Case report

Rare case of idiopathic lymphocytic interstitial pneumonia exhibits good response to Mycophenolate Mofetil

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1. Introduction

Lymphoid interstitial pneumonia (LIP) is a poorly understood lymphoproliferative disorder originating from hyperplasia of bronchus-associated lymphoid tissue (BALT). Peribroncholar and interstitial lymphocytes accumulate in response to various stimuli. LIP is usually found in association with several diseases and conditions, however some remain idiopathic. Up to 25% of LIP cases are associated with Sjögren’s Syndrome. Some studies suggest associations with HIV and Epstein–Barr virus. Although LIP has been regarded as steroid responsive and mainly treated with oral corticosteroids, its response has been unpredictable. Approximately 33–50% of patients die within 5 years of diagnosis, and approximately 5% of cases of LIP transform to lymphoma.1 Previous reports have shown successful treatment with Cyclophosphamide pulse therapy and Rituximab in association with Sjögren’s Syndrome.2,3 Another report shows responsive treatment with Prednisone and Mycophenolate Mofetil (MMF) in association with Systemic Lupus Erythematosus (SLE).4 Here we describe a case of idiopathic LIP that shows clinical improvement to Mycophenolate Mofetil after intolerable side effects of high-dose steroids.

2. Case report

A 35 year-old Guyanese female presented to our hospital with progressive left-sided pleuritic chest pain, intermittent dry cough and exertional dyspnea. She denied history of fever, tuberculosis or known exposure, asthma, seasonal allergy, gastroesophageal reflux disorder, postnasal drip or smoking tobacco. She moved to New York City from Guyana 10 years prior with no recent travel history or sick contact. The patient used to work as a certified nurse aid and owned a rabbit. She denied any history of muscle weakness, joint pain, rash, photosensitivity, color changes in her digits, dysphagia, dry eyes or dry mouth. Chest x-ray showed small infiltrates in the left lung base. D-dimer was elevated and CT thorax showed moderate interstitial and airspace opacities in lower lobes, atelectasis and/or pneumonia with no evidence for pulmonary embolism. She owned a rabbit. She denied any history of muscle weakness, joint pain, rash, photosensitivity, color changes in her digits, dysphagia, dry eyes or dry mouth. Chest x-ray showed small infiltrates in the left lung base. D-dimer was elevated and CT thorax showed moderate interstitial and airspace opacities in lower lobes, atelectasis and/or pneumonia with no evidence for pulmonary embolism. She diagnosed with community acquired pneumonia and discharged home on Moxi...
anti-smith, anti-DNA antibodies and anti-SCL70 levels were negative. C3 and C4 levels were within normal range. Urinalysis was negative for RBC, blood and protein. Bronchoscopy with right lower lobe biopsy showed minute fragments of unremarkable bronchial wall with no lung parenchyma identified. Cytology showed negative malignant cells with numerous benign bronchial cells, lymphocytes, neutrophils and macrophages. Bronchoalveolar lavage showed no growth in mycobacterium and fungal cultures with no pneumocystis on smear. After a couple weeks, she was admitted to our hospital secondary to worsening dyspnea on exertion and left-sided chest pain. Video-assisted thoracic surgery (VATS) was performed for lung biopsy. Lung tissue showed no growth in mycobacterium and fungal cultures with no pneumocystis on smear. Left upper and lower lung pathology suggested lymphoid (BALT) hyperplasia with acute and predominantly chronic fibrous pleuritis (Fig. 1). Methylprednisolone pulse therapy was initiated followed by Prednisone 80 mg daily as an outpatient regimen. After a month, the patient developed an acneiform rash on her face, a cushingoid appearance, restlessness and anxiety. At that point, MMF 1 g daily was initiated. Throughout the next 5 months, MMF was increased to 2 g with a rapid taper of Prednisone to 5 mg daily. There was complete resolution in the patient's symptoms with significant clearing of sub-segmental opacities on serial CT (Fig. 2B).

Repeat labs included negative ANA and CRP levels; however p-ANCA continued to stay in the same range until three months later in which they were negative. At that point the patient was in complete remission on MMF 2 g daily with Prednisone completely tapered off. Repeat pulmonary function testing showed improvement in total lung capacity and normal diffusing capacity.

3. Discussion

LIP is a poorly understood lymphoproliferative disorder with unknown pathogenesis. It is associated with several diseases and conditions including Sjögrens syndrome, HIV and Epstein–Barr virus. Treatment regimens have not been well established. However, LIP is believed to respond to steroid therapy most of the time. In our case, the patient was diagnosed with idiopathic LIP. She had no signs of associated conditions, including Sjögrens syndrome or HIV. Initial low ANA levels were attributed to the inflammatory process in the lungs with no criteria meeting connective tissue disease. Even though serial p-ANCA levels trended in the same
range initially, other labs and tissue pathology did not suggest vasculitis. After initial pulse therapy with Methylprednisolone followed by a high-dose Prednisone regimen, the patient was not able to tolerate the side effects. She developed an acneiform rash on her face, a cushingoid appearance, restlessness and anxiety. By starting the patient on MMF, a rapid taper of Prednisone was allowed with complete remission.

MMF was originally approved by the FDA for prevention of transplant rejection. It inhibits the synthesis of guanosine monophosphate (GMP), preventing the proliferation of T and B lymphocytes. MMF has had an increase use in rheumatic diseases, including maintaining remission in moderately severe SLE. Additionally, other reports have shown responsive treatment with Prednisone and MMF in LIP associated with SLE and MMF alone in connective tissue disease-associated interstitial lung disease.4,6

4. Conclusion

In summary, we report a case of idiopathic LIP. MMF was started secondary to intolerable side effects of high-dose Prednisone, allowing for a rapid taper and resolution of symptoms. There was improvement in radiologic findings on serial HRCT, pulmonary function testing and clinical presentation. This case will further support the previously mentioned report of LIP associated with SLE.4 In cases of failed therapy with steroids or severe side effects, one may consider the use of MMF for further treatment. More research will be needed for fostering treatment guidelines in cases of LIP.

Conflict of interest

All authors have no conflict of interest to disclose.

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