COMPLEXO HOSPITALARIO UNIVERSITARIO
JUAN CANALEJO
A Coruña

SERVICIO GALEGO DE SAÚDE

Club Nefropatologia. Congreso SEAP. 29-31 Mayo.Madrid

Insuficiencia renal y proteinuria.

Resumen clínico:

Paciente de 37 años de edad que ingresa por IR y malestar general.

Antecedentes personales: Hipertensión intermitente sin tratamiento.

Enfermedad actual: Dolor cervical y en hombros y posterior afectación del estado general y edema facial.

Exploración física: T.A. 165/100. Afebril. Edema palpebral bilateral. Extremidades: No edemas.

Exploración-palpación abdominal : ausencia de masas y/o órganomegalias.Datos complementarios: 11.000 leucocitos con fórmula normal; hemoglobina 11,7 gr.;Hematocrito 37%; plaquetas 297.000; glucosa 82 mg.;ácidoúrico 10,8; Colesterol 201; Triglicéridos 660; Proteínas tot. 5.7Albúmina 3.8. Inmunoquímica: IgG 72; IgA 24; IgM 17; C3 141; C4 48.Crioglobulinas (-) ANAS (-) ANCAS (-) Albúmina 3.6Ganmaglobulinas 0.1; **Bence Jones Kappa 4.3 gr./24 h.**

Inmunofijación: **banda monoclonal cadena ligera Kappa**.Serología de hepatitis B, C y HIV (-)

Evolución y comentarios:

Se realizó biopsia renal al día siguiente de su ingreso con el resultado del estudio de la biopsia renal y ante el deterioro dela función renal se comenzó diálisis realizándose biopsia de médula ósea y plasmaféresis.

Se indicó ttmo. quimioterápico

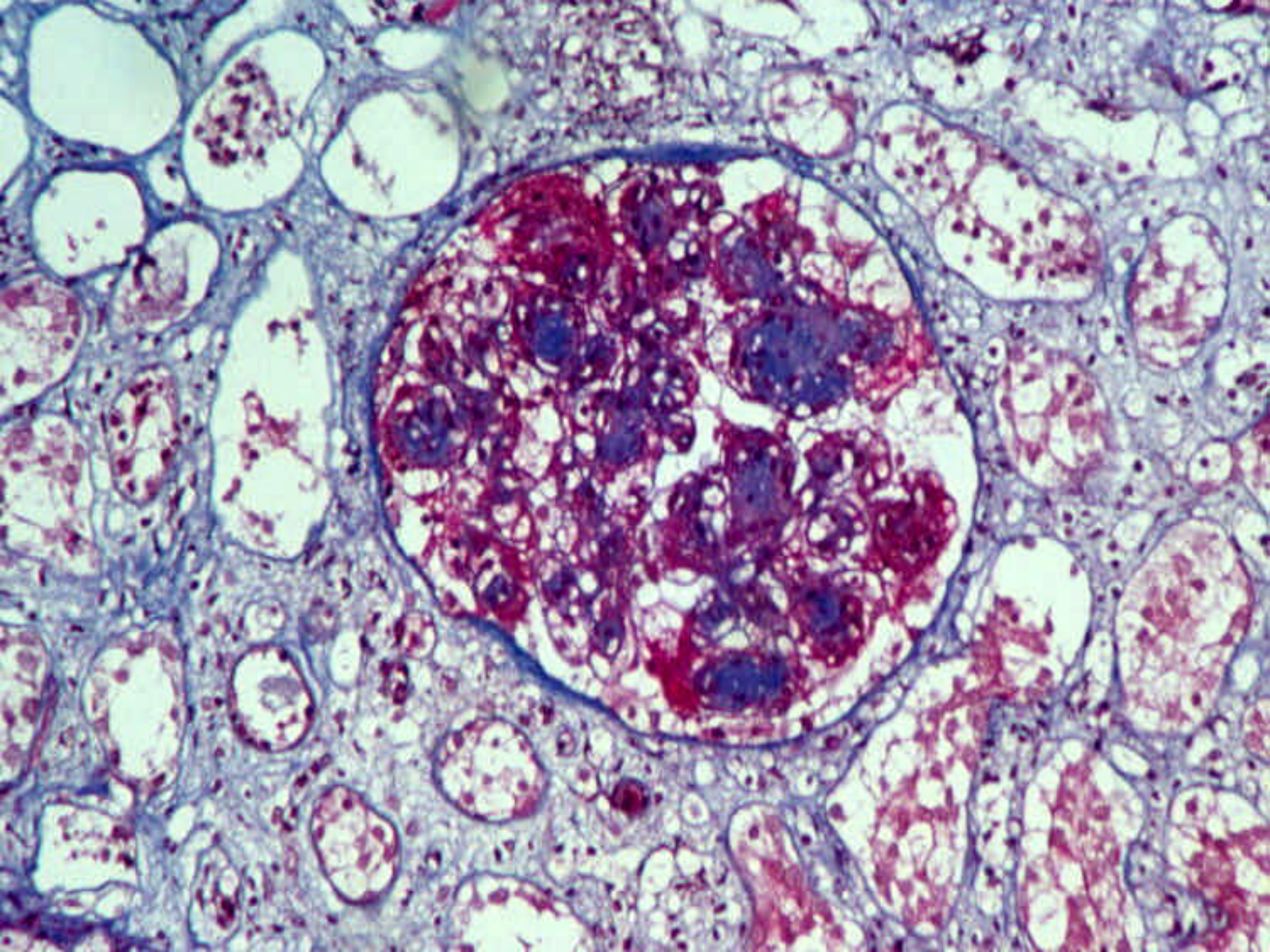
Función renal: Crp 3 mg./dl. Proteinuria 20 gr.

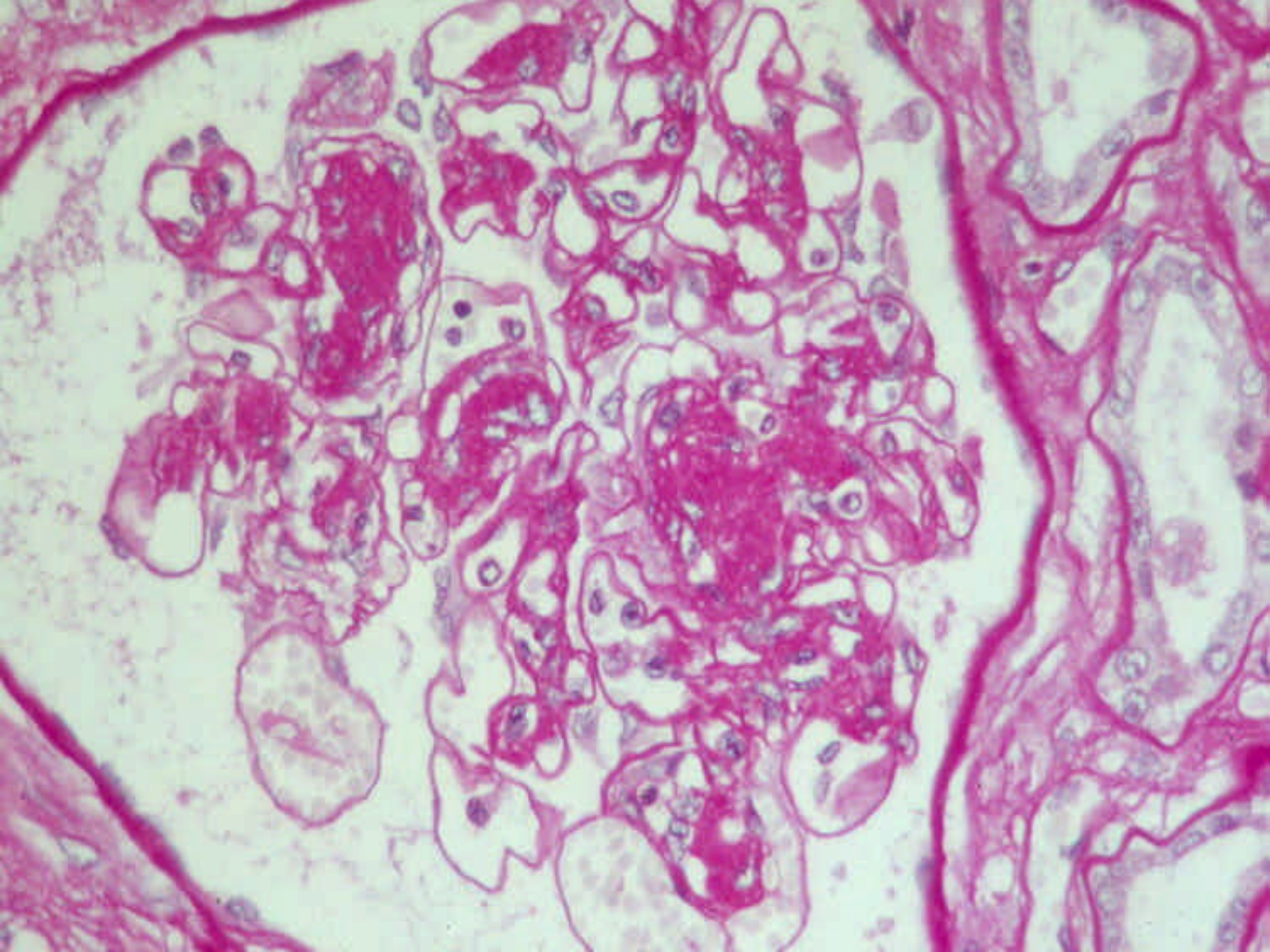
Sedimento 5-10 hem./campo.

