An Unusual Cause of Cavitary Lesions in the Lung: Pulmonary Pyoderma Gangrenosum

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INTRODUCTION: Pyoderma gangrenosum (PG) is a neutrophilic dermatosis that can be associated with irritable bowel disease, arthritis, monoclonal gammopathy, malignancy and myelodysplastic disease. Extracutaneous manifestations are rare and can present in the lung as nodules and cavitary lesions, or as pleural effusions. The main differential diagnoses of cavitary lung disease to consider are granulomatosis with polyangiitis (Wegener’s), lung cancer, and lung abscess.

CASE PRESENTATION: A 46 year old female with past history of PG, endometriosis, and breast adenoma was referred for necrotic lung nodules. She was diagnosed with PG of the right upper extremity 5 years before and treated for years with prednisone and mycophenolate mofetil and on narcotics for pain from PG skin ulcers. Because of an abnormal chest radiograph, a right sided video assisted thorascopic surgery was done. It showed only inflammation and dense adhesions. At the current admission she was febrile, had night sweats and pleuritic chest pain. She recently completed an unsuccessful course of levofloxacin. On exam, she had a 5cm x 5cm right calf hyperpigmented denuded area which did not have fluctuance, erythema, purulence or tenderness. It was consistent with PG. CT chest (below) showed mediastinal and hilar lymphadenopathy with large cavitary lesions bilaterally. Labs revealed a normocytic anemia, mildly positive C-ANCA (1:20) with negative PR-3/MPO antibodies.

DISCUSSION: The large bilateral cavitary lesions in this patient were consistent with a diagnosis of PG. Pulmonary manifestations of PG are rare. Histopathology includes neutrophilic inflammation, fibrosis, granulomas, or necrosis with no evidence of vasculitis. Neutrophilic dysfunction, inflammatory mediators, and genetic predisposition are thought to be involved in the pathogenesis. Treatment has included corticosteriods, azathioprine, cyclosporine and tumor necrosis factor antagonists. Our patient was begun on adalimumab. Recently an imbalance of T-regulatory and Th17 effector cells has been discovered, suggesting that an IL-17 antagonist may be a possible treatment alternative.

CONCLUSIONS: Pulmonary involvement with PG is uncommon and more common causes of pulmonary cavities need to be ruled out before patients start immunomodulating therapies.


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