PathMD™: Board Review Letter
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Medical Renal - Part 1
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1. Nephrotic syndrome is defined by which of the following:

Answer: C. Nephrotic syndrome is defined by massive proteinuria. Other manifestations include hypoalbuminemia (<3 gm/dL) with resultant generalized edema, hyperlipidemia and lipiduria. (*Robins and Cotran: Pathologic Basis of Disease*. Kumar, V. et al., 7th Edition. 2005. p. 978)

2. This glomerular disease is associated with low serum complement levels, the presence of granular immune deposits within the glomeruli, and subepithelial hump-like electron-dense deposits. The best diagnosis is:

Answer: A. These features are characteristic of acute proliferative glomerulonephritis (a.k.a. poststrep. GN). (*Robins and Cotran: Pathologic Basis of Disease*. Kumar, V. et al., 7th Edition. 2005. p. 974-975)

3. A 35 y/o female with an ovarian mass and possible lupus presents with >3.5grams of protein in a 24 hour urine collection. Histology shown in the images (see Website Case #1) reveals increased glomerular capillary wall thickening. The Jones (silver) stain reveals irregular spikes that protrude from the glomerular-basement membrane (must look close at the image). Based on the clinical history and morphologic findings, the best diagnosis is:

Answer: B. This case illustrates the classic appearance of membranous glomerulopathy, which is clinically characterized by nephrotic syndrome. It is the most common cause of nephrotic syndrome in adults. It characteristically has diffuse thickening of the glomerular capillary wall with electron dense "spikes" along the subepithelial side of the basement membrane. In 85% of patients, there is no other disease or condition associated with the renal disease. However, drugs, malignant tumors, infections and other autoimmune disorders have been associated with membranous GN. (*Robins and Cotran: Pathologic Basis of Disease*. Kumar, V. et al., 7th Edition. 2005. p. 979-981)

4. A patient with an h/o multiple myeloma undergoes a renal biopsy. The histology as shown on the website (Case#2) shows interstitial and glomerular deposition of amorphous material. The PAS stain shows weak staining of this material present within the glomeruli. The Jones stain shows focally delicate spikes projecting from the outer aspect of the glomerular capillary wall (must look closely at the image). Based on these findings and the clinical history, the most likely diagnosis is:

Answer: B. AL Amyloidosis is associated with a lambda light chain monoclonal gammopathy. The EM shows 8-12 nm fibrils. AA Amyloidosis, which is also known as secondary amyloidosis usually, occurs in patients with chronic inflammatory diseases. Fibrillary GN shows 15-30 nm characteristic fibrils on EM, and the immunotactoid GN has microtubules that are 20-50nm on EM. Cryoglobulinemic GN can be associated with an underlying B-cell dyscrasia, but has dense deposits with 25-35 nm tubules. (*Non-Neoplastic Kidney Diseases*. Vivette DD, et al. AFIP Non-Tumor Fascicle Series #4. 2005. pp. 199-235)

5. A 20 y/o patient with HIV and recent pregnancy loss presents with HTN, proteinuria, and red cell casts. Serum complement levels are low. Histology (Website Case #3) shows hypercellular glomeruli, and the EM shows podocyte effacement with electron dense subepithelial deposits. IF shows granular deposits of IgG, IgM, and C3 in the mesangium and basement membrane. Based on the findings correlated with the patient's history, the best diagnosis is:

Answer: E. Acute proliferative (post-streptococcal / post-infectious) GN is characterized by clinical presentation of nephritic syndrome with red cell casts, mild proteinura and mild HTN. The serum complement levels are low, and there is granular immune deposits within the glomeruli. The classic EM finding are subepithelial "humps," which is shown as a single hump in the EM photograph in this case. MPGN type II is also known as dense deposit disease characterized by EM findings of dense deposits within the basement membrane. MPGN type 1 is characterized by nephrotic syndrome and subendothelial deposits on EM. FSGS is associated with nephrotic syndrome. (*Robins and Cotran: Pathologic Basis of Disease*. Kumar, V. et al., 7th Edition. 2005. p. 974-976)

6. A 9 y/o male presents with 3.0 grams of proteinuria in 24 hours with no red cell casts. A renal biopsy (Website Case #4) was performed and shows mesangial widening and proliferation. EM (not shown) showed electron dense deposits within the mesangium. IF was performed and the anti-IgA image is shown in this case. Based on the clinical presentation and the renal biopsy findings, the best diagnosis is:

Answer: C. Henoch-Schoenlein purpura is closely associated with IgA nephropathy, and many consider it within the spectrum of IgA nephropathy disease. IgA nephropathy (Berger disease) is characterized by the presence of IgA deposits within the mesangium on IF with these deposits being electron dense on EM. Histologically, there is mesangial proliferation and increased matrix, which is noted on the images on the website. IgA nephropathy is also associated with celiac and liver disease. (*Robins and Cotran: Pathologic Basis of Disease*. Kumar, V. et al., 7th Edition. 2005. p. 986-989) Don't expect them to put IgA as an answer (that would be too easy).

7. A 33 y/o male with a family h/o renal failure in male members presents with acute renal failure after renal transplant. Renal biopsy with histology and IF (Website Case #5) available for review shows focal crescentic glomerulonephritis with interstitial nephritis and mild focal increased mesangial cellularity. The IgG IF image is also available for review. Based on the clinical history and biopsy findings, the best diagnosis for this patient's underlying disease is:

Answer: D. Alport' syndrome, which is characterized by nephritis, nerve deafness, and eye d/o's is X-linked. Most severe symptomatology occurs in men, but women can also have significant disease. In this case, the IF shows a linear pattern of IgG deposition, which immediately makes one consider Goodpasture's syndrome. However, given the history of renal transplant and the fact that Alport's syndrome patients have abnormal type IV collagen, these patients have natural antibodies to the normal type IV collagen present in the renal transplant. This gives an IF pattern identical to that of Goodpasture's syndrome. Interestingly, Goodpasture's antigen involves the type IV collagen molecule. (*Robins and Cotran: Pathologic Basis of Disease*. Kumar, V. et al., 7th Edition. 2005. p. 975-989)

8. Which of the following are associated with tubulo-reticular inclusions within epithelial cells on EM?

Answer: D. Both A and C are correct. Tubulo-reticular inclusions within epithelial cells have been shown to be induced by circulating interpheron alpha, and are associated with both lupus and HIV disease. (*Robins and Cotran: Pathologic Basis of Disease*. Kumar, V. et al., 7th Edition. 2005. p. 994)

9. The following tumor is present in 25-50% of patients with tuberous sclerosis.

Answer: D. Angiomyolipomas (AMLs) are frequently present in patients with tuberous sclerosis. Tuberous sclerosis is also characterized by cerebral cortex lesions which may produce epileptic seizures.

10. Proteus bacteria are associated with which of the following types of renal stones?

Answer: B. Magnesium, Amonium, Phosphate stones (struvite) are formed after infections with urea splitting organisms. Proteus is the most common urease organism associated with UTIs. These lead to the "staghorn calculi" that often form stones that are in the shape of the collecting ducts. A simple way to remember this is that Magnesium, Amonium, Phosphate stones make a "MAP" of the collecting system (struvite stones). (*Robins and Cotran: Pathologic Basis of Disease*. Kumar, V. et al., 7th Edition. 2005. p. 1014)

Notes for question set:[1]

[1] PathMD strives for the highest quality and accuracy. However, the *PathMD: Board Review Letter* is for review purposes and not meant for clinical decision making. It should not be used in place of review of primary reference texts and the current medical literature. If inaccuracies are identified, please notify us so that a correction may be published. (info@PathMD.com)