中文題目：一名患有腎臟輕鏈澱粉樣變性(AL amyloidosis)並隨後發展為多發性骨髓瘤的患者：病例報告

英文題目：A Patient with Renal AL Amyloidosis and Later Developing to Multiple Myeloma: A case report

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Introduction:

Amyloidosis refers to a group of diseases that are caused by protein misfolding and aggregation into amyloid fibrils that deposit in tissues, causing organ damage. Light chain amyloidosis(AL) is the most common type of systemic amyloidosis. It is frequently caused by a clonal expansion of bone-marrow plasma cells that secrete a monoclonal immunoglobulin light chain (LC) depositing as amyloid fibrils in tissues. The incidence of AL amyloidosis has been estimated at 4.5 cases/100,000 population. We report a case of renal AL amyloidosis and later developing to multiple myeloma.

Case presentation:

This is a 62-year-old woman with underlying disease of chronic hepatitis B. She suffered from progressive bilateral low extremity edema for two weeks. She came to our nephrology outpatient department. The urine protein creatinine ratio was 4.3(g/g). Other laboratory data showed WBC:4990(/uL), hemoglobin: 13.1(g/dL), platelet: 278000(/uL) ,GOT 36(IU/L), GPT 34(IU/L), BUN:8.0(mg/dL) creatinine :0.72 (mg/dL), albumin: 2.7 (g/dL), and hyperlipidemia [LDL (low-density lipoprotein): 203mg/dL, HDL(high-density lipoprotein): 87 mg/dL, cholesterol:347 mg/dL] . With the impression of nephrotic syndrome, we arranged further laboratory examination. The laboratory data revealed ANA (anti-neutrophil antibody, <1:40), C3: 10(mg/dL) and C4: 14 (mg/dL). The pANCA (0.2 IU/ml) and cANCA(<0.2EiU/mL) were negative. Serum IgG elevated (1720 mg/dL, normal range 700-1600 mg/dL), but IgA(139 mg/dL) and IgM (67 mg/dL) were within normal range. The free kappa/lambda was 0.52 [free kappa 15.8 mg/L, free lambda 31 mg/L]. However, the immunofixation revealed monoclonal IgGλ gammopathy in serum and urine. Renal biopsy was performed, and the pathology revealed AL amyloidosis, suspicious of multiple myeloma with renal involvement. Therefore, first bone marrow biopsy was also arranged, and the pathology revealed hypocellular marrow without malignancy. Because monoclonal protein was still noted, second bone marrow biopsy was repeated three months later, and the pathology report disclosed 20% plasma cells with slight lambda predominance in marrow. The multiple myeloma was diagnosed, and the

patient received VRD (bortezomib, lenalidomide, dexamethasone)

Discussion

Amyloidosis is the term for a group of protein misfolding disorders characterized by the extracellular deposition of insoluble polymeric protein fibrils in tissues and organs. AL amyloidosis is usually caused by the low-level expansion of an indolent B cell clone that produces an immunoglobulin light chain λ and κ light chains . The heart and the kidneys are the two most frequently affected organs in systemic AL amyloidosis. Renal AL amyloidosis usually manifests as proteinuria, often in the nephrotic range and associated with hypoalbuminemia, secondary hypercholesterolemia and hypertriglyceridemia, and edema or anasarca. Although AL amyloidosis is the result of clonal proliferation of plasma cells, most patients do not meet criteria for multiple myeloma. While patients are diagnosed with monoclonal gammopathy of undetermined significance (MGUS) with abnormal free light chain ratio or smoldering myeloma, amyloidosis should be excluded carefully.

Supportive care is important for patients with renal amyloidosis. For amyloidosis-related nephrotic syndrome, diuretics and supportive stockings can ameliorate edema; angiotensin-converting enzyme inhibitors should be used with caution and have not been shown to slow renal disease progression. We suggested follow-up the serum and urine protein electrophoresis and immunofixation, serum free light chain assay , electrolyte to determine to development of multiple myeloma. For multiple myeloma nephropathy with acute or subacute kidney injury, we suggested supportive care, including intravenous fluid therapy, avoidance of nephrotoxic agent. Besides, bortezomib-based chemotherapy with high-dose dexamethasone might be started as soon as possible to decrease light chain production. Lenalidomide is typically avoided in patients with acute kidney injury (AKI) unless the patient is refractory to other options. Dialysis should be initiated for the conventional indications, such as fluid overload, hyperkalemia, and uremia.

Conclusion

For AL amyloidosis, kidneys are the most frequently involved organ with the presentation of azotemia, nephrotic syndrome, or AKI. Some case will develop into multiple myeloma. Follow-up of serum and urine immunofixation and electrophoresis, serum creatinine level, electrolyte, immunoglobin level is necessary.