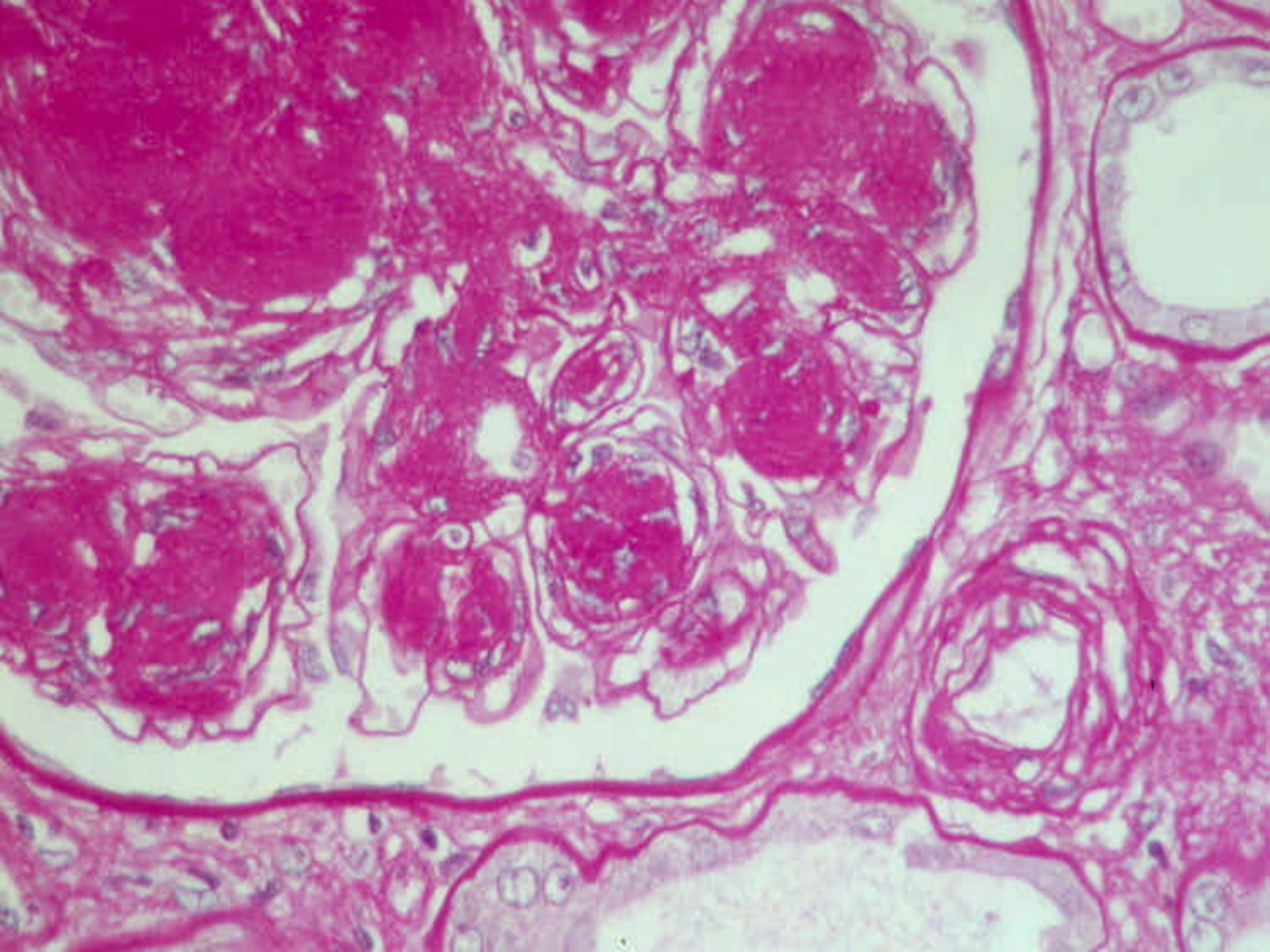
ECG: Hipertrofia ventricular izquierda.

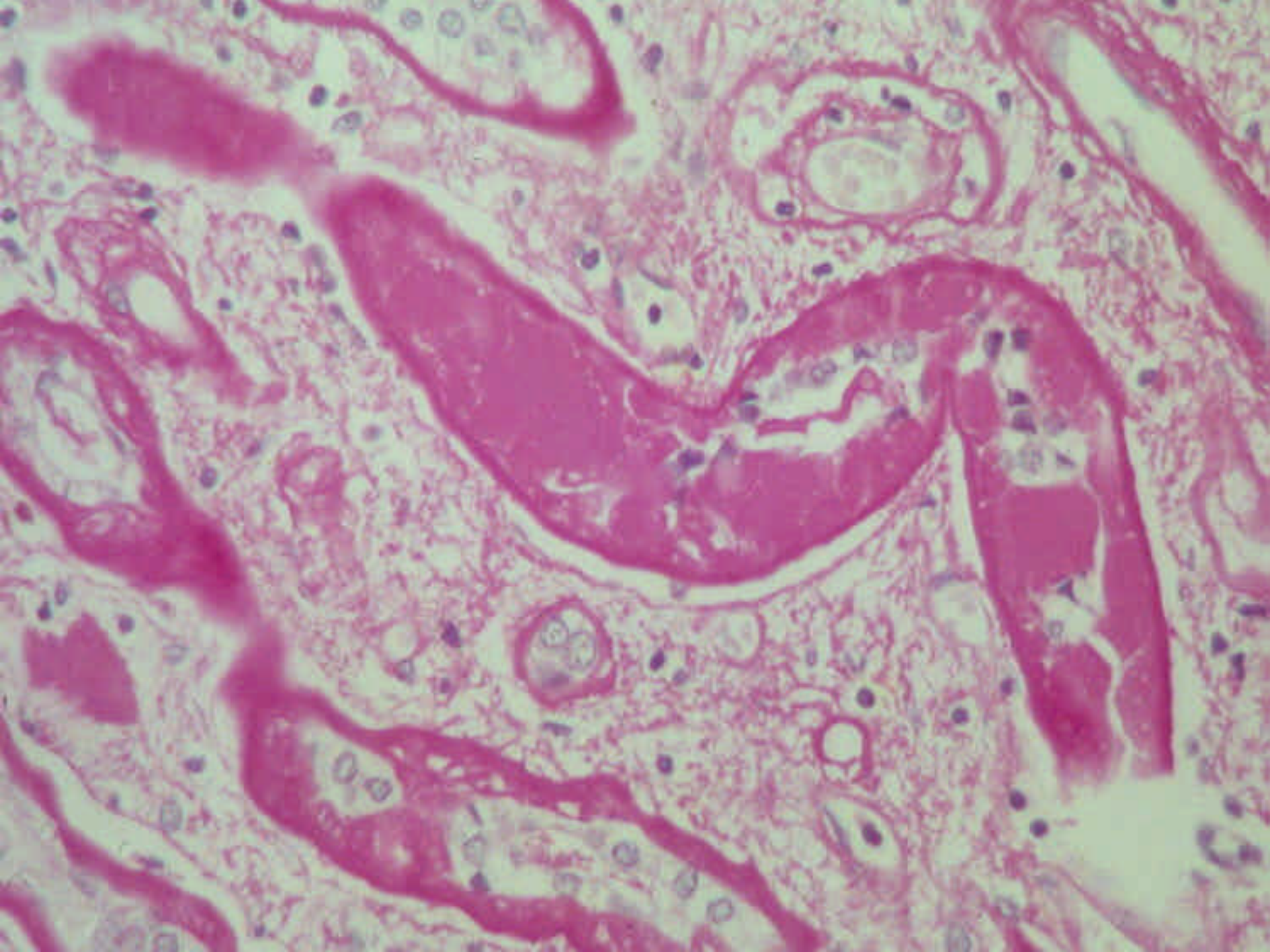
El paciente fallece por arritmia cardiaca y con insuficiencia renal.

