SEAP-IAP
[Sociedad Española de Anatomía Patológica]
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115ª Reunión de la Asociación territorial Valenciana de la SEAP

Caso clínico
Patología hematolinfoide

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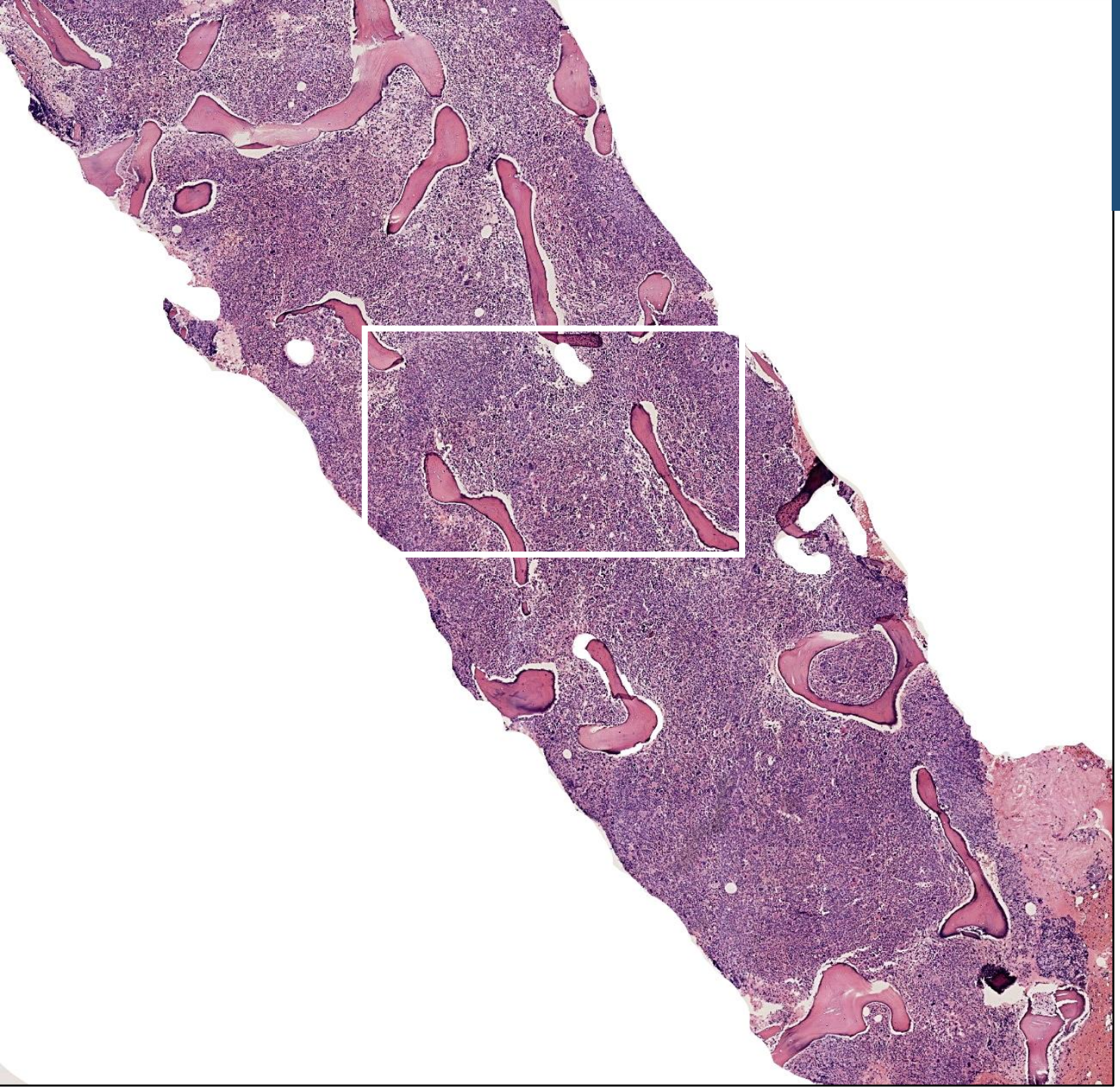


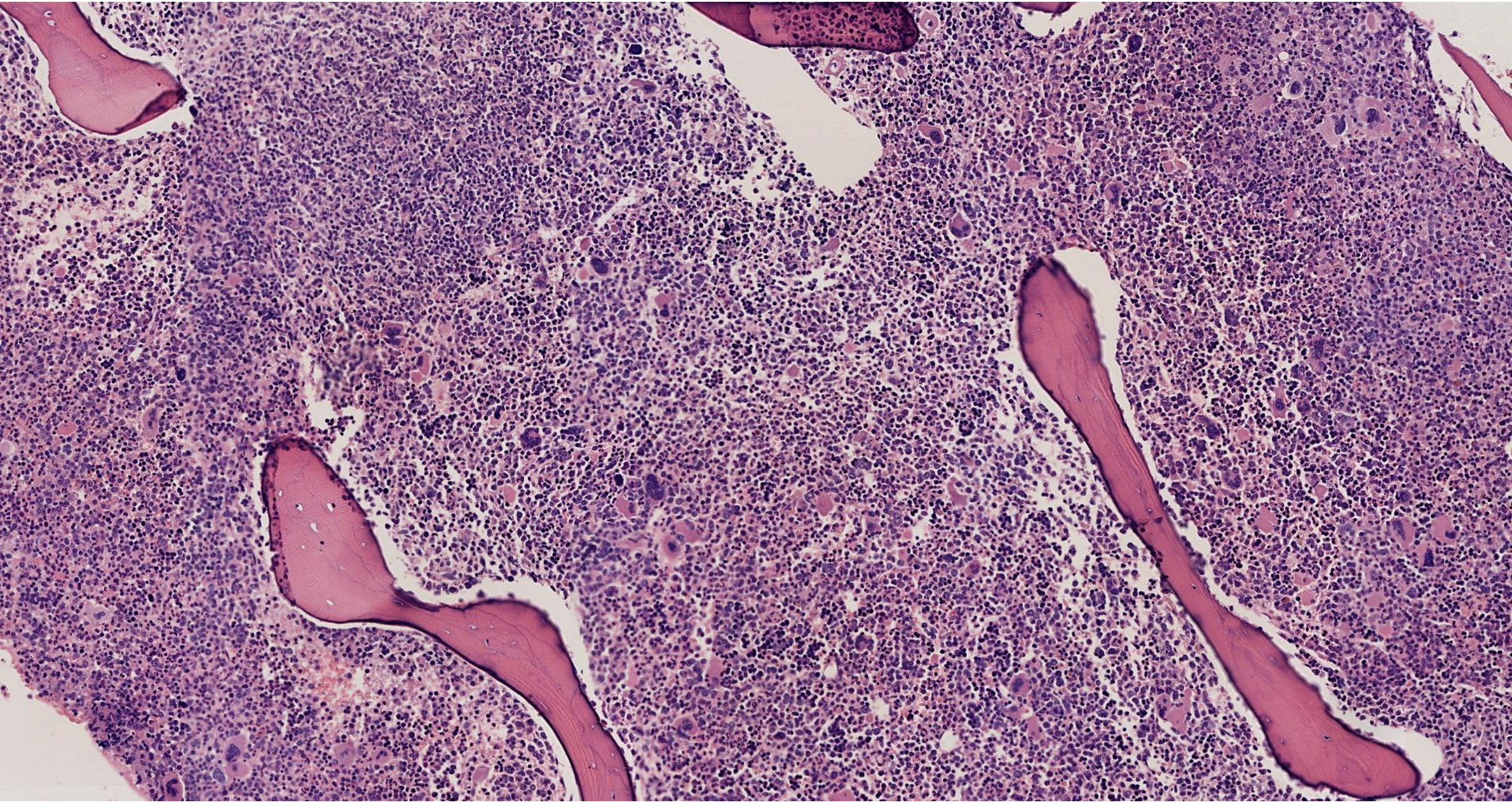
BMO

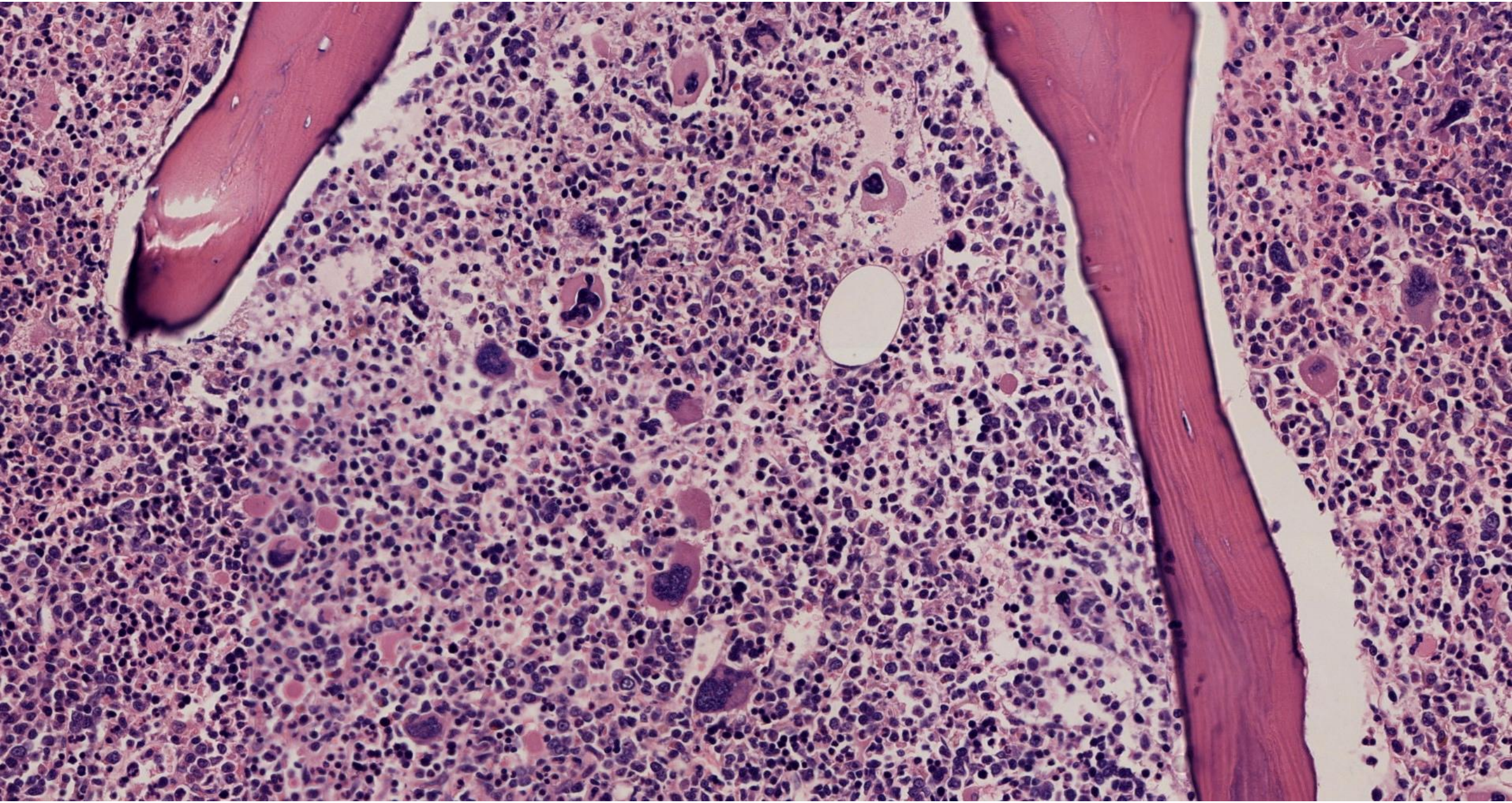
Datos clínicos:

