, with new focus developing on the potential of inhaled GM-CSF.

Methods: Data on all patients with PAP attending our Rare Lung Disease clinic was analysed to establish mode of presentation, diagnosis, CT findings, lung function and treatment.

Results: Shortness of breath (63%) was the most common presenting symptom. 100% of patients had high resolution CT imaging (HRCT) with ground glass changes and 50% had evidence of 'crazy paving' suggestive of PAP. All cases had bronchoscopy and BAL with 50% undergoing biopsy. Mean FEV1 and DLCO were 87.8% (SD +/- 22.9%) and 69.8% (SD +/- 13.5%) respectively. 50% had DLCO $\leq$ 70% predicted. Two patients required long-term oxygen therapy; two have developed fibrosis and two have undergone WLL. Two patients are currently receiving treatment in the open arm of IMPALA-2 trial.

Discussion: Characteristic HRCT findings should prompt the consideration of PAP and include anti-GM-CSF antibody testing.

Disclosures: Nothing to disclose