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A RARE CASE OF DOUBLE POSITIVE RAPIDLY PROGRESSIVE GLOMERULONEPRITIS

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Abstract :

Granulomatous polyangitis is a small vessel vasculitis of elderly age group. C-ANCA being a marker for wegeners granulomatosis, co existence of c-ANCA and anti - GBM antibody positivity is a rare event. In any given individual co existence of c-ANCA and anti GBM indicates the severity of disease and predicts the worst renal outcome.

Keyword :WEGENERS GRANULOMATOSIS, c-ANCA, anti GBM, RPGN, ESRD

Introduction:

Patients presenting with haemoptysis or pulmonary inltrates together with glomerulonephritis, particularly the rapidly progressive cases, are considered to be cases of pulmonary renal syndrome. In many such patients may initially be investigated extensively for malignancy or therapy-resistant infectious disease. The relative degrees of respiratory tract and renal involvement vary, and in some patients the initial symptoms may be conned to one or the other of these organ systems. Pulmonary renal syndrome is encountered in several diseases such as Goodpasture's syndrome, antineutrophil cytoplasmic antibody (ANCA)-associated systemic vasculitis (Wegener's granulomatosis, microscopic polyangiitis) systemic lupus erythematosus , and infection or drug-induced glomerulonephritis.To preserve both renal and respiratory function, it is of vital importance to institute therapy early, in the acute phase.

Case Report

A young 17 year female came to emergency department with complaints of cough with expectoration of 1 month duration with 2 episodes of hemoptysis, worsening breathlessness for 1 week now increased since 1 day, no h/o orthopnea and PND. H/o of on and off fever ,no chest pain or upper abdominal pain . Patient was treated for the same in a private hospital and was treated with ATT for the last one month for sputum negative pulmonary TB.

On admission she was drowsy , dyspnoic , tachypnoic , her tempareture was 98.8 F, PR = 130/min , BP= 80/60 , RR= 56/min . On examination there was pallor present , no clubbing, no cyanosis, no lymphadenopathy and no pedal edema.

Respiratory system –NVBS, bilateral coarse crepitations (R>L).
Cardiovascular system – normal S1S2 heard ; no murmurs.
GI system – soft, nontendor, no palpable organomegaly, no free fluid.
Central nervous system - no focal neurological defecit .

Investigations

CBC - TC- 7200 /cu.mm, DC-N64 , L33 , E3 , HB% 7, Plat count- 165000/cu.mm , ESR- 112mm
Urine Routine **-** Alb - 3+, Rbc 8-10 Cells, Rbc casts +
Urine spot PCR – Prot 515, Creat-233
RFT – RBS 106mg/dl, B.urea-65mg/dl, S.creat- 3.8mg/dl
ABG – metabolic acidosis with respiratory alkalosis.
LFT – TSB- 0.9, SGOT- 241IU/L, SGPT - 401IU/L, SAP- 970.
Sputum AFB – negative.
Urine culture and blood culture – negative
CHEST X- RAY – Bilateral fluffy opacities (fig no 1)

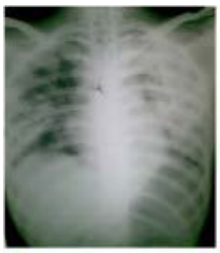

Figure 1 showing Bilateral Fluffy opacities

CT SCAN THORAX – bilateral parenchymal cavities were seen. (Fig no 2)

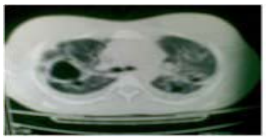

Figure 2 showing bilateral parenchymal cavities

With the above clinical history and investigations revealing raised renal parameters and presence of urinary RBC casts and glomerular protienuria , we worked out a provisional diagnosis as PULMONARY RENAL SYNDROME. Since the closer differential diagnosis were vasculitis ,
connective tissue diseases we further performed battery of investigations to come to a diagnosis.
Serological markers.
ANA – Negative , dsDNA – Negative.
C3 – Low , C4 – Normal .
C-ANCA – Positive , P-ANCA – Negative , anti GBM – Positive.
RENAL BIOPSY – Cresentric glomerulonephritis and immunoflorescence demonstrated linear
Deposits

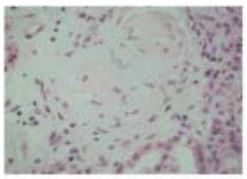

Figure 3 showing sclerosed glomerulous with crescents (arrow)

TREATMENT GIVEN.

Initially patient was symptomatically treated with iv antibiotics, iv fluids inotropic support. Later patient was given immediate hemodialysis for acute renal failure. Tachypnoea , blood pressure and general condition improved. Later once serological marker became positive patient was started on pulse methyl prednisolone for 3 days , cyclophosphamide 750 mg pulse , 7 cycles of plasmapheresis , followed by oral steroids. Due to failure in satisfactory improvement in blood urea and sr.creatinine patient was given maintenance hemodialysis . Inspite of effective renal support given patient remained dialysis dependent and reached END STAGE RENAL DISEASE within 3 months . Recently patient underwent renal transplant and doing well till date.

Discussion

WG was first described by Klinger in 1933, followed by other investigators, including Rossle in 1933,

Wegener in 1936 and 1939, and Ringertz in 1947.

WG is currently characterized as one of the ANCA-associated small vessel vasculitides. It is distinguished clinically by its predilection for affecting the upper and lower respiratory tracts and kidneys and by the histologic presence of necrosis, granulomatous inflammation, and vasculitis. There is a strong and specific association with autoantibodies directed against proteinase 3, a constituent of neutrophil azurophilic granules.The presence of such antibodies is a strong indicator for a diagnosis of WG,(10) but should not be used in place of a tissue diagnosis.

Unexplained constitutional symptoms like fever,weight loss, fatigue(8). The upper airway disease is the most common presenting feature includes pleuritis are the most common pulmonary symptoms sinusitis, oral lesions , otitis media, hearing loss, epistaxis, and saddle nose deformity.

Pulmonary involvement is one of the cardinal features of WG.Cough, hemoptysis, and with X-RAY showing pulmonary infiltrate and nodules.(7) Renal disease can be initial presentation or during the course of the disease. Renal disease may vary from asymptomatic and mild to fulminant glomerulonephritis within days or weeks, resulting in endstage renal failure. Even with appropriate therapy, it may lead to chronic renal insufficiency and renal failure. (6,7)

There have been reports of c- ANCA and anti-GBM positivity and we have found one. Reported cases showed aggressive renal disease in terms of both recovery of renal function and mortality(1,2,3,4,7,9) IT has been suggested anca plays a pathogenic role causing damage which exposes the glomerular basement membrane and its cryptic epitope.(3)

Conclusion

Patients with either ANCA related vasculitis or anti GBM disease diagnosed by histopathology or by immunological marker study should be checked for the presence of second antibody. Thisapproach should throw light on better man agement.

List of abbreviations used.

Anca – antineutrophil cytoplasmic antibody.
Anti GBM – glomerular basement membrane antibody.
RPGN – rapidly progressive glomerulonephritis.
ESRD – end stage renal disease.

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