中文題目:誰是抗嗜中性球細胞質抗體-血管炎造成的肺腎症候群?

英文題目:Who has ANCA-vasculitis related pulmonary-renal syndrome?

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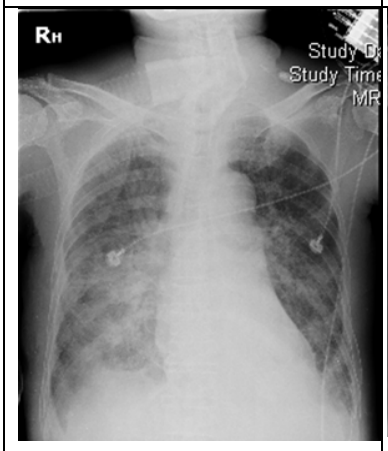
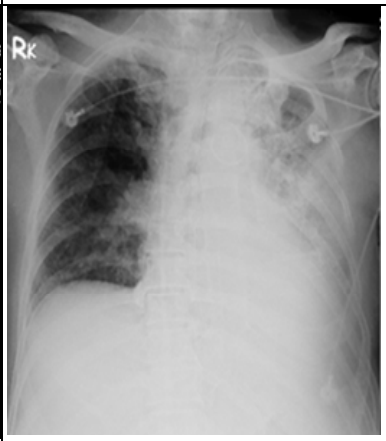
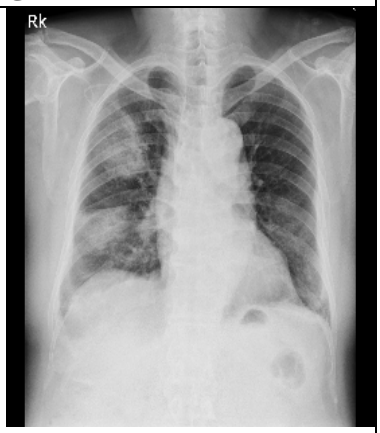
Background: The pulmonary-renal syndrome is defined as a combination of diffuse alveolar hemorrhage and rapidly progressive glomerulonephritis. Up to 60% of these cases are related to antineutrophil cytoplasmic autoantibodies (ANCA)-associated vasculitis, while 20% are caused by anti-GBM disease. The major target antigens of ANCA associated vasculitis are myeloperoxidase (MPO) and proteinase 3 (PR3). MPO-ANCA is related to microscopic polyangiitis, and PR3-ANCA is the marker antibody in Wegerner's granulomatosis.
This time , we will present 3 old male patients with hemoptysis and renal failure, but only one is caused by ANCA- related pulmonary-renal syndrome. Who is he ?

Case reports:

Case A: A 74-year-old male patient with myelodysplastic syndrome presented with hemoptysis and intermittent gross hematuria for 10 more days. His renal function was fair before this admission .Physical examination showed facial edema with poor response to diuretics, but his four limbs were free of edema . No skin rash appeared over his body. His routine blood examination showed a white cell count of 5.94 x10 3 /uL, hemoglobin 7.7 g/dl, ,platelet 14 x10 3/ul, Creatinine:3.0 mg/dl.

Case B: A 86-year-old male patient who has a history of chronic obstructive pulmonary disease and hypertension was admitted due to acute renal failure and urinary tract infection. However, sudden onset of hemoptysis combined with hypoxic respiratory failure developed during hospitalization. His routine blood examination showed a white cell count of 9.34 x10 3 /uL, hemoglobin 10.3 g/dl, ,platelet 89 x10 3/ul, Creatinine:3.8 mg/dl.

Case C: A 69-year-old male patient complained that hemoptysis developed for 1 month. Allergic bronchitis was suspected by ENT but not responsive to the medication. He was then admitted since chest X ray showed 2 consolidations over right lung. Laboratory data showed WBC:5.64K/uL, Hb:9.7g/dl, PLT:176k/ul, Cre:1.2 mg/dl.

A	B	C
		

Result:

Case A: This 74-year-old man had myelodysplasia syndrome related thrombocytopenia induced pulmonary hemorrhage.His poor renal function was related to sepsis and dehydration induced prerenal failure. So he was not ANCA vasculitis related pulmonary renal syndrome.

Case B: While following up the laboratory data, his Hb decreased form 10.3 g/dl to 6.4 g/dl. No immune profile such as ANA,dsDNA,ANCA,anti-GBM ,C3 and C4 abnormality was noted. Bronchoscopy and chest computerized tomography was arranged for case B. From the chest image, bronchiectasis induced hemoptysis,and hemorrhagic hypovolemia induced acute renal ient fialure was suspected. Though case B had hemoptysis related hypoxic respiratory failure and acute renal failure,he was not ANCA vasculitis related pulmonary-renal syndrome.

