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A CASE OF GRANULOMATOSIS WITH POLYANGITIS (GPA) COMPLICATED WITH CEREBROVASCULAR EVENT (ISCHAEMIC WITH HAEMORRHAGIC CHANGES).

Respiratory Medicine

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ABSTRACT

AntiNeutrophilic cytoplasmic antibody(ANCA) associated vasculitis predominantly includes granulomatosis with polyangitis (GPA), eosinophilic granulomatosis with polyangitis(EGPA) or churg strauss syndrome, microscopic polyangitis(MPA).GPA being the most common vasculitis among these mainly involves small vessel and veins . It has associations with both thrombotic and haemorrhagic events which might involve organs like upper airway, lungs, kidney, heart, nervous system.We are presenting here a case of granulomatosis with polyangitis complicated with cerebrovascular event (ischaemic with haemorrhagic changes).

KEYWORDS

GPA, Wegeners granulomatosis, ANCA

Case Report:

A 65 year old female patient with no comorbidities presented with complaints of breathlessness , hemoptysis , fever , dry cough , oral ulceration for 5-6 days with history of chronic sinusitis .Chest radiography taken revealed right side middle and lower zone alveolar infiltrates with costophrenic angle blunting suggestive of pleural effusion(FIG 1) . On routine blood investigation she was found to be anaemic and her serum creatinine was found to be on the higher side (s. creat=5.8) with neither compliants of reduced urine output nor any uraemic symptoms. USG chest was done which showed right sided mild pleural effusion with underlying consolidation. Urine routine suggests red cell cast with no protineuria. Considering clinical history and radiological findings ANCA was sent which turned out to be positive for P-ANCA and nephrologist opinion was taken who advised for haemodialysis due to continuous increase in serum creatinine from 5.8 to 9.3 with preserved urine output. Patient was then treated with blood transfusion, immunosuppressant (Inj.Cyclophosphamide) and glucocorticoids. Consecutively during hospitalization patient developed left sided hemiplegia with slurring of speech ensuing that an MRI brain was done which was suggestive of acute infarct at right basal ganglia capsular regions with midline shift along with multiple loci of GRE blooming noted in bilateral cerebral hemispheres – microhaemorrhages more likely(Fig 2). Unfortunately all management had poor outcome due to severity of the illness and biopsy was not possible since the patients general condition was poor.

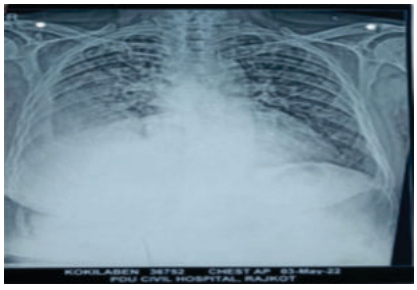

Figure 1: Chest radiography suggestive of right sided midzone and lowerzone alveolar infiltrates with ipsilateral costophrenic blunting suggesting pleural effusion

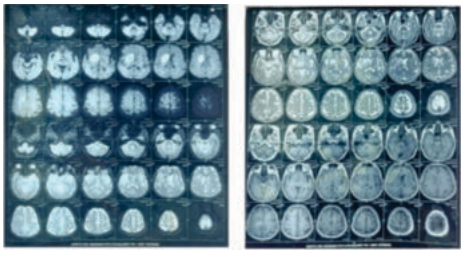

Figure 2: MRI brain suggestive of acute infarct at right basal ganglia capsular regions with midline shift along with multiple loci of GRE blooming noted in bilateral cerebral hemispheres – microhaemorrhages more likely.

DISCUSSION:

GPA is said to be necrotizing granulomatous vasculitis of small sized and medium sized vessels, the most common of the ANCA associated vasculitis which involves upper airways and lower airways predominantly followed by kidneys. In around 50% patients nervous system is involved. The incidence is found to be 3/100000 (1 lakh) with generally occurring at an age of 40-65 years. Other systems involved are found to be heart and eyes. The common presenting compliants of GPA with lung involvement will be cough , dyspnea , chestpain, haemoptysis due to diffuse alveolar haemorrhage. Glomerulonephritis is the most common manifestation of renal involvement of GPA. Interestingly in GPA patient might end up or present with renal failure(end stage) with absence of any symptoms which happened to our case as well.

Usually GPA is associated with positive C-ANCA in about 80-95% but it might be associated with positive P-ANCA in about 5-10% in rare case . There have been many previous case reports in the past where GPA is associated with venous thrombosis and DVT but very few are present in view of GPA complicated with arterial thrombosis . Damage to the arterial wall in GPA can lead to intracranial aneurysmal formation and haemorrhage or vascular occlusion due to inflammatory injury to the endothelium. ANCA is been primarily postulated for the endothelial cell damage by direct cytotoxicity or localized immune complex formation with target antigens bound to the endothelial cell surface. Rodent models of MPO –ANCA associated vasulitis support this hypothesis where ANCA had close association with development of vasculitis and glomerulonephritis. Cerebral vessel occlusion by granulomata had also been reported in the past.

MRI is the key diagnostic tool for CVA where one may find multiple infarcts with or without haemorrhages present in more than one vascular territory with is suggestive of underlying inflammatory process which is true in our case where multiple microhaemorrhages are seen in bilateral cerebral hemispheres.

Finally in view of positive P ANCA which usually positive for Microscopic polyangitis , it is our responsibility to differentiate it from GPA which is supossed to be our case report. With concerned to GPA the positive findings were involvement of upper and lower respiratory tract along with pleural effusion and lead to end stage renal disease with no symptoms requiring haemodialysis complicating with CVA with both ischaemic and haemorrhagic events . Negative points taking into consideration of this case as MPA are presence of pleural effusion which is rare but might occur and involvement of peripheral neuropathy(mononeuritis multiplex) being most common in case of MPA than central nervous system and renaldisease will be precede months before lung involvement .

Yet final diagnosis is to be confirmed histopathologically following a biopsy of kidney or lungs where granulomas will be found In case of GPA and is absent in MPA.

Treatment for both GPA and MPA is found to be same, immunosuppressant preferably cyclophosphamide or rituximab with glucocorticoids and correction of anemia wherever necessary and dialysis in case of end stage renal disease .One RCT have shown that IV pulse therapy of cyclophosphamide consisting of three pulses of 15 mg/kg given 2 weeks apart followed by 15 mg/kg given every three weeks for 6 months is effective in inducing remission . Rituximab is preferred in case of young patients in whom fertility to be preserved . In patients with fulminant disease IV methylprednisoslone 1000mg per day for 3-5 days is to be administered to effectively control inflammation. Plasmapheresis stand out to be a promising treatment modality.

CONCLUSION:

Even when patient doesn't present with DAH and has alveolar infiltrates on chest radiography and with elevated creatinine levels with no renal symptoms all patients should be evaluated for pulmonary vasculitis where prompt administration of immunosuppressant and glucocorticoids might prevent further inflammation and complicating events like CVA or prevents involvement of other systems. Studies suggested that with recent treatment the 5 year survival rate have increased drastically.

Conflict of Interests: Nil.

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