O_10 DOI: https://doi.org/10.5114/reum/219176

Eosinophilic granulomatosis with polyangiitis – one disease with two overlapping mechanisms: a case report

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Key words: eosinophilic granulomatosis with poliangiitis, ANCA antibodies, mepolizumab

Introduction: Eosinophilic granulomatosis with polyangiitis (EPGA) is a rare multisystem inflammatory disease characterised by asthma with eosinophilia, polyneuropathy, and inflammation of small- and medium-sized blood vessels. The EGPA commonly progresses through three phases: allergic, eosinophilic, and vascular. We distinguish two clinical manifestations of Churg-Strauss syndrome: anti-neutrophil cytoplasmic antibodies (ANCA)-positive, typically associated with renal failure and neuropathy and ANCA-negative with heart and lung involvement. ANCA antibodies determined in EGPA can be both c- and p-ANCA. The ANCA-positive phenotype is observed in approximately 30–40% of patients with EGPA. First-line treatment consists of glucocorticosteroids (GCs) nonetheless, in severe cases, immunosuppressants such as cyclophosphamide in combination with immunotherapy targeting the interleukin-5 pathway, like mepolizumab, offer an effective treatment approach.

Case description: A 56-year-old male with a history of asthma presented with chronic cough lasting over 3 months, recent vomiting, peripheral blood hypereosinophilia, macular rash on lower extremities, and renal failure with eGFR 27 ml/min/1,73 m^2, accompanied by proteinuria. An enlarged hilum was found on a chest radiograph. Further diagnostics with computed tomography (CT) confirmed enlarged hilar lymph nodes. Moreover, it revealed subpleural nodules and ground-glass opacities in the apex of the right lung. Biopsy of the lymph nodes disclosed eosinophilic infiltrates. The CT scan of the paranasal sinuses showed thickening of the mucosal membranes. An extensive rheumatological workup was conducted, yielding negative results. Taking all the clinical manifestations into account, the diagnosis of seronegative EGPA was made. Initial treatment with GCs was insufficient. The patient was subsequently treated with cyclophosphamide in combination with mepolizumab, resulting in complete clinical remission.

Conclusions: We chose this case to highlight the overlapping manifestations of ANCA-negative and ANCA-positive EGPA. Although the patient was seronegative, he exhibited symptoms characteristic of both clinical manifestations, including lung and renal involvement. While the mechanisms of organ damage occurring in Churg-Strauss syndrome seem to be well described, this case is a clear example that we should keep looking for the underlying pathophysiological basis for overlapping features between the two clinical manifestations of this rare and heterogeneous disease.