Case C: After the ptient admitted, his hemoptysis progressed and oxygen demand increasing form nasal cannular 3L/h to 6L/h. A chest computerized tomography scan showed right lower lung consolidation patches.Microscopic hematuria was noted,multiple RBC casts was revealed on microscope. Laboratory data showed ANA=1:2560,dsDNA:negative, ANCA:P-ANCA positive,anti-GBM:negative ,C3 and C4 under normal range.Renal biopsy revealed focal segmental necrosis,and pauci-immune glomerulonephritis.After steroid pulse therapy (solumedrol 1g qd x 3 days),his hemoptysis and microscopic hematuria improved.The patient was p-ANCA vasculitis related pulmonary renal syndrome.

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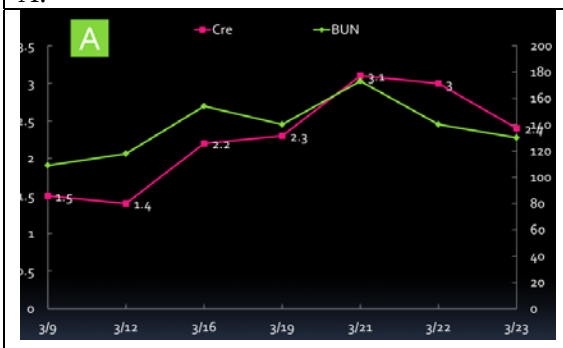
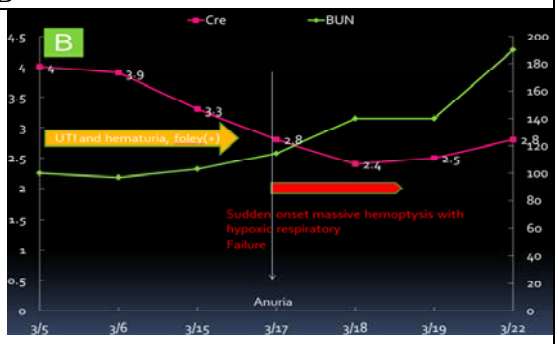
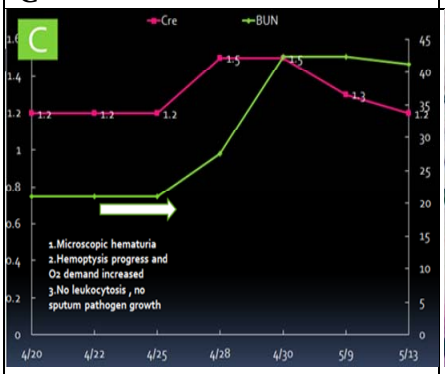
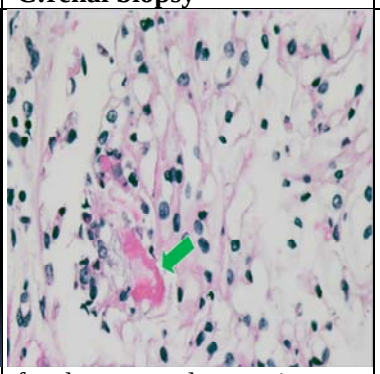
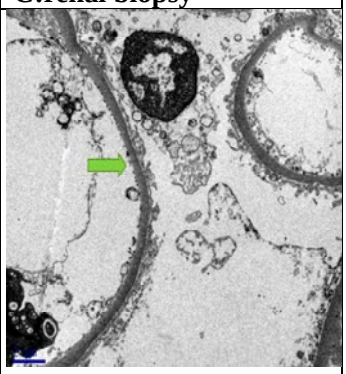
	

C	C:renal biopsy	C:renal biopsy
	 focal segmental necrosis	

Discussion :

Pulmonary-renal syndromes is a combination of diffuse alveolar hemorrhage and glomerulonephritis.However,in clinicle performances,we see many patients with hemoptysis and renal failure,but they are pulmonary-renal syndromes mimics which can cause by severe congestive heart failure (pulmonary edema & prerenal azotemia),sepsis shock,Legionnaire's disease,hantavirus pulmonary syndrome. How should a patient be evaluated when we

suspected him or her have pulmonary-renal syndromes?We provide five steps.

Step 1- clinical presentation. Pulmonary-renal syndromes is male predominance and the age of onset is between 50 to 60.Patients with ANCA vasculiitis can present with different symptoms such as a flu-like illness,arthralgias,skin rash. 40~ 60% of cases may have 3H, hemoptysis,hypoxia,a drop in hemoglobin level.

Step 2- images.Chest x ray may show bilateral consolidation patches in ANCA vasculitis,but it was not spicific. Computerized tomography may showes trees in bud sign,ground glass and crazy paving signs.

Step 3- DLCO test. The DLCO in diffused alveolar hemorrhage increases >130% predicted or increases 30% above baseline. In contrast, DLCO decreases in pneumonitis.

Step 4- Laboratory data. Urine analysis will show RBC casts or dysmorphic RBC. Immune profile such as ANA,anti-dsDNA,Complement,rheumatoid factor ,ANCA,ESR,CRP provide a good tool for diagnosing ANCA vasculitis.

Step 5- Biopsy.Lung biopsy would show neutrophilic infiltration over alveolar, karyorrhexis or extravascular granuloma. Renal biopsy can see focal segmental necrosis, crescentic glomerulonephritis ,fibrinoid necrosis,and pauci immune glomerulonephritis.