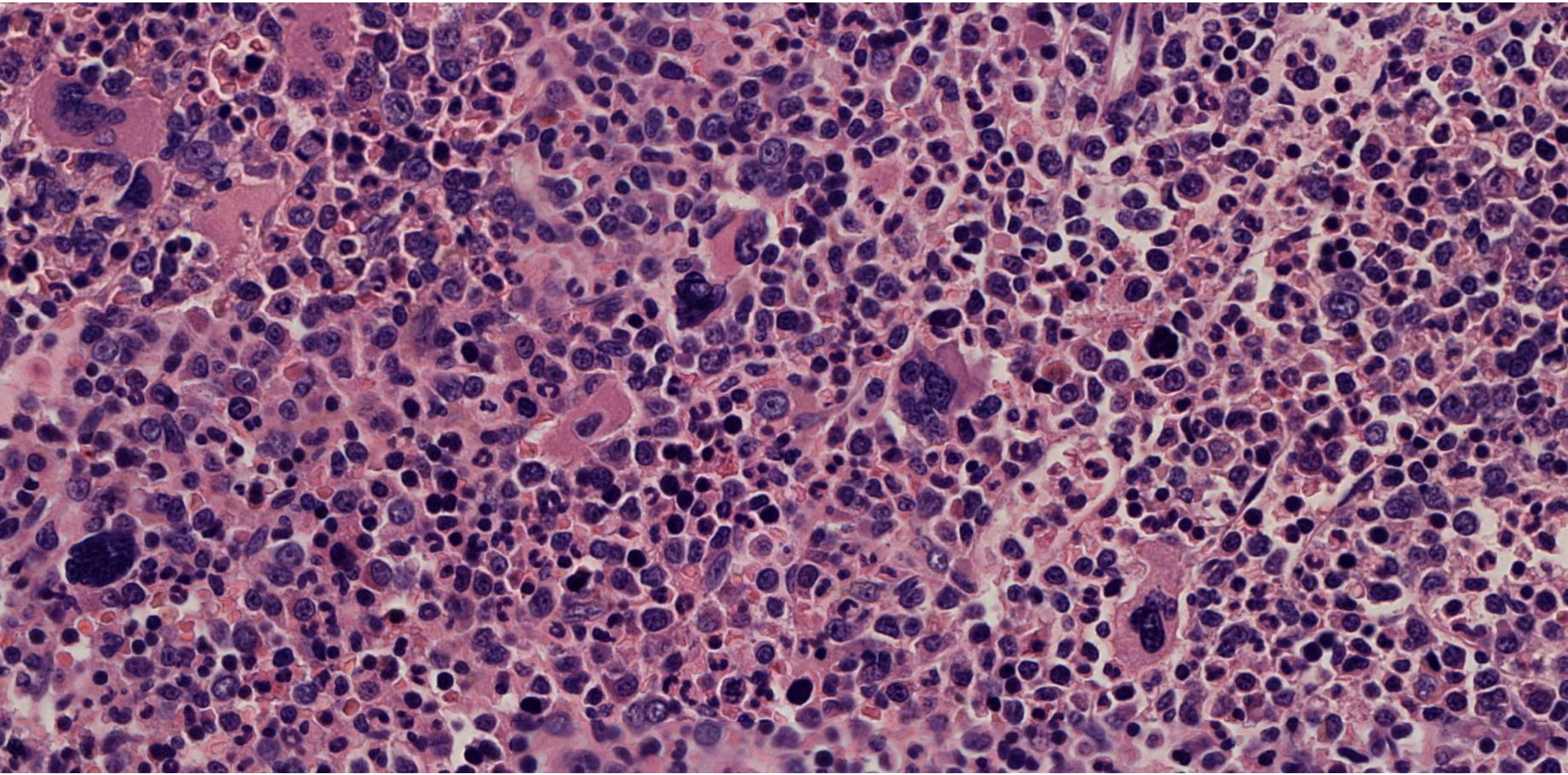
Paciente de 65 años con bicitopenia, cuadro poliadenopático y esplenomegalia

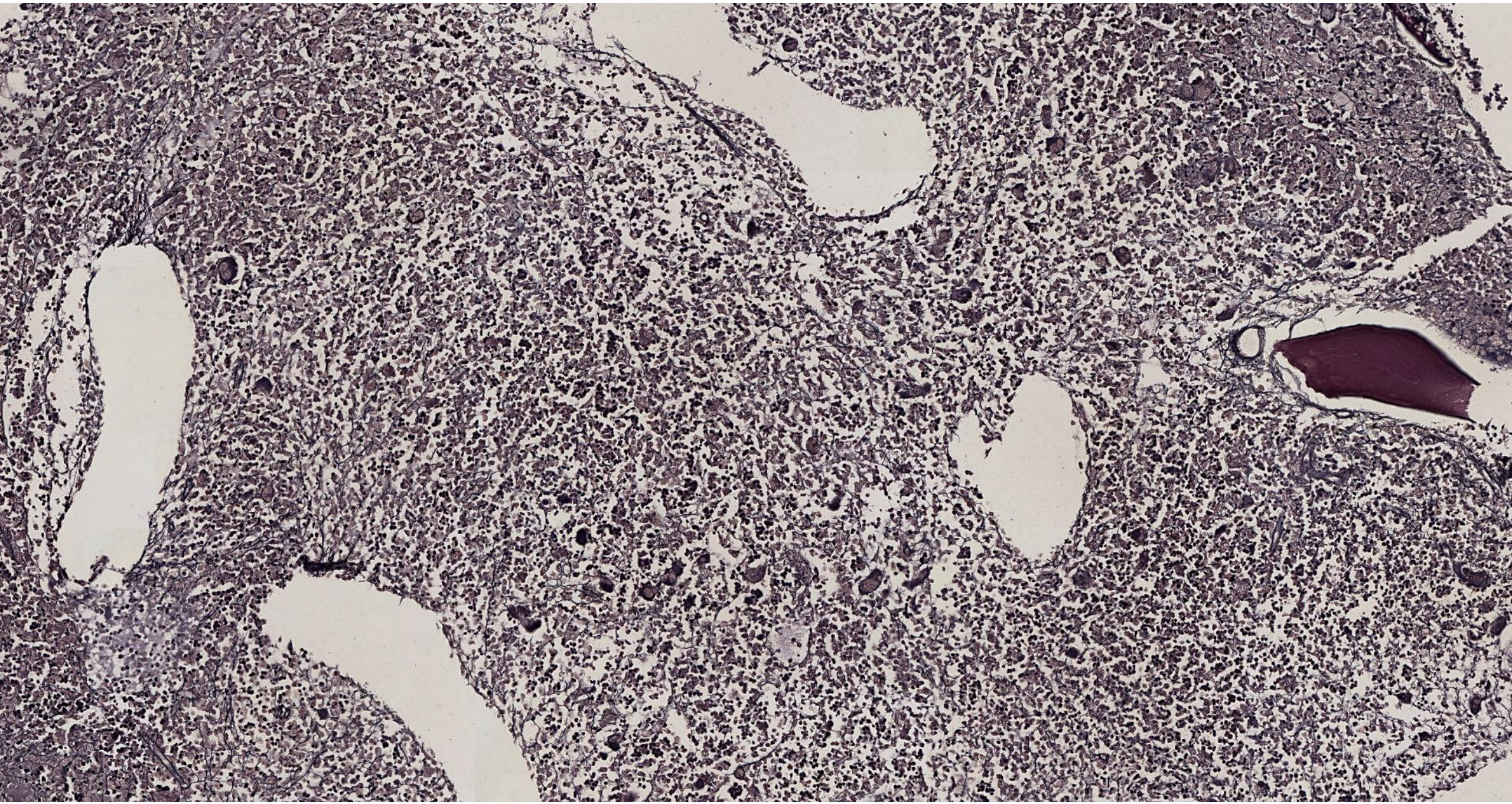
Sospecha diagnóstica:

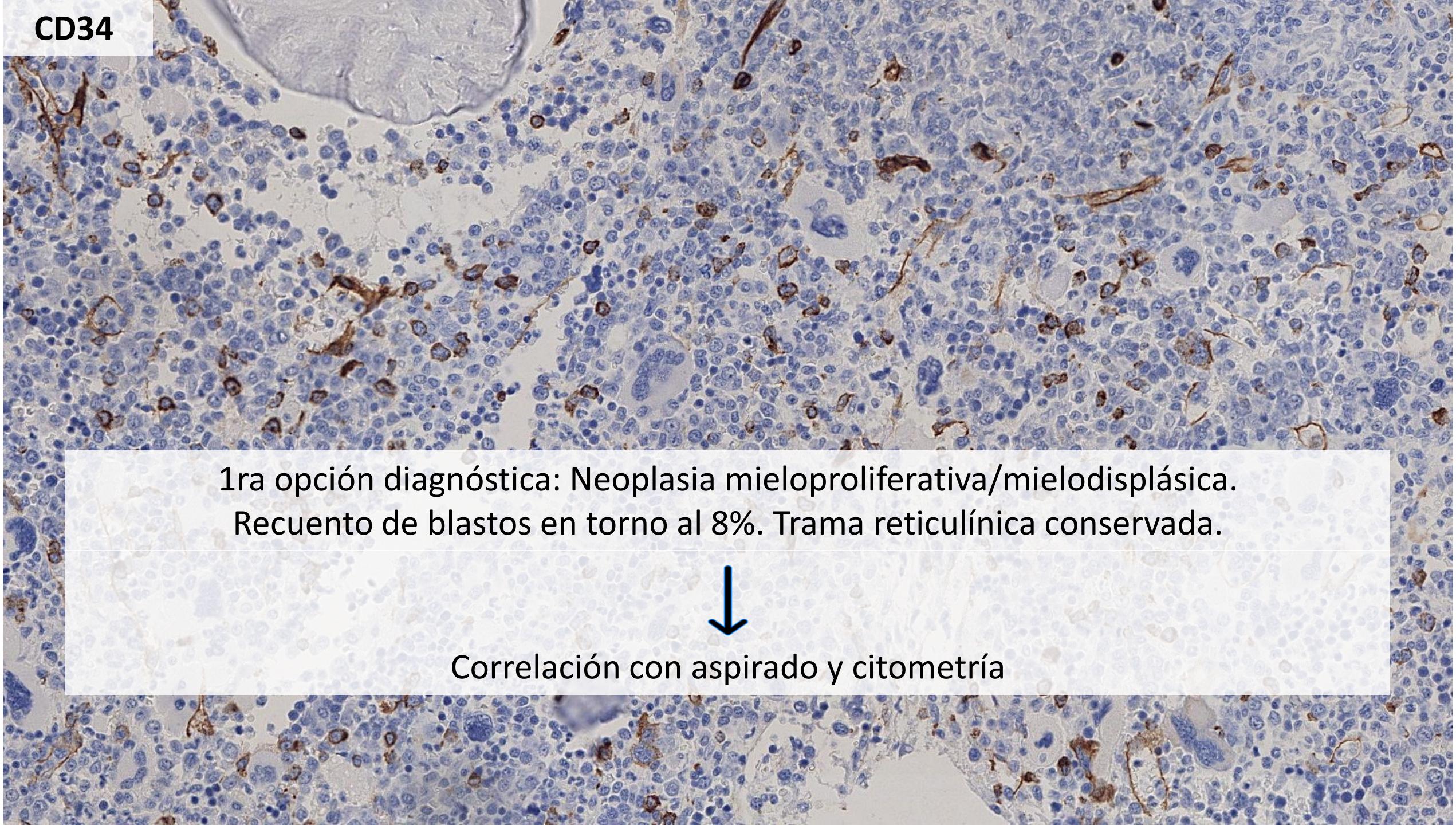
Síndrome mielodisplásico vs síndrome linfoproliferativo











1ra opción diagnóstica: Neoplasia mieloproliferativa/mielodisplásica.
Recuento de blastos en torno al 8%. Trama reticulínica conservada.

↓

Correlación con aspirado y citometría

Presentación clínica

Mujer 65 años
Fumadora, obesidad, HTA y DM.
Trombopenia leve desde 2016 con AMO/BMO normal.

