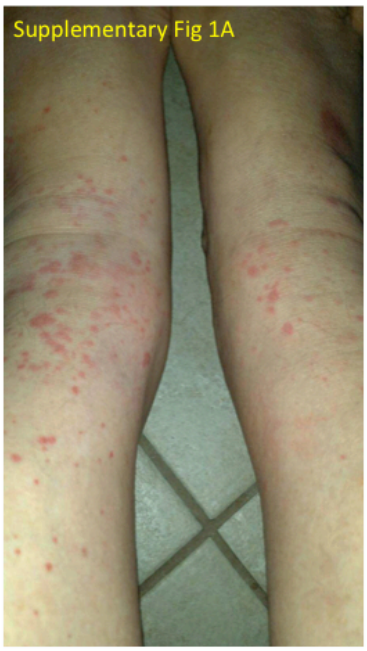
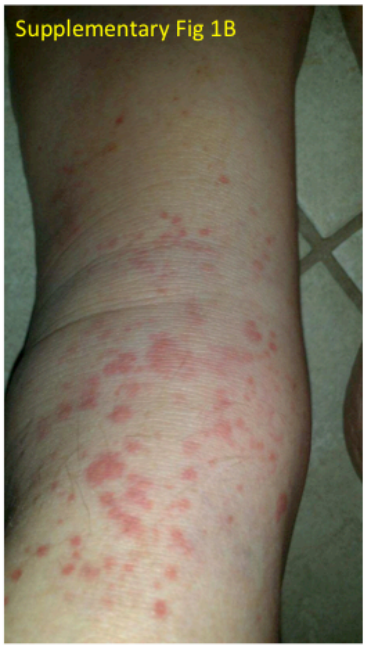
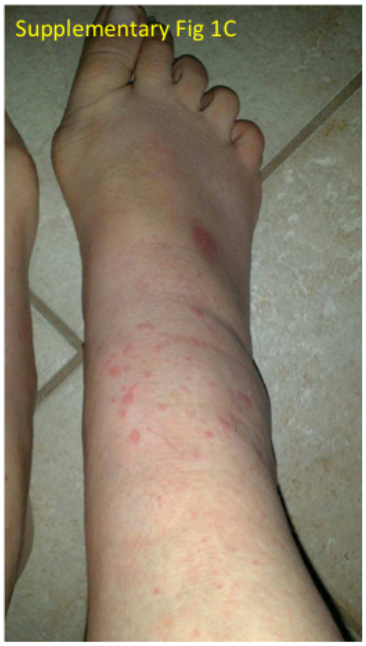
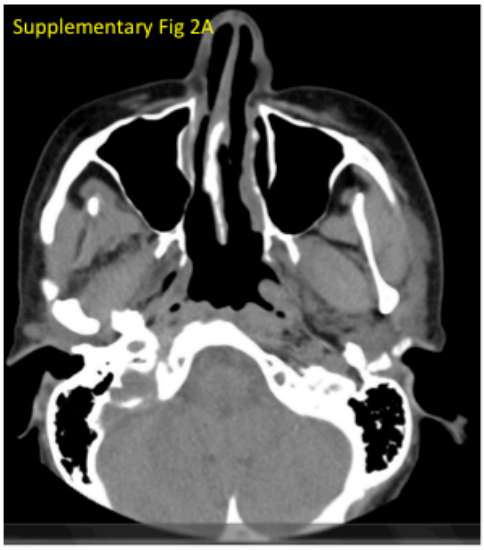
Supplementary Figures and Tables:

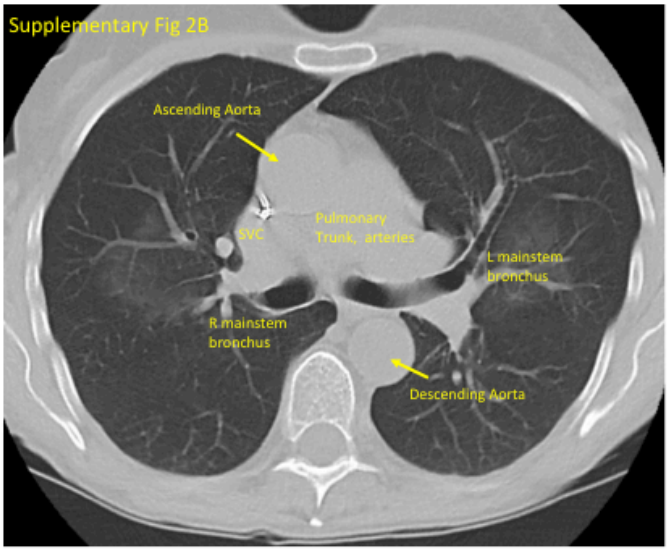






Supplementary Figure 1 (A,B): Leukocytoclastic Vasculitis rash lasting 3 weeks one month prior to hospital admission (2017).
Supplementary Figure 1C: Peripheral edema





