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CASE REPORT

Lambda Light Chain Cast Nephropathy: A Case Report

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Abstract

Excess production of free light chains is due to underlying plasma cell dyscrasia or a lymphoproliferative disease. When the degradation capacity by proximal tubule is exceeded as a result of overproduction, the excess light chains produced reach the distal tubule and cause accumulation, causing deterioration in kidney functions. Although this condition, called cast nephropathy, is most seen in multiple myeloma, it may also be associated with Waldenström macroglobulinemia and lymphomas.

Keywords: Acute kidney failure; cast nephropathy; light chain; plasma cell disease.

Excess production of free light chains is due to underlying plasma cell dyscrasia or a lymphoproliferative disease[1]. When the degradation capacity of the proximal tubule is exceeded as a result of overproduction, the excess light chains produced reach the distal tubule and accumulate, causing deterioration in kidney functions[2]. Although this condition, called cast nephropathy, is most commonly seen in multiple myeloma, it may also be associated with Waldenström macroglobulinemia and lymphomas[3-5].

Case Report

A 65-year-old male patient applied to our clinic with complaints of weakness, fatigue, and muscle pain for 3 weeks. His present complaints were accompanied by polydipsia, pollakiuria, and new-onset itching on the ankle. The patient had a history of coronary artery disease for 1 year and had a history of stent on the left anterior descending and circumflex coronary arteries. There was no smoking or alcohol use and no family history of a disease. Except for a 2/6 systolic murmur heard in the aortic area in his physical examination, no pathology was detected in the cardiovascular system and other system examinations.

In laboratory tests, hemoglobin and platelet values were lower than normal; 11.1 g/dL (n:13.5–17), 147,000 (n:150–439×103), respectively, and BUN and creatinine values were found to be high; 63 mg/dL (n:8–23) and 6.17 mg/dL (n:0.7–1.3), respectively. When the anamnesis was detailed, it was learned that the serum creatinine level had progressed to 1.9 mg/dL 5 weeks ago, and the statin dose was halved by the cardiologist and ramipril treatment was discontinued. In addition, the corrected serum calcium value was 10.35 mg/dL (n:8.5–10.1), alkaline phosphatase (ALP): 153 IU/L (n:39–119), lactate dehydrogenase: 262 IU/L (n:85–227), and above the normal ranges, while creatine kinase, erythrocyte sedimentation rate (ESR), C-reactive protein, and total protein and albumin levels were within the normal range.

Sodium hydrogen carbonate, allopurinol, and hydration treatments were started in the patient whose uric acid level rapidly increased to 8.23 mg/dL (n:3.5–7.2 mg/dL) and had an acidic pH in blood gas. No improvement was observed in the kidney functions of the patient, whose electrolyte imbalance improved with treatment and who had ade-

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quate diuresis. Dialysis was tried in the treatment of the patient whose creatinine values were 6.17, 6.4, and 5.68 mg/dL with 3 consecutive days of follow-up, but no significant improvement was found in creatinine values.

In the complete urinalysis, erythrocyte and protein values were found as 1+. Nephritic level of proteinuria was detected according to the spot urine protein/creatinine ratio (3.39 mg/mg [n: <0.2]). Anti-nuclear antibody and anti-neutrophil cytoplasmic antibody profiles required for nephritic syndrome etiology were negative and complement C3 and C4 levels were found to be normal.

Primary hyperparathyroidism was excluded, with parathormone level being lower than normal level. Malignancies were considered, for the etiology of hypercalcemia.

While the bronchovascular structures were prominent in the chest X-ray, the whole abdominal ultrasound was reported as normal.

Whole-body magnetic resonance imaging was requested from the patient who was suspected of having hypercalcemia due to increased osteoblastic activity, both to evaluate bone and muscle structures and to examine thyroid or prostate tissues for malignancy. No finding of myositis was observed in the imaging, but symmetrically increased osteoblastic activity was observed in the proximal of the flat and long bones, suggesting lymphoproliferative/hematologic malignancy.

In serum protein electrophoresis, gamma-band activity was at the lower limit and monoclonal spike (M-spike) was not detected. It was decided to perform bone marrow biopsy and renal biopsy to arrive at a definitive diagnosis. Renal biopsy was compatible with light chain cast nephropathy, while bone marrow biopsy was evaluated as plasma cell myeloma (monoclonal lambda) (Figs. 1-3). Dexamethasone treatment was started as soon as the biopsy result was obtained.

