DIFFUSE ALVEOLAR HAEMORRHAGE- A RARE CASE OF p-ANCA POSITIVE VASCULITIS
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Abstract: Diffuse alveolar haemorrhage (DAH) is defined as haemorrhage into the pulmonary microvasculature rather than parenchyma or airways with a catastrophe of events leading to haemoptysis, falling haematocrit, hypoxemia, respiratory failure and diffuse pulmonary infiltrates (1). It should be considered as a medical emergency because of high morbidity and mortality. Annual incidence is up to 2 to 4 cases per million population. Pulmonary renal syndromes, connective tissue disorders and drugs make up most of the cases of DAH. Histologically DAH is recognized with pulmonary capillaritis. (2) Most common autoimmune causes include goodpasture syndrome, small vessel vasculitis and SLE. The non-immune causes of DAH are tumors, coagulopathy, and pulmonary thromboembolism. The mean age of presentation of small vessel vasculitis is 40 to 50 years with equal male female preponderance. This case report identifies diffuse alveolar hemorrhage as a presentation of pulmonary renal syndrome in a young female presenting with haemoptysis. However one third of diffuse alveolar hemorrhage cases may not have haemoptysis (3). HRCT findings can vary with time of onset of hemorrhage and the clinical context is crucial in imaging interpretation. In acute stage can range from ground glass opacities to consolidation due to alveolar filling with blood and in subacute stage within 2-3 days, interlobular lines interlobular septal thickening and crazypaving pattern can be seen. Chronic stage shows ill defined centrilobular nodules reflecting intra alveolar accumulation of pulmonary macrophages.

Keyword: Diffuse Alveolar Haemorrhage, p-ANCA, Vasculitis, Pulmonary Renal Syndrome

CASE REPORT
A 15 year old female presented with complaints of haemoptysis, fever, vomiting, puffiness of face, pedal edema for one week duration. On examination the patient was anemic, tachypneic, nasal and oropharyngeal examination was normal, and pulmonary examination showed bilateral crackles. Investigations showed complete blood count 6,200, hemoglobin 7.9gm/dl, ESR ½ hr 14mm,1 hr 20 mm, blood urea 68 mg/dl, serum creatinine 3.2mg/dl. Urinalysis showed proetinuria 3.5gm. Ultrasound abdomen revealed hepatosplenomegaly of bilateral corticomedullary region of both kidneys. Chest radiograph PA view showed heterogeneous alveolar opacities in the right side upper middle lower zones and left side lower zones. HRCT chest showed diffuse consolidation with prominent segmental and sub segmental bronchi, called as dark bronchus sign. This is observed in acute phase of diffuse alveolar hemorrhage. BAL study showed haemosiderin laden macrophages. The renal biopsy was done which showed global glomerulosclerosis with interstitial fibrosis and tubular atrophy. Serum immunological assay for Anti-neutrophilic cytoplasmic antibody showed p-ANCA positivity. Anti-GBM antibodies and anti neutrophilic antibodies were negative. The diagnosis of pulmonary renal syndrome with p-ANCA positive small vessel vasculitis was made. Treatment was given with three cycles of plasmapheresis. Immunosuppressant therapy with cyclophosphamide was initiated following injection methylprednisolone for three days. Patient improved symptomatically with decrease in the blood urea creatinine. She is advised with salt restriction and fluid restriction along with immunosuppressants.

Fig 1 showing Rt UZ/MZ/LZ and Lt Mz opacities.
Fig 2 computed tomography showing consolidation with dark bronchus sign this is seen in acute stage of diffuse alveolar haemorrhage.

DISCUSSION
The above case presented clinically as diffuse alveolar hemorrhage due to pulmonary renal syndrome. The clinical features of pulmonary renal syndromes overlap. Radiological features are infiltrative opacification in chest radiograph. HRCT chest shows features of diffuse pulmonary haemorrhage when bleeding is the alveolar spaces can be varying from ground glass opacities, in acute phase to interlobar septal thickening to crazy paving pattern in 2-3 days. Fig 2 computed tomography showing consolidation with dark bronchus sign this is seen in acute stage of diffuse alveolar haemorrhage.

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features are infiltrative opacification in chest radiograph. HRCT chest shows features of diffuse pulmonary haemorrhage when bleeding is the alveolar spaces can be varying from ground glass opacities, in acute phase to interlobar septal thickening to crazy paving pattern in 2-3 days. In chronic conditions centrilobular nodules seen. There is an increase in diffusion capacity during alveolar haemorrhage thought to be explained by the increased availability of intra alveolar haemoglobin to combine with carbon monoxide.(4) Most of them are ill and cannot undergo pulmonary function testing and so are not much helpful. Complications include organising pneumonia, collagen deposition in small airways, and pulmonary fibrosis. The 10 year survival is upto 70 to 80%.The role of open lung biopsy is generally not helpful in determining the underlying etiology of DAH. Cytoplasmic antibodies provide key to the diagnosis. Histopathological features are shared by Wegener's granulomatosis(WG), Microscopic polyangitis(MPA), churg-strauss syndrome and other causes. Churg-strauss described the syndrome where granulomatous inflammation, necrotizing systemic vasculitis, and necrotizing glomerulonephritis are associated with raised eosinophils.(5) Wegener’s granulomatosis shows predominant c-ANCA positivity with predominant upper respiratory tract granulomas, whereas M PA and churg-strauss syndrome show p-ANCA positivity.(6,7)

1. Complications include organizing pneumonia, collagen deposition in small airways and pulmonary fibrosis. Diffuse alveolar haemorrhage (DAH) occurs as a consequence of pulmonary capillaritis in the ANCA-associated vasculitides, and is an important cause of morbidity and mortality in this condition [8]. The incidence of DAH is 7–45% in Wegener's granulomatosis [9,10] and 10–30% in microscopic polyangiitis [11]. It is rare in Churg–Strauss syndrome , but almost invariable in isolated pauci-immune pulmonary capillaritis. The acute mortality associated with DAH and underlying vasculitis is approximately 60%, six times greater than vasculitis without pulmonary haemorrhage .

CONCLUSION
Diffuse alveolar haemorrhage syndromes are considered as an emergency. The mean age of presentation of small vessel vasculitis is 40 to 50 years with equal male female preponderance. The above case report identifies Diffuse alveolar hemorrhage as a presentation of pulmonary renal syndrome in young female presenting with haemoptysis which was proved to be p-ANCA positive vasculitis. Complications include an organising pneumonia, collagen deposition in small airways and pulmonary fibrosis. Treatment varies according to the underlying cause. Adequate suspicion and prompt diagnosis by radiological and immunological assay help to prevent the morbidity and mortality in such rare diseases with common presentation of haemoptysis.

REFERENCES