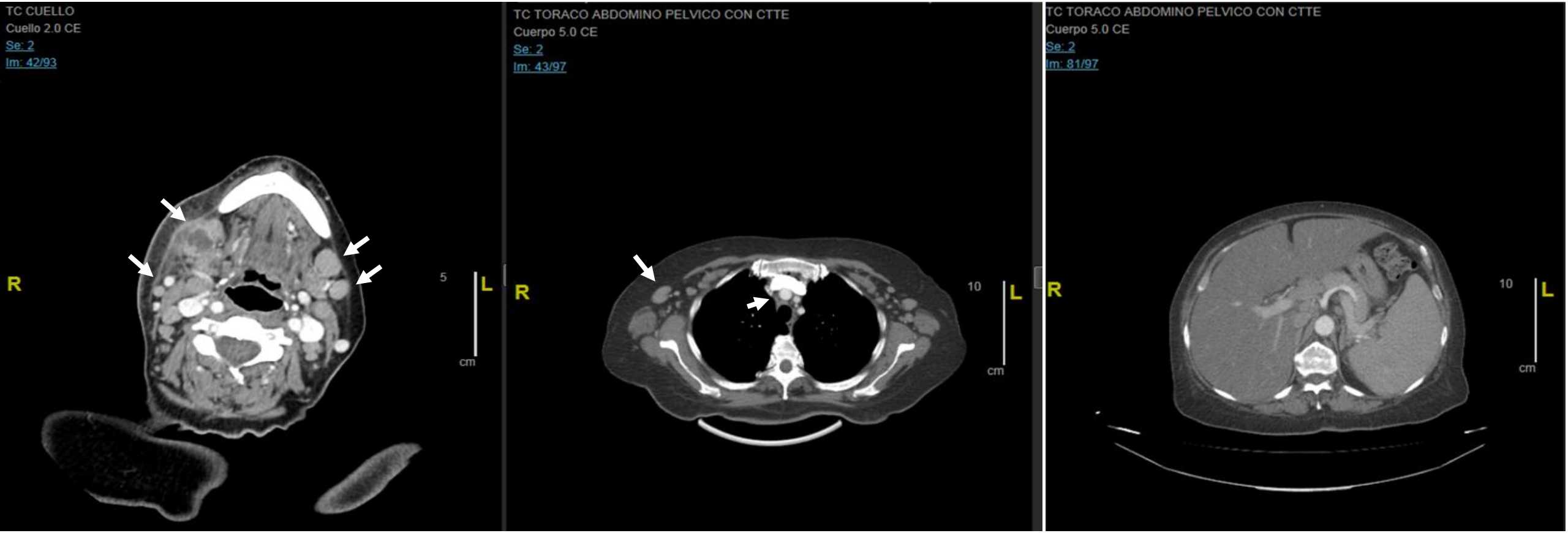
Ingresa en maxilofacial por **adenopatía cervical** abcesificada que requirió drenaje + exodoncia.

- **Analítica urgente:**

Hemograma: Hb 7,6 g/dL, VCM 91fL
Leucocitos 17,77x10^9/L: (Neu 11,38x10^9/L, **Mon 3,65x10^9/L**)
Plaquetas 54x10^9/L.

Pérdida de peso aprox 20 Kg en 6 meses y astenia progresiva.

Presentación clínica



TAC: múltiples adenopatías supra e infradiafragmáticas **sugestivas de síndrome linfoproliferativo**

Presentación clínica

Mujer 65 años
Fumadora, obesidad, HTA y DM.
Trombopenia leve desde 2016 con AMO/BMO normal.

Ingresa en maxilofacial por **adenopatía cervical** abcesificada que requirió drenaje + exodoncia.

Biopsia de adenopatía cervical

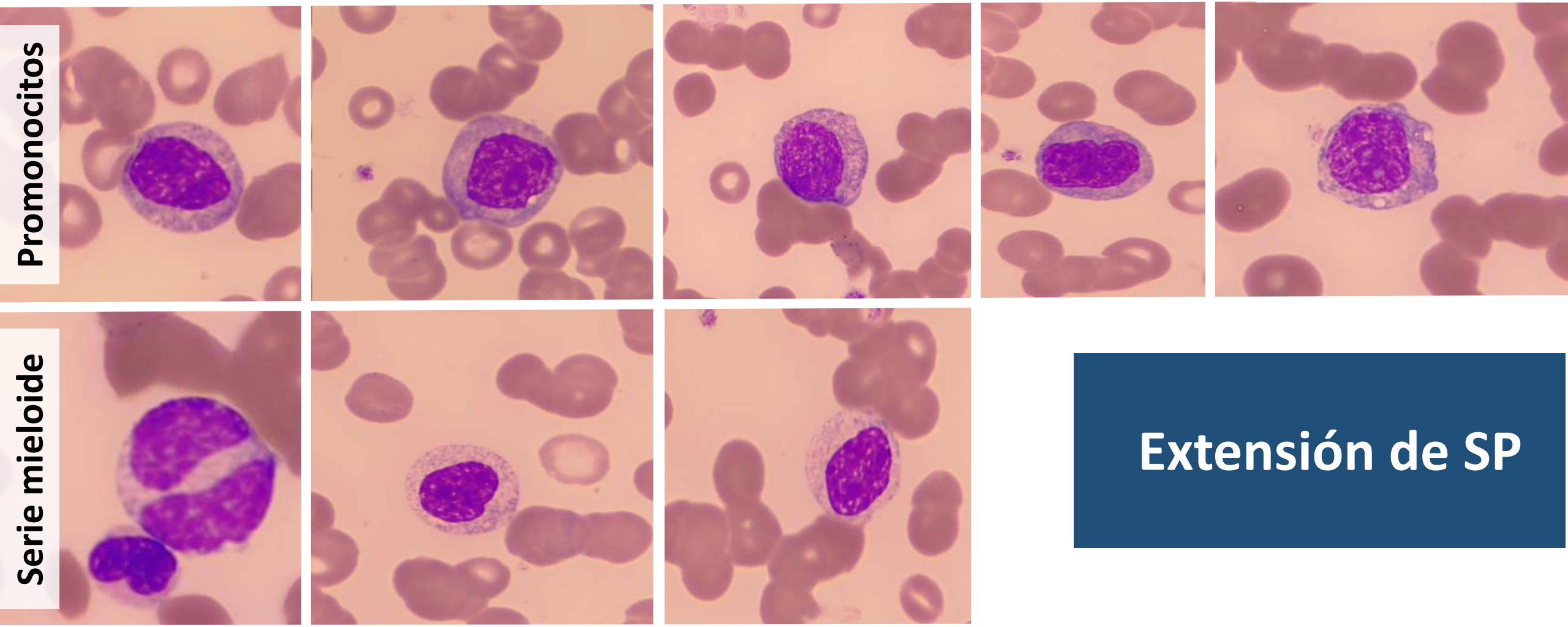
Dx. Linfadenitis reactiva

Biopsia y aspirado de médula ósea

- **Analítica urgente:**

Hemograma: Hb 7,6 g/dL, VCM 91fL
Leucocitos 17,77x10^9/L: (Neu 11,38x10^9/L, **Mon 3,65x10^9/L**)
Plaquetas 54x10^9/L.

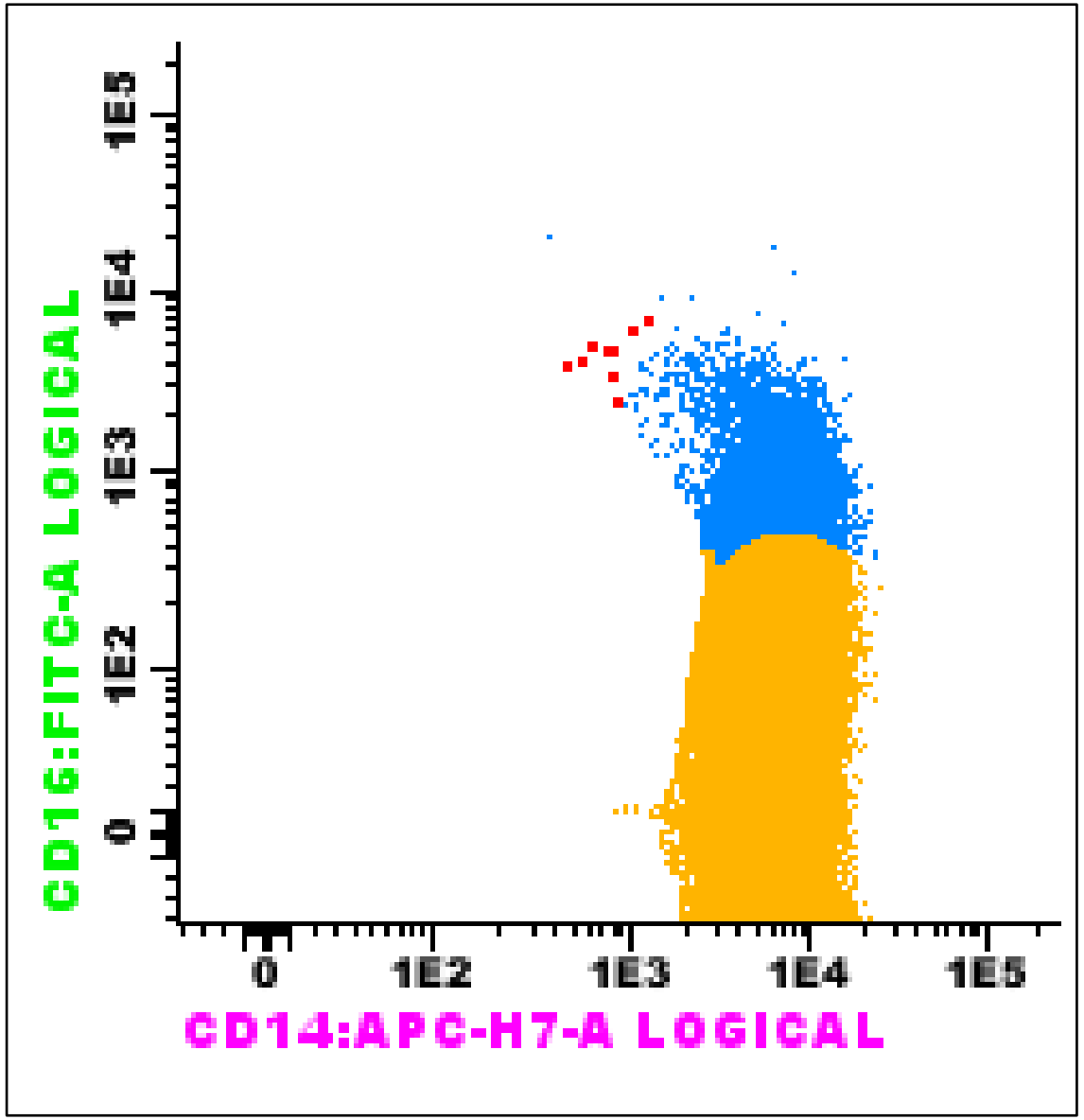
Pérdida de peso aprox 20 Kg en 6 meses y astenia progresiva.



Extensión de SP

Inmunofenotipo de monocitos en SP

- Monocitos clásicos (CD14+/CD16-): 95,11%
- Monocitos intermedios (CD14+/CD16+): 3,52%
- Monocitos no clásicos (CD14+d/CD16+): 0.13%



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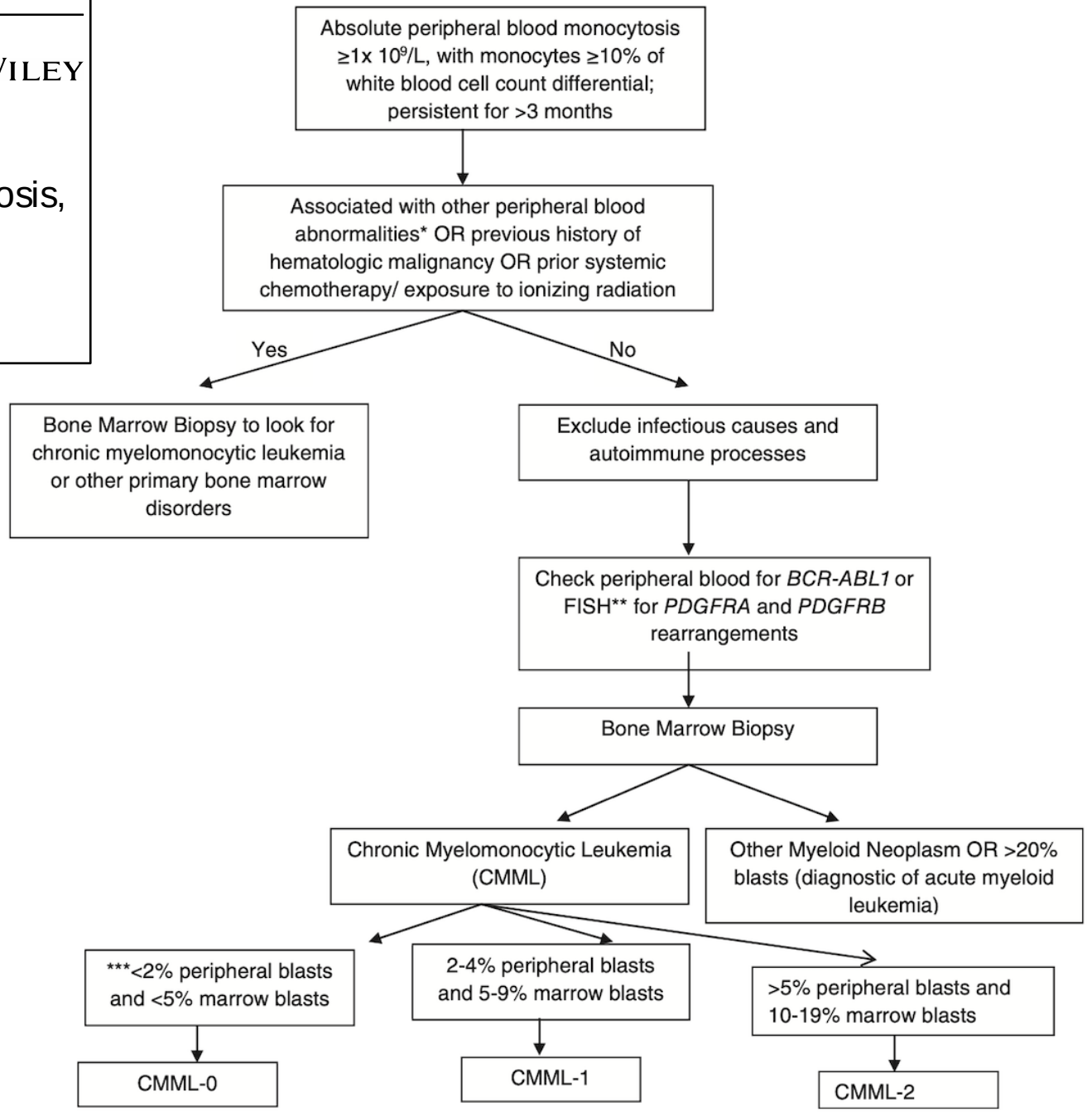
ANNUAL CLINICAL UPDATES IN HEMATOLOGICAL MALIGNANCIES

