Case Report

Focal segmental glomerulosclerosis, secondary amyloidosis and multiple myeloma

Farzaneh Ashrafi[1], Mojgan Mortazavi[2], Navid Manouchehri[3], Noushin Afshar Moghaddam[4], Hamid Nasri[5], Amir Hossein Sarrami[6]

ABSTRACT

Focal segmental glomerulosclerosis (FSGS) is a kind of glomerulonephritis characterized by scar tissue that forms in parts of the kidney called glomeruli. Association of FSGS with hematologic malignancies such as plasma cell disorder is an uncommon condition. We report a 58-year-old male with FSGS who developed the manifestation of amyloidosis in his course of disease. Following urine protein electrophoresis and bone marrow study confirmed the diagnosis of multiple myeloma.

KEY WORDS: Amyloidosis, FSGS, Multiple Myeloma.

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INTRODUCTION

Focal segmental glomerulosclerosis (FSGS) is one of the most common types of glomerulonephritis in Iran.[1] It has various etiologies such as genetic abnormalities, metabolic disorders, infections and drug abuse.[2] However, association of FSGS with hematologic malignancies such as lymphoproliferative, myeloproliferative and plasma cell disorders is an uncommon condition. The authors present a case of FSGS, secondary amyloidosis and multiple myeloma.

1. Farzaneh Ashrafi,
Department of Hematology and Medical Oncology,
2. Mojgan Mortazavi,
Isfahan Kidney Diseases Research Center,
Isfahan University of Medical Sciences, Isfahan, Iran.
3. Navid Manouchehri,
4. Noushin Afshar Moghaddam,
5. Hamid Nasri,
6. Amir Hossein Sarrami,
3, 6: Medical Students' Research Center,
4, 5: Department of Pathology,
1-6: Isfahan University of Medical Sciences,
Isfahan, Iran.

Correspondence:

Mojgan Mortazavi.
E-mail: m_mortazavi@med.mui.ac.ir

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CASE REPORT

A 58-year-old male was referred to nephrology department (Alzahra Hospital, Isfahan University of Medical Sciences) for evaluation of proteinuria. His proteinuria had been found incidentally in routine screening tests. The patient had no complaint nor had any history of underlying disease. Physical examination revealed blood pressure 160/90 mmHg and two plus pitting edema in his lower limbs. Laboratory studies were creatinine 1.1 mg/dL, blood urea nitrogen (BUN) 20 mg/dL and proteinuria 2200 mg per 24 hours. Further diagnostic workup for proteinuria did not show any abnormal finding. An angiotensin converting enzyme inhibitor (ACEI) was administered and a renal biopsy was taken. The histopathological study revealed a tip variant FSGS (Fig.1).

In the follow-up his high blood pressure was controlled, but the proteinuria and serum level of creatinine increased gradually. After nine months, the patient complained of drooling and he developed macroglossia within a month (Fig.2). Considering the probability of primary amyloidosis he underwent a periumbilical fat biopsy. Positive staining of amyloid deposition with Congo red confirmed the diagnosis of amyloidosis (Fig.3). In an abdominal ultrasonography there was no evidence

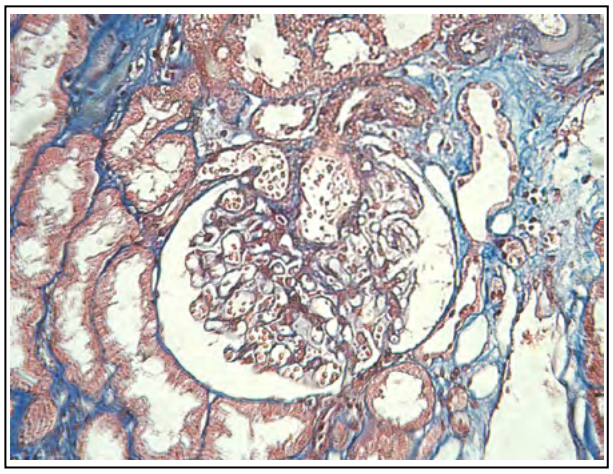

Fig.1: Photomicrograph showed tuft-capsule fusion at tubular pole (Masson's trichrome staining 400×).

of organ enlargement. His cardiac evaluation revealed no abnormality. For further investigations a urine protein electrophoresis and bone marrow aspiration and biopsy were performed. There was positive light chain in immunofixation of urine.

In the study of bone marrow aspiration there were more than 40 percent plasma cells that all were kappa positive in immunohistochemistry study of bone marrow biopsy specimen (Fig. 4 and 5). Clinical and pathological findings were compatible with the diagnosis of multiple myeloma. After three days the patient developed acute renal failure and hypercalcemia. He was treated with hydration, thalidomide and dexamethason plus pamidronate. After one week of treatment his calcium and creatinin reduced. In the following month his drooling improved and macroglossia decreased. The next plan for his treatment is autologous bone marrow transplantation.

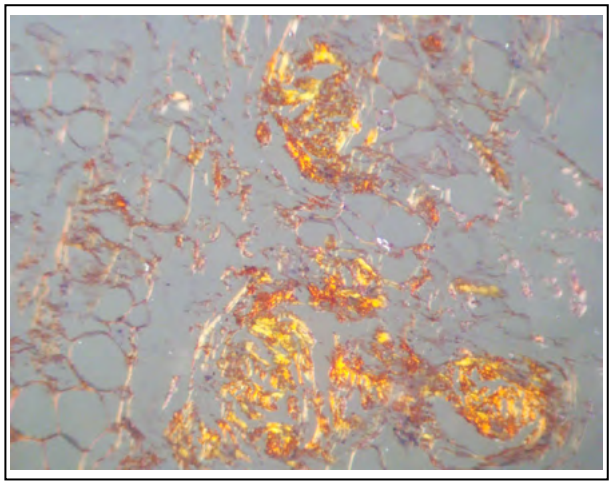

Fig.3: Congo red staining under polarized filter revealed positive birefringence (400×).

