BIOPSIA RENAL EN PACIENTE CON COLANGITIS BILIAR PRIMARIA

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20/10/2023

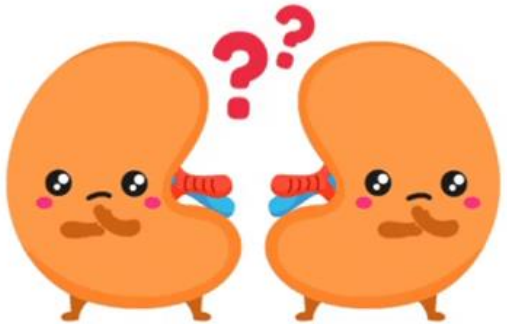
apmur

Arrixaca
Hospital Clínico Universitario
Virgen de la Arrixaca

PRESENTACIÓN DEL CASO CLÍNICO

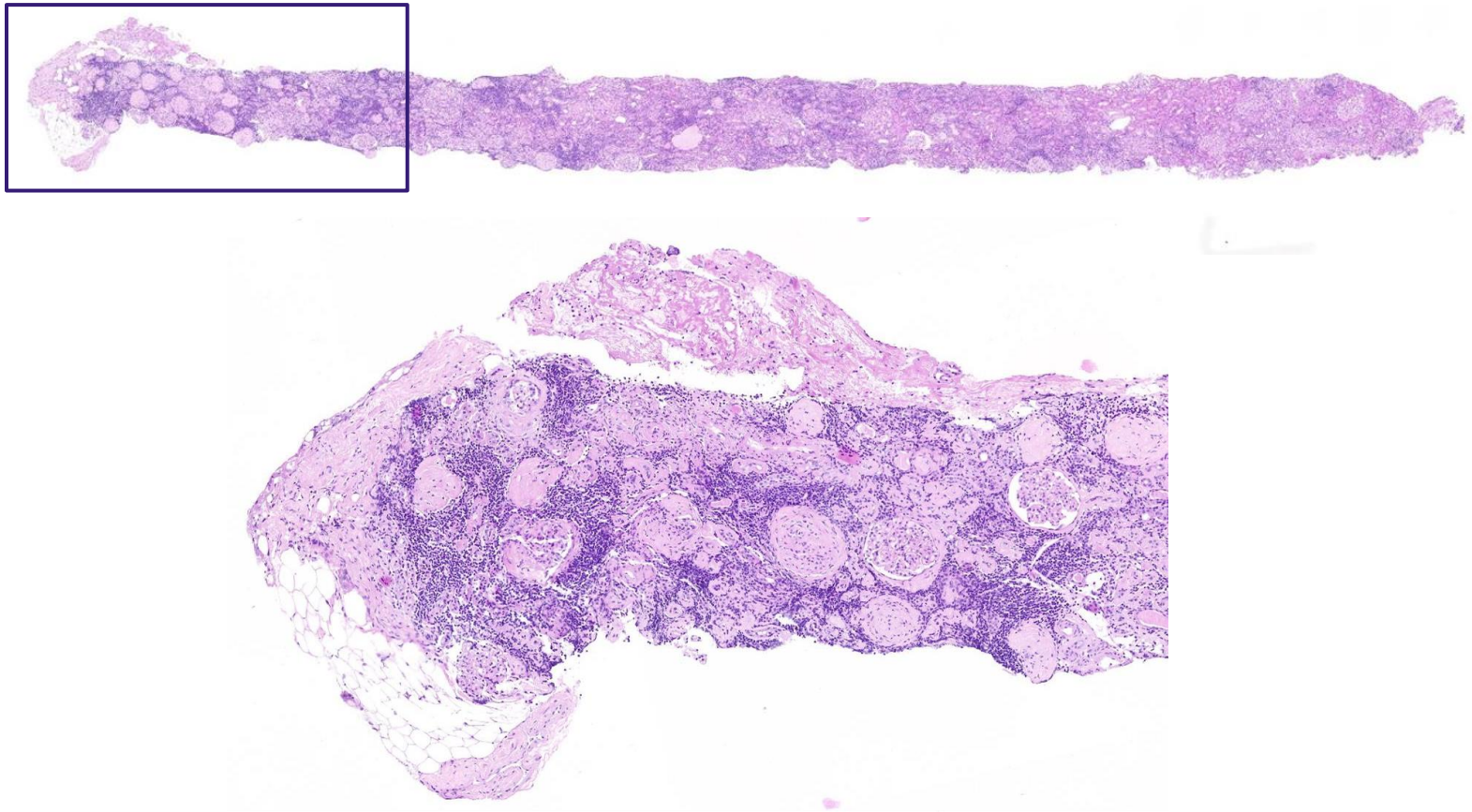
Varón de 51 años

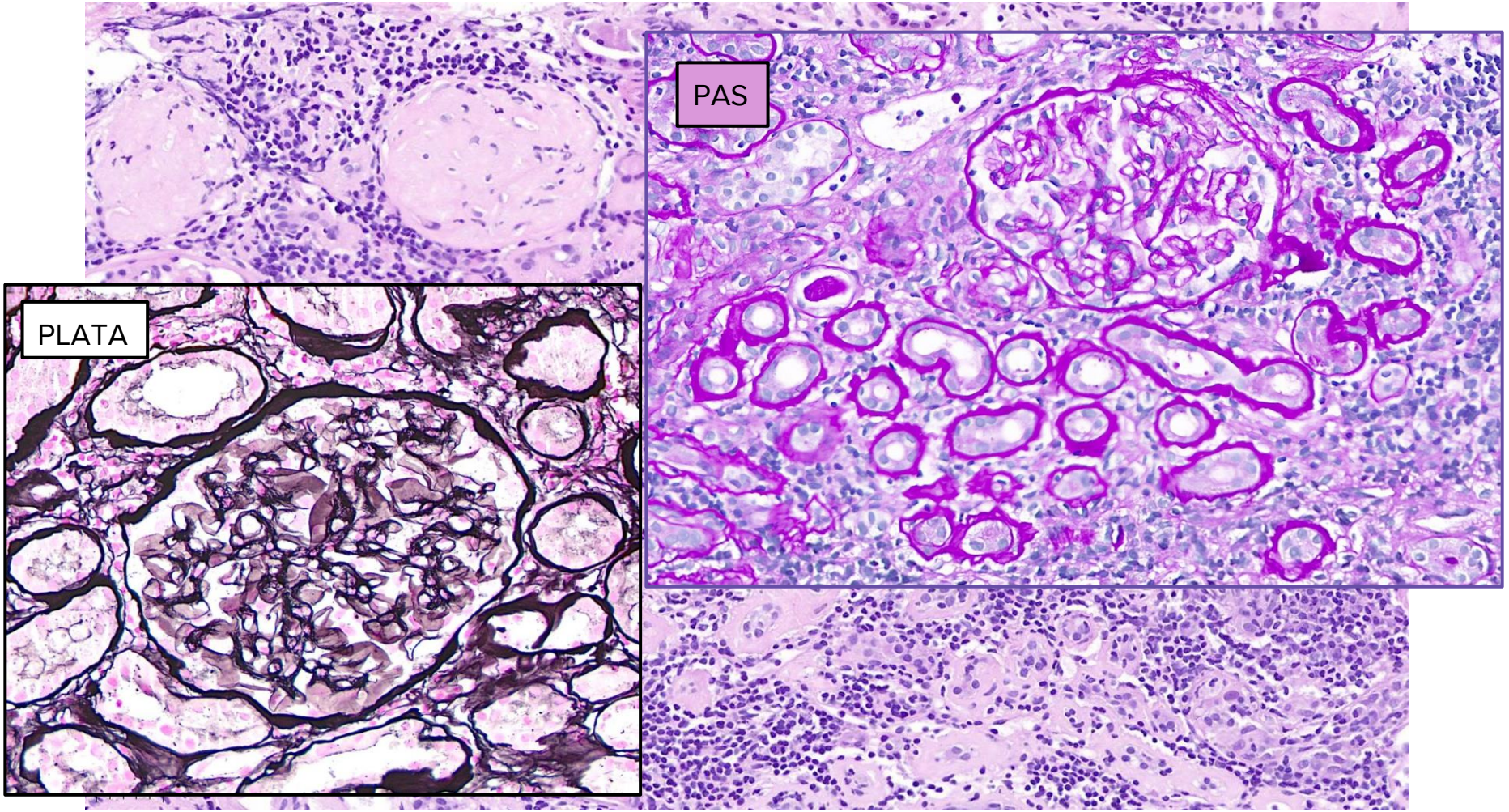
- Diabetes Mellitus tipo 2 (2003)
- Colangitis biliar primaria
  - AMAs (1/640) ✓
  - Elevación fosfatasa alcalina ✓
  - Sin confirmación de biopsia hepática