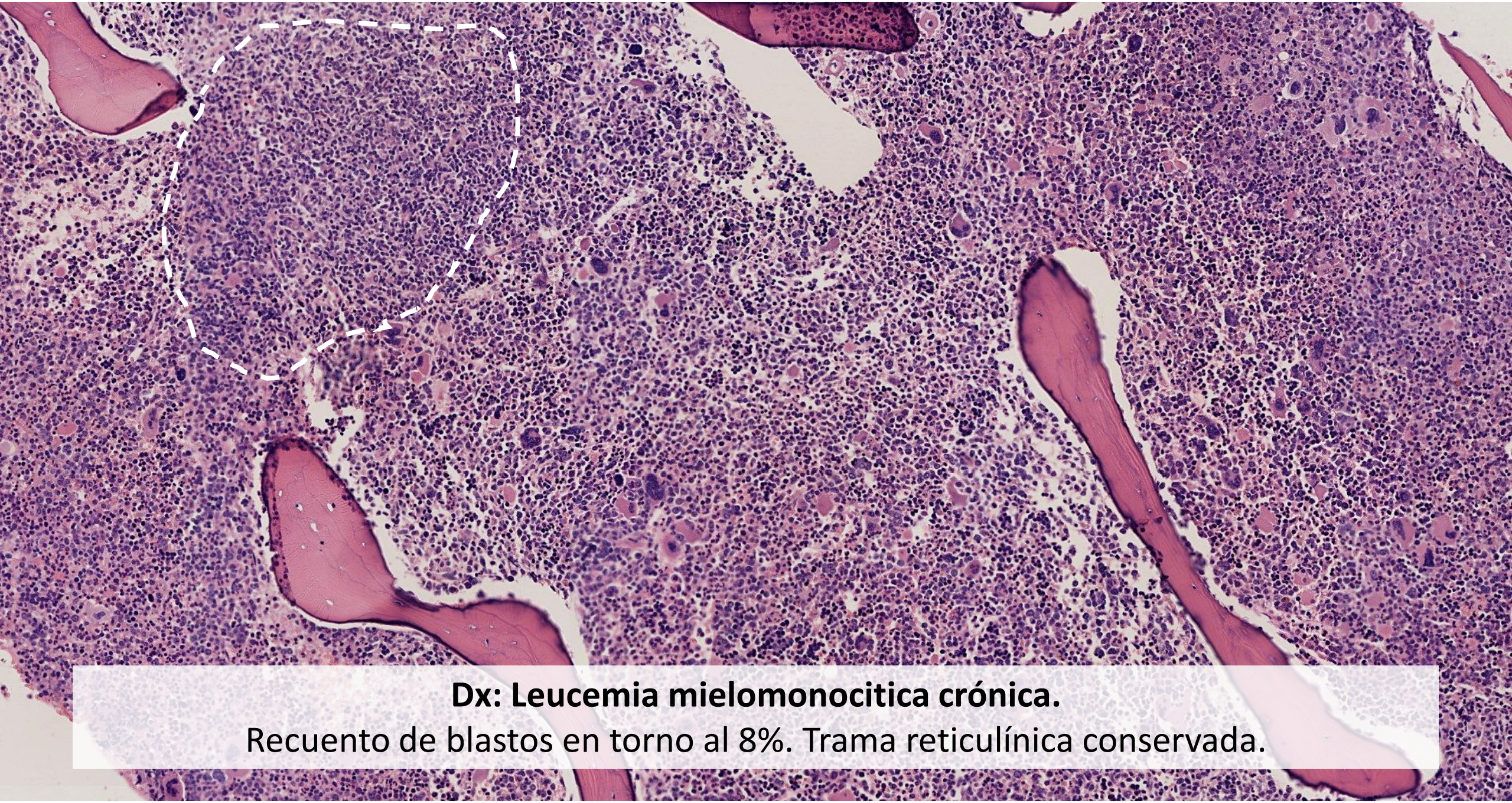
Chronic Myelomonocytic leukemia: 2020 update on diagnosis, risk stratification and management

Mrinal M. Patnaik | Ayalew Tefferi

Disease overview: Chronic myelomonocytic leukemia (CMML) is a clonal hematopoietic stem cell disorder with overlapping features of myelodysplastic syndromes and myeloproliferative neoplasms, with an inherent risk for leukemic transformation (~15% over 3-5 years).

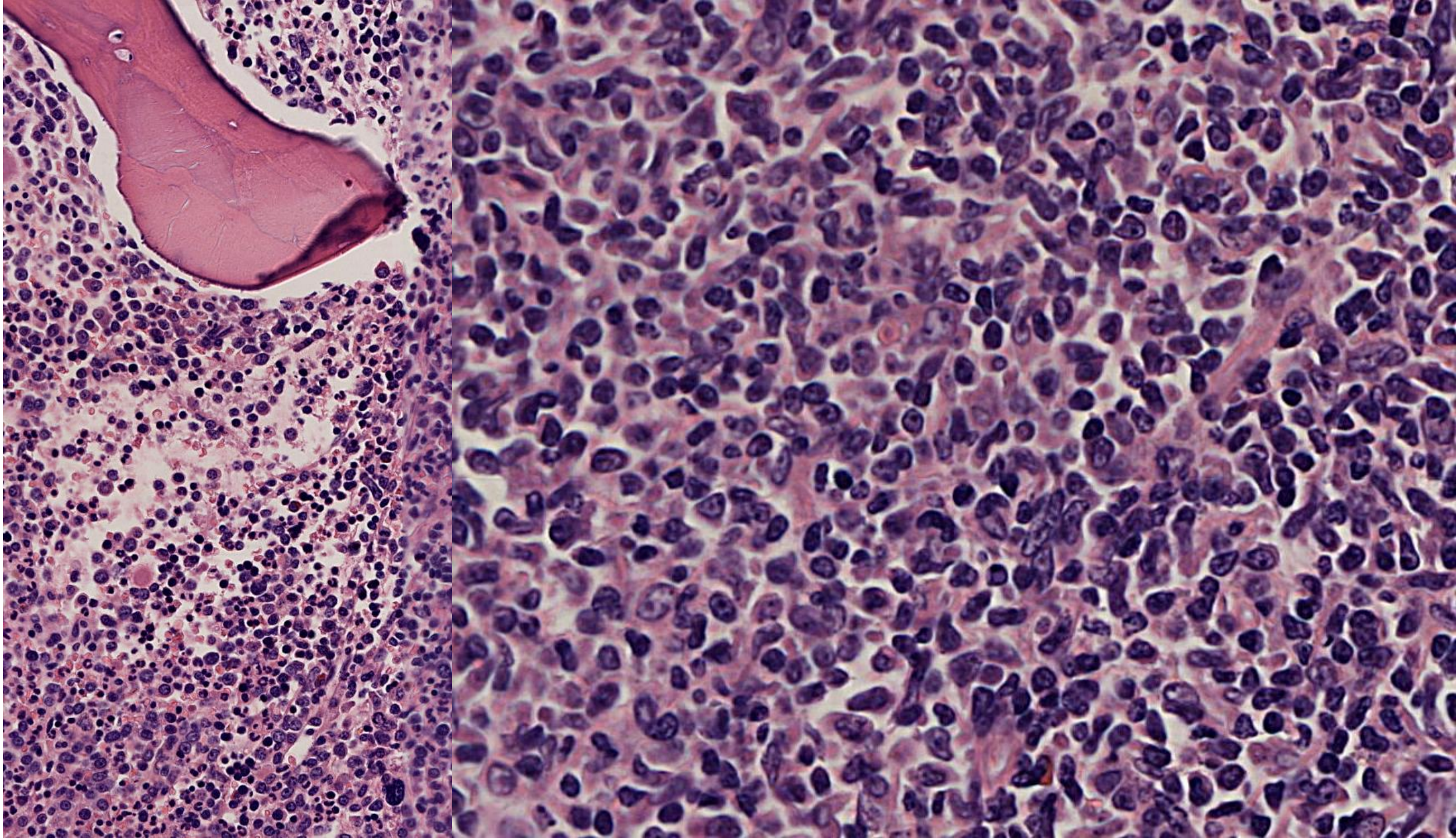
Diagnosis: Diagnosis is based on the presence of sustained (>3 months) peripheral blood monocytosis (≥1 × 10^9/L; monocytes ≥10%), along with bone marrow dysplasia. Clonal cytogenetic abnormalities occur in ~ 30% of patients, while >90% have gene mutations. Mutations involving TET2 (~60%), SRSF2 (~50%), ASXL1 (~40%) and the oncogenic RAS pathway (~30%) are frequent; while the presence of ASXL1 and DNMT3A mutations and the absence of TET2 mutations negatively impact over-all survival.





Dx: Leucemia mielomonocitica crónica.

Recuento de blastos en torno al 8%. Trama reticulínica conservada.



Human Pathology (2013) 44, 512–520

ELSEVIER

Human PATHOLOGY

www.elsevier.com/locate/humpath

Original contribution

Benign lymphoid aggregates in the bone marrow: distribution patterns of B and T lymphocytes

Kaveh Naemi DO[a], Russell K. Brynes MD[b], Niloufar Reisian MD[a], Abbey Johnston MD[a], Ramandeep Dhillon MS[a], Vighnesh Walavalkar MD[a], Xiaohui Zhao MD, PhD[a], Sherif A. Rezk MD[a,*]

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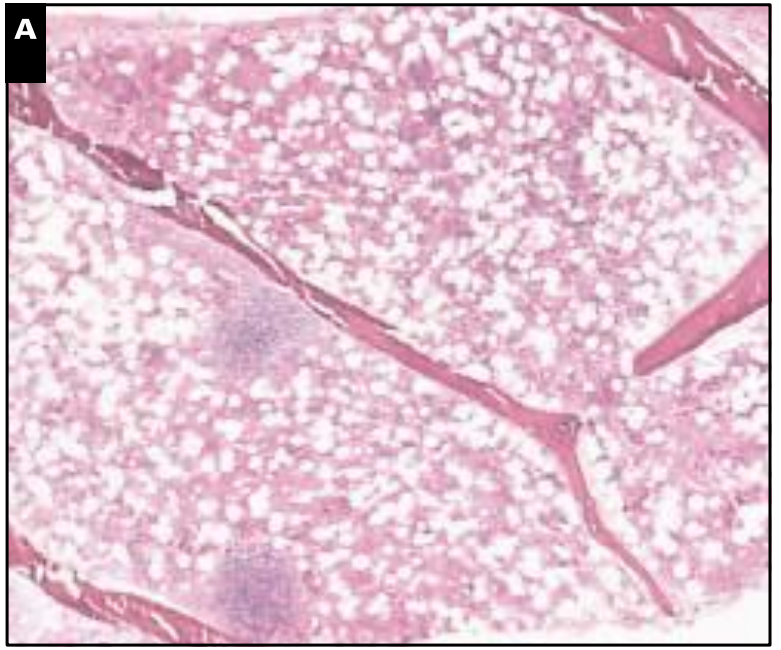
[b]*Department of Pathology, University of Southern California Keck School of Medicine, Los Angeles, CA 90033, USA*

Summary Benign lymphoid aggregates are seen in only a minority of bone marrow specimens, but their distinction from non–Hodgkin lymphoma, particularly B-cell lymphomas, can represent a diagnostic challenge. Although criteria have been proposed to help distinguish between benign and malignant aggregates, a detailed description of the distribution patterns of B and T lymphocytes within benign lymphoid aggregates has not been investigated. One hundred thirty-seven cases of bone marrow specimens containing benign aggregates were studied with a panel of immunostains. A subset of these cases was also examined for immunoglobulin gene rearrangements by polymerase chain reaction. The aggregates were categorized based on size, location (paratrabecular or random), presence of infiltrating edges, and distribution of lymphoid cell populations. In addition, we examined 40 cases of bone marrow biopsies with documented malignant lymphoid aggregates for comparison purposes. We report that the distribution of B and T lymphocytes within lymphoid aggregates may serve as a useful criterion to aid in the separation between benign and malignant aggregates. When aggregates exhibit a predominance of T cells, consist of a central core of T cells surrounded by a rim of B cells, or have a mixed distribution of B and T cells, they are more likely to be benign. On the other hand, an increased likelihood of malignancy occurs when aggregates exhibit a predominance of B cells or consist of a central core of B cells surrounded by a rim of T cells (excluding germinal center formation), and assessing other features worrisome of malignancy (large aggregate size, presence of infiltrative edges, cellular atypia, and paratrabecular location, among others) is warranted.

