Rapidly Progresive Glomerulonephritis

Just like acute glomerulonephritis, but in fast forward: rapid decline in renal function , and subsequent end-stage renal failure within days or weeks.
LUCKILY ITS RARE. 2 to 4% of GN are rapidly progressive.

PATHOLOGICAL HALLMARKS:
Cellular crescents
surounding the glomeruli.
- these are made of endothelial cells, mononuclear infiltrate and recruited fibroblasts.

ALSO:
- linear deposition of immunoglobulins all along the GBM in 20%
- granular (blobby) deposition of these Ig's in the GBM in 30%.
- In the remainder of pts, no immune deposits of any sort are detectable.

Natural History

- **INSIDIOUS ONSET:**
  - **Malaise, lethargy, microscopic hematuria**
  - **Proteinuria in ~30% of patients**
- **KNOW TO LOOK FOR RARE DISEASES KNOWN TO BE ASSOCIATED WITH RAPIDLY PROGRESIVE GN:**
  - **a VASCULITIS of some sort, be it**
    - **WEGENER'S GRANULOMATOSIS,**
    - **MICROSCOPIC POLYANGIITIS, or**
    - **CHURG-STRAUSS SYNDROME**
  - **CRYOGLOBULINAEMIA**
  - **SYSTEMIC LUPUS ERYTHEMATOSUS**
  - **GLOMERULAR BASEMENT MEMBRANE ANTIBODIES**
  - **GOODPASTURE'S SYNDROME (also haemoptysis)**

MANAGEMENT is AGGRESSIVE and DETERMINED.

Kick-start with **IV corticosteroids and cyclophosphamide**
Monitor progress: if response is limited move on to **PLASMA EXCHANGE**
(thats if you can identify an antibody as the culprit)

Renal survival is most closely related to serum creatinine titres at presentation.
Only 40% of patients escape dialysis at 1 year of follow-up.