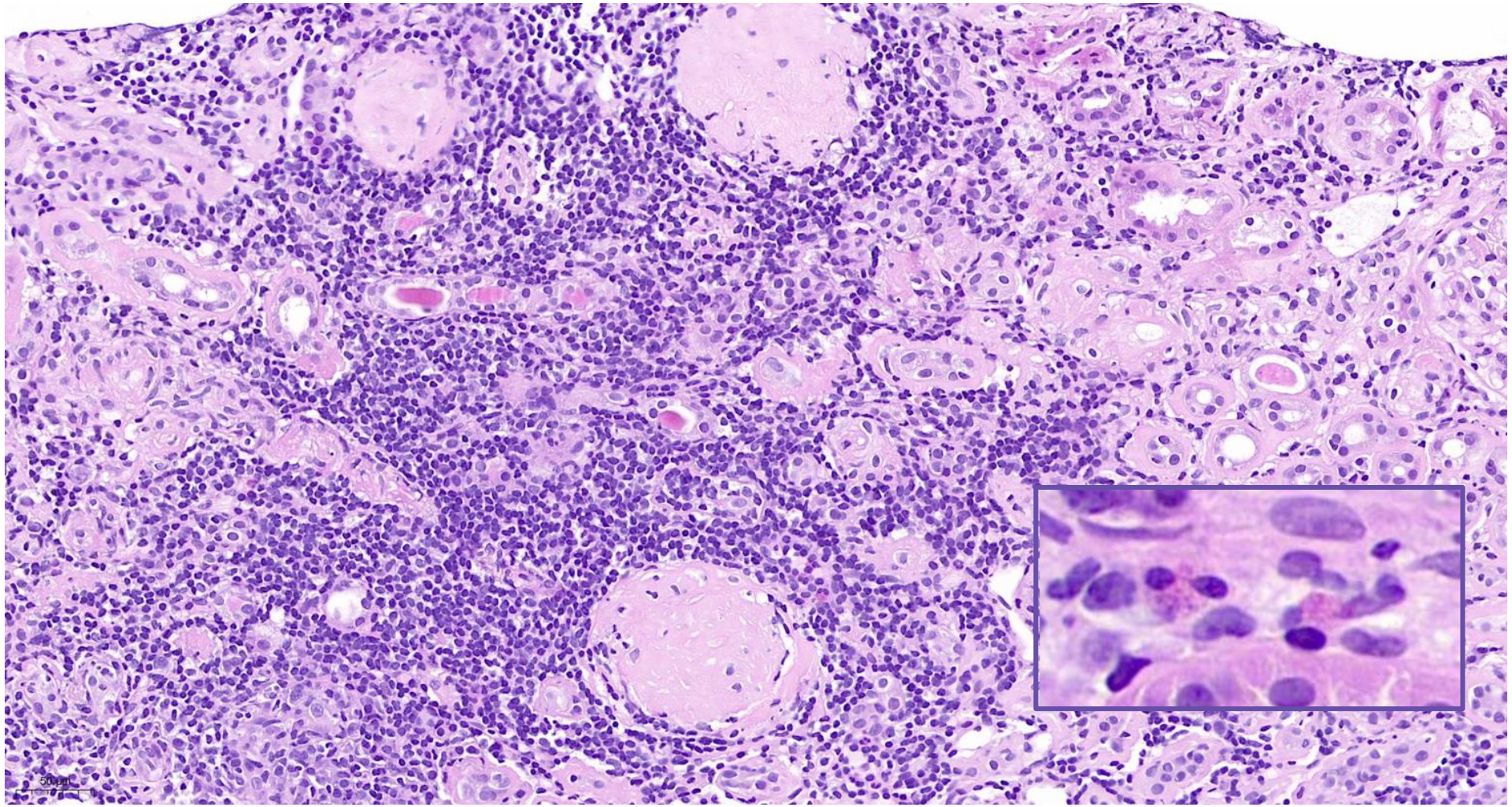


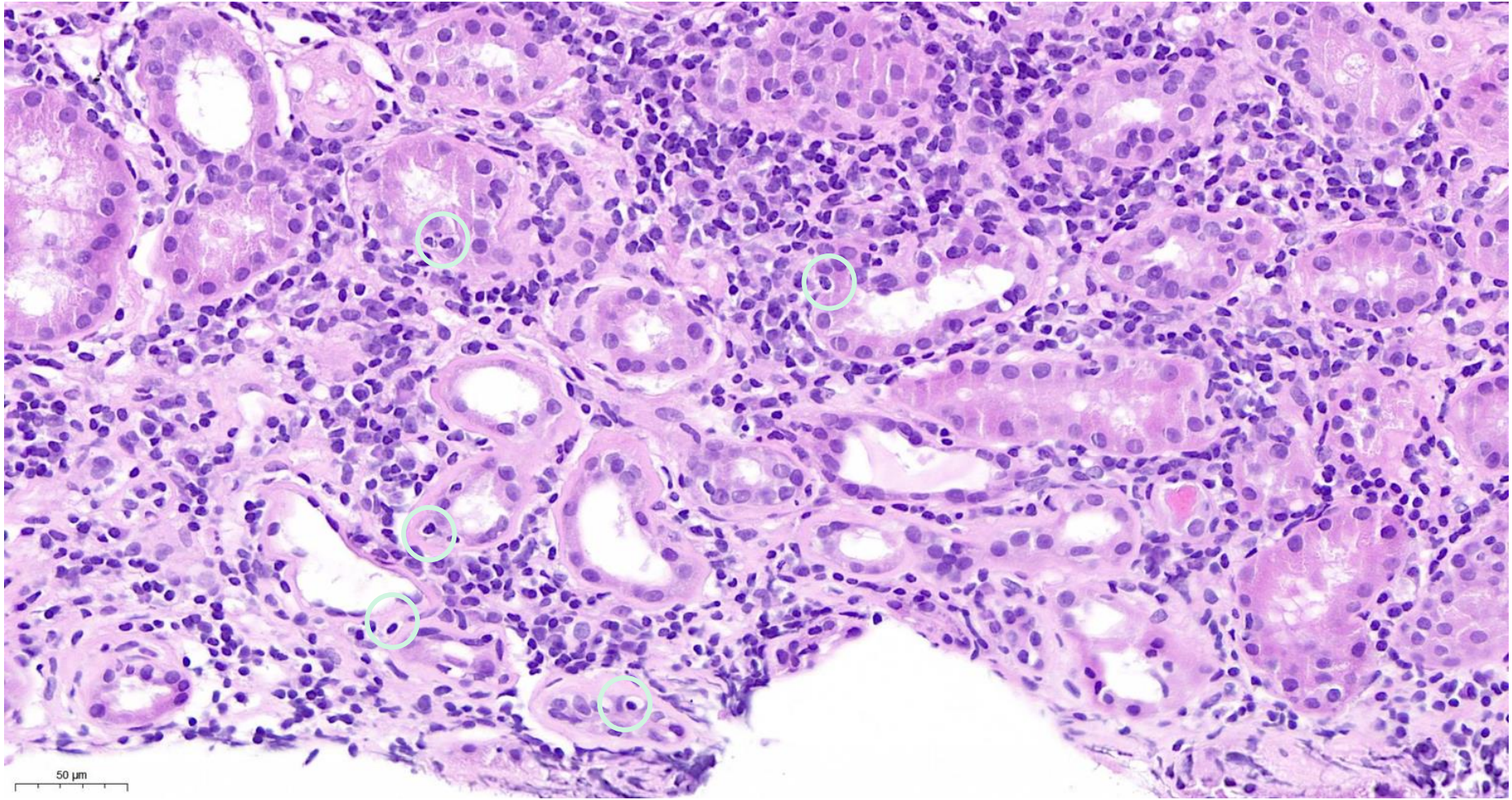
- Consulta por síndrome constitucional
- Ingreso para biopsia renal por **alteración en el sedimento** con datos de **disfunción tubular proximal** y sospecha de **nefritis tubulointersticial**

