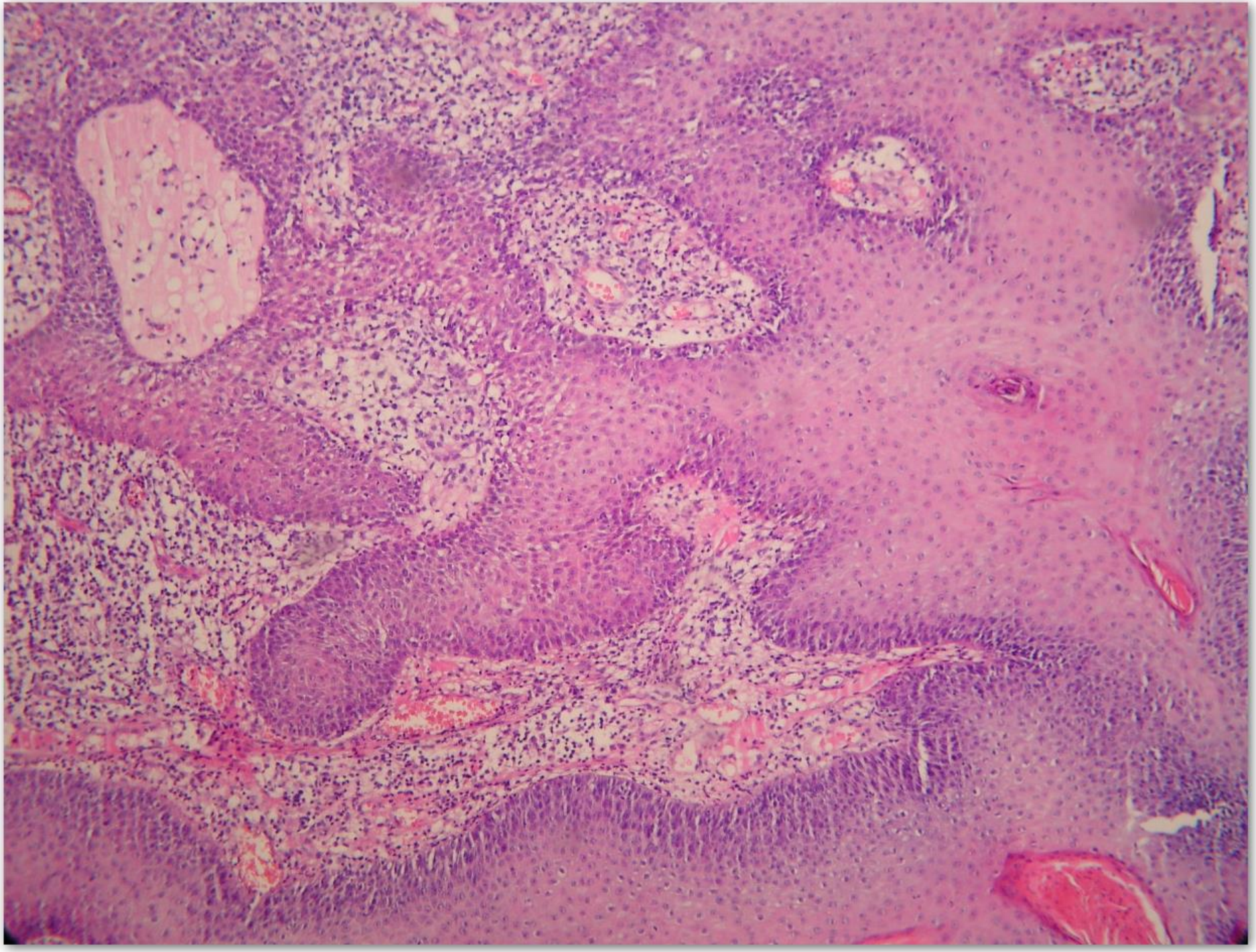
Prípad SD-IAP 637

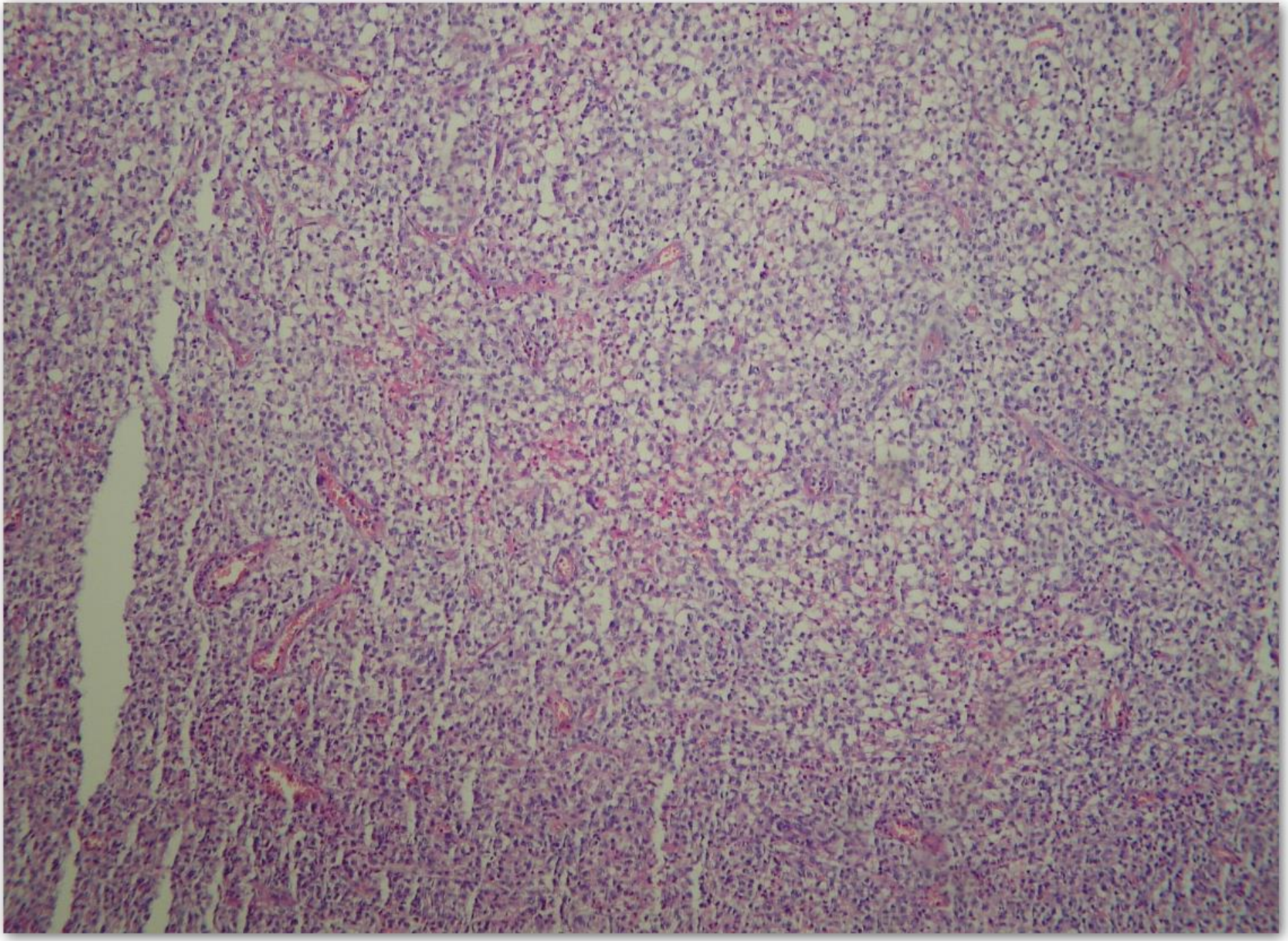
P Talarčík
Cytopathos

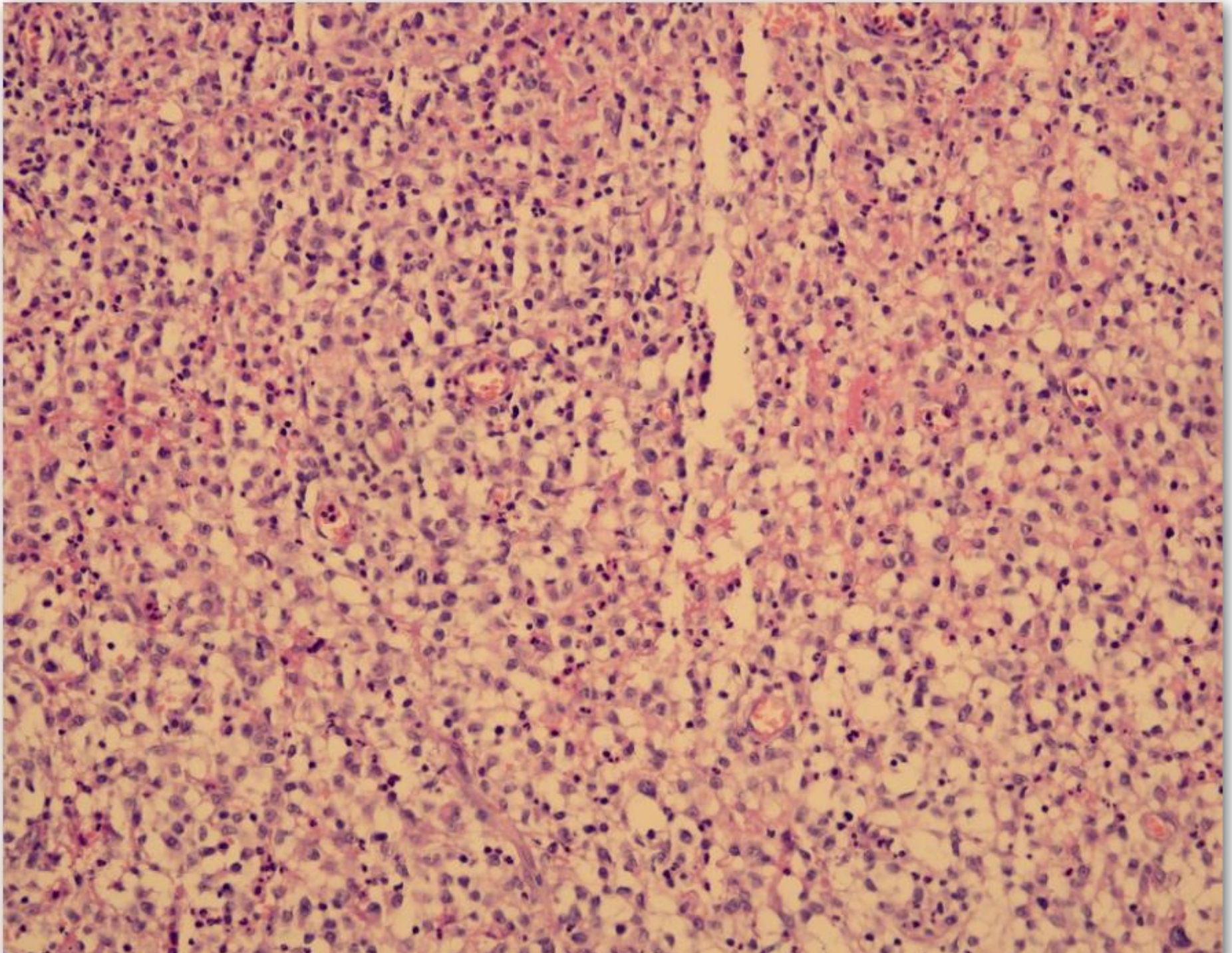
Vek 85 rokov, muž