Follicular Pattern of Bone Marrow Involvement by Follicular Lymphoma

Emina Torlakovic, MD,[1] Goran Torlakovic, MD,[1] and Richard D. Brunning, MD[2]

Key Words: Follicular pattern; Bone marrow; Follicular lymphoma



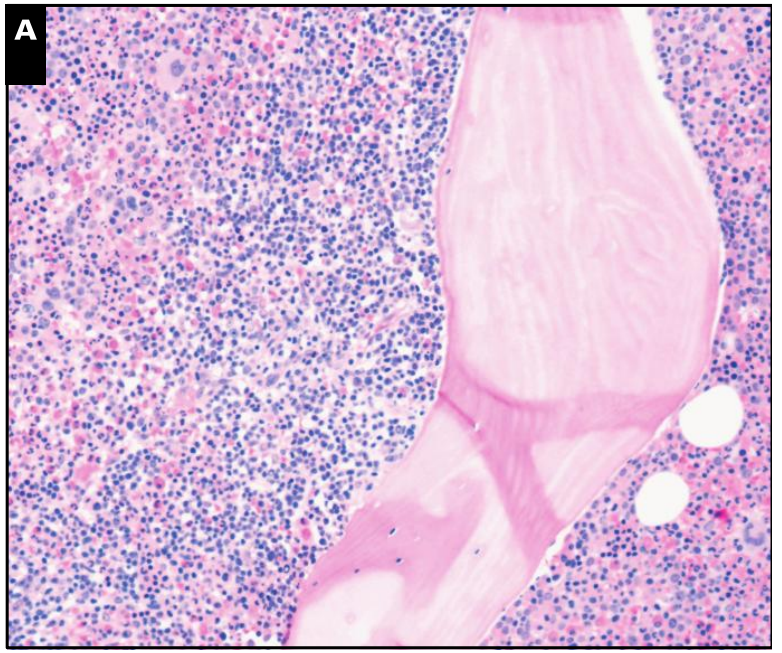
Lymphoplasmacytic Lymphoma and Marginal Zone Lymphoma in the Bone Marrow

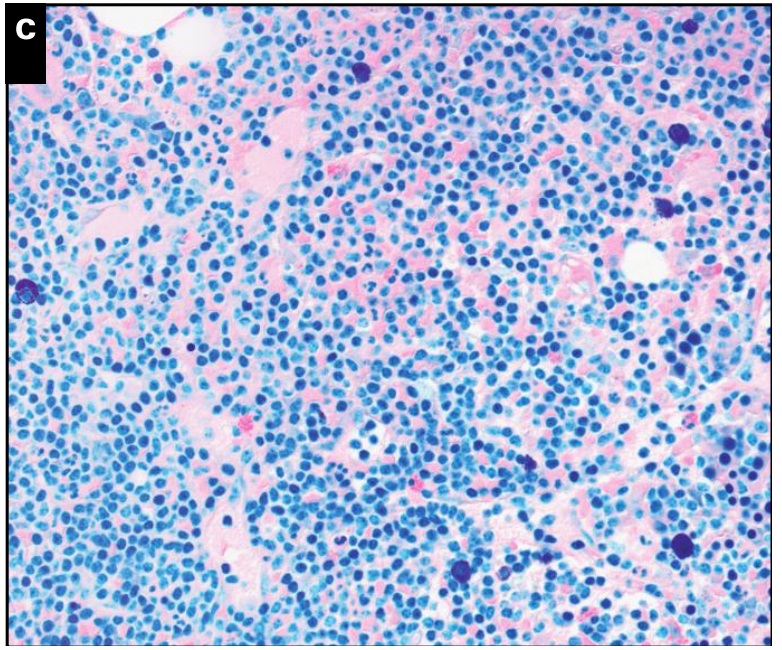
Paratrabecular Involvement as an Important Distinguishing Feature

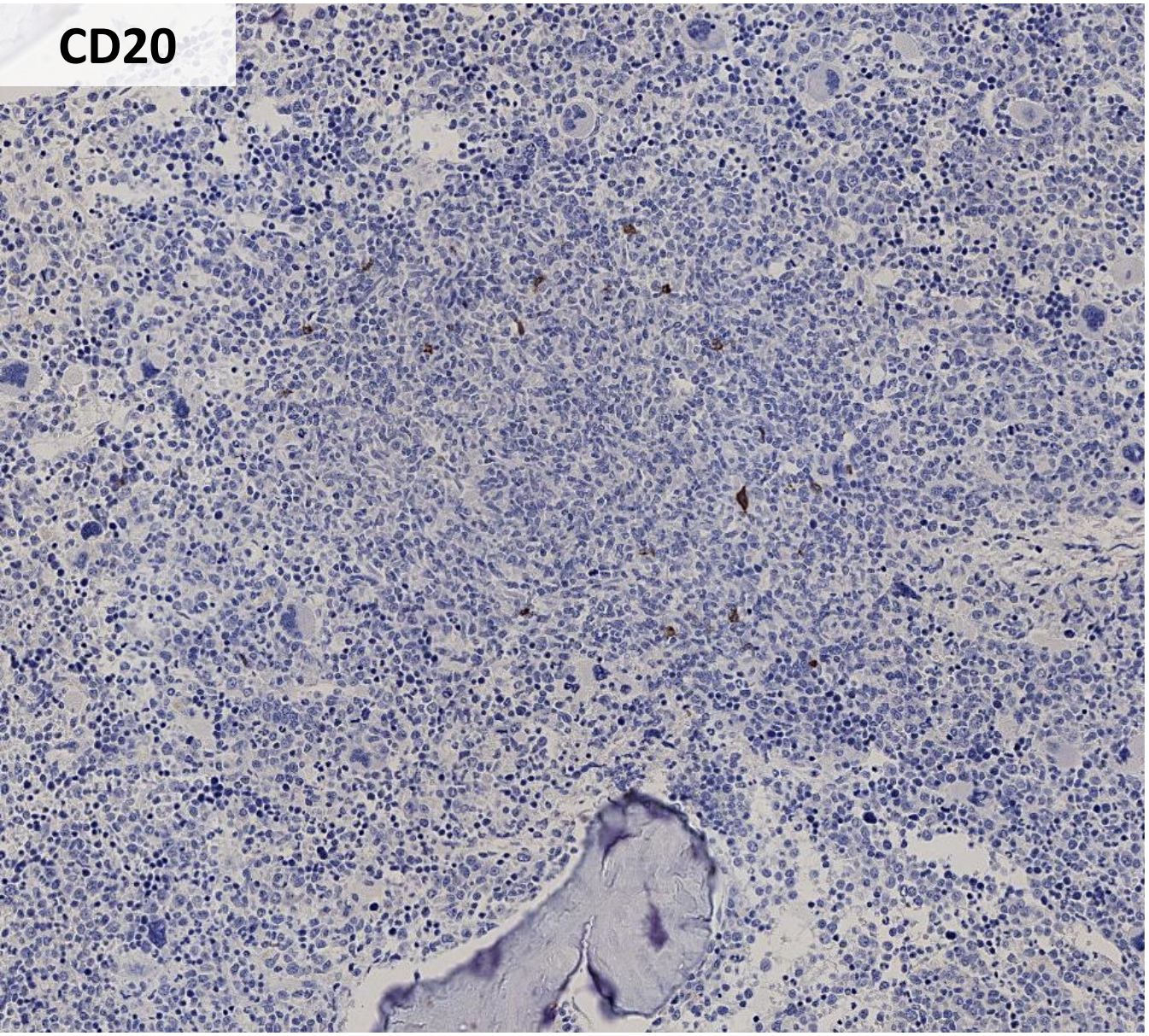
Assia Bassarova, MD, PhD,[1,2] *Gunhild Trøen, PhD,*[2,3] *Signe Spetalen, MD, PhD,*[3] *Francesca Micci, PhD,*[4,5] *Anne Tierens, MD, PhD,*[6] *and Jan Delabie, MD, PhD*[2,6]

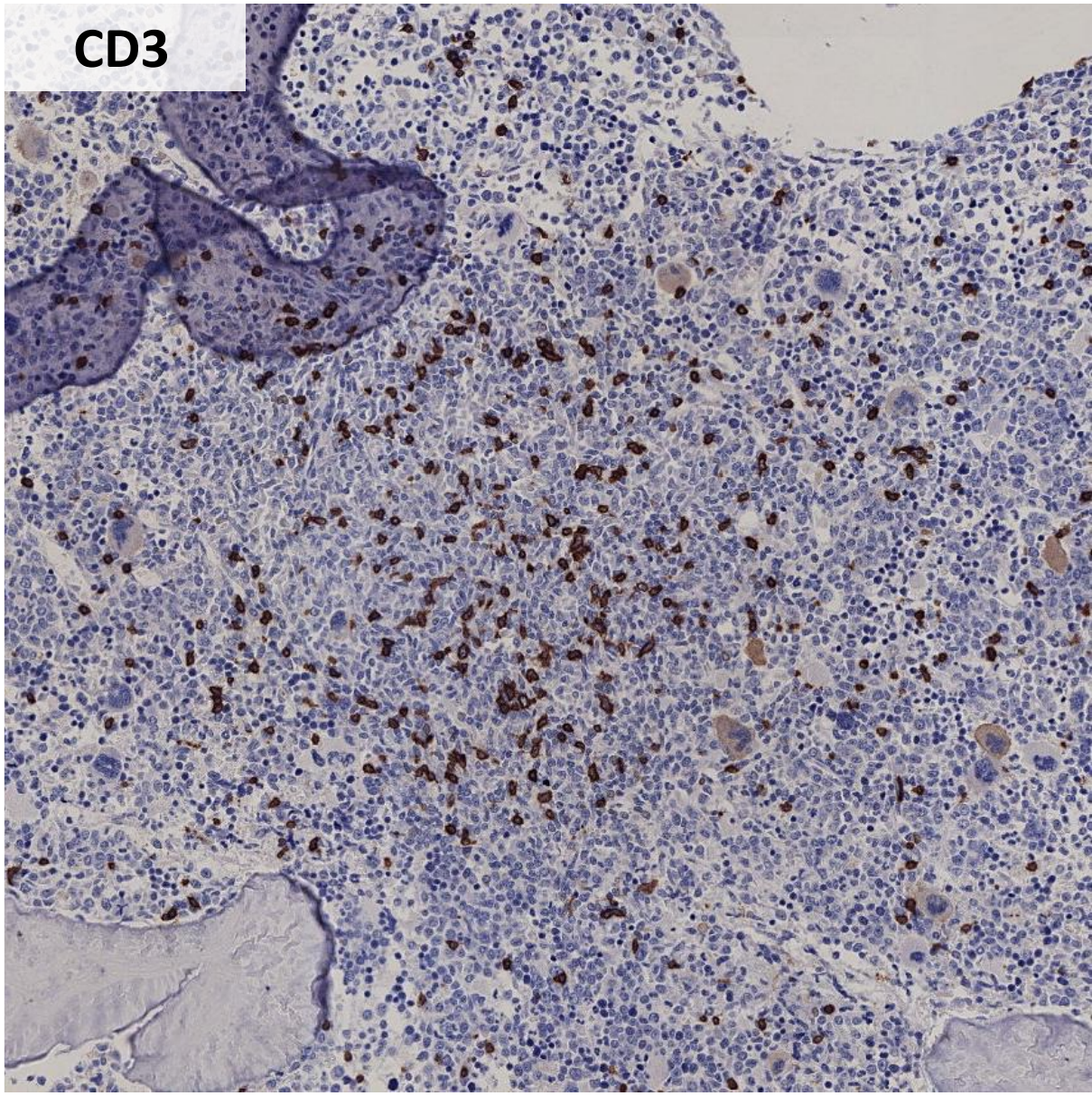
From the [1]Department of Pathology, Akershus University Hospital, Oslo, Norway; [2]University of Oslo, Oslo, Norway; [3]Department of Pathology and [4]Section for Cancer Cytogenetics, Institute for Cancer Genetics and Informatics, The Norwegian Radium Hospital, Oslo University Hospital, Oslo, Norway; [5]Centre for Cancer Biomedicine, University of Oslo, Oslo, Norway; and [6]Department of Pathology, University Health Network, Toronto, Canada.