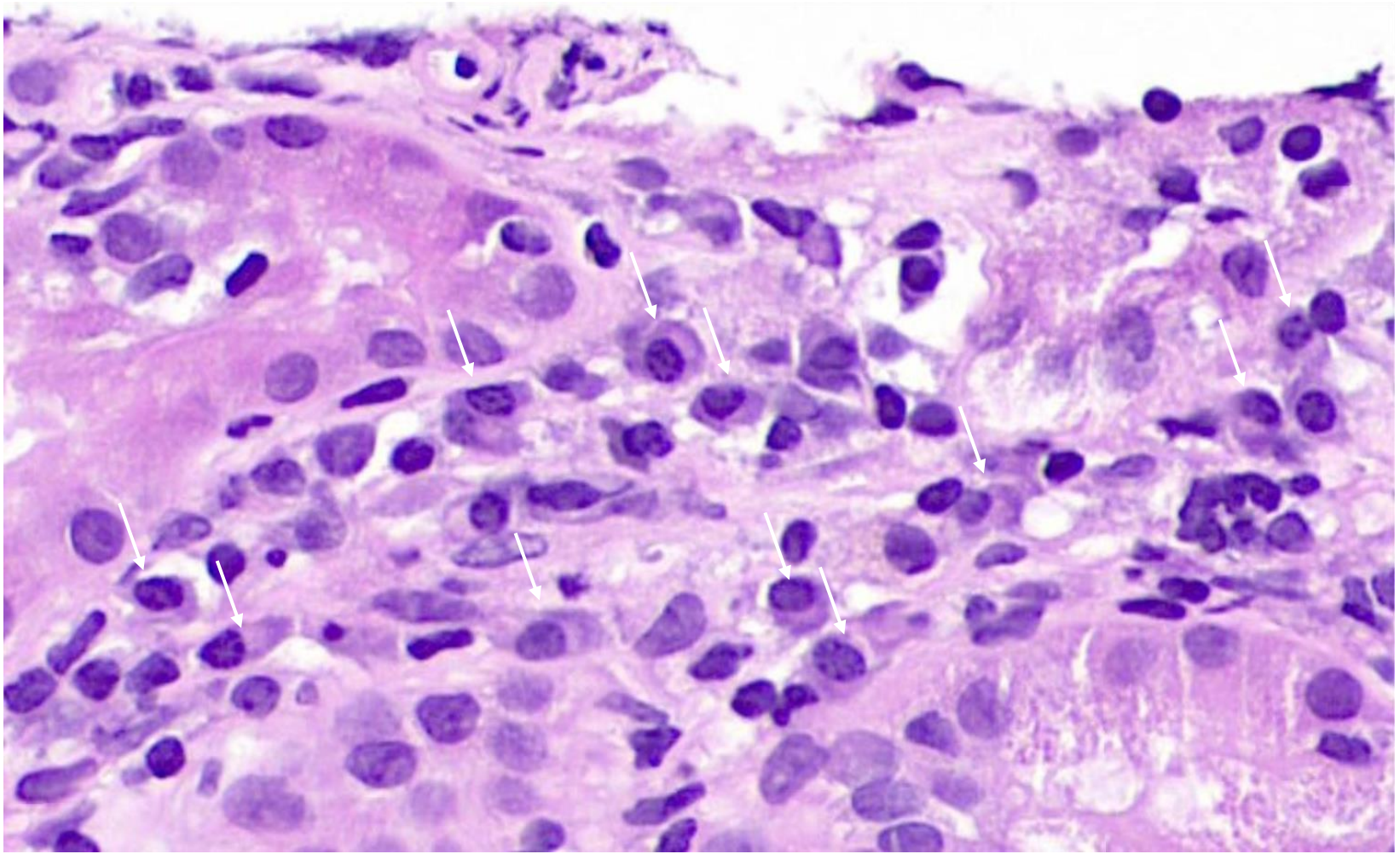
BIOPSIA RENAL

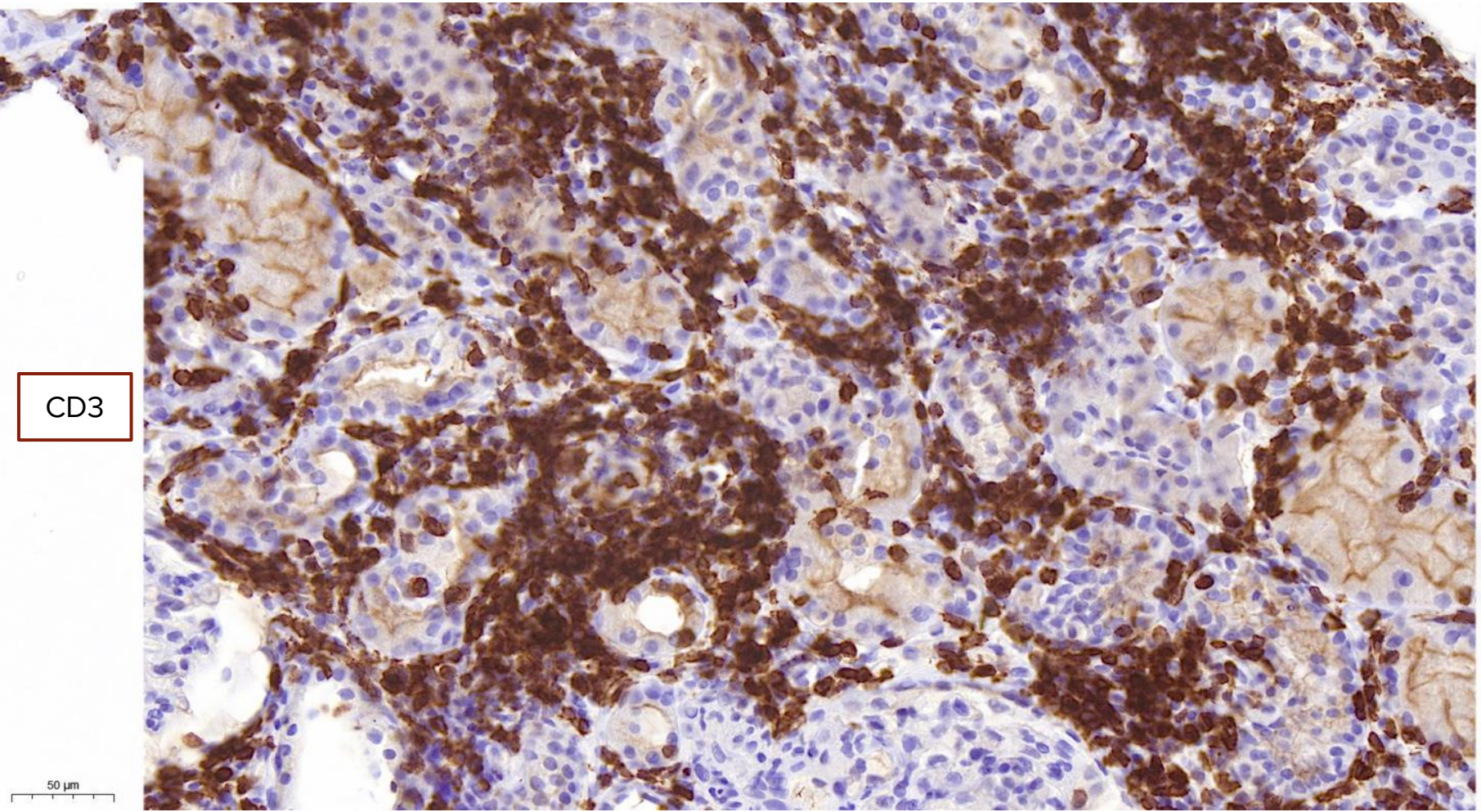



PAS
PLATA

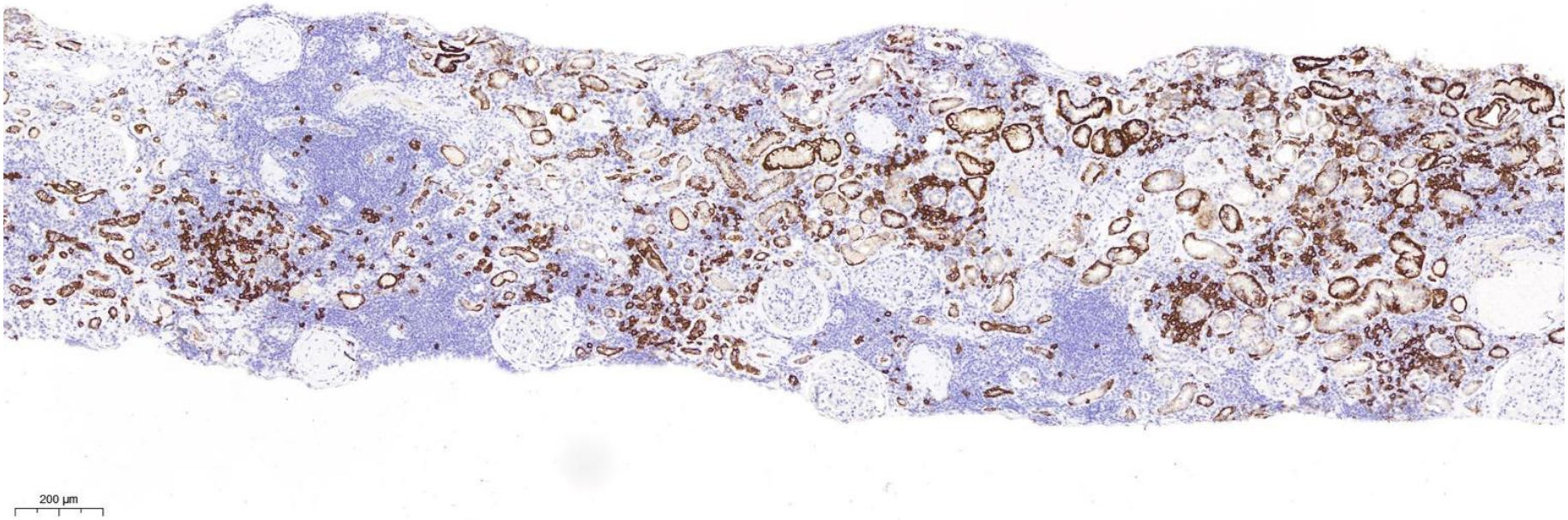

50µm

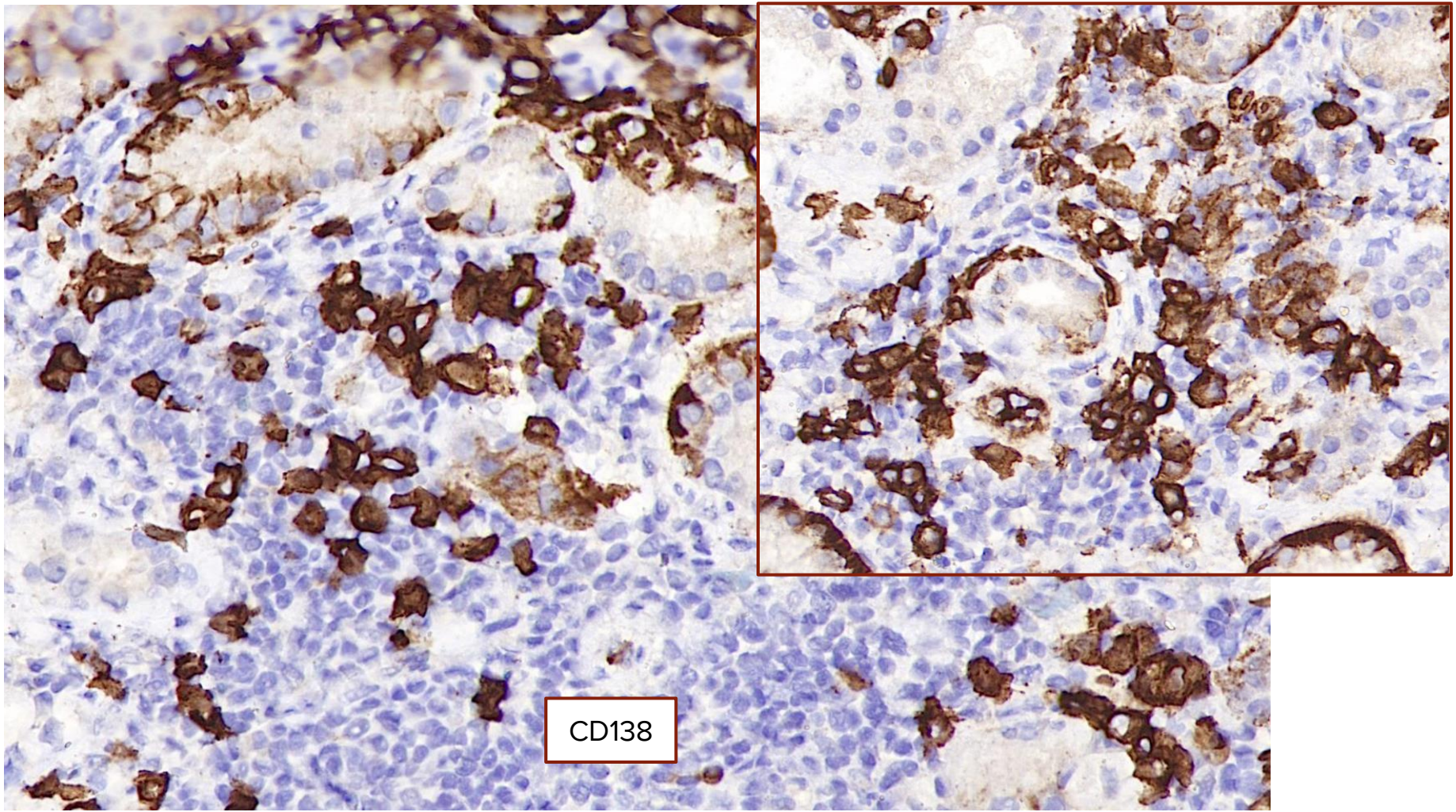

50 µm




CD3
50 µm