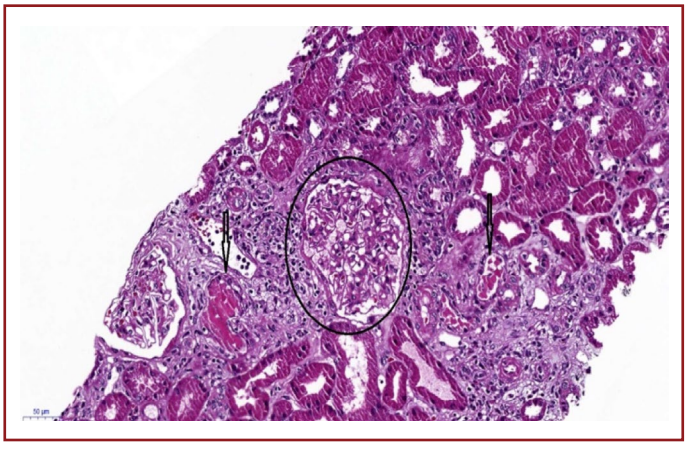

Figure 1. Cast nephropathy in kidney biopsy.

The immunofixation results were reported 5 days after the biopsy result. In immunofixation electrophoresis, Ig A, G, M, and kappa light chain levels were found to be low, while lambda light chain levels were found to be 2.42 g/L and high (n:0.9–2.1) (Fig. 4). When the serum-free lambda light chain level was >14000, the diagnosis was confirmed.

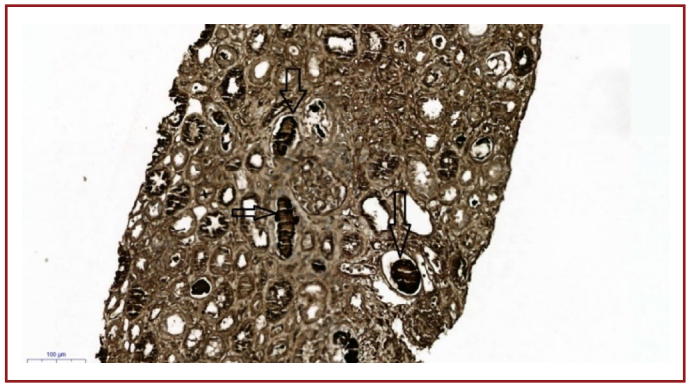

Figure 2. Lambda light chain staining in renal biopsy.

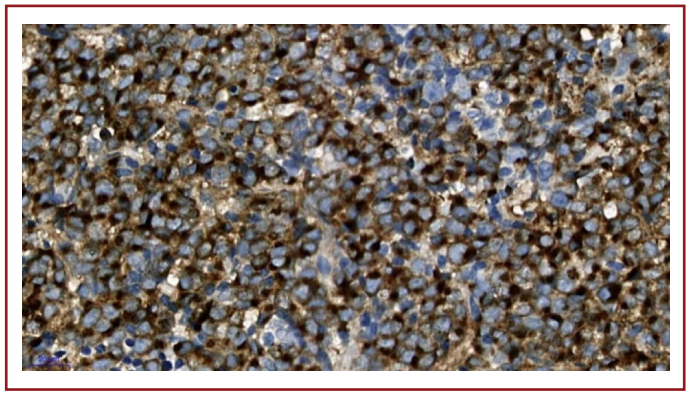

Figure 3. Plasma cell infiltration in bone marrow biopsy, lambda light chain immunostaining.

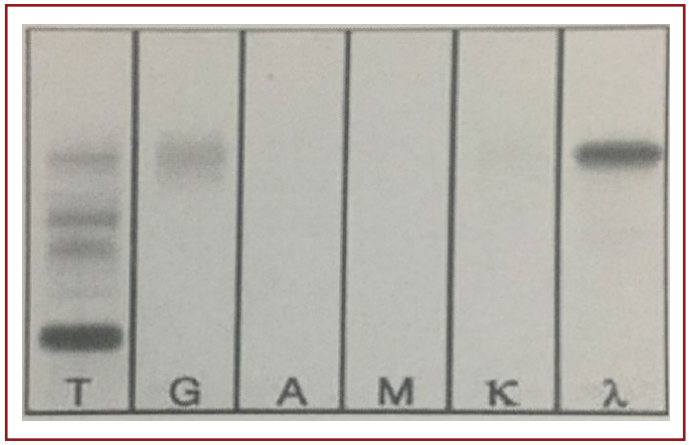

Figure 4. Free lambda band observed in serum immunofixation electrophoresis.

