中文題目: 血管炎導致多重器官出血

英文題目: Vasculitis related multiple organ hemorrhage

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Background : In clinical practice, vasculitis has heterogenic presentation in patients. Vasculitis involves different size of vessel characterize with inflammation and necrosis. For ambiguous presentation , doctor should always keep vasculitis as different diagnosis.

Abstract : A 53 year old woman lived in U.S.A was diagnosed as uncertain autoimmune disease (suspect allergic vasculitis, rheumatoid arthritis, Sjogren syndrome). She had irregular follow up and medication since 2008. She came back Taiwan for medical care sine 2015 March. Her chief complaint was hematuria, dysuria, incontinence for 6 months. She also complaint dry eye, dry mouth for more than 3 months. Physical examination revealed pale conjunctiva, left side knocking pain, bilateral lower leg skin rash, left ankle tenderness. She was admitted in Ren-Ai hospital on 2015/08/03. Abdominal CT on 8/4 showed bilateral lobar nephronia, irregular thickening of urinary bladder wall. After admission, she received antibiotic treatment as Cefazolin. She received TUR-biopsy and pathology showed chronic cystitis. PES on 8/14 showed 1. Superficial gastritis and erosive gastritis 2. Duodenitis, suspect ischemic enteritis, with some blood clot or necrotic tissue like material, s/p biopsy for Stool OB EIA (+). Gastric tissue pathology report hemorrhagic necrosis, consistent with ischemic change. Her rheumatology serology was p-ANCA < 0.2 IU/ml, c-ANCA = 0.2 IU/ml, ANA Negative (Cytoplasma positive), RA = 205 IU/ml, Anti-DNA = 30.5 WHO units/mL, Beta 2 microglobin = 11.3 mg/L, C3 = 50.0 mg/dl, C4= 1.7mg/dl, Ig E = 369 IU/ml, ESR = 99 mm/hr, Anti GBM Ab= 0.8 U/ml, SCL-70 < 0.3 U/ml, Anti-Ro > 240 U/ml, Anti-La = 0.3 U/ml. Sjogren's syndrome or rheumatoid arthritis flare up with vasculitis was impressed at that time, we add Imuran *(Azathioprine 50mg)* 1tab BIDPC (8/13~8/15), then mini-pulse therapy (Methylprednisolone IV form 500mg QD). She had acute respiratory failure for pulmonary hemorrhage on 8/15 received endotracheal tube insertion, then transfer to MICU. Bronchoscope showed inflammation of mucosa is possible and right lower lobe , left lingual lobe seem more severely. Due to family' request and uncontrolled vasculits, she was transferred to NTUH on 8/20 for further care.

We keep follow up this patient in NTUH. During admission in NTUH, she had tried Endoxan 450mg, self paid IVIG, Arheuma, Enbrel. She can not weaning ETT for persistent pulmonary hemorrhage . Bronchoscope on 8/26 showed active oozing from BR4 and BR6 was suspected. For immune compromised status, she had bacteremia of *Ralstonia mannitolilytica, Burkholderia cepacia complex and Candida tropicalis.* Antibiotic based on culture sensitivity report was used. Doctors kept titrate steroid dosage for infection status. For septic shock status, her family agreed with DNR on 8/30. She expired on 9/15 3:18 am.