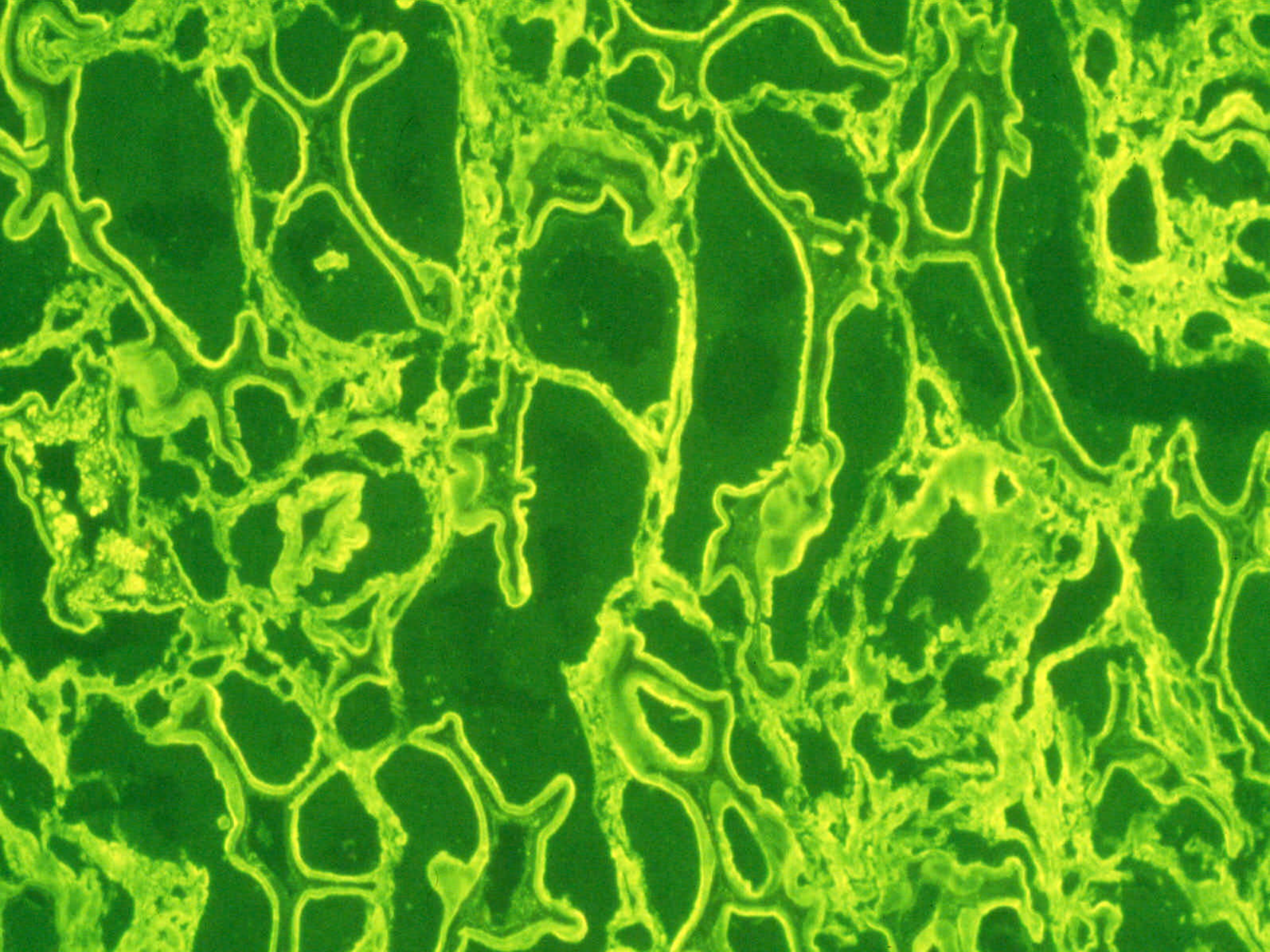
Hospital Universitario Juan Canalejo

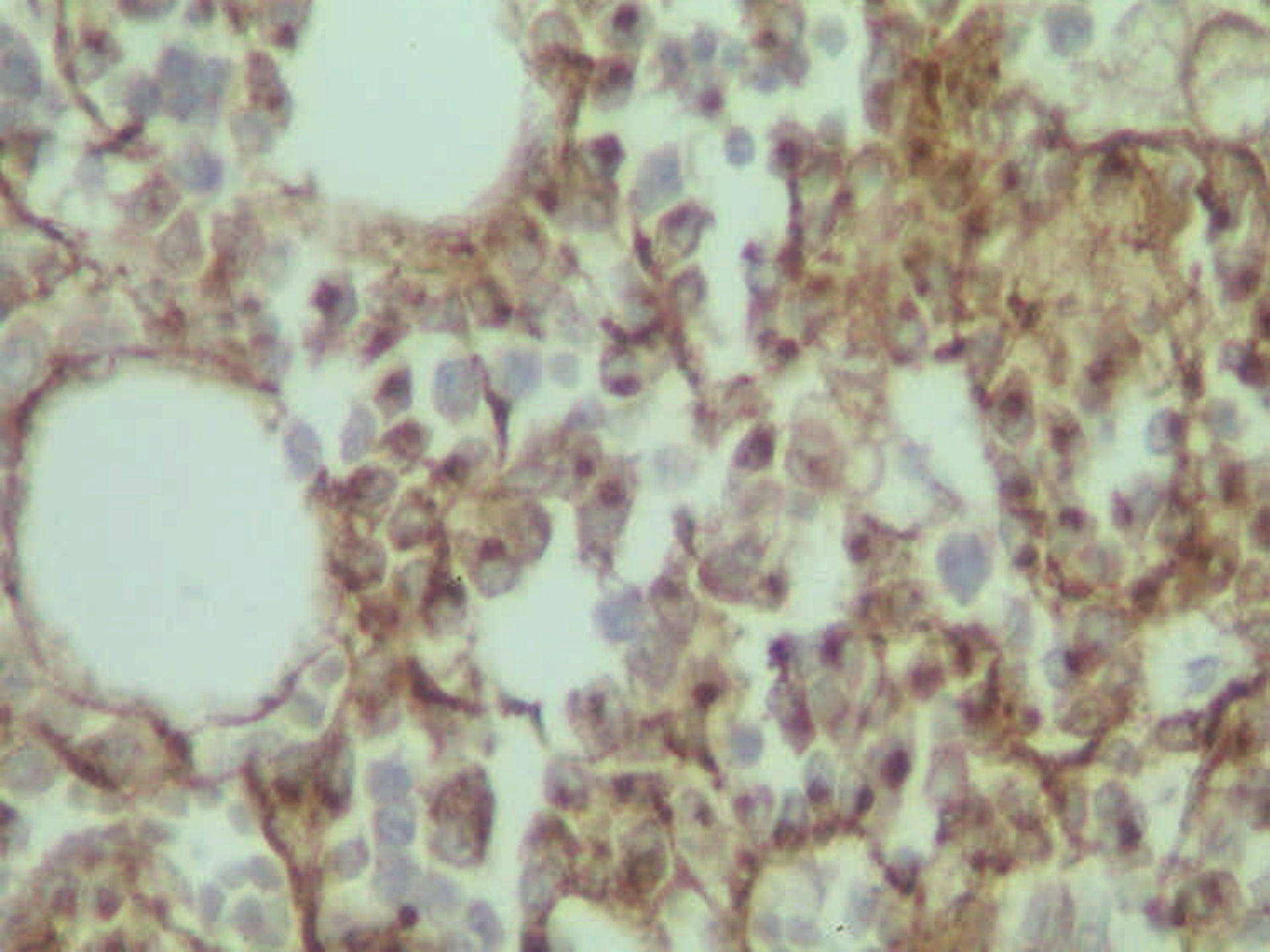


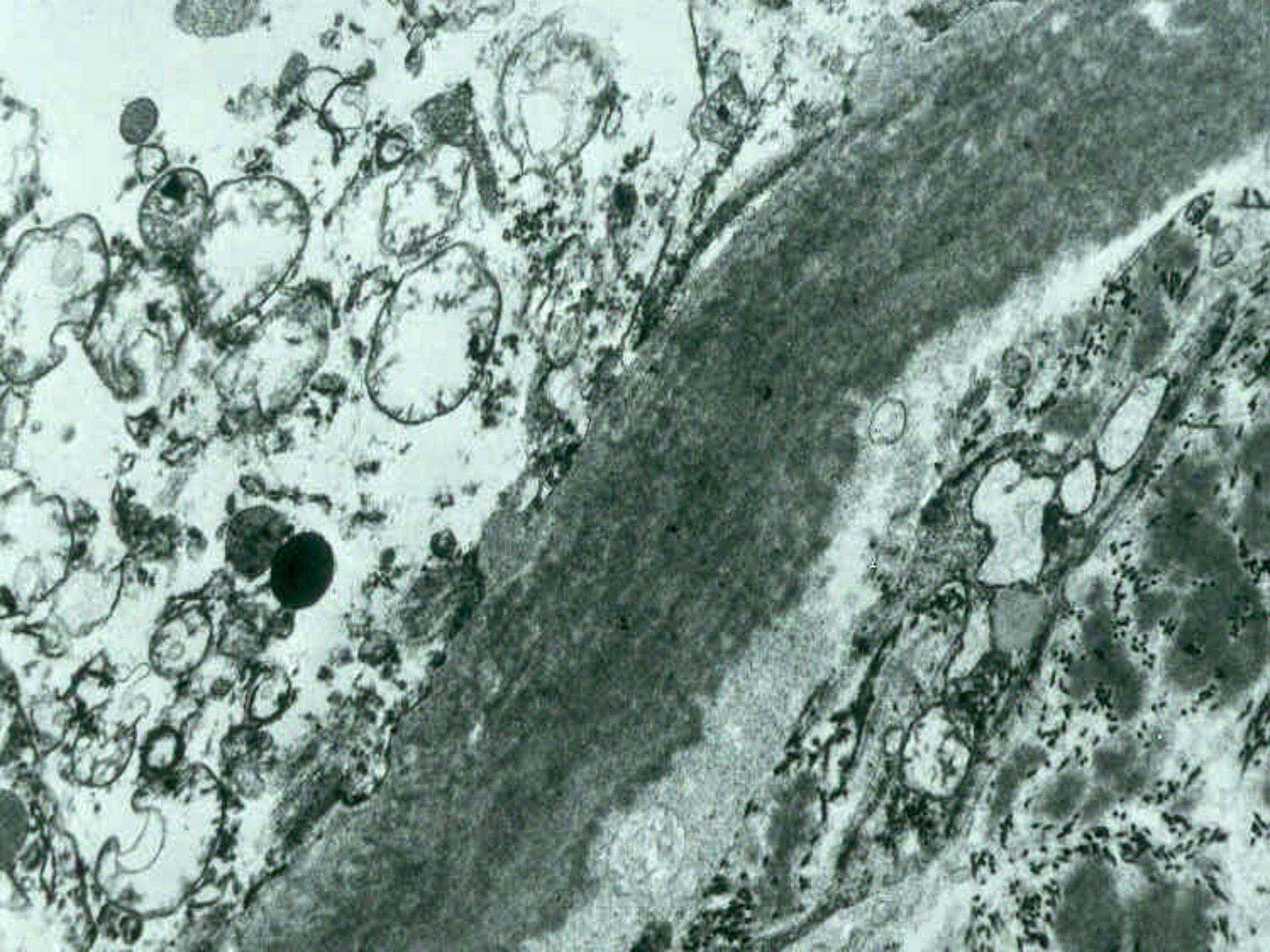












Enfermedades Disproteinémicas

- Se caracterizan por la proliferación clonal celulas plasmáticas o celularidad linfoide, que producen un excesivo incremento de una inmunoglobulina (paraproteina).
- La identificación de esta paraproteina en suero u orina así como en la MBG,MBT, es clave para el diagnóstico.

Enfermedad Renal y Disproteinemias

- Amiloidosis y variantes
- Enf. Cadenas ligeras (EDCL)
- Enf. Cadenas Pesadas
- Crioglobulinemias
- Gn. Fibrilar