Supplementary Figure 2A: CT sinus without contrast: clear paranasal sinuses, well aerated. No evidence of sinusitis.
Supplementary Figure 2B: CT chest without contrast: no evidence of mediastinal lymphadenopathy or mass. Lungs demonstrated patchy areas of interstitial infiltrates which may be inflammatory (vasculitis) or may represent edema. (SVC: Superior Vena Cava)

Supplementary Table 1: Pathology Report: 2007 Kidney Biopsy

A: Surgical Pathology Report 2007

Microscopic Description:

- approximately 7 glomeruli are present, 4 of the glomeruli are obsolescent. The non-obsolescent glomeruli show areas of segmental sclerosis. Foam cells are also present. A PAS stain was performed.

- Light microscopic changes suggestive of focal sclerosing glomerulopathy. Full diagnosis deferred, pending special outside studies

B: External Pathology Consultation:

- light microscopic examination is performed with H and E and PAS stained slides. 3 glomeruli and 4 small dense glomerular scars are identified. 2 of the glomeruli are abnormal with greater than 50% degree of segmental sclerosis with adhesions onto Bowman's capsule. No cellular proliferative changes are present and no foci of necrosis, thrombosis or crescent formation.

There is no tubular necrosis. There are few foci of inactive chronic interstitial inflammatory infiltrate. No granulomata, neoplasm or atypical cellular infiltrates are present

Immunofluorescence:

-the frozen sections onctain 3 glomeruli and 5 glomerular scars. This study was negative including IgG, IgA, C3, C1q, kappa and lambda

Electron microscopy:

- At the ultrastructural level, 3 glomeruli were examined. One glomerulus had early segmental collapse and sclerosis of capillaries. In particular, no electron dense deposits of the type associated with entrapment of immune complexes were identified and no fibrillary deposit of amyloid were present.

EM diagnosis:

- kidney core needle biopsy: focal segmental sclerosing glomerulopathy

Supplementary Table 2: Pathology Report: 2017 Kidney Biopsy

Light Microscopy:

- Sections stained with H&E, PAS and silver methenamine reveal 3 portions of renal cortical tissue, 1 with capsule and 1 portion of renal cortico-medullary tissue.

- Glomeruli: On serial sections, at least 12 glomeruli are present, 9 of which are globally sclerotic. The remaining glomeruli show mesangial normocellularity. One glomerulus contain cellular crescent with segmental, small fibrinoid necrosis. Another 2 glomeruli show segmental scarring with cellular crescent. Fibro-crescent is seen in globally sclerotic glomeruli.

- Tubules and Interstitium: The tubular atrophy and interstitial fibrosis are severe. Overall tubular loss estimated to be 45-55% of the renal cortex sampled. There is moderate interstitial inflammation with mainly lymphocytes and some mononuclear cells in areas of tubular atrophy and interstitial fibrosis.

- Erythrocyte casts are seen in some proximal and distal tubules.

- Vessels: Arteries up to intra-lobular size are present and unremarkable. No active arteritis seen.

Immunofluorescence:

- The specimen processed for immunofluorescence consists of 1 portion of renal cortical tissue with at least 15 glomeruli, 13 of which are globally sclerotic.

- By immunofluorescence, there is diffuse, granular mesangial staining for IgM (3+). There is diffuse, pseudolinear glomerular capillary loops staining for albumin (1 +). No specific glomerular staining for IgG, IgA, C3, C1q, kappa light chain or lambda light chain is noted.

- Tubular casts stain lor IgA (3+), kappa light chain (3+) and lambda light chain (3+).

- The negative control is negative.

Electron Microscopy:

- Electron microscopic examination was performed at UF Health Pathology Laboratories, Gainesville FL.

- Tissue submitted for electron microscopy consists ot 1 portion of renal cortical tissue with 1 glomerulus, which shows normal mesangial cellularity in 1 micron section.

- No cellular crescent or necrosis is seen.

- By EM, the glomerular basement membranes show normal trilaminar structure and mean thickness within normal range. No immune deposit or tubule-reticular inclusion is identified.

- The majority of podocyte foot processes remains intact. Some vacuoles are noted in proximal tubules.

- Atrophic tubules are also seen along with interstitial fibrosis.

Note: In present biopsy, there are severe tubular atrophy and interstitial fibrosis, along with diffuse global glomerulosclerosis. The remaining glomeruli still shows cellular crescent, 1 of which also has segmental small fibrinoid necrosis.

No significant immune complex is detected by immunofluorescence.

In views of clinical presentation with ANCA positive, these findings are consistent with pauci-immune crescentic necrotizing glomerulonephritis.

FINAL DIAGNOSIS

-Pauci-immune crescentic necrotizing glomerulonephritis

-Severe tubular atrophy and interstitial fibrosis.