**Abstract:** Granulomatosis with polyangitis (GPA) or Wegener's granulomatosis is an anti-neutrophil cytoplasmic antibody (ANCA) associated small vessel vasculitis. It is characterized by granulomatous inflammation of the respiratory tract and necrotizing glomerulonephritis. Clinical presentation is variable according to the underlying organ involvement. Simultaneous involvement of lower respiratory tract with associated diffuse alveolar hemorrhage and rapidly progressive glomerulonephritis leads to pulmonary renal syndrome. As pulmonary renal syndrome always imply a life threatening condition, rapid evaluation of etiology and appropriate treatment is essential to prevent significant mortality and morbidity. However, the etiology is diverse, which makes the diagnosis is challenging in a resource poor setting.

Here we report a rare case of GPA presenting with acute febrile illness, arthralgia, myalgia and red eye mimicking leptospirosis. A 39-year-old male with recent history of sinusitis and right sided nasal polypectomy was presented with acute febrile illness of three days duration with associated dry cough, arthralgia, myalgia and red eye. His symptoms deteriorated and sudden onset shortness of breath developed with radiological evidence of diffuse pulmonary hemorrhage. Later he developed rapidly progressive glomerulonephritis as evidenced by reduced urine output, glomerular hematuria and rapidly rising serum creatinine. The possibility of systemic vasculitis should be considered in patients presenting with acute febrile illness with pulmonary- renal syndrome even though infection like Leptospirosis being the commonest culprit.

While GPA is primarily associated with c-ANCA in 65-75% of cases, rest of the patients with clinical GPA have the alternative ANCA. If untreated often it has a rapidly progressive and fatal disease course.