



Hospital Clínic
Universitari de València

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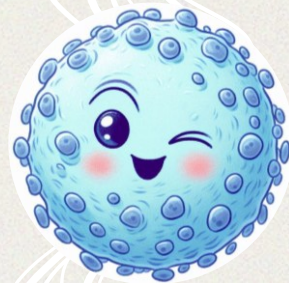
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Yassin El Bouzaydy El Gueddari
Antonio Ferrández Izquierdo - María Carolina Martínez Ciarpaglini

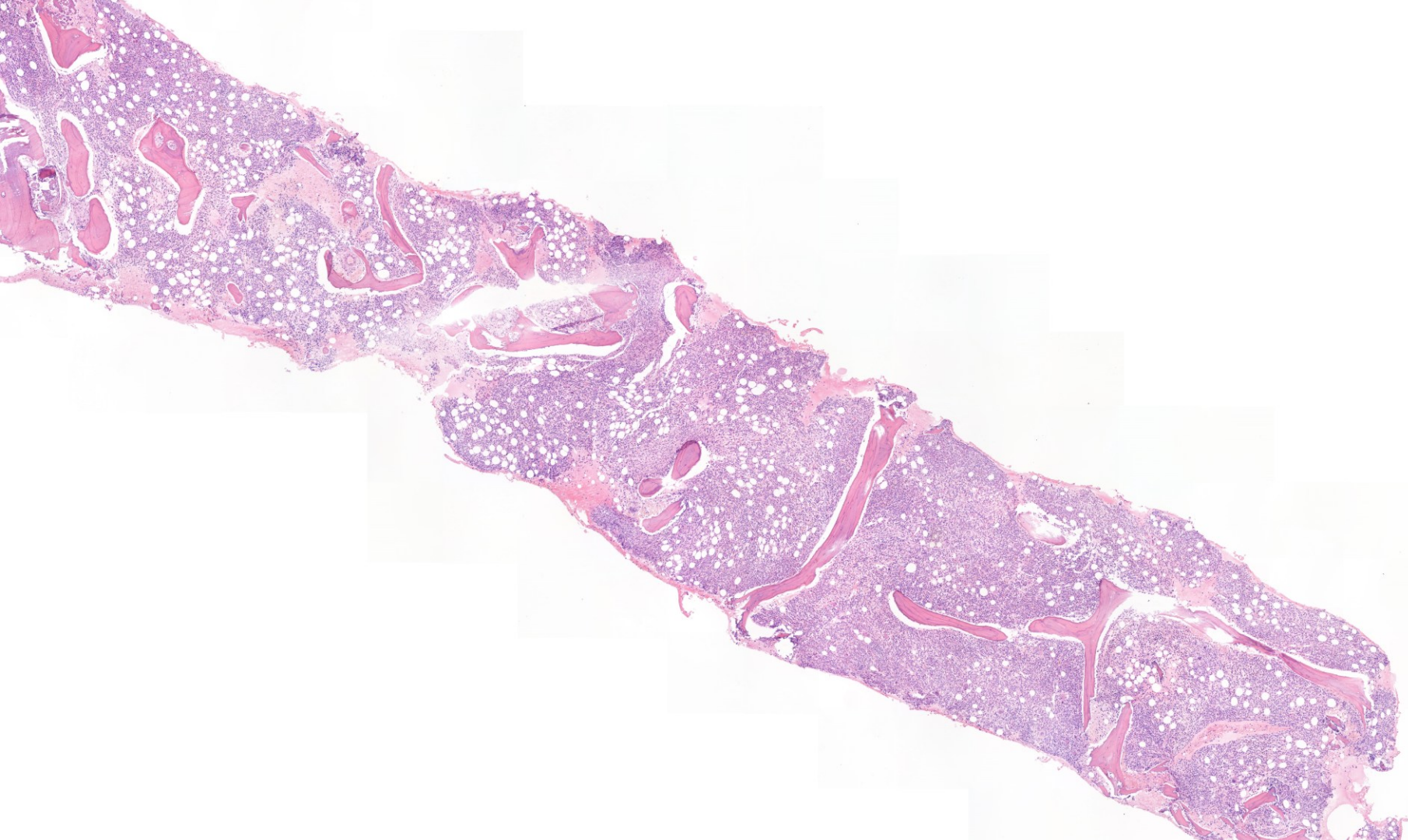
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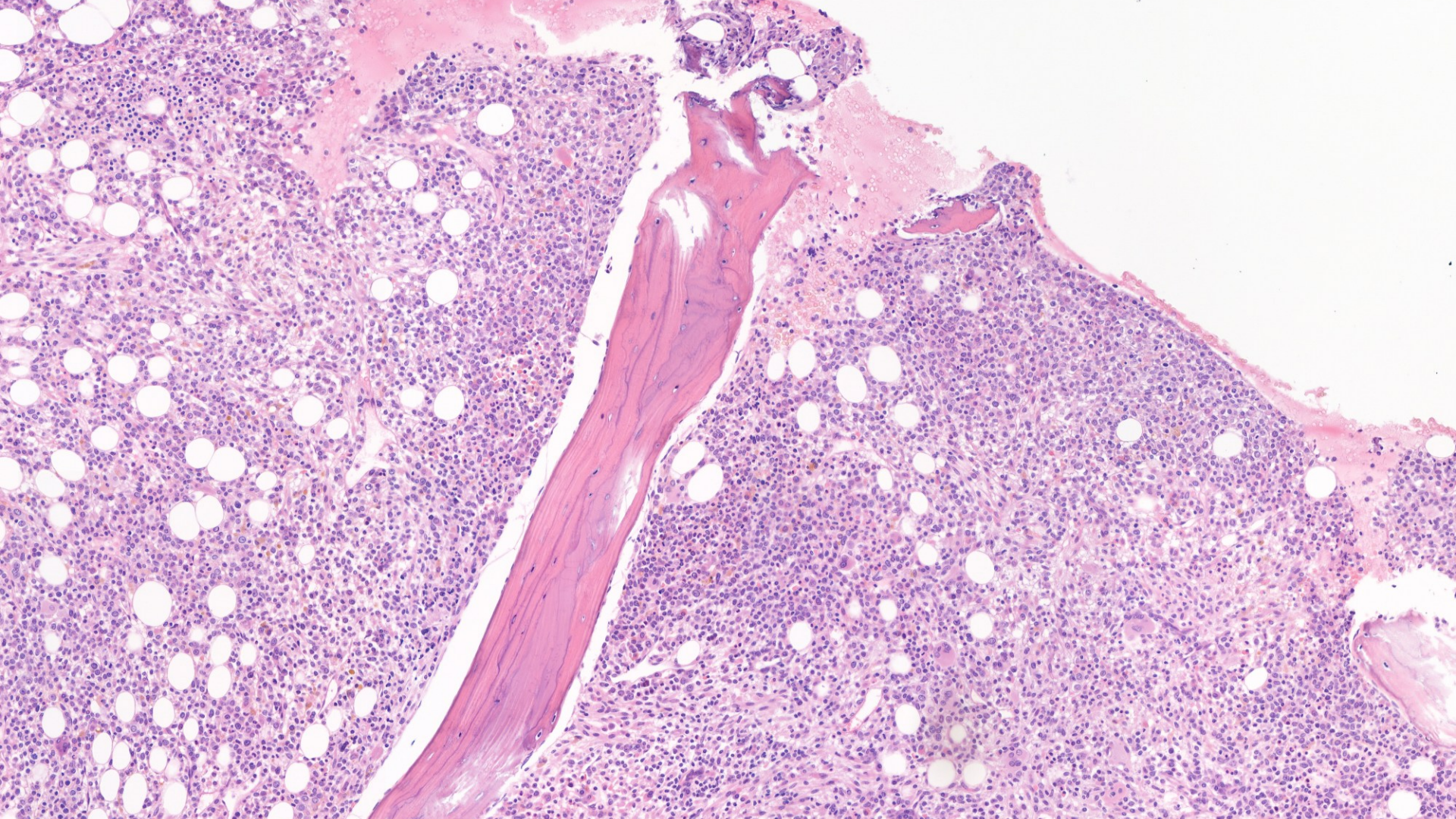