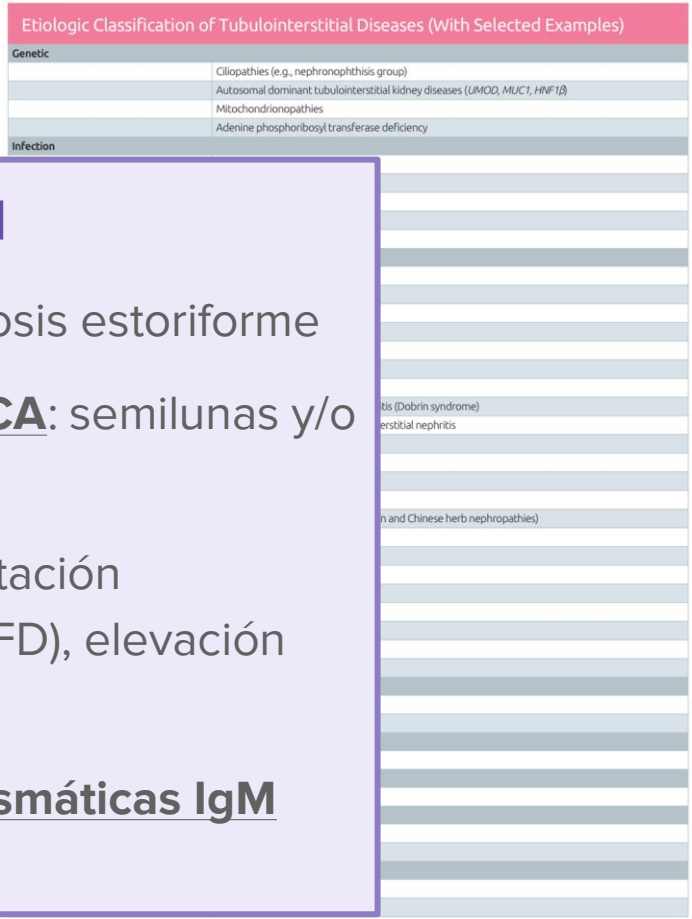
CD138




CD138

DIAGNÓSTICO DIFERENCIAL

- Células plasmáticas → **Autoinmunidad**
  - **Enfermedad por IgG4**: células IgG4+, fibrosis estoriforme
  - **Nefritis tubulointersticial asociada a ANCA**: semilunas y/o glomerulonefritis necrotizante
  - **Macroglobulinemia de Waldeström:** afectación glomerular, depósitos IgM glomerulares (IFD), elevación IgM MONOCLONAL
  - **Nefritis tubulointersticial con células plasmáticas IgM positivas**



