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Title: Pulmonary alveolar proteinosis due to monocytopenia: Lung transplant or haematopoeitic stem cell transplant?

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Body: Pulmonary Alveolar Proteinosis (PAP) is characterised by the accumulation of periodic acid Schiff-positive, lipid-rich material in alveoli and progressive respiratory failure. It occurs in two distinct forms: auto-immune (primary PAP) or secondary to an associated systemic disorder. Treatment options include whole lung lavage, and Granulocyte Macrophage-colony stimulating factor (GM-CSF). We describe a 22-year-old female presenting with breathlessness, pulmonary infiltrates, recurrent pulmonary sepsis and severe Type 2 respiratory failure requiring oxygen therapy and non invasive ventilation (NIV). HRCT scan and VATS biopsy confirmed a diagnosis of PAP. Anti-granulocyte-macrophage colony-stimulating factor (GM-CSF) autoantibody levels were low, and response to whole lung lavage (WLL, 50 litres) transient. An extensive search identified a monocytopaenia and family history of haematological malignancies prompting bone marrow and skin biopsy identifying a deficiency in circulating monocytes, B lymphocytes and NK lymphocytes, and tissue dendritic cells. The pattern was consistent with an autosomal dominant and sporadic monocytopenia with susceptibility to infection and PAP (Vinh et al Blood 2010). A matched unrelated-donor allogeneic stem cell transplant was performed. Progressive respiratory improvement resulted with no current symptoms of breathlessness, weaning from NIV and LTOT, improved lung function and reduction of alveolar infiltrates. Blood monocyte count now in low normal range. We wish to highlight haematopoietic stem cell transplant as treatment for a rare secondary form of PAP due to DC, monocyte and lymphoid (DCML) deficiency (Bigley et al. J Exp Med 2010).