- Gn. Inmunotactoide

Patologia renal y Disproteinemia

- **Amiloidosis AL**
- **Enfermedad Depósitos Cadenas Ligeras (EDCL)**
- **Tubulopatía del mieloma**
- **Enfermedad con Depósitos Cadenas Pesadas**
- **Gn Crioglobulinémica: Gn MGC**
- **Gn macroglobulinemia de Waldenstrom**
- **Sindrome de Fanconi Y EDCL**

Enfermedad Renal Fibrilar No Amiloidótica

- Enf. Monoclonal de cadenas pesadas
- Enf. Monoclonal de cadenas ligeras
- Enf. Monoclonal mixta
- Enf. Policlonal: LES,crioglobulinemias

Enferm. Renal y Disproteinemias. Afectación renal

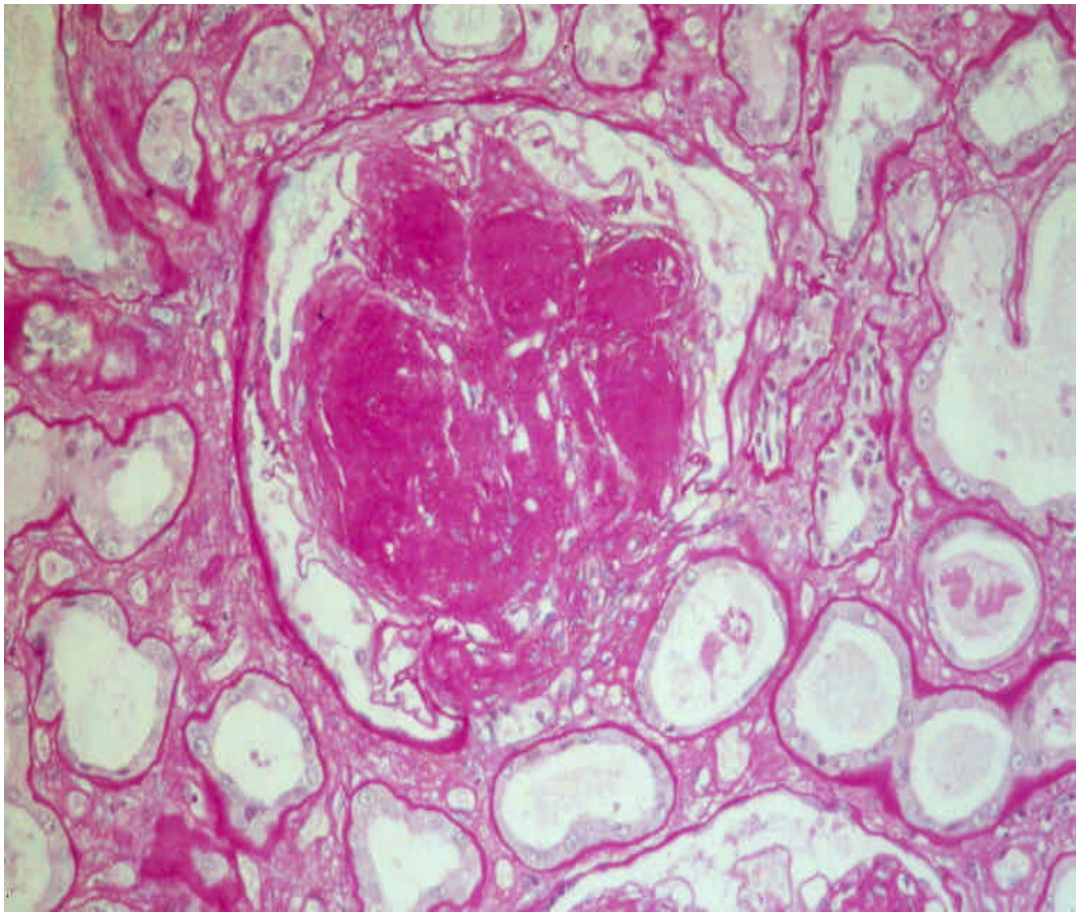
- **Afectación glomerular**.: Glom Nodular