Characteristic	No./Total No. (%)		
	Lymphoplasmacytic Lymphoma	Marginal Zone Lymphoma	*P* Value
Infiltration pattern[a]			
Paratrabecular	10/27 (37)	0/16 (0)	<.001
Nodular nonparatrabecular	0/27 (0)	12/16 (75)	<.001
Paratrabecular and nonparatrabecular	15/27 (56)	0/16 (0)	<.001
Intrasinusoidal	10/27 (37)	6/16 (38)	1
Diffuse	0/27 (0)	4/16 (25)	.015






CD20


CD3

Received: 29 November 2018 | Accepted: 5 December 2018
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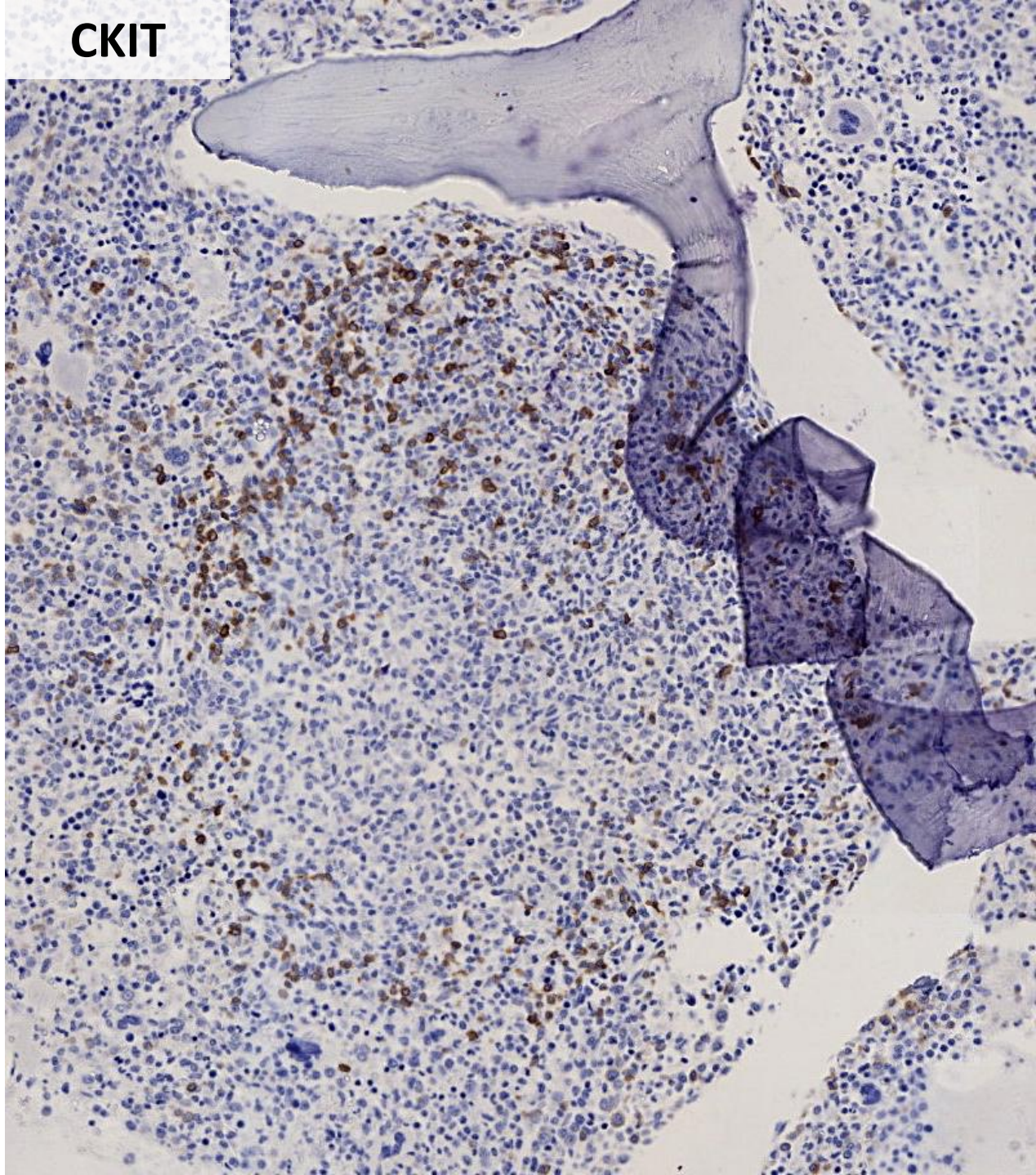
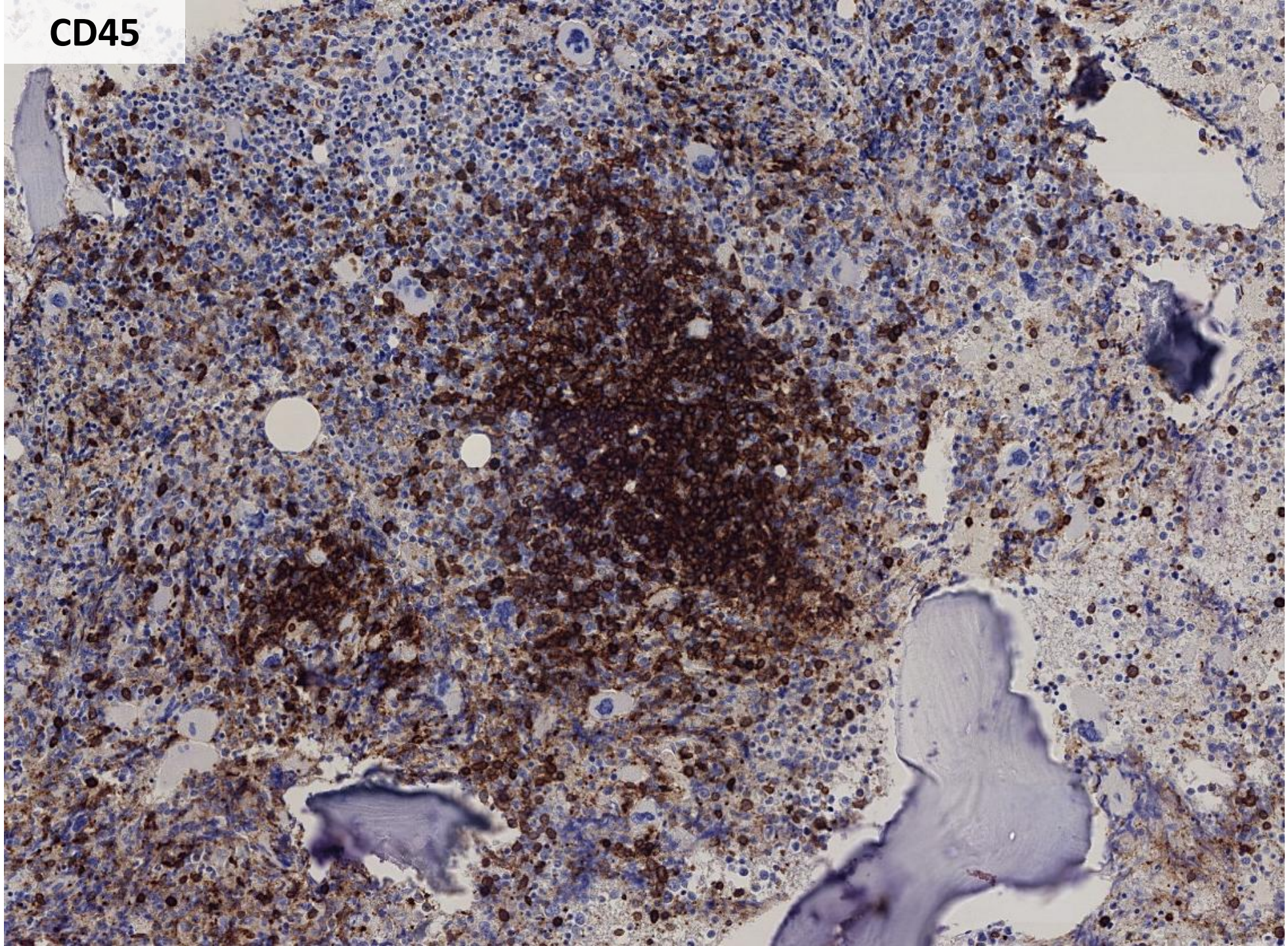
WILEY AJH

ANNUAL CLINICAL UPDATES IN HEMATOLOGICAL MALIGNANCIES

Systemic mastocytosis in adults: 2019 update on diagnosis, risk stratification and management

In general, the pathognomonic multifocal dense MC aggregates, frequently in perivascular and/or paratrabecular BM locations (major diagnostic criterion), may not be readily recognized by standard dyes such as Giemsa, particularly when MC exhibit significant hypogranulation or abnormal nuclear morphology, or in cases with extensive BM involvement by a second hematological neoplasm (eg, AML), or when significant reticulin fibrosis is present. Among the immunohistochemical markers, tryptase is the most sensitive, given that virtually all MC, irrespective of their stage of maturation, activation status, or tissue of localization express this marker, and consequently allows for detection of even small and/or immature MC infiltrates.[56–58] It must be




