JOURNAL OF CLINICAL AND TRANSLATIONAL NEPHROLOGY
JCTN (2024) Volume 3: Article no: 2

RARE RENAL MANIFESTATION OF PLASMA CELL DISORDER

Shamira Shahar, Christopher Thiam Seong Lim, Haidi Mohamad, Fauzah Abd Ghani, Bak Leong Goh
Nephrology Unit, Department of Medicine, Hospital Sultan Abdul Aziz Shah, Universiti Putra Malaysia.
Nephrology Department, Hospital Sultan Idris Shah, Serdang.
Pathology Department, Hospital Sultan Idris Shah, Serdang.
Department of Pathology Faculty of Medicine and Health Sciences, Universiti Putra Malaysia.

Correspondence:
Shamira Shahar
Nephrology Unit, Department of Medicine, Hospital Sultan Abdul Aziz Shah, Universiti Putra Malaysia
Email: shamira@upm.edu.my

CASE PRESENTATION

A 56 year old lady presented with 2 weeks history of nausea, vomiting, and unintentional weight loss, with reduced effort tolerance. She has underlying type 2 diabetes mellitus, and hypertension. On examination, blood pressure was 140 / 80mmhg, pallor conjunctivae, and other physical examination was normal. Her investigations showed creatinine of 1214 umol/L, urea 24.3 mmol/L, eGFR of 2 ml/min/1.73m^2, albumin 35 g/L, total protein 71g/L, globulin 34g/L, albumin /globulin ratio 1.08, haemoglobin 6.8 g/dl, platelet 312x 10^3/uL, total white cell 7.97 x 10^9/L, calcium 2.44 mmol/L, phosphate 2 mmol/L, and erythrocyte sedimentation rate (ESR) 100 mm/hr. Urine microscopy shows 1+ protein, no RBC presence and urine protein creatinine index of 12.9g/day. Immunology screening revealed normal ANA, C3, C4, and ANCA. Kidney ultrasound shows a left kidney size of 10.3cm, and right kidney of 11.7cm with preserved corticomedullary differentiation bilaterally. She was initially diagnosed with ESKD, and long-term kidney replacement therapy was planned for her.

DIFFERENTIAL DIAGNOSIS:

Initial differential diagnoses upon presentation were diabetic kidney disease, multiple myeloma, and secondary membranous nephropathy.

HISTOPATHOLOGICAL EXAMINATION

Given normal kidney size and lack of chronic features on ultrasound, she underwent renal biopsy. Histology analysis reveals a malignant plasma cell infiltration in the interstitial, occasional fractured cast in the tubules with diffused acute tubular injury and interstitial fibrosis. Immunohistochemistry of the malignant cells shows a positive value for CD 138 with a Ki67 proliferation index of about 50-60%. Otherwise, histology showed normal glomeruli. This finding was later confirmed with serum immunofixation, which revealed IgD lambda paraproteinemia, 2.7g/L and serum-free light chain lambda of 6380 mg/L, with a free light chain ratio of 0.002.

FINAL DIAGNOSIS:

Lambda restricted IgD multiple myeloma with light chain proximal tubulopathy and plasma cell infiltration.

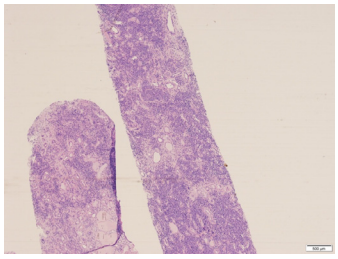

Figure 1: Dense infiltrate of plasma cells in the interstitial, compressing the tubules and microvasculature.

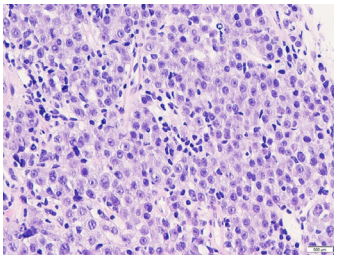

Figure 2: Atypical plasma cells can be appreciated with atypical hyperchromatic nuclei.

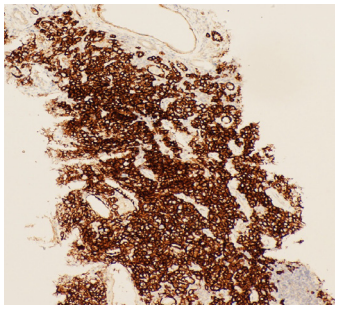

Figure 3: Immunostaining for CD138 highlighting the plasma cells.

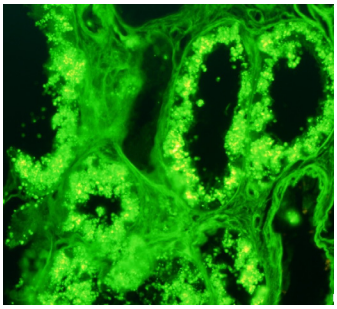

Figure 4: Immunofluorescence image showing deposits of kappa along the tubular basement membrane.

LEARNING POINTS

1. The association of kidney failure, and anaemia; with or without hypercalcemia should raise suspicion of multiple myeloma.
2. The occurrence of plasma cell infiltration in the kidney is extremely rare, and usually with the presence of other myeloma kidney manifestation.
3. Proteinuria with presence of albuminuria suggests the presence of myeloma cast nephropathy as well as light chain deposition disease which cause disruption of the filtration barrier or glomerular basement membrane.
4. Histology findings of acute tubular injury and acute tubulointerstitial nephritis should raise a ‘red flag’ for potential injury from high levels of free light chains in patients with multiple myeloma.