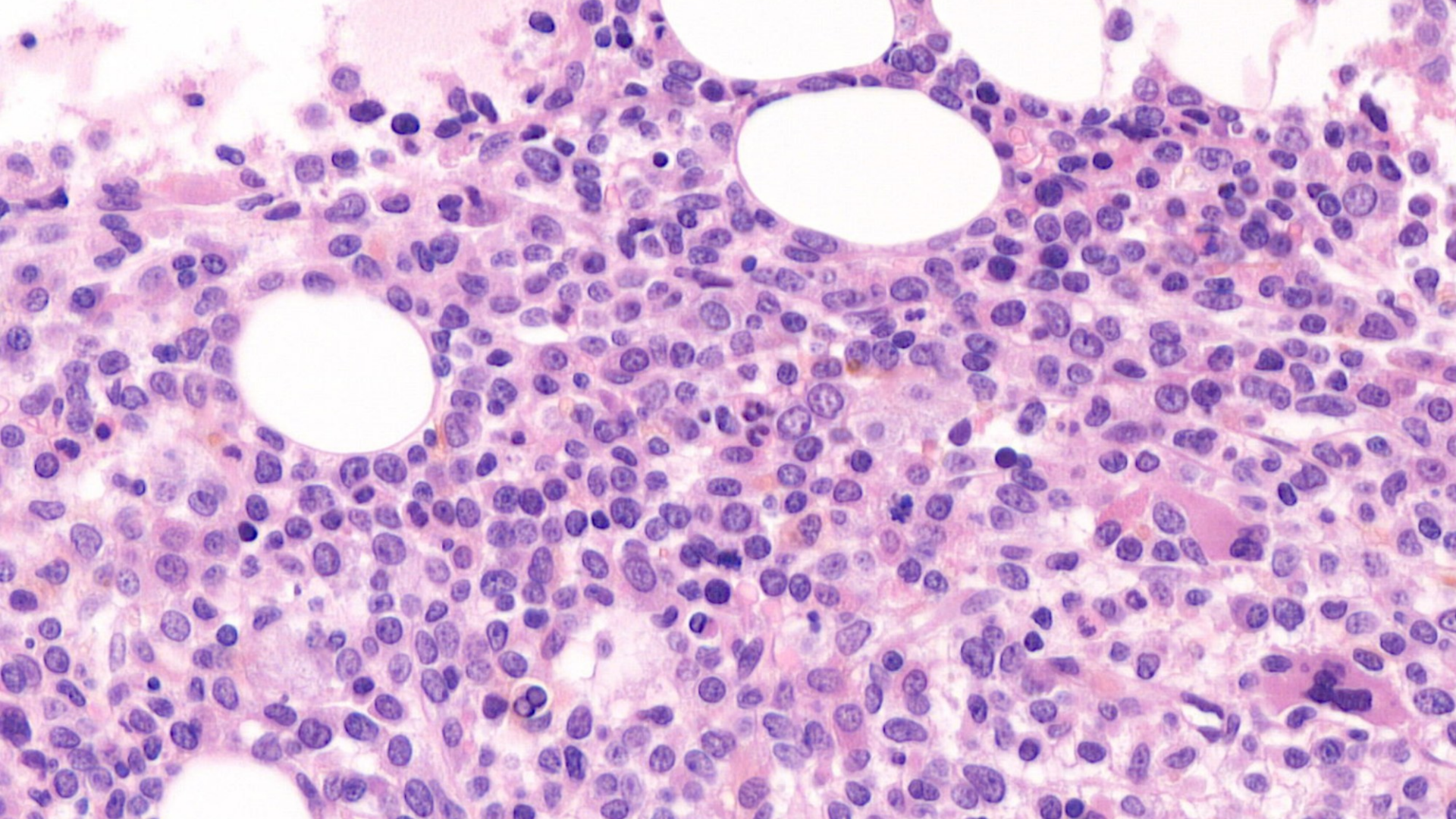
Nuestra paciente

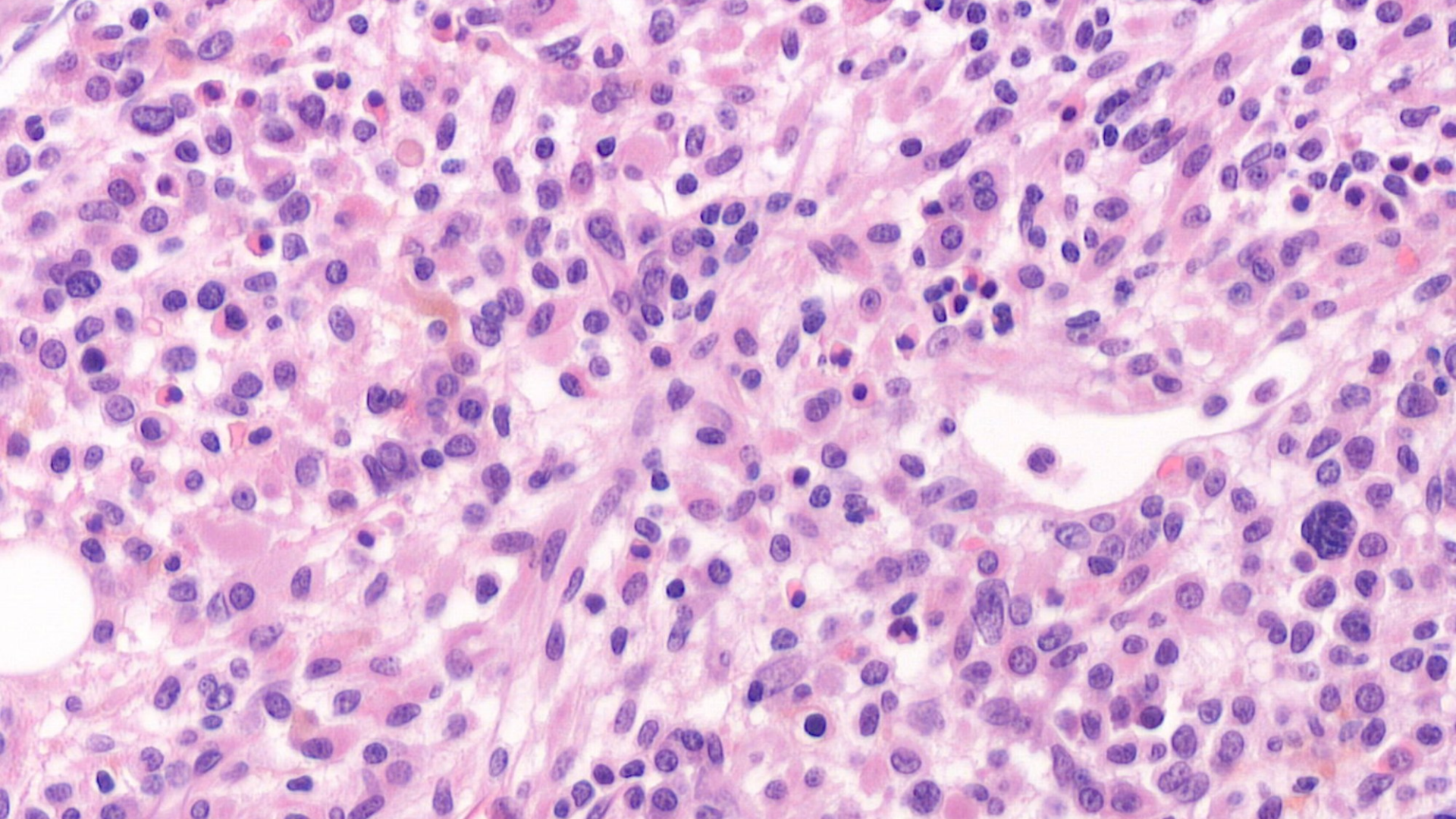
- Mujer de 68 años
- Fumadora. Mastocitosis cutánea diagnosticada hace 4 años
- Astenia y pérdida de peso
- Anemia, trombopenia y leucocitosis