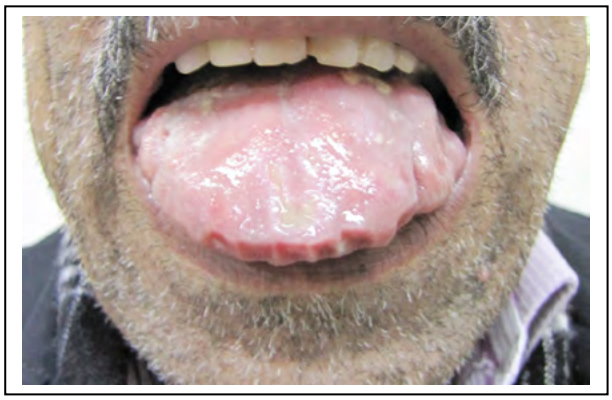

Fig.2: Macroglossia.

DISCUSSION

Multiple myeloma is an uncommon disease characterized by clonal proliferation of malignant plasma cells in the bone marrow.[3,4] The multiple myeloma cells may produce an excessive amount of a monoclonal immunoglobulin detected on serum or urine protein electrophoresis.[5] Symptomatic multiple myeloma may present with anemia, hyper calcemia, bone disease and renal dysfunction. Renal dysfunction in multiple myeloma may be as a result of various conditions including of precipitation of monoclonal light chains in renal tubules, dehydration, hypercalcemia, hyperuricemia and administration of nephrotoxic drugs.[6]

Our patient presented with proteinuria and hypertension which was diagnosed due to FSGS. FSGS is a clinicopathologic entity characterized by scar tissue that forms in parts of the kidney called glomeruli. FSGS has various histological variants. Tip variant FSGS which was seen in our patient has a higher renal survival and remission rate compared with other variants.[7] FSGS has various etiologies and may be as a consequence

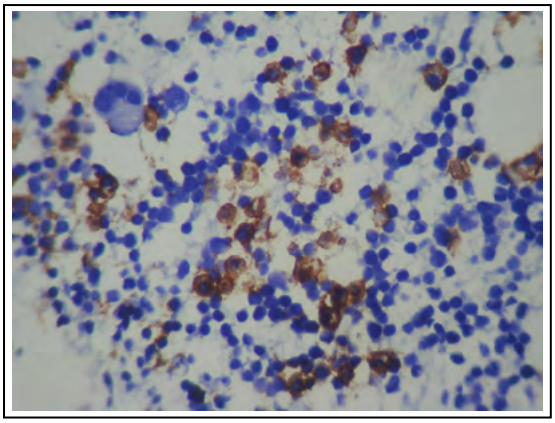

Fig.4: Bone marrow immunostaining for CD 138 revealed immunoreactive plasma cell (400×).

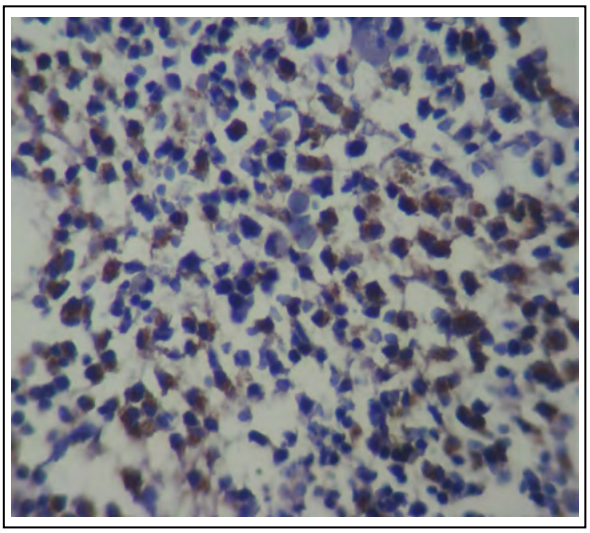

Fig.5: Immunostaining for kappa light chain revealed positive reaction in most of the plasma cells (400×).

of multiple pathways of renal injury.[2] Association of FSGS with hematologic malignancies such as lymphoproliferative, myeloproliferative and plasma cell disorders is an uncommon condition.[2,8]

Shah et al reported a case of collapsing FSGS in multiple myeloma resolved after chemotherapy.[8] In a retrospective study Dingli et al identified four patients with multiple myeloma which also developed FSGS.[2] They suggested that before labeling the FSGS as idiopathic, it is better to exclude a plasma cell proliferative disorder. They also showed that the occurrence of these two conditions may not be as a result of chance.[2]

In our patient macroglossia was a diagnostic signal for consideration of amyloidosis. When the diagnosis of amyloidosis was confirmed with positive Congo red staining, we evaluated the patient for finding the probable underlying disorder. Bone marrow study and urine protein electrophoresis established the diagnosis of multiple myeloma.

Early diagnosis and treatment of the underlying plasma cell disorder may reduce the renal complications of FSGS.[2] The response of renal disorders to therapy also determines the prognosis of multiple myeloma.[9]

In conclusion, this report may put emphasize on the advantage of a urine or serum protein electrophoresis in patients with idiopathic FSGS and may be an evidence for association of multiple myeloma and FSGS.

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