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Case Report

Acute renal failure due to light chain cast nephropathy (A Case Report)

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Abstract. Myeloma cast nephropathy, also known Bence Jones cast nephropathy or myeloma kidney is the most common form of renal disease associated with multiple myeloma. Clinically, it may present as progressive renal insufficieny or acute renal failure. We describe a case of multiple myeloma who presented with acute renal failure due to light chain cast nephropathy and had a primary diagnosis made by renal biopsy. A renal biopsy was performed for acute renal failure in a 44 year old female patient. The renal biopsy showed numerous dilated tubules filled with pink eosinophilic fractured casts surrounded at places by multinucleated giant cells and accompanied by moderate interstitial infiltrate consisting of lymphomononuclear cells. There was tubular atrophy with interstitial fibrosis. The renal biopsy is occasionally the first test indicative of myeloma in a patient with acute renal failure of seemingly unknown origin. Thus the microscopic appearance of the tubules in this biopsy is easy to appreciate and is diagnostic of light chain cast nephropathy.

Key words: Multiple myeloma, light chain cast nephropathy, acute renal failure

1. Introduction

Myeloma cast nephropathy, also known Bence Jones cast nephropathy or myeloma kidney is the most common form of renal disease associated with multiple myeloma. Myeloma cast nephropathy is sometimes associated with acute renal failure. Multiple myeloma (MM) is characterized by the proliferation of a malignant clone of plasma cells that produce large amounts of immunoglobulins. This can be detected electrophoretically as a sharp monoclonal or M spike in serum or urine and is a hallmark of MM (1-3). In rare situations, this M spike is absent and can lead to problems in diagnosis. Renal involvement is seen in about 10–20% of all MM cases and most commonly takes the form of acute renal failure (ARF) due to myeloma cast nephropathy (2). There is little information in the literature about the nature of renal involvement in nonsecretory myeloma. Clinically, it may present as progressive renal insufficiency or acute renal failure, which may be precipitated by dehydration, hypercalcemia, intravenous infusion of contrast media, or antibiotic toxicity.

We describe a case of multiple myeloma who presented with acute renal failure due to light chain cast nephropathy and had a primary diagnosis made by renal biopsy.

2. Case report

A renal biopsy was performed for acute renal failure in a 44 year old female patient. The serum creatinine was 14.49 mg/dl, calcium 5.8 mg/dl, uric acid 7.5 mg/dl, albumin 3.8 g/dl, globulins 2.7 g/dl, blood urea nitrogen (BUN) 110 mg/ dl, creatinine 14.49 mg/dl, total protein 6.5 g/dl, Na 133 mEq/l; K, 5.25 mEg/l and alkaline phosphatase 5 King-Armstrong units. Urine total protein excretion was 1.9 g/24h.

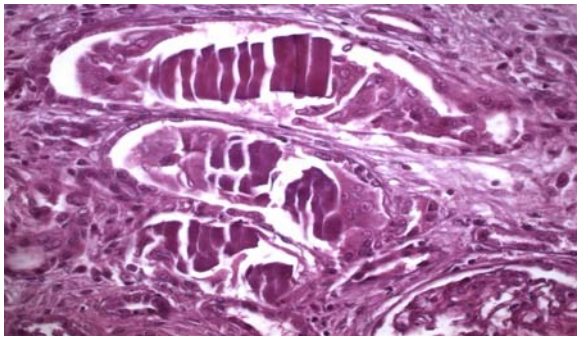

Fig. 1. Photomicrograph showing a dilated tubule containing eosinophilic hyaline cast with characteristic fractures and surrounding giant cells. The interstitium shows infiltration with lymphomononuclear cells (H&E x400).

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Renal ultrasound showed the right and left kidneys to be normal. Renal biopsy had 18 glomeruli. The focal moderate mesangial expansion was present. The renal biopsy showed numerous dilated tubules filled with pink eosinophilic fractured casts surrounded at places by multinucleated giant cells and accompanied by moderate interstitial infiltrate consisting of lymphomononuclear cells (Fig. 1). There was severe tubular atrophy with interstitial fibrosis (Fig. 2).

