Abstract:

Granulomatosis with polyangiitis (GPA), previously known as Wegener's granulomatosis, is a rare autoimmune disease characterized by systemic vasculitis, primarily affecting small- to medium-sized blood vessels. GPA typically involves the upper and lower respiratory tracts and kidneys, with a wide array of clinical features that can confound physicians during the diagnostic process [1].

We report an intriguing case of a 59-year-old female with a history of type 2 diabetes and a recent nasal septal abscess who presented with a 3-month history of intermittent fever, weight loss, and malaise. High inflammatory markers, negative cultures, and various negative serological tests led to an extensive evaluation.

The diagnosis of GPA was confirmed based on the patient's history of nasal septal abscess, rapid renal decline, elevated inflammatory markers and positive c-ANCA. Renal biopsy revealed rapidly progressive glomerular nephritis, which is a characteristic feature of GPA.

The patient was initiated on high-dose IV methylprednisolone as an initial treatment for GPA, with plans for Rituximab therapy, which was temporarily delayed due to a positive COVID-19 diagnosis. The timing of Rituximab administration should be tailored to the patient's clinical status and infection control.

This case emphasizes the significance of early recognition and appropriate management of GPA, as delayed diagnosis can lead to disease progression and severe complications. And through this case report, we aim to shed light on the intricate nature of diagnosing GPA, especially when it presents as a fever of unknown origin.