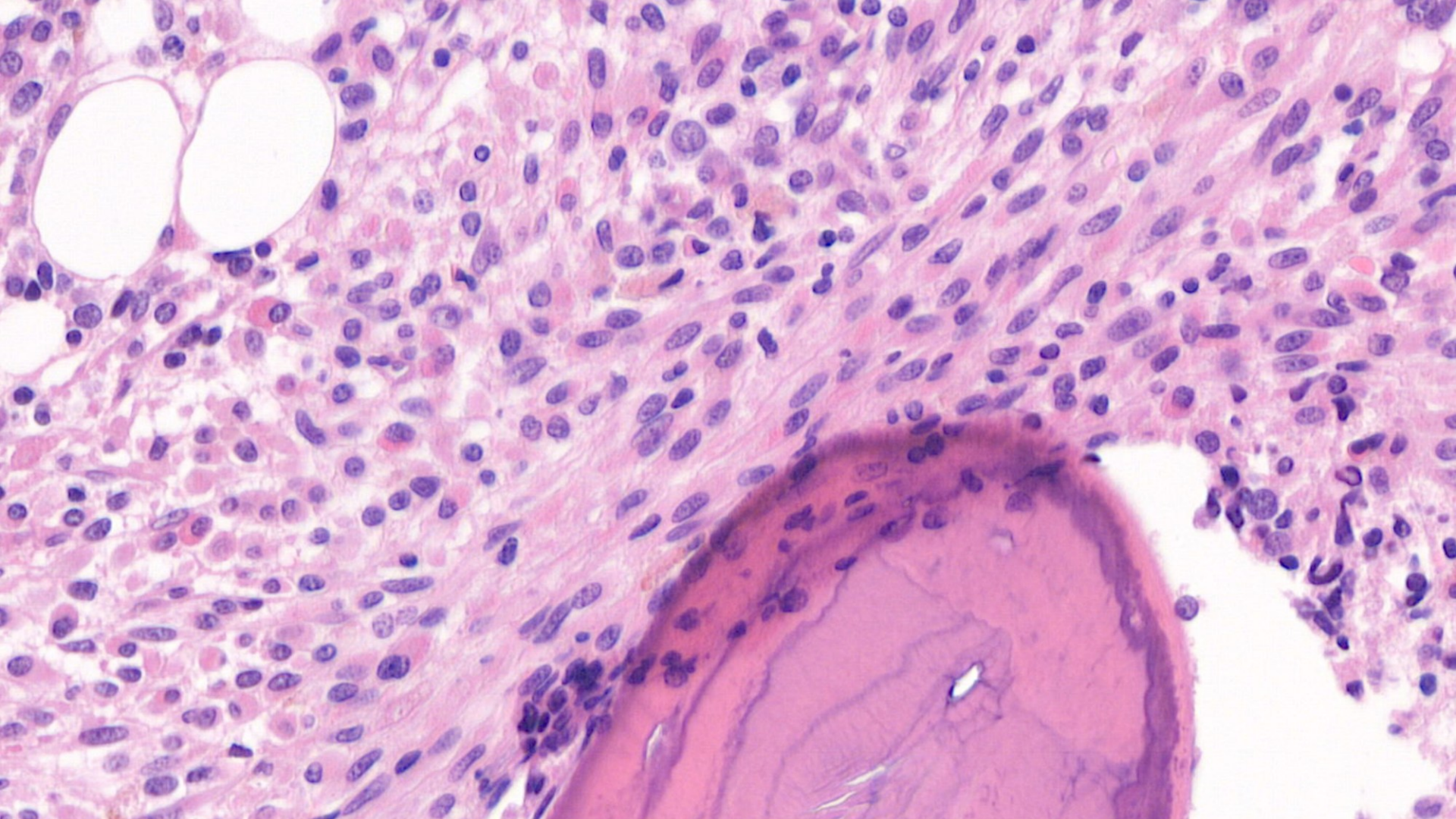


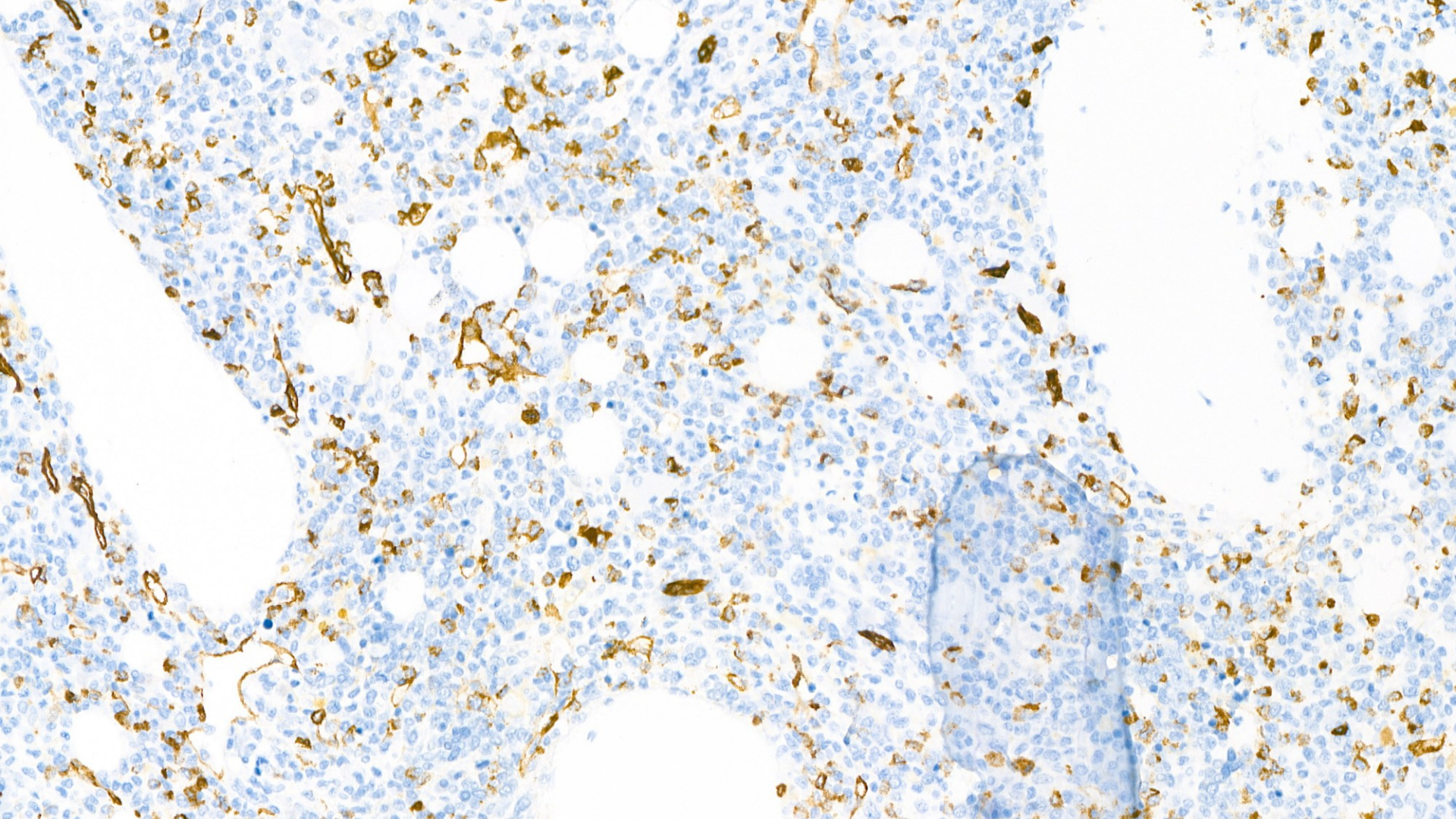


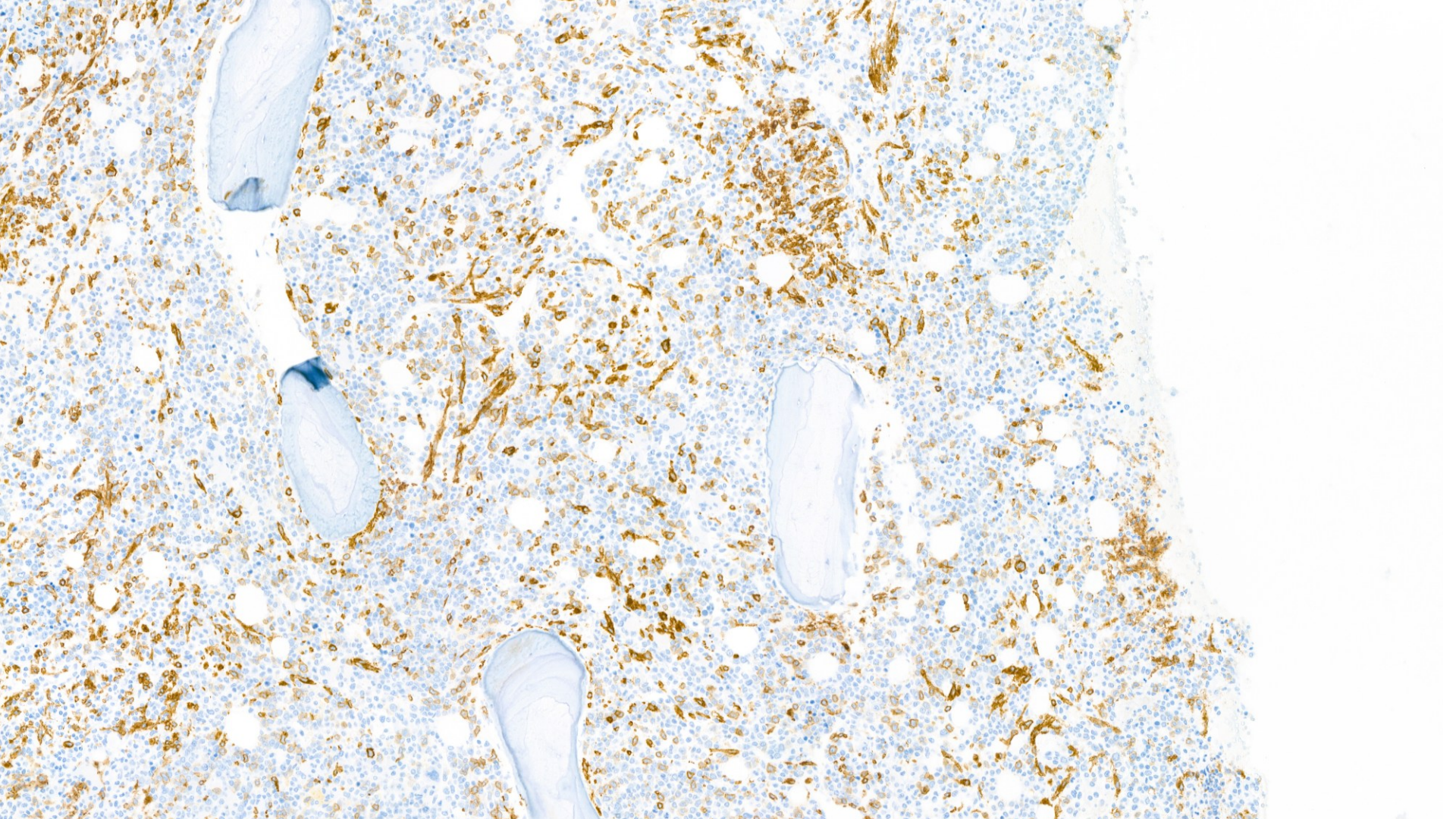


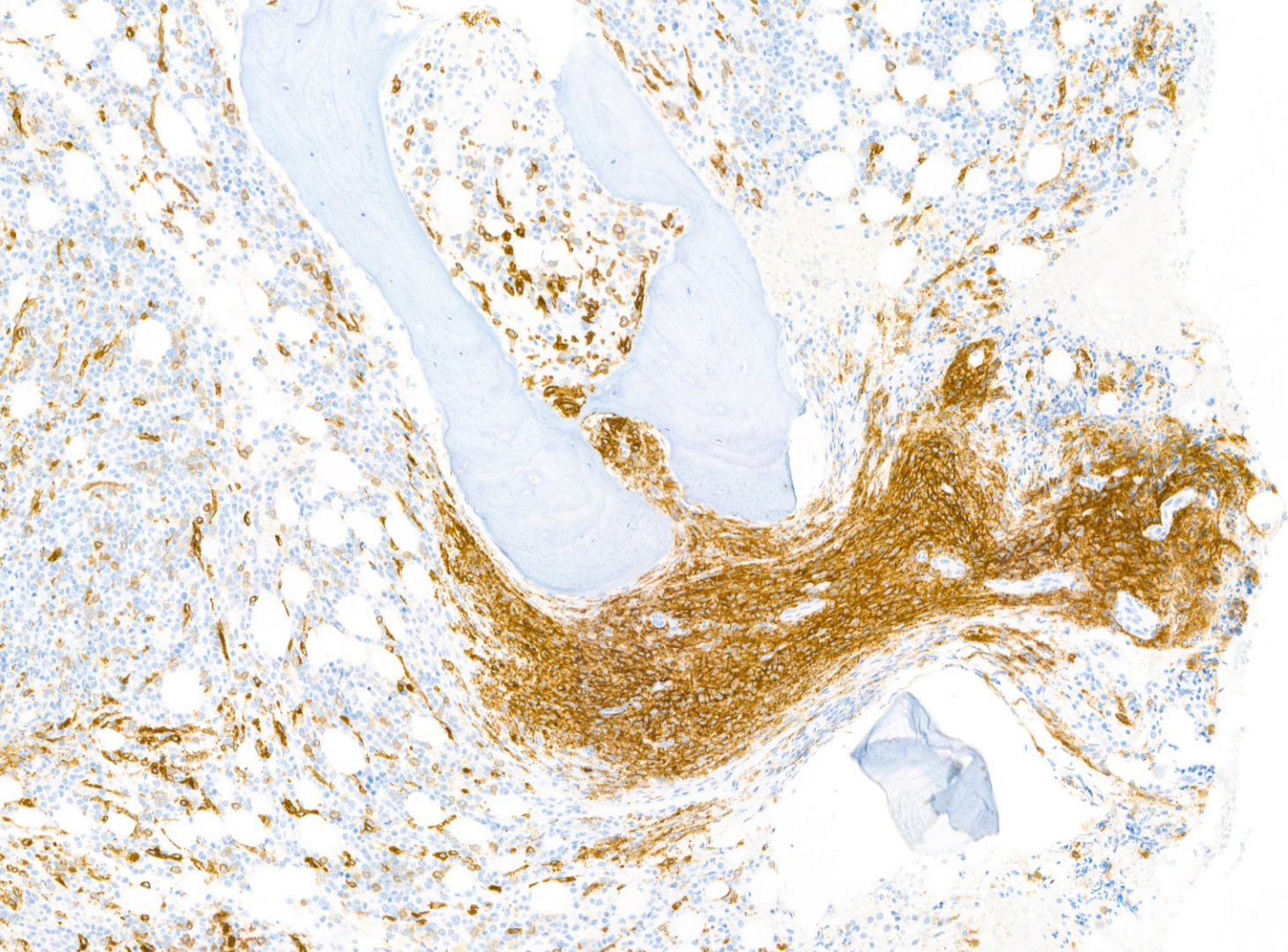


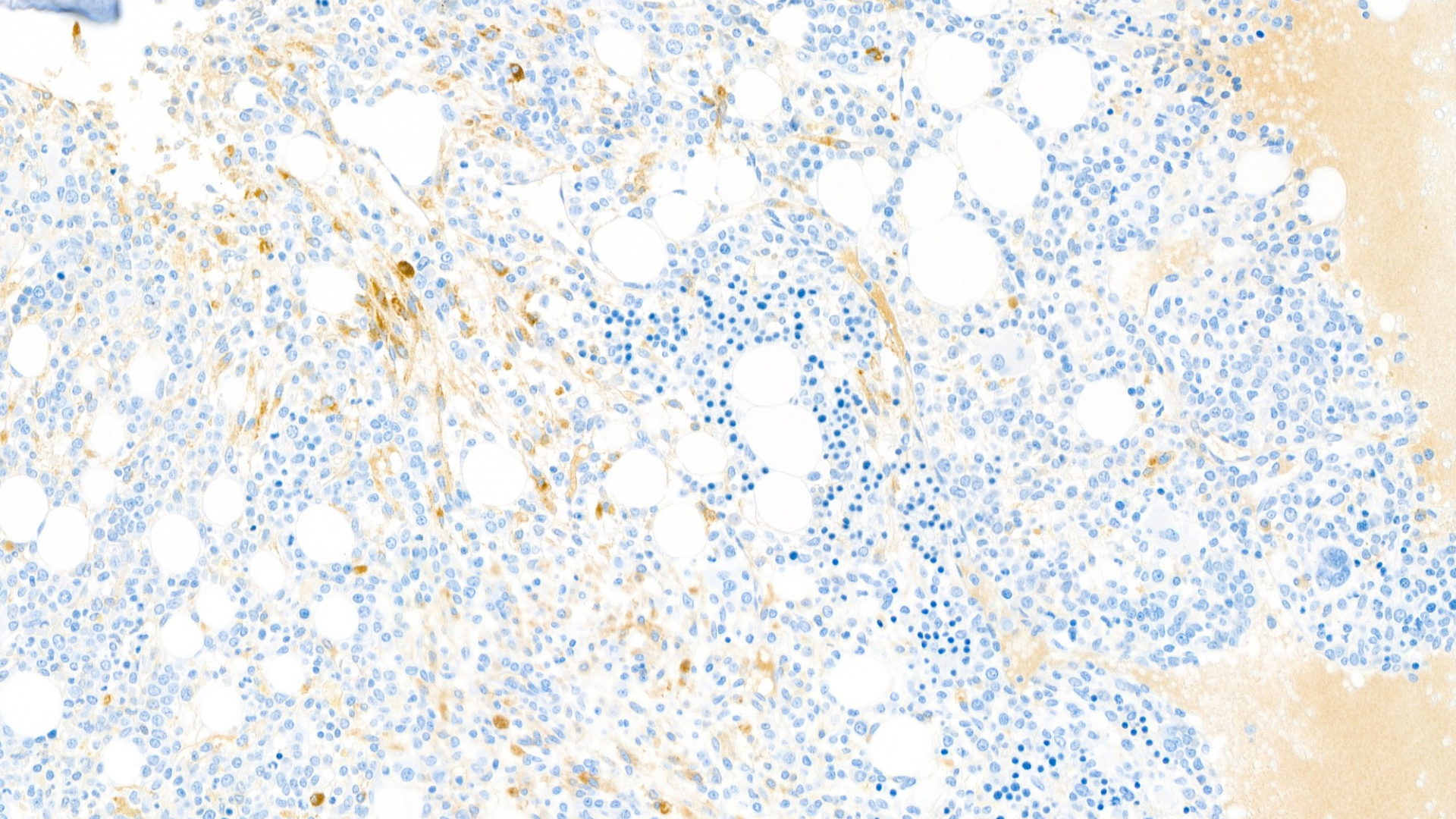


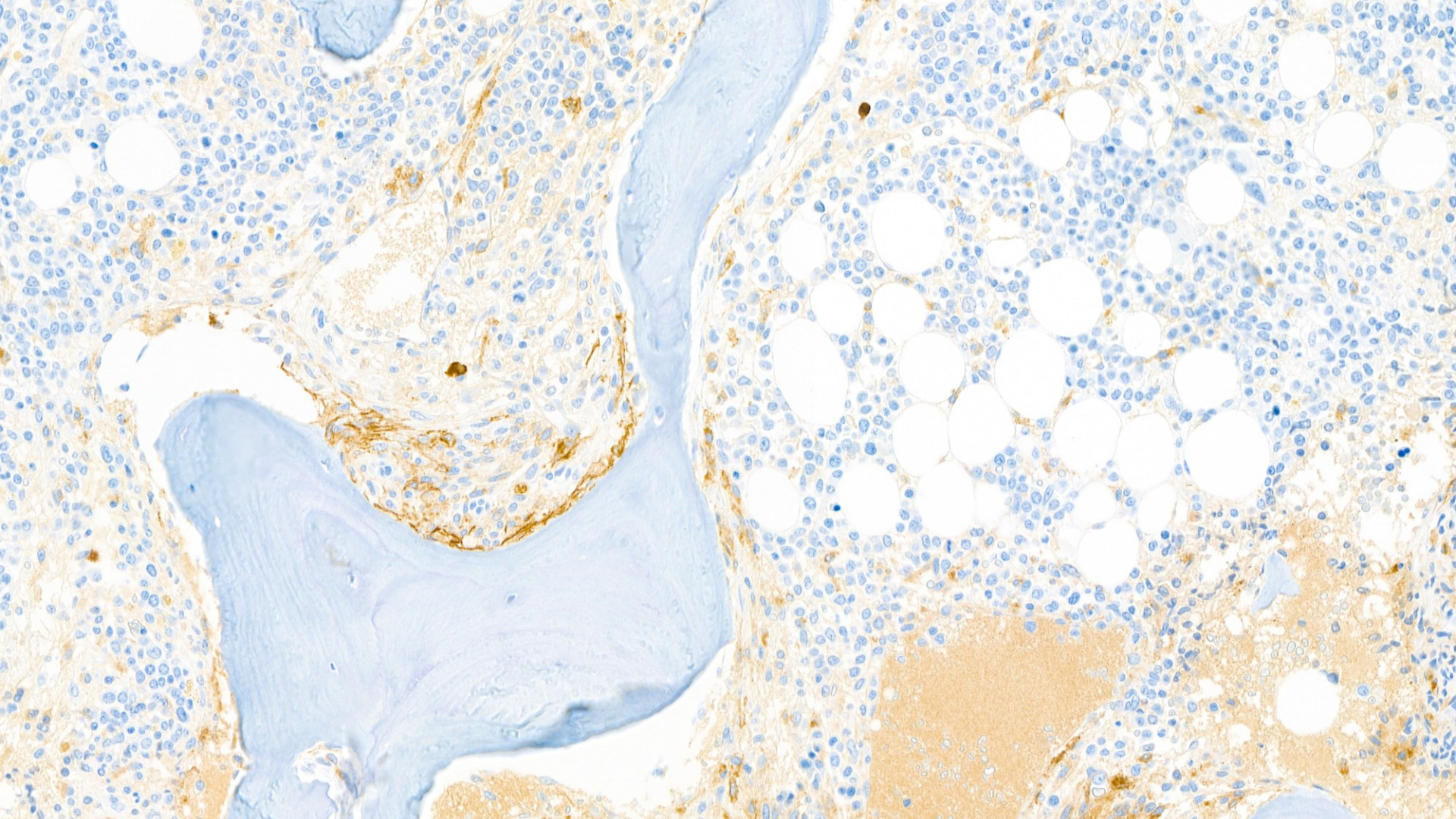












MASTOCITOSIS SISTÉMICA CON NEOPLASIA HEMATOLÓGICA / MIELOIDE ASOCIADA



Diferentes visiones

| ICC 2022 | Proposed WHO 5th Edition |
|---|---|
| <ul style="list-style-type: none"> • Cutaneous mastocytosis • Urticaria pigmentosa/maculopapular cutaneous mastocytosis • Diffuse cutaneous mastocytosis • Mastocytoma of skin • Systemic mastocytosis • Indolent systemic mastocytosis (includes bone marrow mastocytosis) • Smoldering systemic mastocytosis • Aggressive systemic mastocytosis • Mast cell leukemia • Systemic mastocytosis with an associated myeloid neoplasm • Mast cell sarcoma | <ul style="list-style-type: none"> • Cutaneous mastocytosis • Urticaria pigmentosa/maculopapular cutaneous mastocytosis <ul style="list-style-type: none"> • Monomorphic • Polymorphic • Diffuse cutaneous mastocytosis • Cutaneous mastocytoma <ul style="list-style-type: none"> • Isolated mastocytoma • Multilocalized mastocytoma • Systemic mastocytosis • Bone marrow mastocytosis • Indolent systemic mastocytosis • Smoldering systemic mastocytosis • Aggressive systemic mastocytosis • Systemic mastocytosis with an associated hematologic neoplasm • Mast cell leukemia • Mast cell sarcoma |



