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Delayed diagnosis of Eosinophilic Granulomatosis Polyangiitis (EGPA) presenting with follicular bronchiolitis and siderosis

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Introduction: This case highlights an unusual case of EGPA presenting with non-classical lung biopsy and initially negative autoimmune screen.

Case: A 48 year old gentleman presented to our institution reporting pleuritic chest pain, shortness of breath and polyarthralgia of three weeks duration. This presentation was on a background history of decreased exercise tolerance for three years. He had been investigated for shortness of breath two years previously and lung biopsy showed follicular bronchiolitis with siderosis. He was then assessed for an underlying rheumatological condition but immunology screen was negative and there were no clinical manifestations disease. On examination he had bilateral knee effusions, synovitis of right wrist, right elbow and bilateral MCP joints. There was a palpable purpuric rash on the distal limbs. CT thorax was suggestive of pneumonitis. CT sinuses reported mucosal thickening with normal biopsy. PFTs showed FEV1/FVC of 37% with low DLCO. Echo showed normal pericardium. He had a positive pANCA titre (1/180) and MPO titre 32(normal <5). PR3 was negative. The remainder of bloods were normal. Diagnosis of EGPA (Churg Strauss) was reached two years after initial investigations.

Discussion: This case highlights the diagnostic difficulties associated with atypical presentations of EGPA. Delayed diagnosis allows increased disease progression and potentially irreversible end organ damage. In this case the diagnosis secured two years after initial presentation. Despite the early immunologic screen being negative and unusual lung biopsy results this patient was subsequently diagnosed with EGPA. This case stresses the importance of pursuing clinical suspicion and repeating the autoimmune screen.

Biography

Sharon Cowley is a graduate of National University of Ireland, Galway, graduating with a 1.1 Honours degree in Medicine in 2013. She has also completed her Irish Clinical Medical Memberships examinations with the MRCPI and is currently on the Basic Specialist Training Scheme for General Internal medicine in Dublin, Ireland.

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