IgG4

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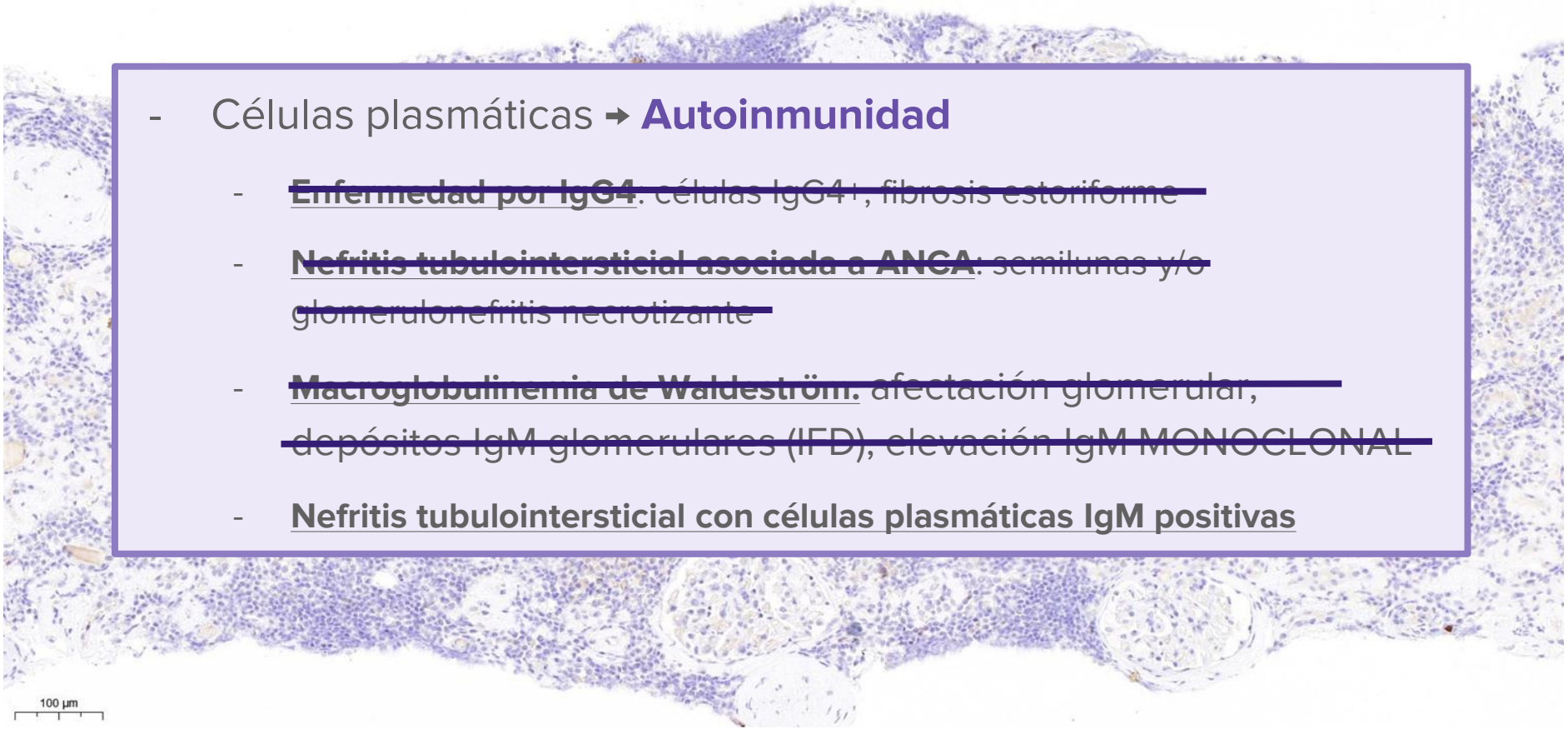
