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Difficulties in maintaining treatment of granulomatosis with polyangiitis

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Introduction: Granulomatosis with polyangiitis (GPA) is an autoimmune vasculitis which predominantly affects the respiratory system and kidneys. Immunosuppressive therapy effectively induces remission, but also may cause life-threatening complications. Consequently, maintaining balance between disease control and the risk of treatment-related adverse events is crucial in the therapeutic process.

Case description: In January 2020, a 62-year-old woman was admitted to a local hospital with unintentional weight loss, malaise, ascites and oliguria ongoing for several weeks. Chest high-resolution computed tomography revealed lung infiltrates with cavitary lesions, suggestive of *Staphylococcus aureus* pneumonia (Fig. 1). Unsuccessful treatment with antibiotics, together with oral and labial ulcerations, raised the suspicion of GPA. The diagnosis was confirmed by the presence of elevated levels of proteinase 3 antineutrophil cytoplasmic autoantibody (PR3-ANCA). Progressive renal failure required hemodialysis (HD) via a tunneled catheter. Plasmapheresis, intravenous prednisolone, and cyclophosphamide were initiated.

In June 2023, the patient was readmitted with suspected pneumonia and radiological progression of pulmonary lesions. Bronchioalveolar lavage cultures yielded *Acinetobacter baumannii* and *Candida krusei*, prompting initiation of targeted therapy with amikacin, colistin, and micafungin. Echocardiography revealed a thrombus in the right atrium and intravenous catheter distal tip thickening. Although positron emission tomography suggested fungal endocarditis, repeated blood and catheter cultures were negative. However, continuation of antifungal therapy resulted in clinical improvement. After creating an arteriovascular shunt on the right arm, HD was continued. Persistent pulmonary progression, elevated PR3-ANCA levels and hearing loss prompted qualification for rituximab.

In September 2023, the patient developed right arm cellulitis due to the right subclavian vein thrombosis, which was treated with piperacillin-tazobactam. Endovascular embolisation of the lateral rami of the cephalic vein contributed to the reduction of cellulitis. After subsidence of inflammation, rituximab treatment was initiated in December 2023.

Conclusions: The underlying disease and its treatment can cause severe infections and thrombotic events, contributing to the complexity of GPA management. This case shows that systematic reassessment and individualised therapy are essential to maintaining the effectiveness of the treatment, along with minimising treatment-related morbidity.

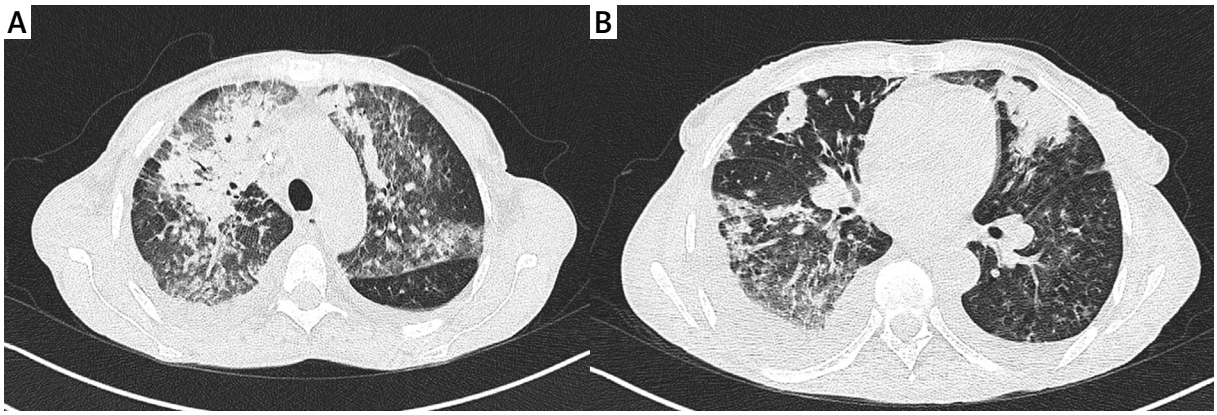

Fig. 1. High-resolution computed tomography scans of pulmonary lesions at the time of diagnosis in Ferbuary 2020.