PathMD™: Board Review Letter

Author: Philip Ferguson, M.D. Renal Medical - Part 3 Volume 1, Number 29

1. Which of the following is most associated with an increase in minimal change disease?

    D. Hodgkin's disease

Answer: D. Hodgkin's lymphoma is associated with an increased incidence of minimal change disease. (p. 981) The association of minimal change disease is also associated with other leukemias and lymphomas, but less frequently.

2. This lesion can easily be overlooked on renal biopsy, especially if there are too few glomeruli for evaluation. It may also be mistaken initially for minimal change disease. Which of the following diagnoses best fits this description?

    B. Focal Segmental Glomerulosclerosis

Answer: B. FSGS is the most common cause of nephrotic syndrome and adults in the United States. It can be difficult to separate from minimal change disease if a limited number of glomeruli are present on the biopsy. Just as in minimal change disease, immunofluorescence is negative and electron microscopy shows effacement of foot processes. (Kumar, p. 982-983)

3. This disease is characterized by nephritis which progresses to chronic renal failure in patients who also have nerve deafness and various eye disorders. The hereditary pattern is X-linked.

    C. Alport's syndrome

Answer: C. Alport's syndrome is characterized by nephritis, which progresses to chronic renal failure, and also is associated with various eye disorders. This d/o follows an X-linked inheritance pattern with males expressing the full syndrome, and females usually only having hematuria (Kumar, p. 988)

4. This disease is characterized by familial asymptomatic hematuria, which is usually found incidentally. Renal biopsy shows a glomerulobasement membrane 150-250 nm in thickness. The best diagnosis is:

    D. Thin Basement Membrane Disease

Answer: D. Thin basement membrane disease. Normal glomerulo-basement membrane thickness is 300-450 nm in adults. There may be mild to moderate proteinuria in these patients, but the overall prognosis in these patients is excellent. (Kumar, p.988-989)

5. This syndrome is characterized by skin lesions, usually involving the extensor surfaces of the arms and legs, abdominal pain (nausea, vomiting, GI bleeding), and joint pain in addition to renal pathology. The renal manifestations includes nephrotic syndrome. Renal biopsy shows IgA deposition in the mesangium. The skin lesions are noted to have a necrotizing vasculitis in the small vessels. The best diagnosis is:

    D. Henoch-Schonlein purpura

Answer: D. Henoch-Schonlein purpura is a syndrome which includes skin lesions on the extensor surfaces in addition to abdominal manifestations, joint pain and renal abnormalities. This syndrome is closely related to IgA nephropathy and many believe that they are a spectrum of the same disease (Kumar, p. 990)

Cases Associated Website Images

Case #1 After undergoing a renal transplant, a 32-year-old is found to have pyelonephritis. A renal biopsy is performed and represented images including a special immunohistochemistry stain are shown. Based on the history and histologic findings, what virus is immunohistochemistry stain specific for?

B. Polyoma Virus

Answer: B. Polyoma virus can cause pyelonephritis in allograft recipients. It is been known to cause allograft failure in 1-5% of renal transplant patients. Histologically, polyoma virus characterized by nuclear enlargement and intranuclear inclusions visible by light microscopy. In the images for this case, they have a somewhat ground glass appearance. Just as in CMV, polyoma virus has immunohistochemistry stains available for detection. This immunohistochemistry stain is illustrated in the last image for this case. Treatment includes reduction of immunosuppression. (Kumar, p. 1000)

Case #2 A 60-year-old man with hypercalcemia is found to have a tubulointerstitial nephritis. An image of the immunofluorescence for kappa is shown. The immunofluorescence highlights like chain deposition in the glomerular basement membranes and tubular basement membranes. Based on these findings what is the patient's most likely diagnosis?

C. Multiple Myeloma

Answer: C. this case represents light chain deposition disease and is one of the representative pathologies in the kidney that patients with multiple myeloma may have. The important multiple myeloma pathologies of the kidney to know are: cast nephropathy induced by Bence-Jones proteins, amyloidosis, light chain deposition disease, and hypercalcemia/hyperuricemia (nephrocalcinosis). (Kumar, p. 1004-1006)

Case #3 A HIV infected patient presents with nephrotic syndrome. A renal biopsy is performed and representative light microscopic images are shown. Low-power examination shows focal cystic dilation of tubule segments and scattered inflammation. Jones stain highlights the glomerular pathology. Electron microscopy (not shown) shows tubuloreticular inclusions within the endothelial cells. Based on these findings, what is the most likely diagnosis?

E. Focal Segmental Glomerulosclerosis-Collapsing Variant

Answer: E. This case is the collapsing variant of focal segmental glomerulosclerosis which is associated with up to 5-10% of HIV-infected patients. It is characterized by focal dilation of tubules, inflammation, and fibrosis on light microscopy. Electron microscopy may show numerous tubuloreticular inclusions within endothelial cells. In this case the Jones stain highlights the collapsing glomerular structures. When the presence of tubuloreticular inclusions are noted on electron microscopy this should raise suspicion for HIV and SLE. (Kumar, p. 982-984)

Case #4 A 60-year-old patient with multiple myeloma presents with renal failure. A renal biopsy is performed, and represented images are shown. Based on the findings, what is the best diagnosis?

D. Nephrocalcinosis

Answer: D. This case represents nephrocalcinosis. It is characterized by calcium deposits usually underneath the tubular epithelium. Amyloidosis is characterized by hyalinized pink material which has apple green birefringence with Congo red staining. Cast nephropathy has intratubular collections of Bence-Jones proteins and have a varying pink to blue amorphous appearance. Light chain deposition disease can only be characterized by immunofluorescence. (Kumar, p. 1005-1006)

Case #5 A biopsy from a 44 y/o patient with a significant past medical history shows nodular glomerulosclerosis in which the nodules in the image (trichrome stain) for this case are also PAS positive within the glomerular lobules. The surrounding capillary loops are patent. These features are most characteristic of:

B. Kimmelstiel-Wilson Nodules

Answer: B. This is an example of Kimmelstiel-Wilson nodules which is a feature of diabetic glomerulosclerosis. (Kumar, p. 990-993)

References:

Robins and Cotran: Pathologic Basis of Disease. Kumar, V. et al., 7th Edition. 2005.

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)