  Variantes de GNMGC
  Gn Focal
  Gn mesangial

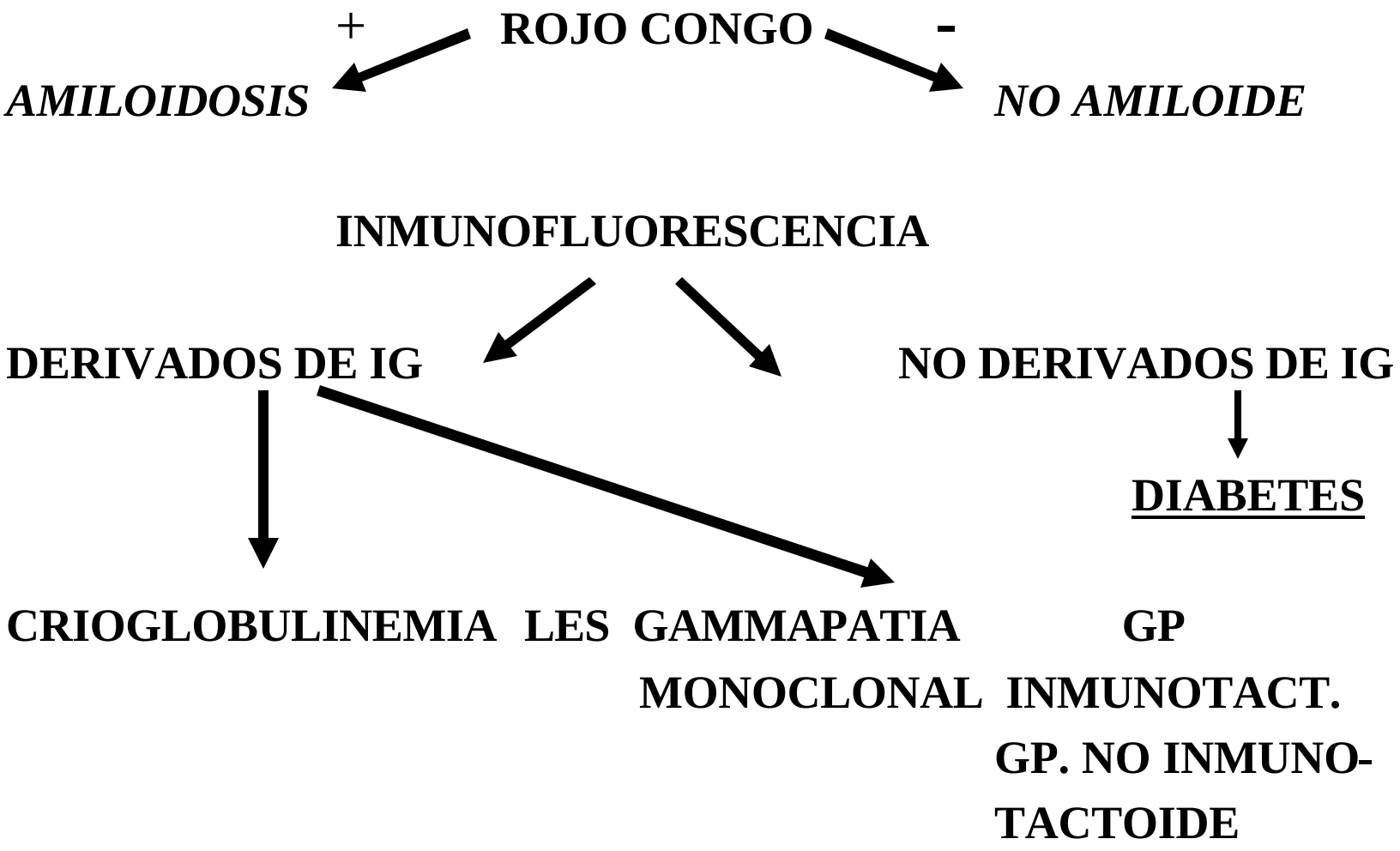
- **Afectación Tubular**: tubulopatia del mieloma

Enfermedad por Depositos Cadenas Ligeras

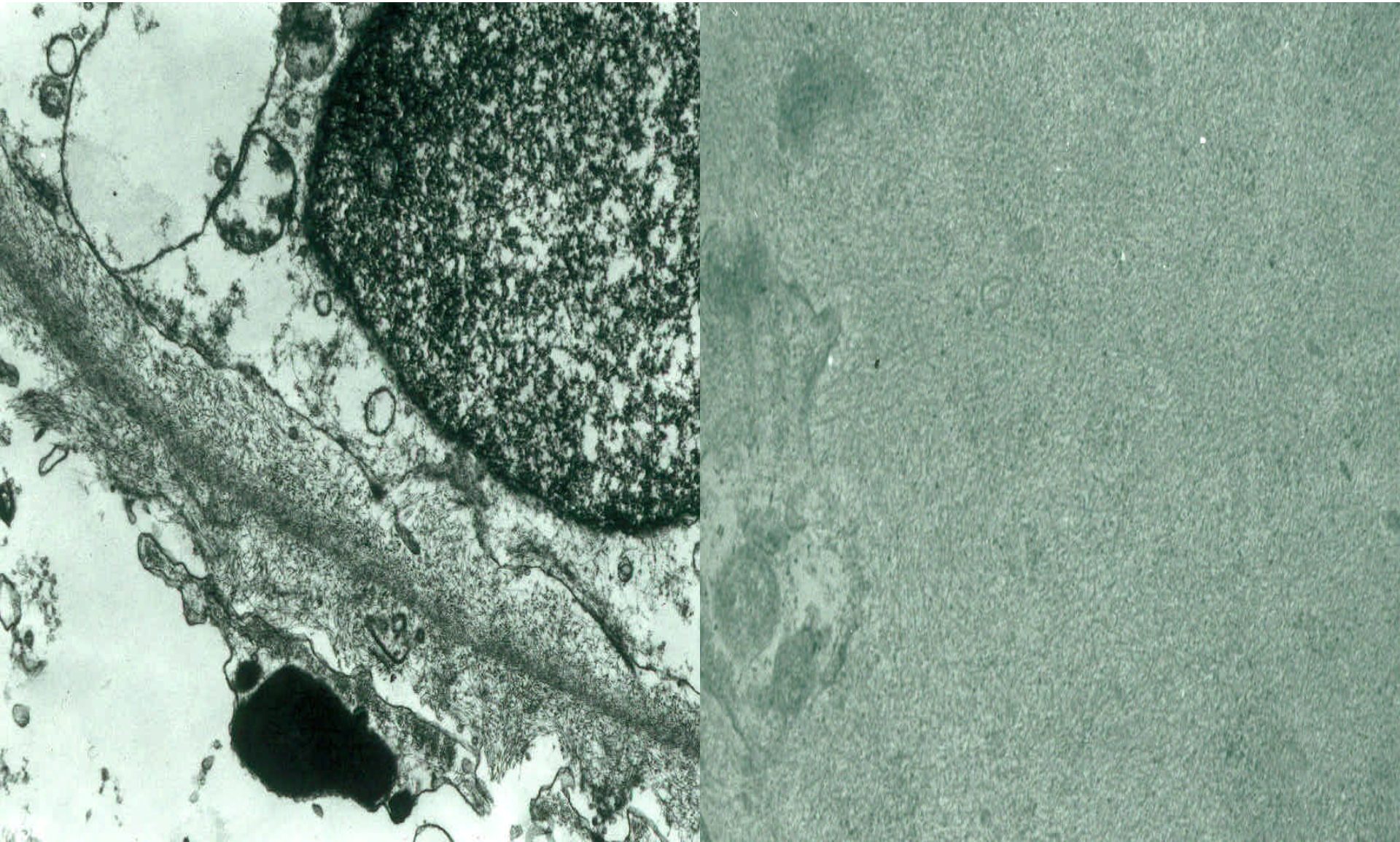
- Afectación renal 90%
- Proteinuria 80%
- Microhematuria 20%



