Pulmonary-renal syndrome: Dx

Definition: Pulmonary-renal syndromes or lung-kidney syndromes are clinical syndromes defined by a combination of diffuse alveolar hemorrhage (DAH) and glomerulonephritis. Pulmonary-renal syndromes are not a single entity, but are caused by a wide variety of diseases, including various forms of primary systemic vasculitis (especially Wegener's granulomatosis and microscopic polyangiitis), Goodpasture's syndrome (associated with autoantibodies to the alveolar and glomerular basement membrane) and systemic lupus erythematosus.

Diagnosis: rests on the identification of particular patterns of clinical, radiologic, pathologic and laboratory features. Serologic testing is important in the diagnostic work-up of patients presenting with a pulmonary-renal syndrome. The majority of cases of pulmonary-renal syndrome are associated with ANCAs, either c-ANCA or p-ANCA, due to autoantibodies against the target antigens proteinase-3 and myeloperoxidase respectively. The antigen target in Goodpasture's syndrome is type IV collagen, the major component of basement membranes. Diffuse alveolar haemorrhage is characterized by the presence of a haemorrhagic bronchoalveolar lavage (BAL) in serial BAL samples. In the clinical setting of an acute nephritis syndrome, percutaneous renal biopsy is commonly performed for histopathology and immunofluorescence studies.

Rx: corticosteroids and immunosuppressive agents such as cyclophosphamide (as induction therapy) or azathioprine (as maintenance therapy once remission has been achieved). Recent evidence suggests that patients with severe ANCA-associated vasculitis, defined by the presence of diffuse alveolar hemorrhage and/or severe renal involvement (creatinine concentration > 5.7 mg/dl), might benefit from plasma exchange in combination with cyclophosphamide and corticosteroids.

Question:

A 73 year-old female is admitted with acute desaturation, diffuse alveolar hemorrhage and acute kindey injury (AKI). Her creatinine is 7 mg/dL. The best approach for treatment is to:

a. Perform dialysis
b. Plasma exchange in combination with steroids
c. Broad spectrum antibiotics
d. Avoid doing lung or renal biopsies

Answer: B - Recent evidence suggests that patients with severe ANCA-associated vasculitis might benefit from plasma exchange in combination with cyclophosphamide and corticosteroids