CD45

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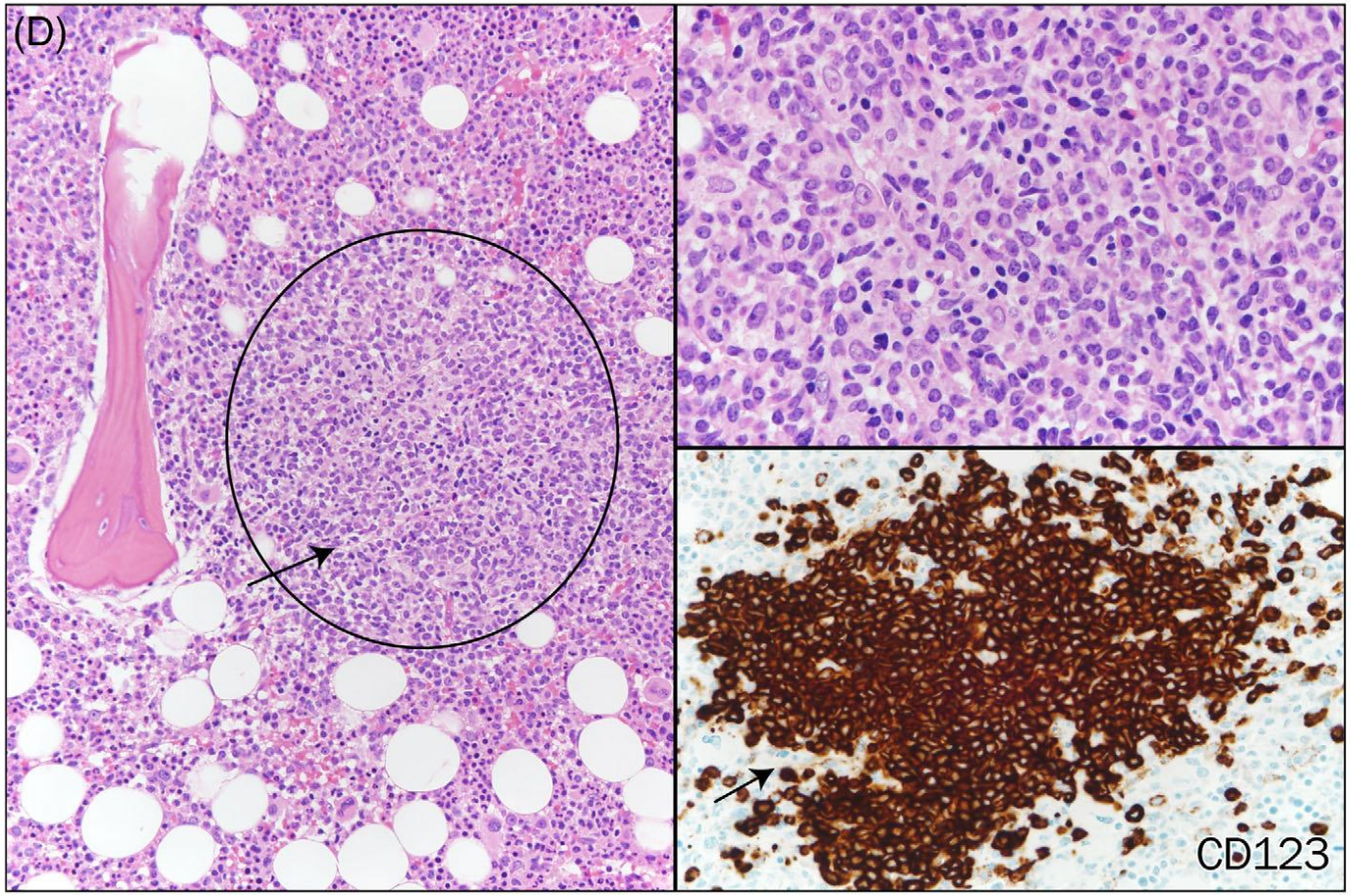
ANNUAL CLINICAL UPDATES IN HEMATOLOGICAL MALIGNANCIES

Chronic Myelomonocytic leukemia: 2020 update on diagnosis, risk stratification and management

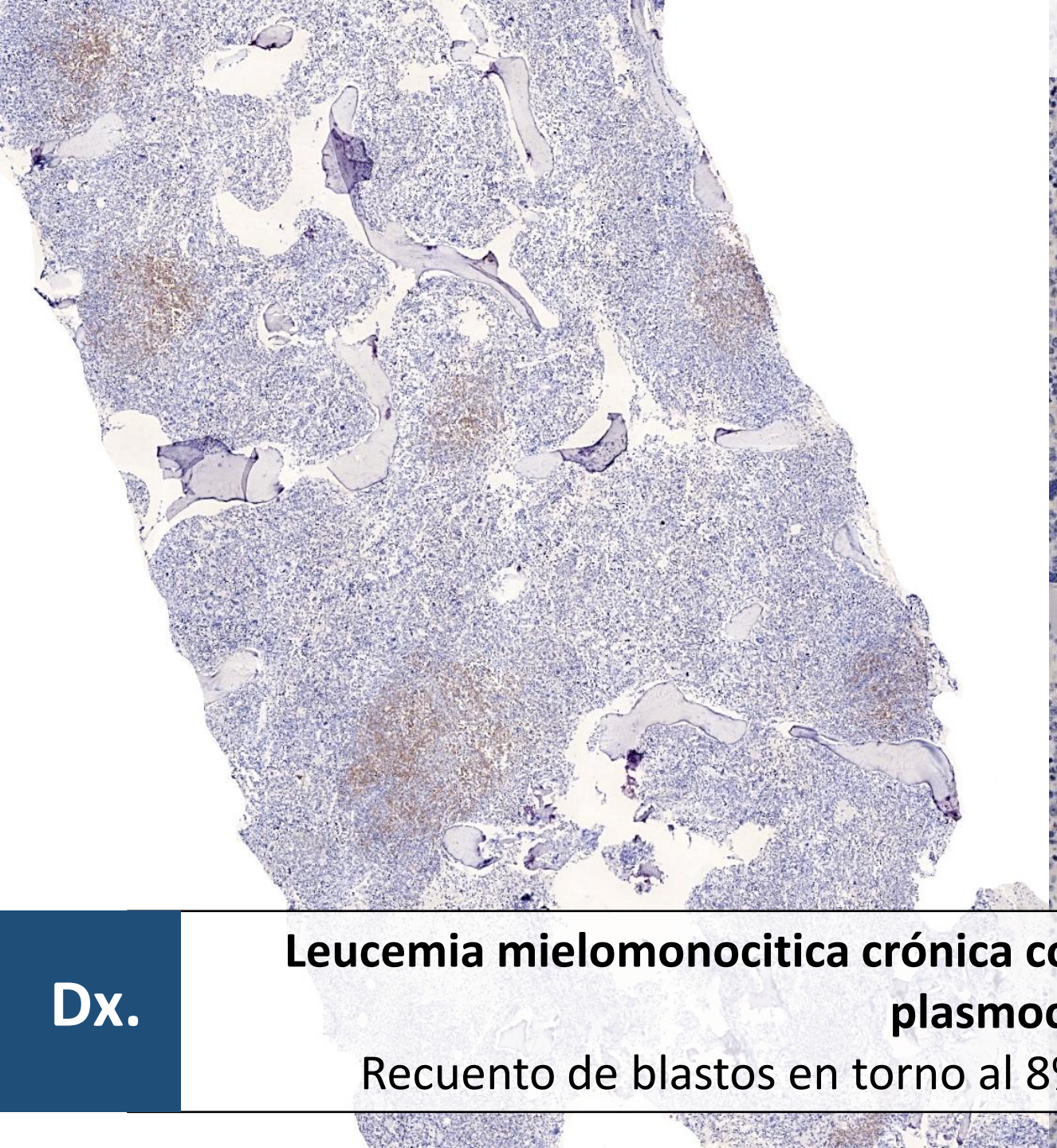
Mrinal M. Patnaik | Ayalew Tefferi

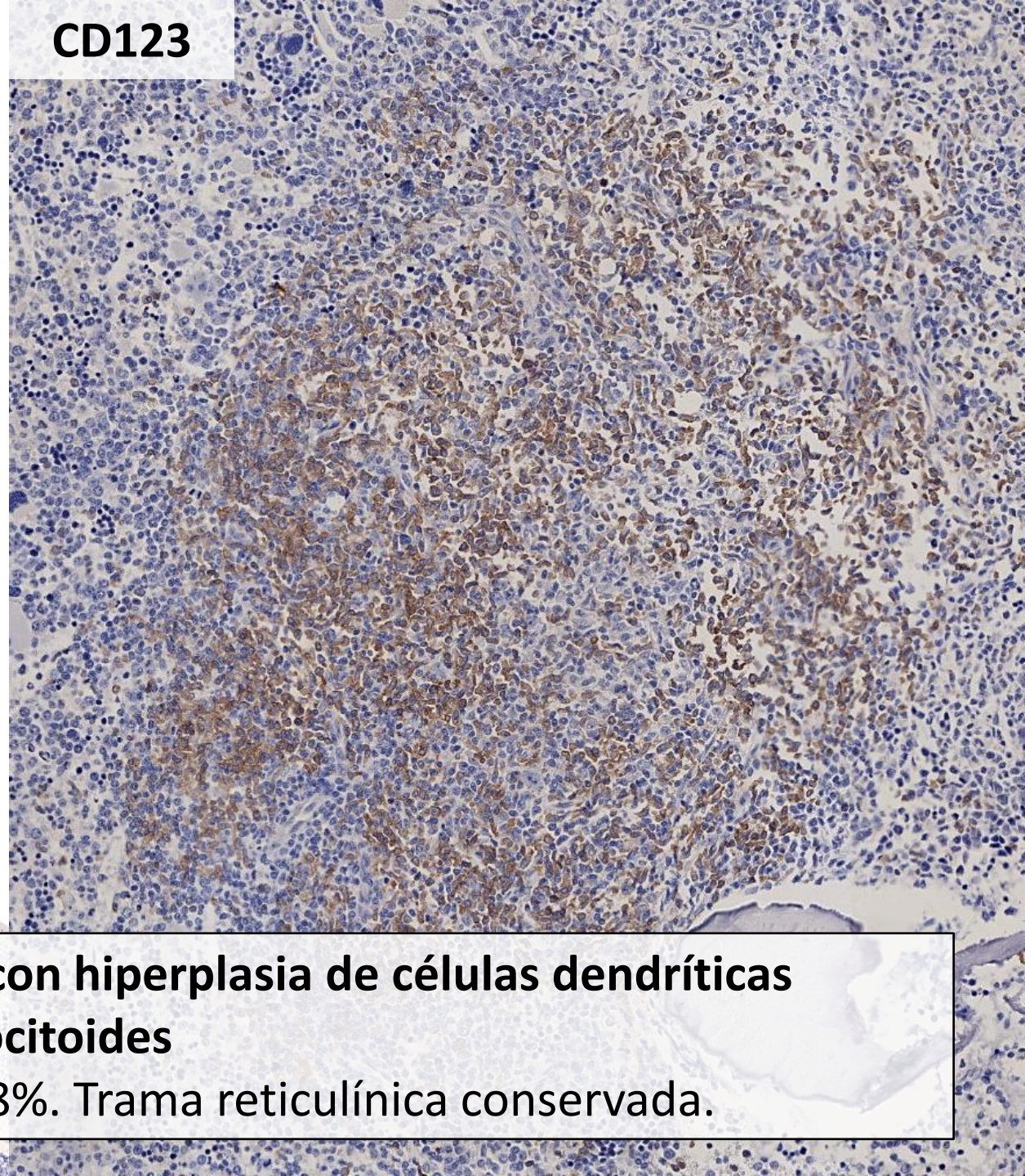


ncrease in BM reticulin fibrosis (Figure 2C and 3D,E).[33] Approxi-
mately 30% percent of patients can demonstrate nodules composed
of mature plasmacytoid dendritic cells that are clonal [CD123+, line-
age negative, CD45+, CD11c-,CD33-, HLA-DR+, BDCA-2+ and
BDCA-4+], often have *RAS* pathway mutations and predict for an
inferior leukemia-free survival (LFS) (Figure 2D).[34] The identification

30% de los casos de LMMC presentan agregados de células dendríticas plasmocitoides en MO.

Estas células son CD45+ y CD123+





Dx.

Leucemia mielomonocitica crónica con hiperplasia de células dendríticas plasmocitoides

Recuento de blastos en torno al 8%. Trama reticulínica conservada.

