SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
FORTALEZA 2019
PROMOÇÃO
70 anos Sociedade Brasileira de Reumatologia
REALIZAÇÃO
SCR SOCIEDADE CEARENSE DE REUMATOLOGIA

PULMONARY ALVEOLAR PROTEINOSIS IN A PATIENT WITH C-ANCA POSITIVE REFRACTORY TO CONVENTIONAL THERAPY AND GOOD RESPONSE TO RITUXIMAB: CASE REPORT

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BACKGROUND

Pulmonary alveolar proteinosis (PAP) is a rare entity characterized by alveolar accumulation of surfactant. Three distinct etiologies are described: idiopathic, genetic and secondary, being included in the latter hematological neoplasias, occupational exposure to dust or other toxic materials such as silica, immunosuppressants or autoimmune diseases. The diagnosis is based on compatible clinical-radiological picture with pulmonary image with mosaic pavement, milky appearance of bronchoalveolar lavage and periodic staining with positive acid-Schiff (PAS). The available therapies are the bronchoalveolar lavage washes with large amounts of serum, granulocyte-stimulating factor of macrophages colonies (GM-CSF), and as a third line treatment, Rituximab (RTX). We describe the case of a patient diagnosed with alveolar proteinosis with no response to conventional therapy, with arthritis of large joints and c-ANCA positive with dramatic remission of PAP after RTX treatment.

CASE REPORT

GSF, 48 years old, male, ex-smoker 30 years-old pack, with exposure to dust in the work environment for 2 years. In 2016, he started progressive dyspnea until medium-term efforts, dry cough, arthritis of large joints, weight loss of 12 kg in 4 months and fever. He sought external service in 2017 receiving diagnosis of pneumonia without improvement with antibiotic therapy, evolved with dyspnea progression, hypoxemia and need for intermittent noninvasive ventilation. He was diagnosed with PAP due to pulmonary imaging, bronchoalveolar lavage with a characteristic milky appearance and PSA positivity in the material. Secondary infections were excluded by culture. Prednisone 10mg and hydroxychloroquine 400mg were prescribed due to active arthritis. In the complementary investigation, the patient presented c-ANCA positive several times, with titers up to 1/160. Five therapeutic bronchoalveolar lavages were performed with no improvement. An association with autoimmune disease was suspected, since there is no description of arthritis as a PAP clinical manifestation and due to high titers of c-ANCA. RTX was introduced and after a single dose 2g, patient presented important clinical, radiographic and functional improvement.

CONCLUSION

The association of PAP and autoimmune diseases is rare, and some cases described in the literature are related to vasculitis, rheumatoid arthritis and dermatomyositis. We emphasize the importance of the association of rheumatologic signals and presence of autoantibodies that could help to choose specific treatment, as RTX for this disease.