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A Rare Case of an Asian Woman with Systemic Light Chain Amyloidosis with Multiple Myeloma

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Case Study : Amyloidosis is a rare heterogeneous, acquired or hereditary disease, that results from the abnormal deposition of beta-sheet fibrillar protein aggregates in various tissues, the heart, liver, and kidney. The most common subtype is light chain (AL) amyloidosis. AL amyloidosis has an incidence of 1 case per 100,000 person-years in Western countries. In the United States, there are approximately 1275 to 3200 new cases per year. In Asia, its epidemiology is still unknown. Our case is a 66-year-old female from the Philippines, presented with anemia, bipedal edema, progressive decline renal function accompanied with nephrotic range proteinuria and severe hypoalbuminemia within six months. She denied bone pains. Oral diuretics were initiated and diagnostic workups showed normal electrolytes, ASO and C3 with negative ANA. Serum plasma electrophoresis was indicative of hypogammaglobulinemia with Kappa/Lambda free light chain ratio of 0.17. Normal kidney size was seen on ultrasonography with hepatomegaly and perihepatic ascites. Reduced ejection fraction was also noted on echocardiography. Renal biopsy revealed positive for Congo red stain. Working diagnosis was systemic light chain amyloidosis. Cardiac transthyretin imaging was done and had equivocal results which pointed to AL amyloid or early cardiac amyloid. Liver biopsy showed benign hepatic tissue with extracellular eosinophilic deposits. A bone marrow biopsy was requested for prognostication revealing positive results for AL amyloidosis overlapping with multiple myeloma. Daratumumab, Bortezomib, Cyclophosphamide and Dexamethasone were planned as chemotherapeutic regimens. However, on the 12th hospital day, she developed febrile episodes with decreasing sensorium and declining urine output. She was diagnosed with septic encephalopathy secondary to hospital acquired pneumonia. Patient was then intubated and was prepared for hemodialysis, however a few hours later, she had ventricular arrhythmia which led to her demise.

Congo red stain.png

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