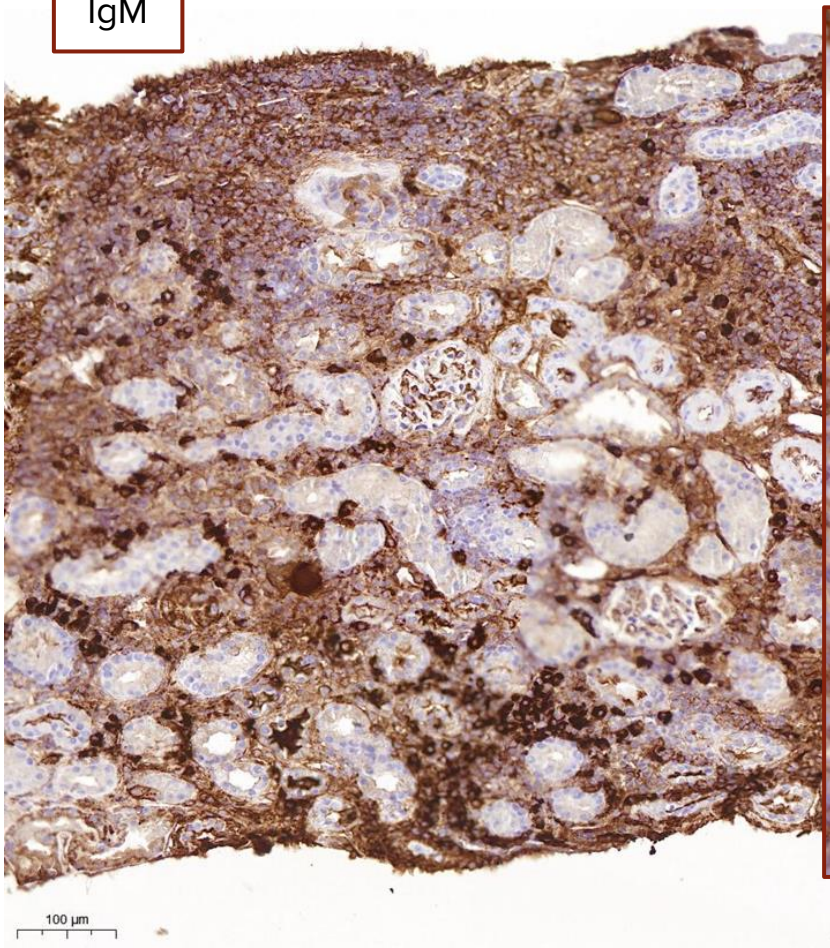
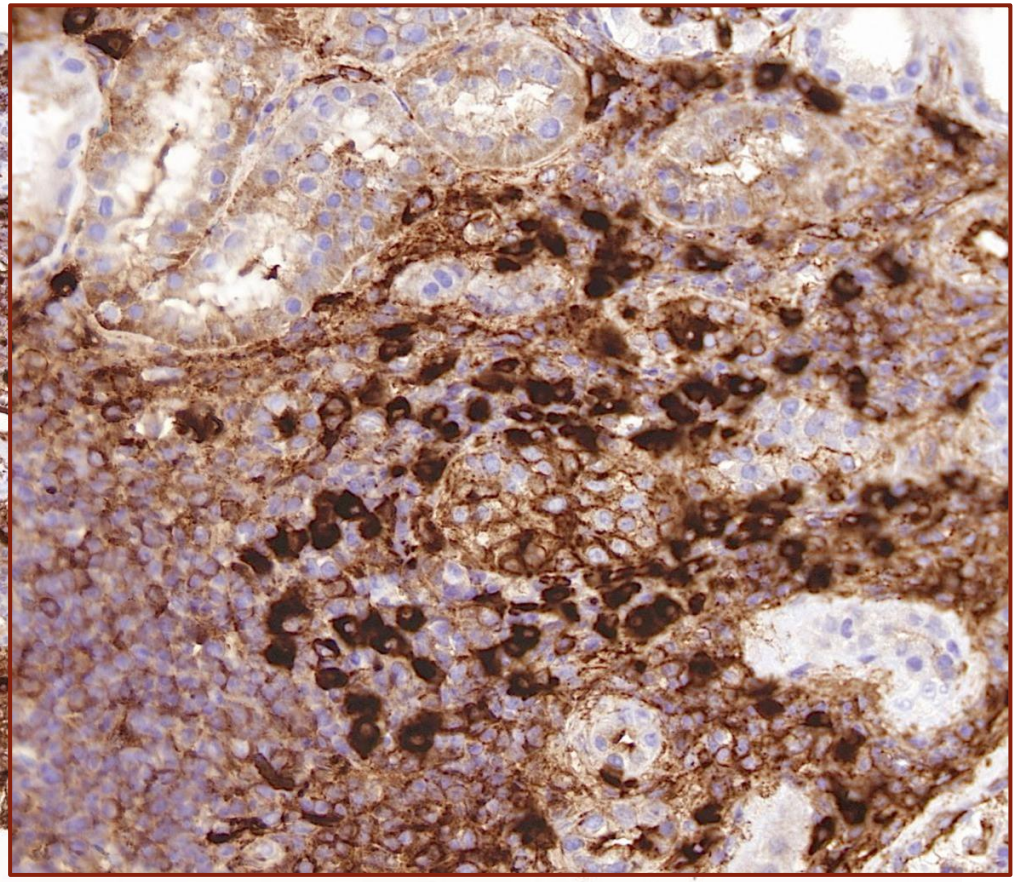
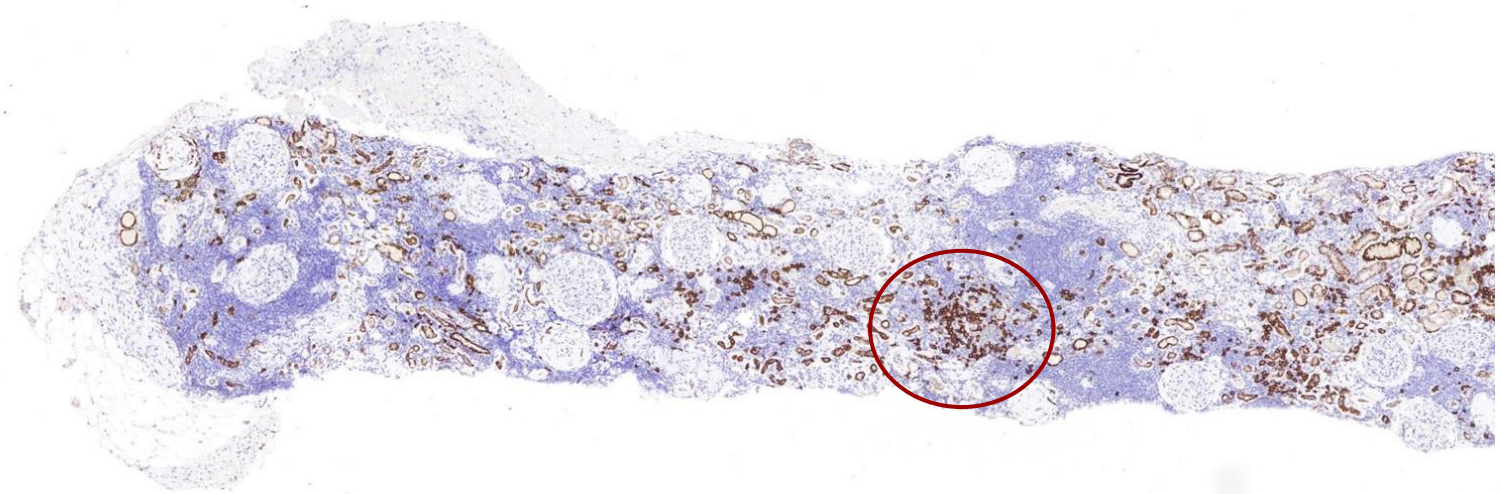


