An unusual presentation of Wegener's granulomatosis with Splenic infarct

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INTRODUCTION

Wegener's Granulomatosis is a rare multisystem disorder causing necrotizing granulomatous inflammation and pauci immune vasculitis in small and medium sized blood vessels.The prevalence of granulomatosis with polyangiitis (GPA) ranges from 2.3 to 146.0 cases per million persons, with an incidence of 0.4 to 11.9 cases per million person-years.[1]The most common site of involvement are upper and lower respiratory tract, Kidney. Splenic infarct is rarely reported in patients with Wegener's granulomatosis.

CASE REPORT

A 21-year-old male, driver by occupation presented with complaints of breathlessness, cough with blood stained sputum and fever for 15 days. He is non-smoker and non-alcoholic with no comorbidities.On examination his vitals were stable and room air saturation was 95%. Routine blood investigation were done which showed neutrophil predominant leucocytosis . In view of fever Blood and Urine culture were done, non-fermenting gram negative bacilli growth was noted in blood for which he was started on antibiotics as per the culture sensitivity report. Chest radiography showed left upper zone homogenous opacity (fig1), further evaluated with Computed Tomography thorax- bilateral multiple nodular opacities and few of them are cavitating nodules with diffuse ground glass opacities (fig 2). During his course of stay in the hospital he developed abdominal pain, more in the left upper quadrant for which he was evaluated with bedside Ultrasonogram of the abdomen which showedperisplenic collection.General Surgeon consultation was sought and advised contrast enhanced CT abdomen which showed non-enhancing spleen suggestive of splenic infarct (fig 3). Echocardiography was suggestive of minimal pericardial effusion. Otolaryngology and ophthalmologic evaluation were normal. Diagnostic bronchoscopy showed pale, unhealth mucosa with bilateral upper lobe bronchial narrowing. Bronchoalveolar lavage (BAL) andTransbronchial biopsy was performed from the left upper lobe segmental bronchi.BAL fluid Cytology showed features of acute inflammation with microbiology profile negative . Transbronchial lungbiopsy was suggestive of granulomatous inflammation. He was suspected to have Wegener's Granulomatosis, further evaluated with ANCA profile. C -ANCA was strongly positive (345.01 RU/mL).

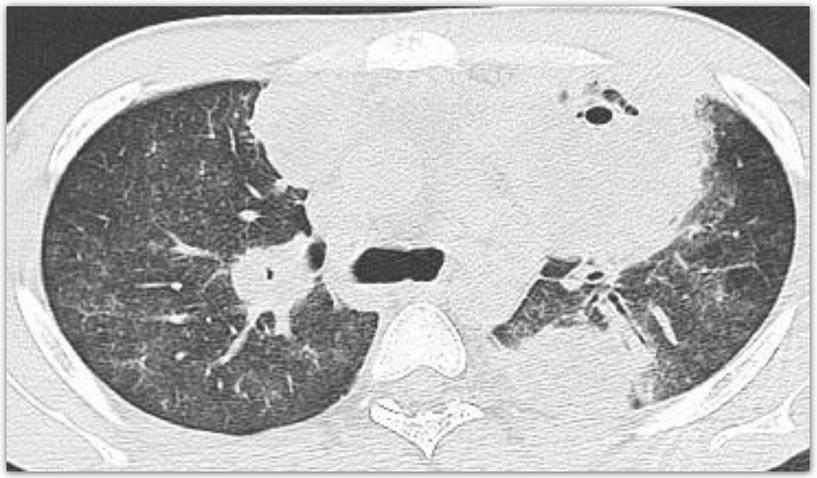
Finally he was diagnosed with Wegener's Granulomatosis with Splenic infarct .Methylprednisolone pulse therapy was given following which patient showed dramatic improvement.Patient was discharged on maintenance therapy with immunomodulators and Co-trimoxazole prophylaxis.

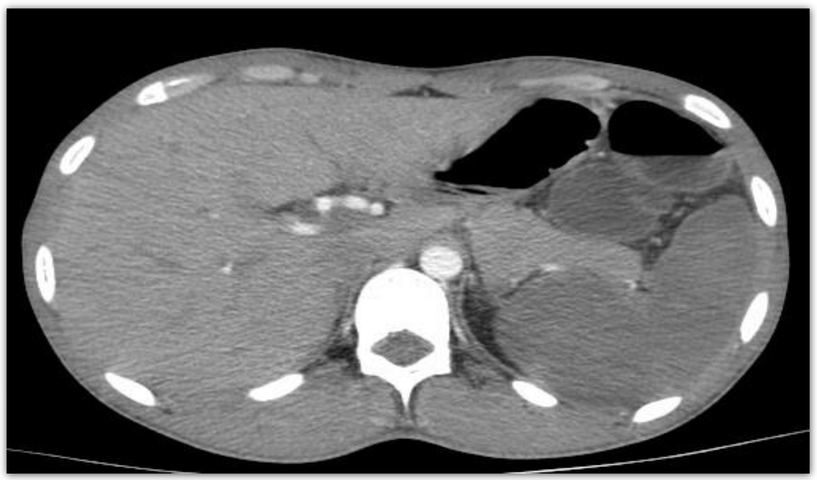
Discussion

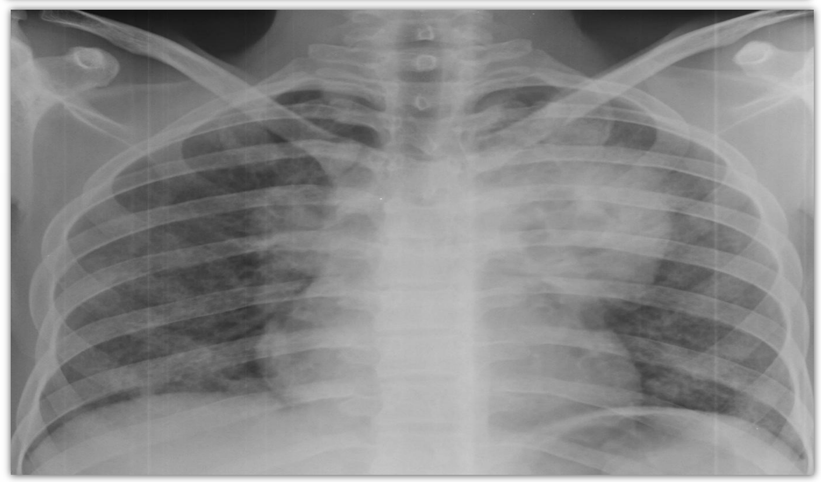
Wegener's Granulomatosis is a systemic disease with variable presentation.Patients with splenic infarcts typically present with left upper quadrant pain or left shoulder tip pain however up to 50% may be asymptomatic.[2] Haemorrhagic and infective involvements are usually symptomatic, while infarcts are asymptomatic, which may result in an underestimate of prevalence of splenic involvement in WG.[3] Splenic infarcts in WG are usual noted in young patients with ENT and Eye involvement. The involvement of spleen without otorhinolaryngology and Eye involvement makes this case unusual.Splenic lesions may lead to complications like splenic abscess, hemorrhage hence a high index of suspicion should be maintained for splenic infarct in patients with presence of abdominal symptoms. In our case patient was treated conservatively for splenic infarct as he did not develop any complications. Patient improved symptomatically after initiation of treatment with steroids.

CONCLUSION

Wegener's Granulomatosis is a systemic disease that can lead to multi organ failure when remain untreated. We encountered a rare unusual presentation of Wegener's granulomatosis with splenic infarct in the absence of renal otorhinolaryngology and eye involvement.







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Conflict of interest : NIL