Hemodialysis was planned for the patient 3 days a week and was transferred to the hematology department to receive bortezomib/Endoxan/dexamethasone treatment.

Discussion

Although cast nephropathy always presents as acute kidney injury/failure, it can sometimes be seen alone or in combination with proximal renal tubular acidosis/Fanconi syndrome. It is also the most common (50%) type of kidney disease in patients with monoclonal gammopathy.

In the cast nephropathy diagnostic algorithm, it is recommended to evaluate serum protein electrophoresis and urinary albumin excretion together in suspected paraprotein-related renal injury (Fig. 5). Renal biopsy is not always necessary if immunofixation electrophoresis and light chain levels can be measured quickly. In our case, there was no evidence of monoclonal gammopathy, with albumin, globulin, and ESR results contributing to demonstrating paraprotein-related disease. These data, when evaluated clinically, necessitated renal biopsy and concomitant bone marrow biopsy due to increased osteoblastic activity in the bone marrow.

Light chain deposition disease (LCDD) is a histopathological diagnosis. Immunofluorescence is the most sensitive method to demonstrate light chain deposits[6,7]. Although LCDD is often associated with kappa light chains in the literature, it was associated with increased lambda light chain production in our case[6]. In addition, although LCDD is characterized by proteinuria in the nephrotic range, in our case, the diagnosis was made within a week before proteinuria reached the nephrotic range.

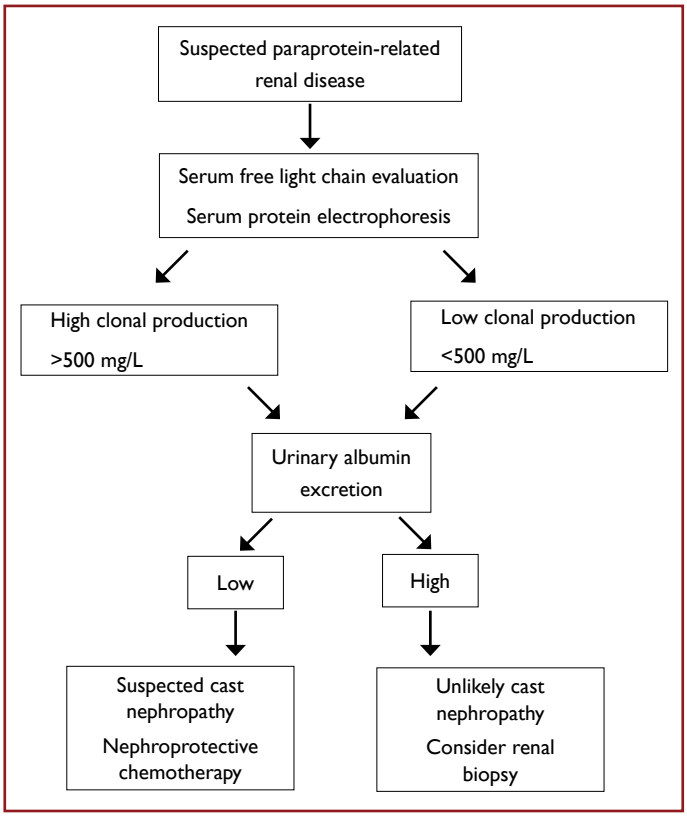

Figure 5. Cast nephropathy diagnostic algorithm.

It has been shown that the development of ARF in plasma cell diseases reduces the 1-year life expectancy. In addition, recovery of renal function is superior to response to chemotherapy on survival. Prognosis improves if renal damage can be reversed with treatment.

Each year, 4.3 patients per million suffer from end-stage renal disease (ESRD) due to plasma cell diseases, and 3-year mortality rates were found to be higher compared to other diseases that cause ESRD.

Histopathological diagnosis guides the diagnosis and treatment of the underlying disease, thus supporting the importance of early diagnosis.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

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