IgM

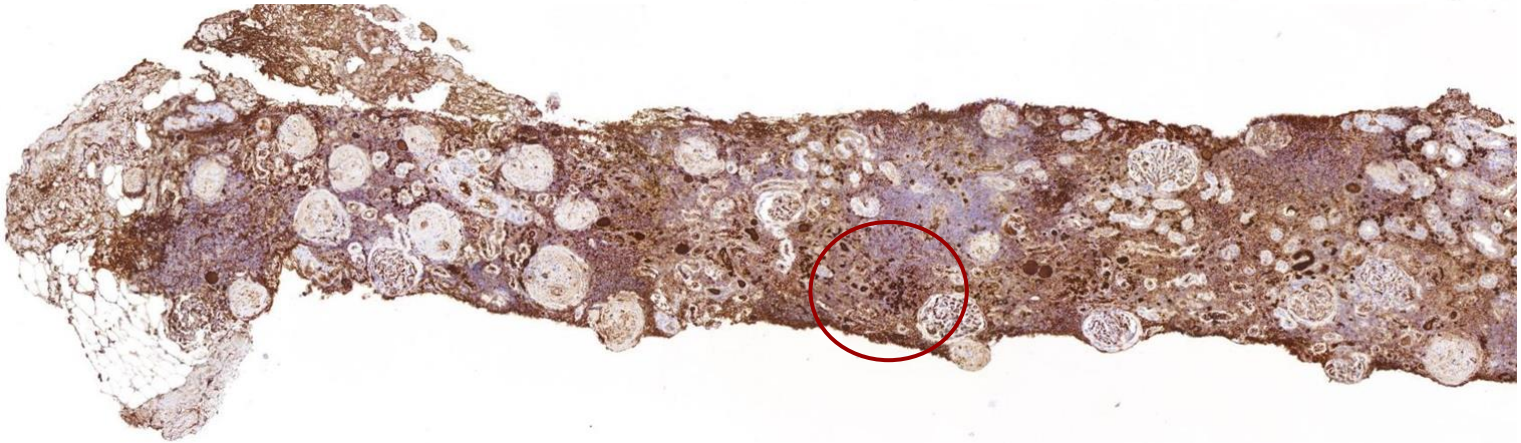




CD138



IgM



NEFRITIS TÚBULO-INTERSTICIAL CON CÉLULAS PLASMÁTICAS IgM POSITIVAS

NEFRITIS TUBULOINTERSTICIAL CON CÉLULAS PLASMÁTICAS IgM POSITIVAS

Clinical Features

We examined the clinical data from a total of 21,786 patients from whom renal biopsy samples were collected and identified 13 patients whose renal biopsies revealed IgMPC-TIN (13 of 21786, 0.06%). The clinical details for these 13 patients at the time of renal biopsy are summarized in Table 1. The patients were predominantly women (12 of 13, 92%) with an average age of 51±9 years (range 38–66) at the time of renal biopsy.

Figure 2, A, B, D, and E). The infiltrates included numerous diffusely distributed IgM-positive cells and CD138-positive plasma cells (Figure 2, A, C, D, and F), with a few IgG-positive cells (Figure 2, B and E) and with a small number of IgG4-positive cells (Supplemental Figure 2E). These plasma cells stained positively with both anti-λ), confirmed. The averaged number of infiltrating IgM-positi it IgMPC-TIN was markedly higher than from patients with N with IgG4-related kidney disease, three with TIN with AN r with antibody-mediated rejection, two with chronic pyelon o Figure 3, Supplemental Figure 2I). Receiver operating char ed infiltrating IgM-positive plasma cells was 13 per hpf, with te 1.007; $P<0.001$) (Supplemental Figure 4). The sensitivity a 9 98.6%), respectively. Moreover, immunohistochemical dua positive plasma cell fraction among total CD138-positive cells was 73.5%±10.5% (range 58.3%–89.2%), which conf cells were plasma cells (Figures 2, G and H, and 4, Table 1).

Se **asocia** a:

- Síndrome de Fanconi
- **Colangitis biliar primaria/** Síndrome de Sjögren
- Acidosis renal tubular distal
- **IgM sérica elevada**
- **Anticuerpos antimitocondriales**

Tubulointerstitial Nephritis with IgM-Positive Plasma Cells

2017

Naoki Takahashi, Takako Saeki, [...], and Masayuki Iwano

IDEAS PARA LLEVAR A CASA

1. Entidad muy nueva e infrecuente (2017)

2. Amplía el diagnóstico diferencial de las nefritis tubulointersticiales

3. Importancia de la historia clínica (pacientes complejos)

4. Buena comunicación con el nefrólogo

BIBLIOGRAFÍA

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MUCHAS GRACIAS