The international consensus classification of mastocytosis and related entities

Review | Published: 10 October 2022

Volume 482, pages 99–112, (2023) [Cite this article](#)

Table 4 Diagnostic criteria for systemic mastocytosis with an associated myeloid neoplasm (SM-AMN)

-
- Meets the diagnostic criteria for systemic mastocytosis
 - Meets the criteria for an associated myeloid neoplasm, e.g., chronic myelomonocytic leukemia (CMML) or other myelodysplastic/myeloproliferative neoplasm (MDS/MPN), myelodysplastic syndrome (MDS), myeloproliferative neoplasm (MPN), acute myeloid leukemia (AML), or other myeloid neoplasm^a
 - The associated myeloid neoplasm should be fully classified according to established criteria^b
-

The international consensus classification of mastocytosis and related entities

Table 1 Systemic mastocytosis: diagnostic criteria

Major criterion

- Multifocal dense infiltrates of tryptase- and/or CD117-positive mast cells (≥ 15 mast cells in aggregates) detected in sections of bone marrow and/or other extracutaneous organ(s)^a

In the absence of the major criterion, at least three of the following four minor criteria must be present

- In bone marrow biopsy or in section of other extracutaneous organs, $> 25\%$ of mast cells are spindle shaped or have an atypical immature morphology^b
 - Mast cells in bone marrow, peripheral blood, or other extracutaneous organs express CD25, CD2, and/or CD30, in addition to mast cell markers
 - *KIT* D816V mutation or other activating *KIT* mutation detected in bone marrow, peripheral blood, or other extracutaneous organs^{a/c}
 - Elevated serum tryptase level, persistently > 20 ng/mL. In cases of SM-AMN, an elevated tryptase does not count as a SM minor criterion
-



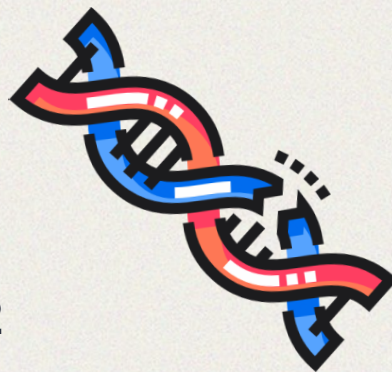
RUNX1

TET2

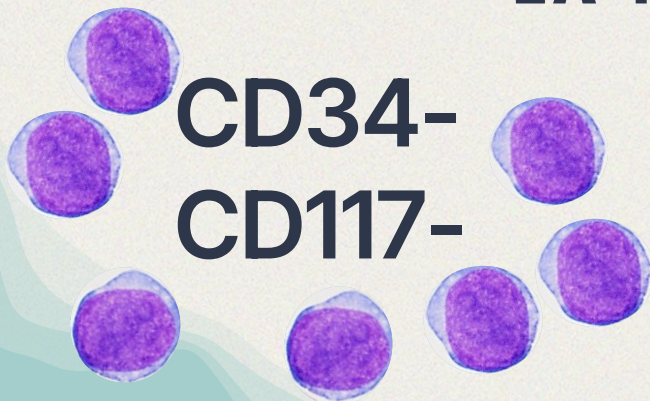
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DNMT3A

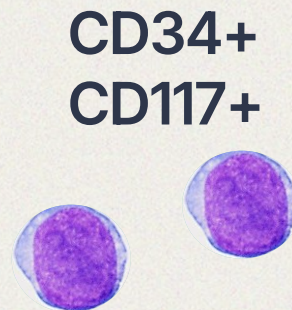
GATA2



LMA CON MUTACIONES RELACIONADAS CON LA MIELODISPLASIA



**CD34-
CD117-**



**CD34+
CD117+**

¿Diagnosticado?

Table 4 Diagnostic criteria for systemic mastocytosis with an associated myeloid neoplasm (SM-AMN)

- ✓ • Meets the diagnostic criteria for systemic mastocytosis
- ✓ • Meets the criteria for an associated myeloid neoplasm, e.g., chronic myelomonocytic leukemia (CMML) or other myelodysplastic/myeloproliferative neoplasm (MDS/MPN), myelodysplastic syndrome (MDS), myeloproliferative neoplasm (MPN), acute myeloid leukemia (AML), or other myeloid neoplasm^a
- ✓ • The associated myeloid neoplasm should be fully classified according to established criteria^b



Systemic mastocytosis with an associated myeloid neoplasm

Report from the XVI European Bone Marrow Working Group Course and Workshop, Barcelona 2023

Roos J. Leguit, MD,¹ Maria Rozman, MD, PhD,²

Hans Michael Kvasnicka, MD, PhD,³ Robert P. Hasserjian, MD,⁴

Leonor Arenillas, MD, PhD,⁵ Estella Matutes, MD, PhD,²

Fina Climent, MD, PhD,⁶ Attilio Orazi, MD^{7,}*

Diagnóstico diferencial

| LEUCEMIA MIELOMASTOCÍTICA | LEUCEMIA BASOFÍLICA |
|------------------------------|------------------------|
| No cumple criterios de SM | CD117- |
| No hay mutación KIT D816V | CD123+ |



KEY POINTS

- The presence of an AMN in a patient with SM may be difficult to identify. Flow cytometry results and the presence of non-*KIT* mutations may support a diagnosis of SM-AMN.
- The SM may be obscured by an AMN, especially in AML (“occult” SM). Immunostaining for CD117 and tryptase is recommended in all patients with a myeloid neoplasm associated with a *KIT* mutation.
- It may be impossible to distinguish ASM from ISM within a case of SM-AMN, unless the observed “C” findings can be attributed to the SM.



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