INTRODUCTION: Plasma cell neoplasms represent 1.4-2% of all malignancies. Primary plasma cell leukemia (pPCL) is a rare and aggressive malignancy of plasma cells with a poor prognosis, constituting only 1-4% of all plasma cell neoplasms. pPCL is characterized by the presence of >2x10^9/l peripheral blood plasma cells or plasmacytosis accounting for >20% of total leucocyte count. Direct renal involvement in pPCL is rarely reported with only few cases in the English literature. We report a case of a 41-year-old female who presented with acute renal failure and heavy proteinuria and was found to have primary PCL invading the kidney coupled with light-chain cast nephropathy.

CASE REPORT: A 41-year-old female presented to the outpatient department with fever, cough, and breathlessness since 3 days and multiple joint pains and fatigue since 1 month. Laboratory data showed total leucocyte count 32.2 x 10^3/µL, haemoglobin 5.4 g/dL, platelet count 20 x 10^9/L, BUN 34 mg/dL, creatinine 1.2 mg/dL, and calcium 11.5 mg/dL. Initial urinalysis showed 2+ blood and 4+ protein. Peripheral blood smear revealed leucocytosis with 40% plasma cells and thrombocytopenia. No lytic lesions were noted in the imagology. Serum protein electrophoresis demonstrated myeloma band. Serum flow cytometry with kappa chain was positive. Beta-2 microglobulin levels were 8448ng/ml which is 12 times more than normal. Bone marrow aspiration and biopsy showed >45% plasmacytosis and immunohistochemistry with CD 138 showed cytoplasmic positivity and confirmed the diagnosis. As patient was in renal failure, dialysis was done. Percutaneous renal biopsy revealed cast nephropathy. Chemotherapy with bortezomib was started. Despite an initial favourable response, the patient died within two weeks due to an infectious complication.

DISCUSSION: Acute kidney disease has various aetiologies and are divided into pre-renal, renal, and post-renal causes. Renal biopsy plays a major role in the diagnosis and management of Acute kidney disease. In this case, the presence of acute kidney disease coupled with heavy proteinuria, the biopsy revealed kidney infiltration by PCL coupled with light-chain cast nephropathy. Treatment includes induction therapy with alkylating agents.

CONCLUSION: pPCL has short unfavourable outcome, requiring the achievement of better data to improve the disease course.