A Case of Plasma Cell Leukemia Presenting as Renal Failure with Heavy Proteinuria

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Introduction

- Plasma cell dyscrasias (such as multiple myeloma, Waldenstrom's macroglobulinemia, and amyloidosis) are often associated with renal dysfunction through the production of monoclonal immunoglobulins.

Case Presentation

- A 48-year-old woman with history of stage II breast cancer was admitted with acute kidney injury with creatinine (Cr) 4.4 (baseline normal).
- Work-up revealed nephrotic-range proteinuria with urine protein/Cr ratio of 6.37. Serum protein electrophoresis showed hypogammaglobulinemia with normal immunofixation; however serum kappa free light chains (FLC) were immeasurably high at >562 mg/dL and urine kappa FLC was 1290 mg/dL.
- Renal biopsy demonstrated light chain cast nephropathy with numerous large casts that had positive immunoreactivity with antibodies to kappa light chains identified on immunofluorescence (Figure 1A and B).
- Peripheral blood smear revealed 22% circulating plasma cells (Figure 2) and bone marrow biopsy showed 100% cellularity with 95% replacement by plasma cells (Figure 3), confirming the diagnosis of plasma cell leukemia (PCL).
- Myeloma FISH later showed gain 1q, deletion 13q, tp53 deletion, and MYC rearrangement.
- She was started on dialysis and plasma exchange for renal failure secondary to cast nephropathy along with CyBorD (cyclophosphamide, bortezomib, dexamethasone).

Figure 1: Renal Biopsy

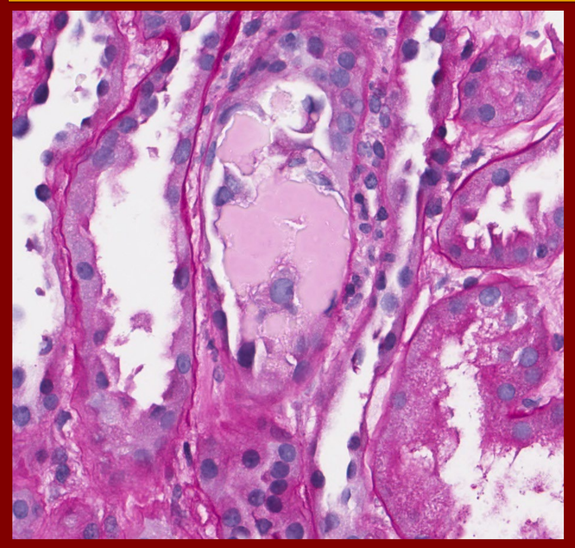

Figure 1A: Periodic-acid-Schiff stain showing light chain casts in tubules

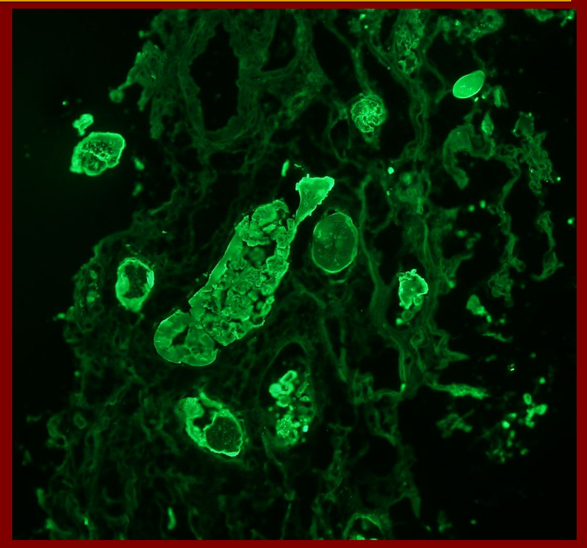

Figure 1B: Direct immunofluorescence with antibodies to kappa light chains

Figure 2: Peripheral Smear

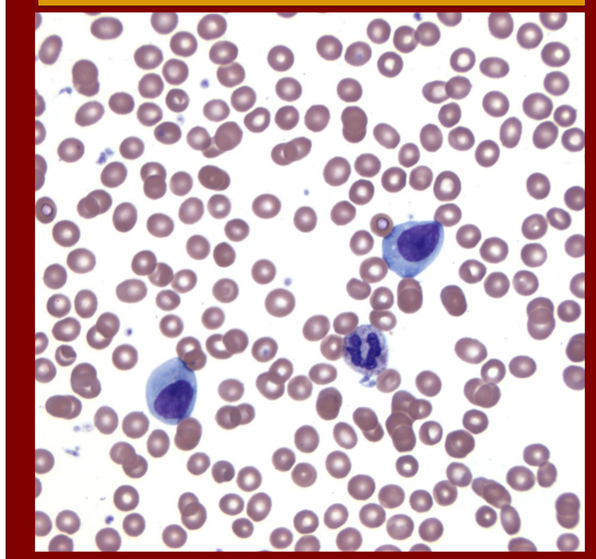

Figure 2: Peripheral smear showing circulating plasma cells

Figure 3: Bone Marrow

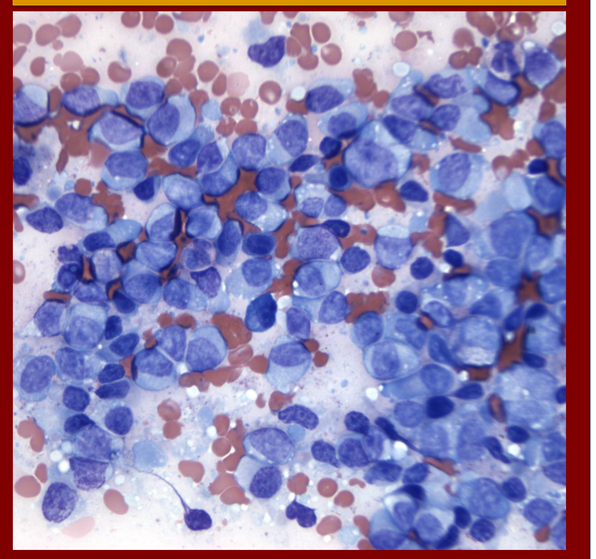

Figure 3: Bone marrow touch imprint with many plasma cells

Case Resolution

- Serum kappa FLC improved to 84 mg/dL after 3 sessions of plasma exchange and renal function eventually improved back to normal.
- She was switched to VDT-ACE (bortezomib, dexamethasone, thalidomide, doxorubicin, cyclophosphamide, etoposide) plus daratumumab after 2 cycles of CyBorD due to disease progression with non-secretory plasmacytomas of the spine.
- Repeat bone marrow after 1 cycle of VDT-ACE-dara showed morphologic remission with MRD negativity. She will soon undergo autologous stem cell transplant (ASCT).

Discussion

- Patients presenting with acute onset renal failure and heavy proteinuria require prompt evaluation for plasma cell dyscrasias.
- Plasma cell leukemia is a particularly aggressive malignancy characterized by >20% circulating plasma cells. Although more commonly seen with multiple myeloma, PCL can also rarely cause cast nephropathy and significant proteinuria.
- The use of plasmapheresis for cast nephropathy is controversial and should be decided on a case-to-case basis.
- While awaiting renal recovery, CyBorD is a good option as these drugs do not require adjustment for renal impairment.
- PCL often requires more intensive induction treatment such as VDT-PACE, which proved to be the case here. ASCT is then recommended in first remission.