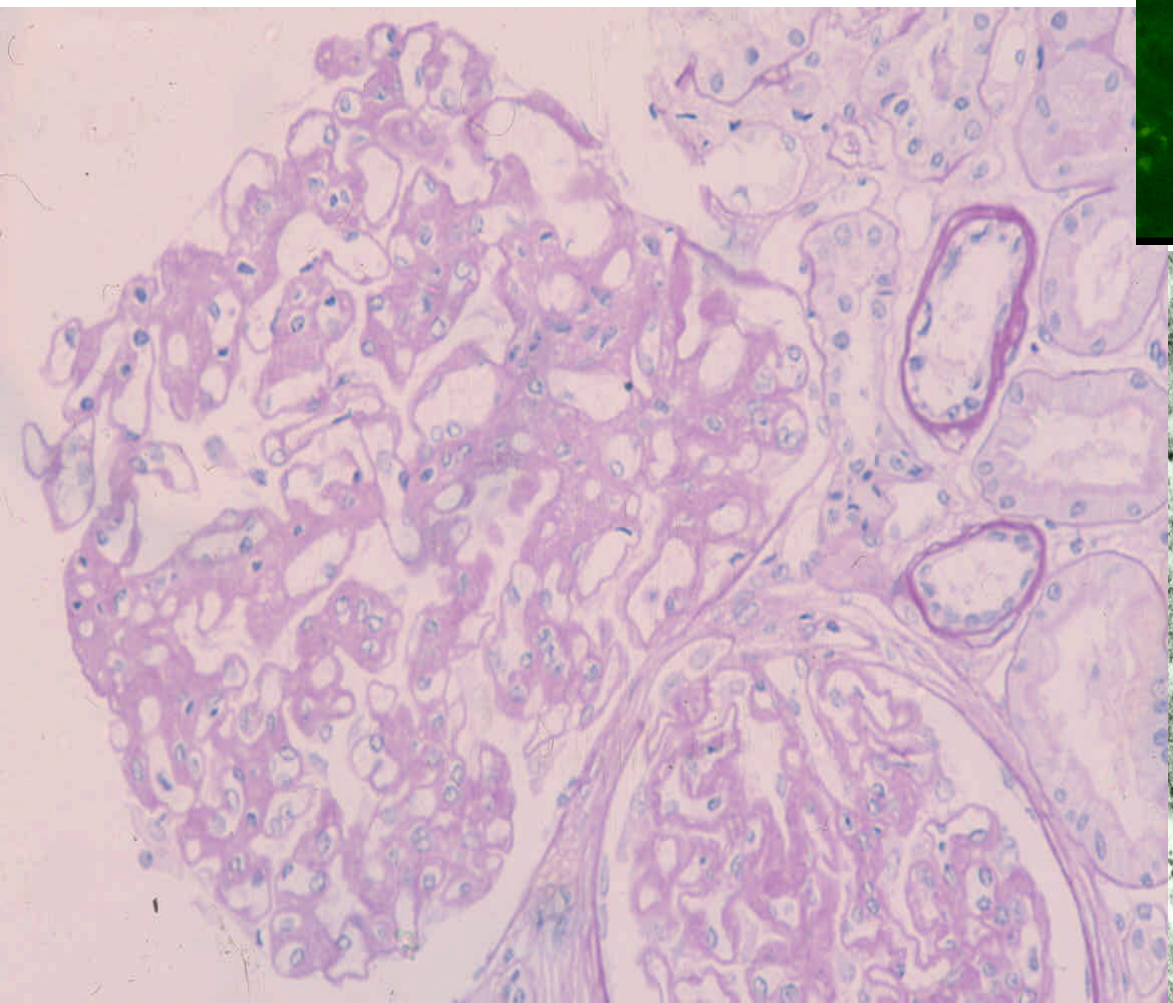
GLOMERULOPATÍAS FIBRILARES
METODOLOGIA DIAGNOSTICA

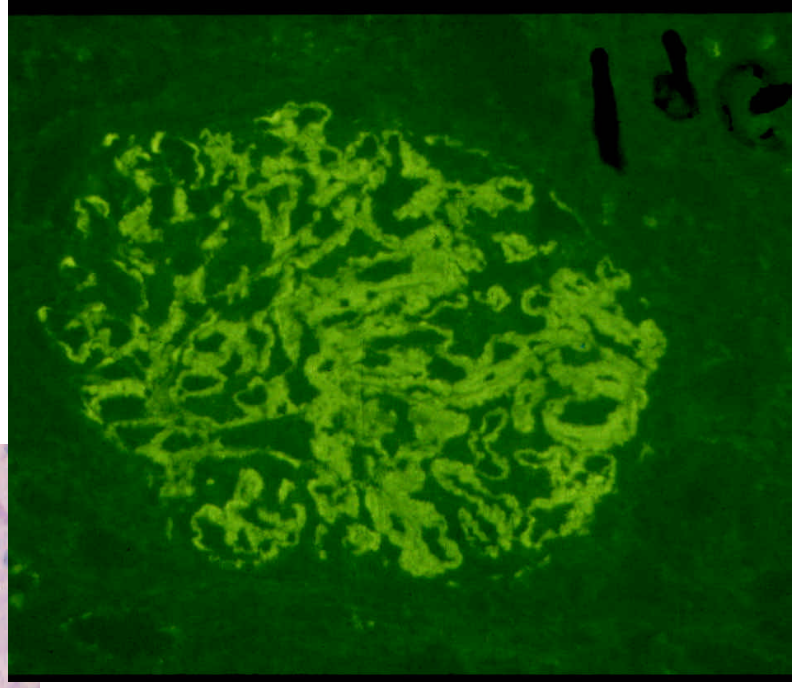


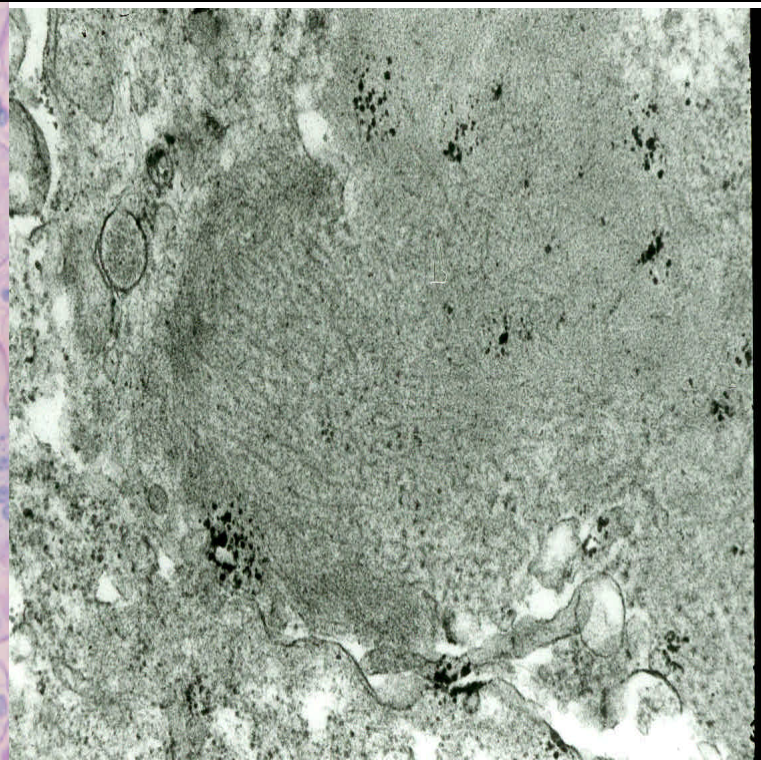
AMILOIDOSIS



GN FIBRILAR







Abbott KC. Agodoa LY. Multiple myeloma and **light chain**-associated **nephropathy** at end-stage renal **disease** in the United States: patient characteristics and survival. [Journal Article]
Clinical Nephrology. 56(3):207-10, 2001 Sep.

Abstract

AIMS: The patient characteristics and clinical course of **nephropathy** associated with multiple myeloma/**light chain disease** (MMN) has not been described for a national sample of end-stage renal **disease** patients. **METHODS:** 375,152 patients in the United States Renal Data System were initiated on ESRD therapy between January 1, 1992 and June 30, 1997, and were analyzed in a retrospective registry study of MMN (PDIS=2030A, 2030B, 2030Z, and 203Z). **RESULTS:** Of the study population, 3298 (0.88%) had MMN. Patients with MMN were disproportionately male (59.5% vs. 53.2%) and Caucasian (76.2% vs. 64.1%, $p < 0.01$ by Chi-square for both comparisons) and older (68.00+/-11.78 vs. 60.69+/-16.55 years, $p < 0.01$ by Student's t-test). In logistic regression analysis, patients with MMN were more likely male and Caucasian, were older,

In Cox regression, MMN was independently associated with decreased all-cause patient survival ($p < 0.01$, hazard ratio for mortality=2.52, 95% CI 2.38-2.67). **CONCLUSIONS:** MMN was associated with Caucasian race, male gender, and older age, compared with other ESRD patients. Patients with MMN had evidence of poorer medical condition on initiation of dialysis compared to other patients. MMN was associated with decreased patient survival after initiation of dialysis,

Br J Haematol 1999
Dec;107(4):835-43

pH-dependent fibrillogenesis of a VkappaIII Bence Jones protein.