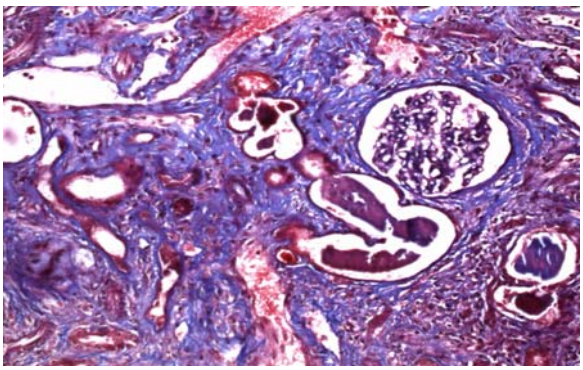

Fig. 2. Showing severe tubular atrophy and interstitial fibrosis (Masson's Trichrome x200).

Tubules showed foci of atrophy with pronounced thickening and lamination of the basement membrane, especially with the periodic acid Schiff stain. Artery walls were thickened with intimal fibrosis and showed modest luminal narrowing. Arteriolar walls were often thickened with muscular hypertrophy and, in many, insudative lesions (''hyalinization''). All glomeruli were a normal structural appearance. The immunofluorescence (IF) microscopy was performed using antisera to human IgG, IgA, IgM, Clq, C3. Tubular casts stained for IgG and C1q. Congo red stain for amyloid was negative. Immunoperoxidase staining using antibodies to lambda and kappa chains showed strong positivity for kappa chains in the casts (Fig. 3).

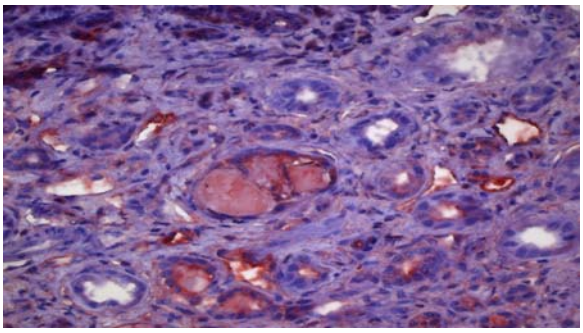

Fig. 3. Photomicrograph showing strong positivity for kappa light chain in the intra-tubular cast (Immunoperoxidase x 400).

After renal biopsy, Xray of skull and aspiration of bone marrow were performed. Xray of skull showed multiple lytic lesions. Aspiration of bone marrow was showed sheets of plasma cells.

Serum immunoelectrophoresis showed no monoclonal spike. Tests for Bence-Jones protein were consistently negative.

3. Discussion

The presence of an ''M'' peak on serum and/or urine electrophoresis, lytic lesions on skeletal radiographs and marrow plasmacytosis are characteristic features of multiple myeloma. The band may not be detectable in about 1–5% of all cases wihich are dubbed ''nonsecretory myeloma (NSMM)'' (3–5).

As seen in this case, ARF can be the presenting feature and antedates the diagnosis of MM in a large number of cases with the classic disease (6). However, our case showed definite ''cast nephropathy'' on biopsy.

Three major renal lesions result from deposition or accumulation of abnormal monoclonal immunoglobulin light chains. These are light chain cast nephropathy, light chain deposit disease, and AL (light chain associated) amyloid. The first is regularly associated with multiple myeloma, whereas the others may or may not be (7-10). Any of these lesions may be the first manifestation of the plasma cell dyscrasia; they often occur alone, but two or all may coexist.

Light chain cast nephropathy is characterized by prominent casts in renal tubules; the casts are usually large and ''brittle,'' have fracture lines, or are broken into many fragments, often with geometric shapes, and are surrounded by tubular epithelium, neutrophils, and typically by multinucleated giant cells of foreign body type. Although they are more common in distal tubules, they may be found in any segment of the nephron, including Bowman's space. The casts have reasonably typical tinctorial properties: PAS negative, brightly eosinophilic, fuchsinophilic with Masson's trichrome, and, infrequently, Congo red positive. This is in contrast to Tamm-Horsfall protein casts (11). The myeloma casts are composed primarily of the abnormal light chain (12). Tubular basement membranes are discontinuous, thereby allowing free communication between interstitium and tubular lumina; it is through these gaps that monocytes and other inflammatory cells migrate from the interstitium (13). This constellation of light microscopic abnormalities, especially the morphology and tinctorial properties of the casts and the surrounding giant cells, is sufficiently

distinctive to be diagnostic of multiple myeloma. The renal biopsy is occasionally the first test indicative of myeloma in a patient who presents with acute renal failure of seemingly unknown origin (14). Thus, the microscopic appearance of the tubules in this biopsy is easy to appreciate and is diagnostic of light chain cast nephropahty.

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