A 74-year-old female patient with rheumatoid arthritis was diagnosed with Pneumocystis jiroveci pneumonia (PcP) following therapy with methotrexate and prednisone. Although bactrim treatment was initiated and PcP was not detected by a control bronchoalveolar lavage, the patient died. The precise cause of death remains unknown. As this case illustrates, PcP must be considered as a differential diagnosis in immunocompromised patients with rheumatic disease. The typical course, diagnosis, prophylaxis and treatment of PcP in this patient group are discussed.