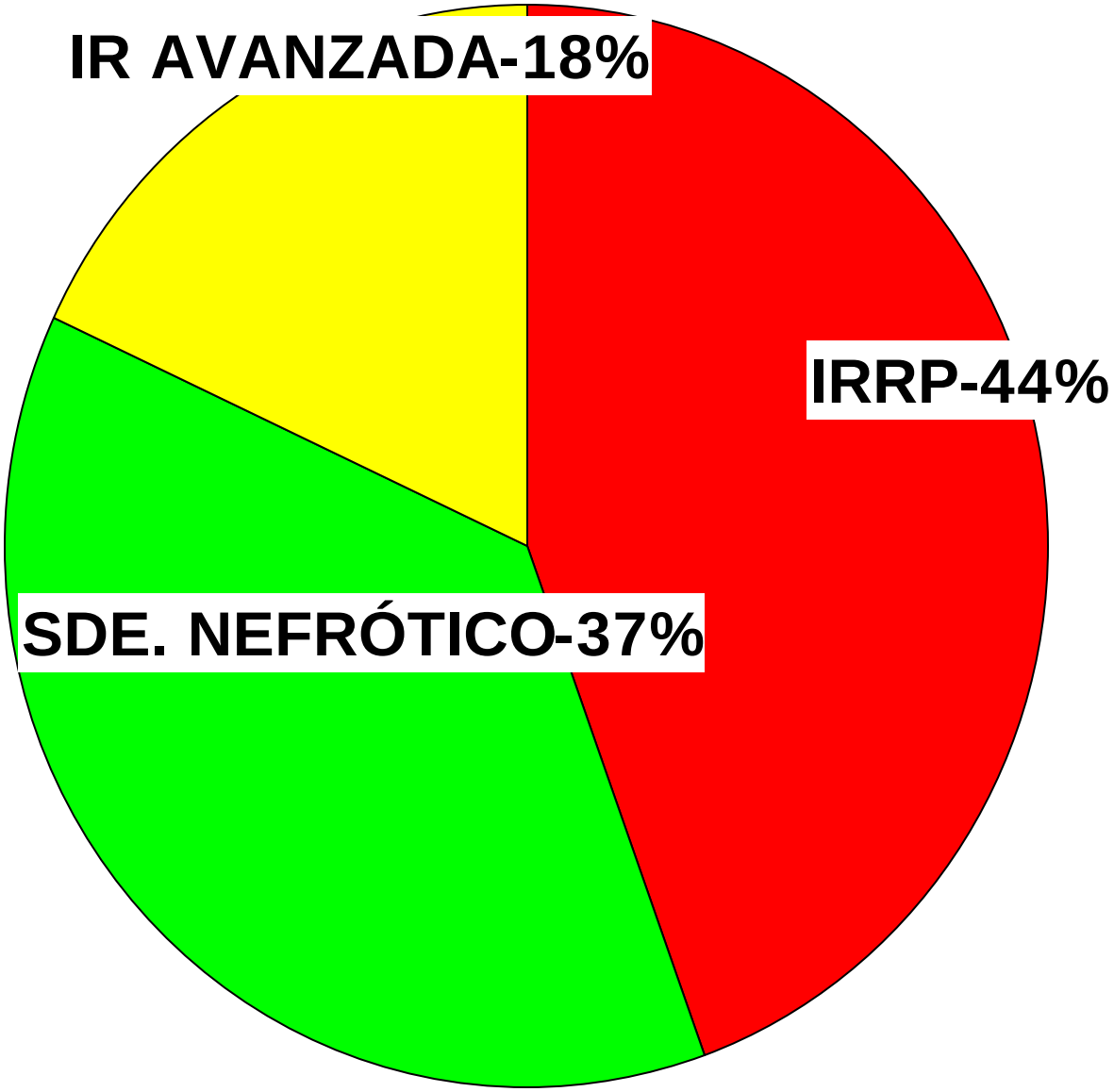
Rostagno A, Vidal R, Kaplan B, Chuba J, Kumar A, Elliott JI, Frangione B, Gallo G, Ghiso J.

Department of Pathology, New York University School of Medicine, New York, N.Y., USA.

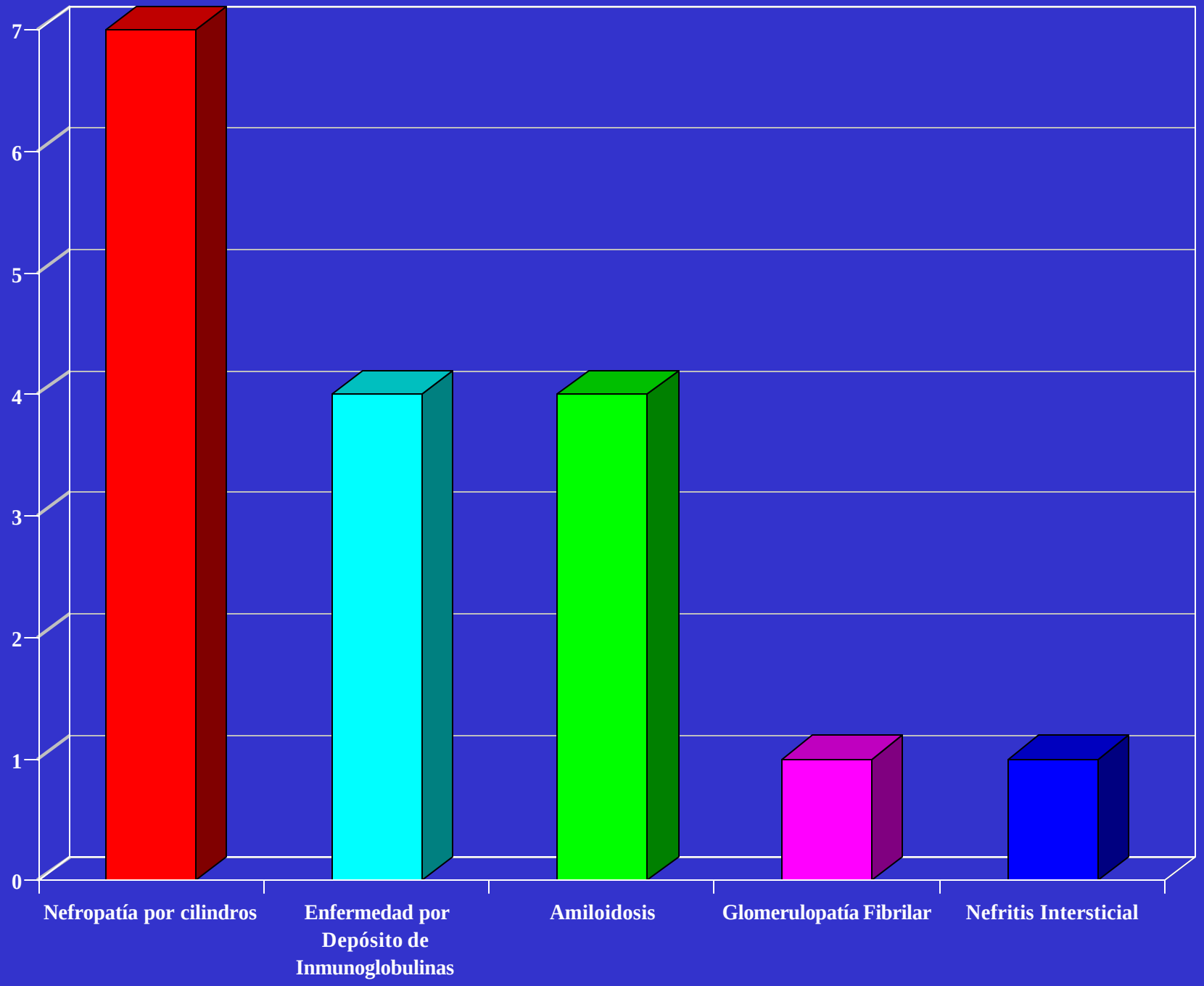
DISPROTEINEMIAS HJC-LA CORUÑA

- **17 pacientes**
- **8 mujeres y 9 hombres**
- **Bx m. o.:**
  - 11 mielomas
  - 6 normal
- **Bx renal**
  - Nefritis Intersticial 1
  - Nefropatía por Cilindros 5
  - Nef. Cilindros + EDIM kappa 2
  - Enf. por Depósito de Proteinas 9