ARTICLE

Myelodysplastic syndrome

Biology and prognostic impact of clonal plasmacytoid dendritic cells in chronic myelomonocytic leukemia

Nolwenn Lucas[1,2,3] · Matthieu Duchmann[1] · Philippe Rameau[4] · Floriane Noël[5] · Paula Michea[5] · Véronique Saada[3] · Olivier Kosmider[6,7] · Gérard Pierron[4] · Martin E Fernandez-Zapico[8,9] · Matthew T. Howard[8,9] · Rebecca L. King[8,9] · Sandrine Niyongere[10] · M'boyba Khadija Diop[1,4] · Pierre Fenaux[11] · Raphael Itzykson[11] · Christophe Willekens[1,3] · Vincent Ribrag[1,3] · Michaela Fontenay[6,7] · Eric Padron[10] · Vassili Soumelis[5] · Nathalie Droin[1,4] · Mrinal M Patnaik[12] · Eric Solary[1,2,3]

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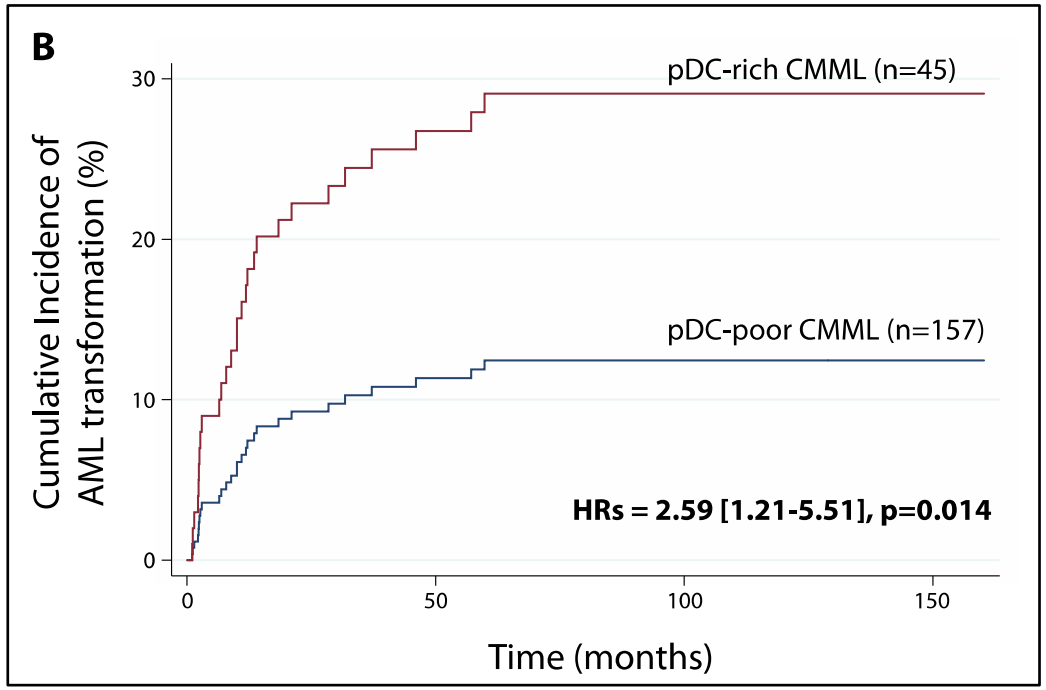
Abstract

Islands of CD123^{high} cells have been commonly described in the bone marrow of patients with chronic myelomonocytic leukemia (CMML). Using a multiparameter flow cytometry assay, we detected an excess of CD123^{+} mononucleated cells that are lineage-negative, CD45^{+}, CD11c^{-}, CD33^{-}, HLA-DR^{+}, BDCA-2^{+}, BDCA-4^{+} in the bone marrow of 32/159 (20%) patients. Conventional and electron microscopy, flow cytometry detection of cell surface markers, gene expression analyses, and the ability to synthesize interferon alpha in response to Toll-like receptor agonists identified these cells as bona fide plasmacytoid dendritic cells (pDCs). Whole-exome sequencing of sorted monocytes and pDCs identified somatic mutations in genes of the oncogenic RAS pathway in the two cell types of every patient. CD34^{+} cells could generate high amount of pDCs in the absence of FMS-like tyrosine kinase 3-ligand (FLT3L). Finally, an excess of pDCs correlates with regulatory T cell accumulation and an increased risk of acute leukemia transformation. These results demonstrate the FLT3L-independent accumulation of clonal pDCs in the bone marrow of CMML patients with mutations affecting the RAS pathway, which is associated with a higher risk of disease progression.



Células dendríticas plasmocitoides en MO en LMMC se asocia a trastornos autoinmunes que sugieren desregulación inmune generalizada.

Indican mayor riesgo de progresión de leucemia aguda.

Asociado con mutaciones de la vía RAS.

Perfil mutacional:

- **ASXL1** p.(Gly646Trpfs*12) exon 13 VAF 38.4 %
- **SRSF2** p.(Pro95Arg) exon 1 VAF 42.1 %
- **KRAS** p.(Gly12Arg) exon 2 VAF 30.9 %
- **NRAS** p.(Gly12Val) exon 2 VAF 11.9 %

Tratamiento

- Múltiples comorbilidades previas.
- Ingreso por insuficiencia cardiaca asociada a hipoalbuminemia.

Azacitidina + soporte transfusional + bolus de metilprednisolona

Median overall survival of CMML patients ranges between 15 and 30 months.
25% of these patients die from disease transformation into acute myeloid leukemia (AML)

Gracias

Carolina Martinez Ciarpaglini

Hospital Clinico Universitario de Valencia.
Fundación biosanitaria INCLIVA. Universidad de Valencia
Departamento de Patologia. Hospital Quiron Valencia.

@carolpath

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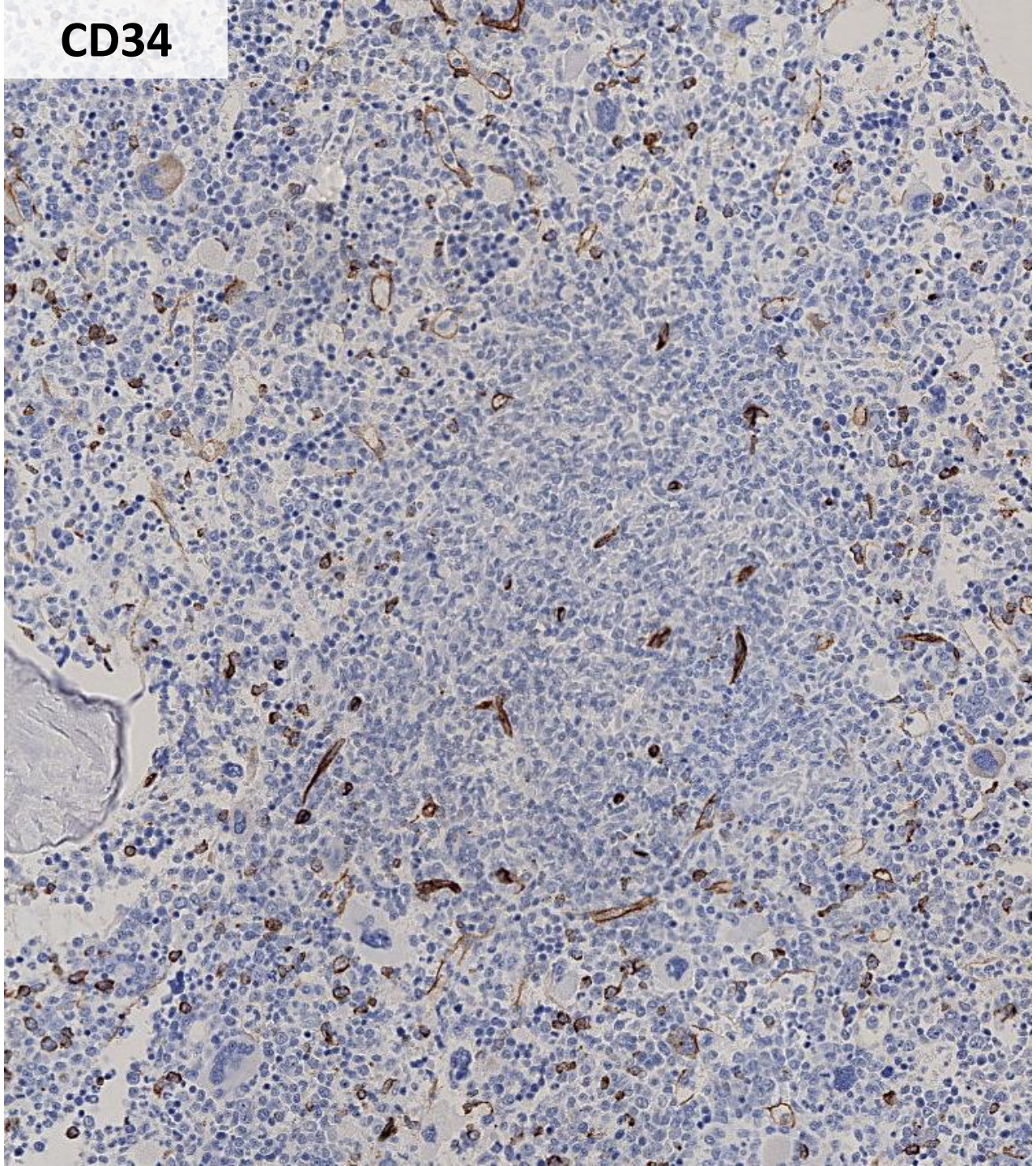
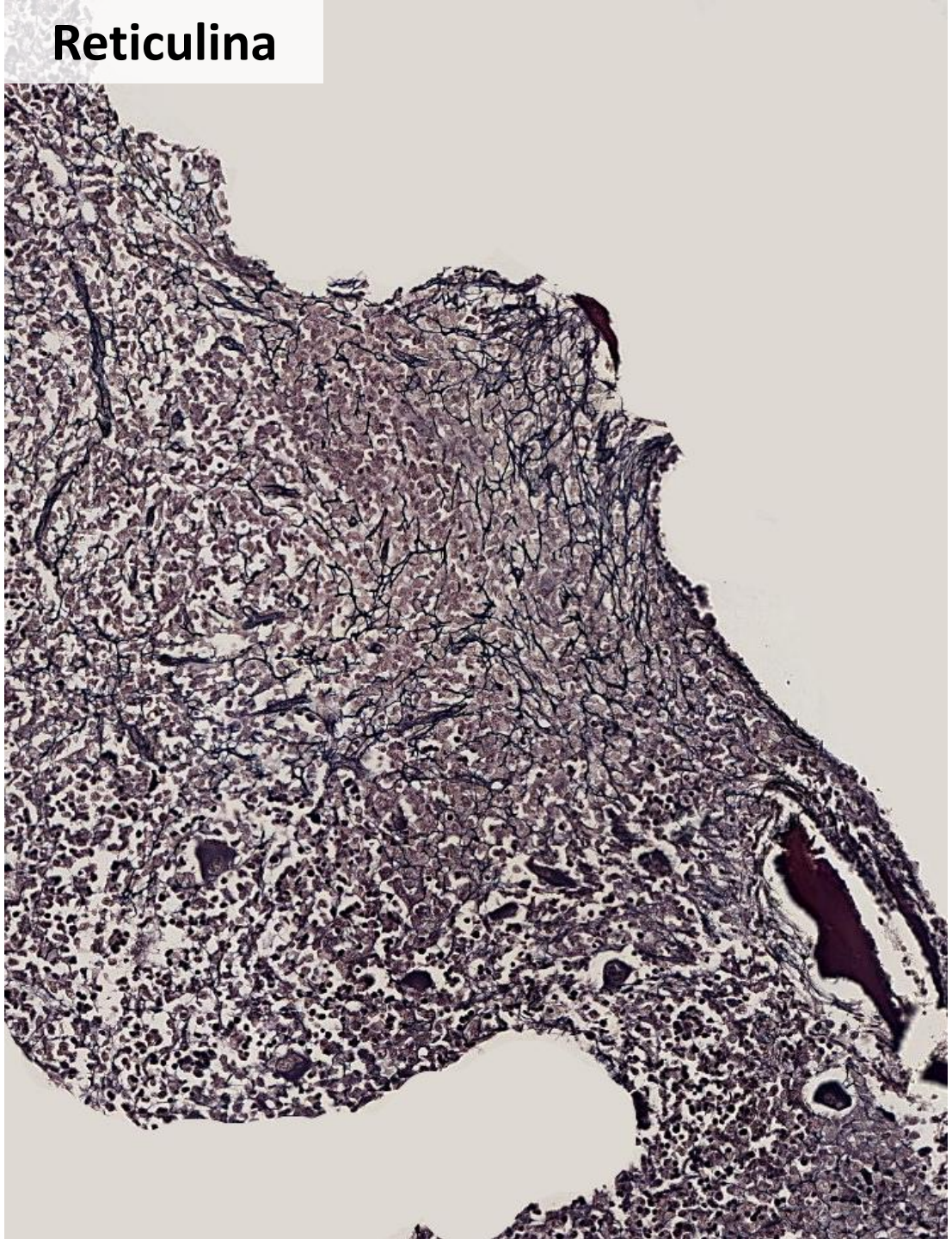

CD34


Reticulina

Published in final edited form as:
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Hairy cell leukemia diagnostic criteria and differential diagnosis

THOMAS A. SUMMERS and **ELAINE S. JAFFE**
Hematopathology Section, Laboratory of Pathology, Center for Cancer Research, National Cancer Institute, National Institutes of Health, Bethesda, MD, USA

Histologically, tumor cells are commonly encountered within the bone marrow and spleen, and less commonly within the liver and lymph nodes. Bone marrow involvement is variable and displays a broad spectrum of histological appearances, ranging from hypocellular, aplastic-appearing infiltrates to diffuse sheets of monotonous cells replacing marrow elements. The most common pattern observed within the bone marrow is an interstitial infiltrate, which tends to infiltrate throughout the marrow cavity preserving the marrow adipose tissue, rather than a nodular growth pattern [Figure 2(A)]. At low power the