The Final Hit, Fibrillary Glomerulonephritis and End Stage Renal Disease

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LEARNING OBJECTIVES

1. To recognize the signs and symptoms of end stage renal disease
2. To learn the diagnostic criteria for Fibrillary Glomerulonephritis

CASE PRESENTATION

Patient is a 61 year old female with a past medical history of a congenital atrophic kidney, Stage V CKD, CAD, HTN, HLD, who presented to the ED for abnormal labs that revealed worsening renal function. She also reported generalized weakness, poor appetite and dysuria.

Labs about 1 week prior to arrival revealed a Creatinine of 8.09, compared to 1.81 one year prior to that.

The patient was being worked as an outpatient for etiology of her renal failure. A biopsy of her kidney contained 3 glomeruli which was a suboptimal specimen. 2 of the glomeruli were globally sclerotic. The 3rd glomeruli had linear staining with IgG consistent with Fibrillary Glomerulonephritis. During this workup, the patient was also found to have a polyclonal gammopathy.

The patient was started on dialysis urgently.

PHYSICAL EXAMINATION

- Vitals: BP: 165/86, HR: 63, Temp: 97.6, RR: 20, SpO2 97%
- AAO x3, Heart RRR, no MRG, Lungs CTAB, Abdomen soft, NTND, No peripheral edema

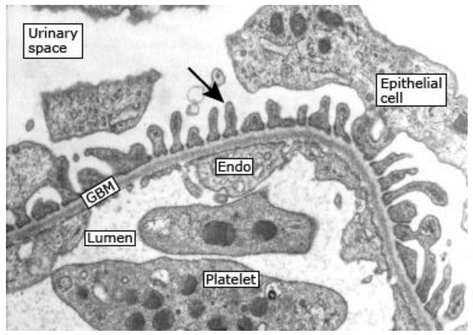
Laboratory Data	
Hemoglobin	8.2 (12-16 g/dL)
WBC	6.7 (10*3/uL)
MCV	97.0 (80-100 fL)
Sodium	135 (136-145 mmol/L)
Potassium	3.9 (3.5-5.2 mmol/L)
Chloride	103 (96-110 mmol/L)
Carbon Dioxide	14 (24-31 mmol/L)
BUN	90 (5-25 mg/dL)
Creatinine	8.53 (0.44-1.00 mg/dL)
Calcium	7.5 (8.5-10.5 mg/dL)
GFR	6 (>60)
Troponin	1.65 (<0.04 ng/mL)
Magnesium	3.0 (1.3- 2.5 mg/dL)
Phosphorus	9.0 (2.5-4.6 mg/dL)

PATHOPHYSIOLOGY

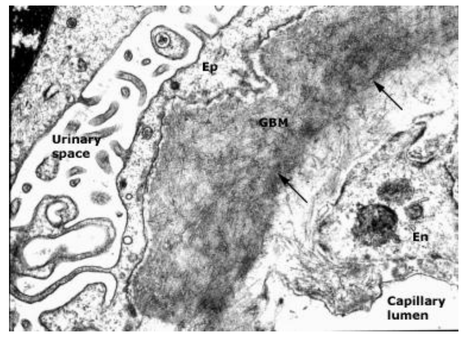
- Fibrillary glomerulonephritis is a rare disorder characterized by immunoglobulin deposition in the glomerular basement membrane.
- Immunofluorescence microscopy is positive for IgG, C3, kappa and lambda light chains.
- The fibular deposits in the mesangium and glomerular capillary walls are clearly distinct from those seen in amyloidosis. The fibrils are randomly arranged and they typically do not stain Congo red.
- Initially, fibrillary glomerulonephritis was thought to be idiopathic however approximately 30-50% of patients have a history of malignancy, monoclonal gammopathy, autoimmune disease, or infections.

PATHOPHYSIOLOGY, CONTINUED

- DNAJB9, a protein involved in the endoplasmic reticulum stress response, has been identified as a highly specific biomarker.



Normal glomerular basement membrane (GBM)



Disrupted GBM in Fibrillary Glomerulonephritis

CONCLUSIONS

- The diagnosis of fibrillary glomerulonephritis should be considered in patients with renal failure.
- The diagnosis should be distinguished from other types of glomerular disease via electron microscopy and immunofluorescence staining for DNAJB9.
- Patients with the condition should be screened for secondary causes such as malignancy, monoclonal gammopathy, autoimmune disease and HCV infection.
- There are no therapies that have been shown to be beneficial.
- Patients may benefit from treatment of an underlying disorder.
- The focus of treatment in patients with idiopathic disease is determined by severity and complications of kidney disease.