Causas de biopsia renal

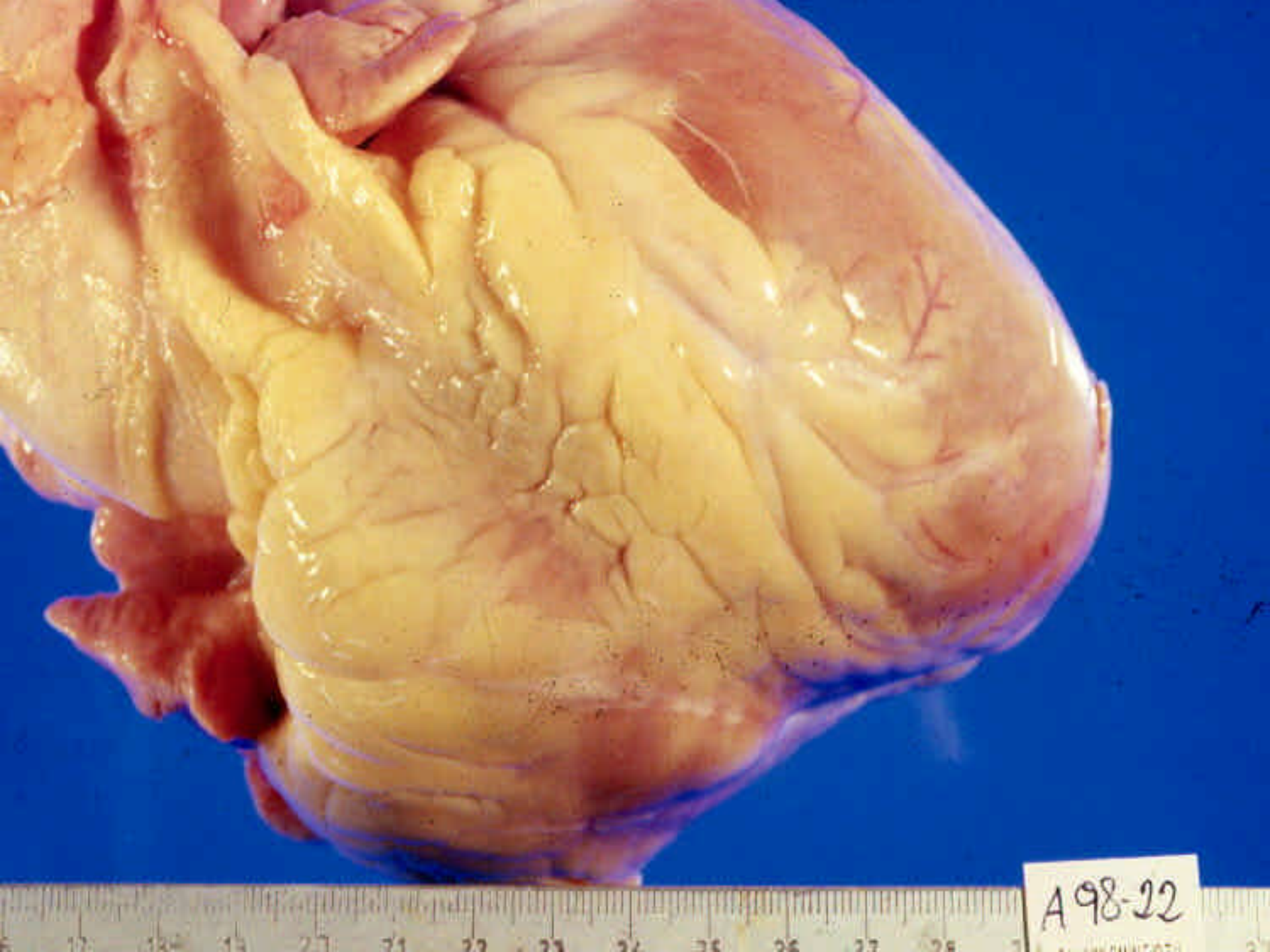


ENFERMEDAD RENAL EN LOS PACIENTES CON DP



DISPROTEINEMIAS HJC-LA CORUÑA

- **Enf. por Depósito de Proteínas: 9**
  - EDIM: 4
    - Kappa 3
    - IgG 1
  - Amiloidosis: 4
    - Lambda 1
    - IgM+Lambda 1
    - Kappa 2
  - Fibrilar: 1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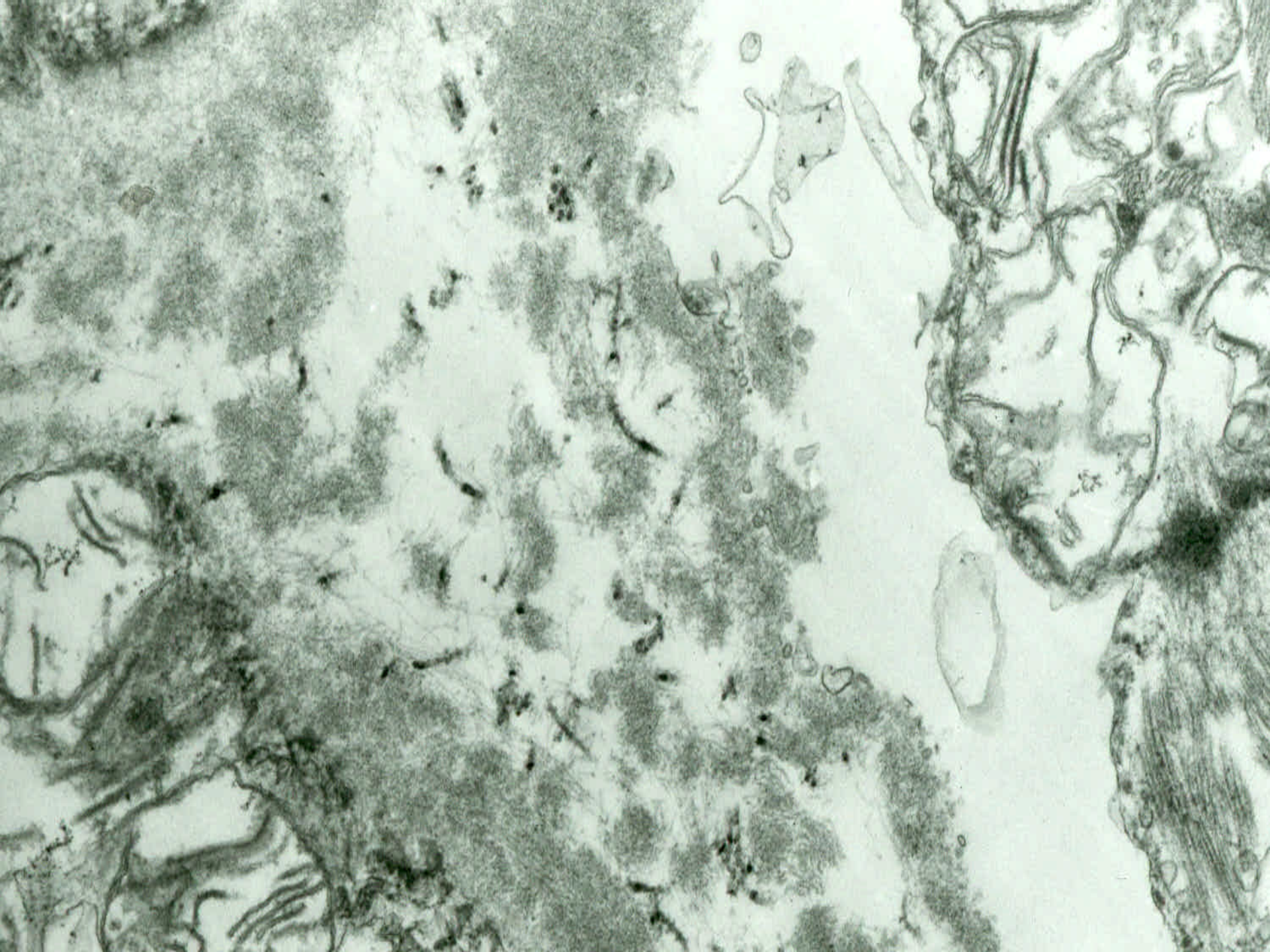

Arch Pathol Lab Med 1988
Aug;112(8):844-6

Light chain cardiomyopathy associated with small-vessel disease.

Peng SK, French WJ, Cohen AH, Fausel RE.

Department of Pathology, Harbor-UCLA Medical Center, Torrance 90509.

Am J Pathol 1996 May;148(5):1397-406

Light chain cardiomyopathy. Structural analysis of the light chain tissue deposits.

Gallo G, Goni F, Boctor F, Vidal R, Kumar A, Stevens FJ, Frangione B, Ghiso