Focal Segmental Glomerulosclerosis

Focal segmental glomerulosclerosis (FSGS) is the most common cause of nephrotic syndrome in Hispanics and African Americans. This disease can occur as primary disease but also occurs in association with other conditions including sickle cell disease, HIV infection, interferon therapy and heroin abuse. Microscopically, as the name implies, lesions are characterized by sclerosis of some but not all of the glomeruli, making the inflammation focal as opposed to diffuse. Additionally, the lesions within the glomeruli only affect a section of the capillary making the damage segmental. On light microscopy, the glomeruli that are affected demonstrate segmental sclerosis and hyalinosis while the unaffected glomeruli appear relatively normal. Electron microscopy demonstrates focal damage of visceral epithelial cells, while immunofluorescence staining is usually negative. Clinically, FSGS typically has a high incidence of microscopic hematuria, reduced GFR, hypertension, nonselective proteinuria, and poor response to corticosteroid therapy. This disease has a relatively poor prognosis with many individuals progressing to chronic kidney disease and more than half of the patients developing end-stage kidney disease in 10 years.

PLAY PICMONIC

Most Common Cause of Nephrotic Syndrome in Hispanic People and African Americans
#1 Foam-finger on Nerd-frog Hispanic and African American
Nephrotic syndrome is a group of symptoms including massive proteinuria defined as a daily loss of 3.5 gm or more of protein, hyperlipidemia, generalized edema, and hypoalbuminemia which results from renal pathology. Nephrotic syndrome is caused by several diseases including membranous glomerulonephritis, minimal change disease, and focal segmental glomerulosclerosis. FSGS is the most common cause of nephrotic syndrome in Hispanic peoples and African Americans.

Associated with Sickle Cell Disease and HIV
Sickle and HIV Band-AID
Primary FSGS is idiopathic but the secondary form of the disease may occur in association with other conditions such as HIV or sickle cell disease.

Associated Interferon Therapy and Heroin Abuse
Interferon-ray-gun and Heroin Abuser
Primary FSGS is idiopathic but the secondary form of the disease may occur in association with heroin abuse or interferon therapy.

Histopathology

LM Segmental Sclerosis and Hyalinosis
(LM) Light-bulb over Segway skull-roses with Highlighter
On light microscopy (LM), the glomeruli that are affected demonstrate segmental sclerosis and hyalinosis while the unaffected glomeruli appear relatively normal.

EM Focal Damage of Visceral Epithelial Cells
Electrons in an Atom and Visor with Epithelial Cell
Electron microscopy (EM) shows focal damage of visceral epithelial cells. This epithelial damage is a hallmark of FSGS. Furthermore, EM shows effacement of foot processes, similar to minimal change disease.

IF Negative
Negative IF-glowsticks
FSGS generally appears negative on immunofluorescence microscopy (IF) because the focal and segmental lesions may involve only a minority of the glomeruli. However, IgM, C1, and C3 may be present in the sclerotic areas and or in the mesangium.

Signs & Symptoms

Microscopic Hematuria
Microscope in Red-urinal
Small amounts of blood can be seen on urinalysis.

Nonselective Proteinuria

Protein-urinal

The proteinuria includes all blood proteins including clotting factors, albumin, and immunoglobulin.

Considerations

Poor Prognosis

Gravestone

This disease has a relatively poor prognosis with many individuals progressing to chronic kidney disease, with more than half of the patients developing end-stage kidney disease in 10 years.