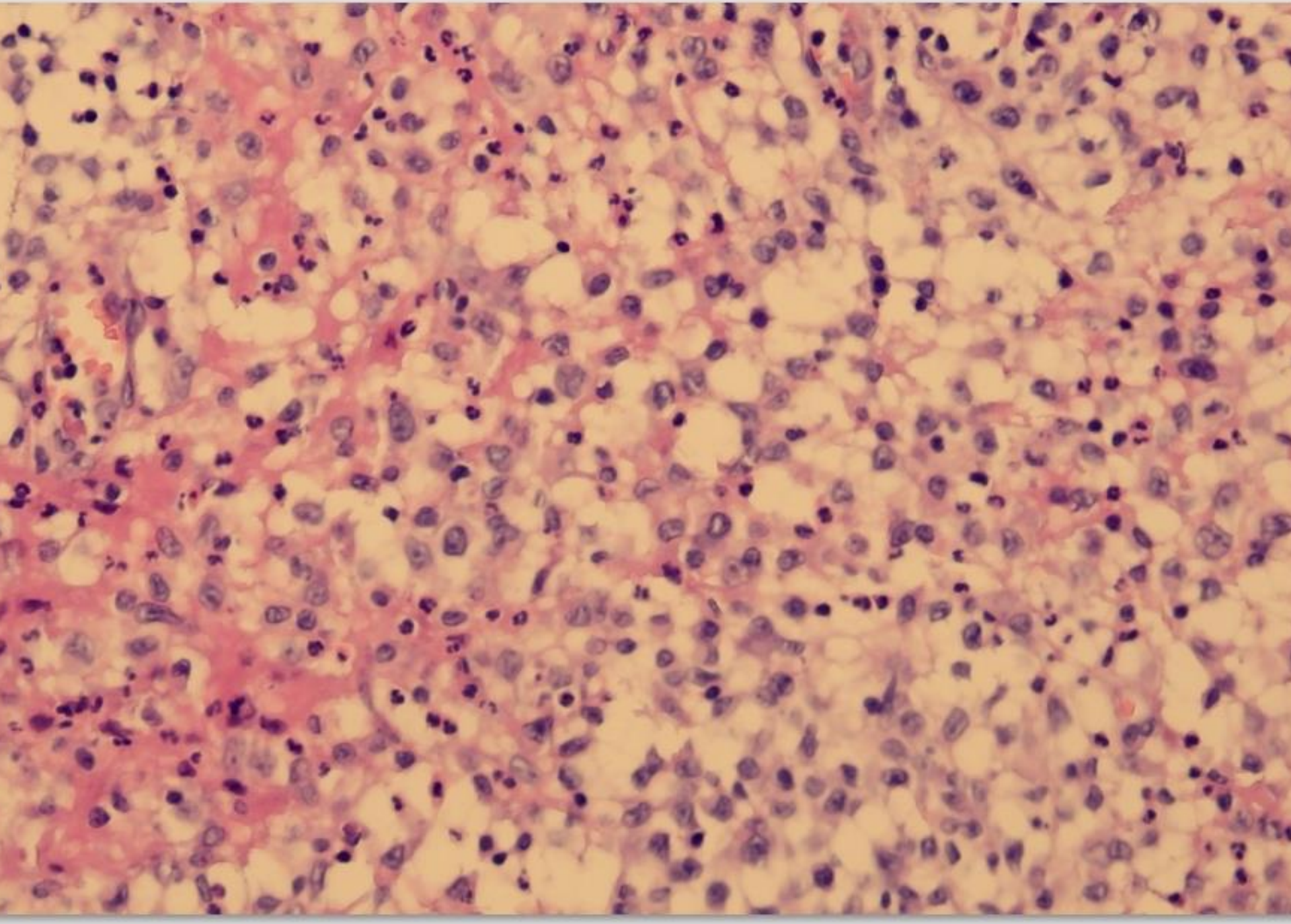
Klinika: pacient s diagnózou v anamnéze „hematodermická dermoplázia“ , myeloproliferatívny syndróm NOS

Makro: TU cutis pectoris l.dx., verukózna lézia pr. 21mm.









Základná diagnóza v rámci HE.

- leukémia cutis/ lymphoid,myeloid/
- myeloidná leukémia cutis/MLC
- MLC najčastejšie pri AML/mono/
- incidencia MLC 1-20%
- incidencia MLC pri AML až 50%
- incidencia pri CMML – 10%

Imunohistochémia:

Základná imunopozitivita:
Difúzne:
CD4+/CD56+/CD43+/CD33+/CD5/
/S100+/CD1a+/lyzozým
Slabo fokálne pozit: CD13, CD14, CD11c

Imunohistochémia:

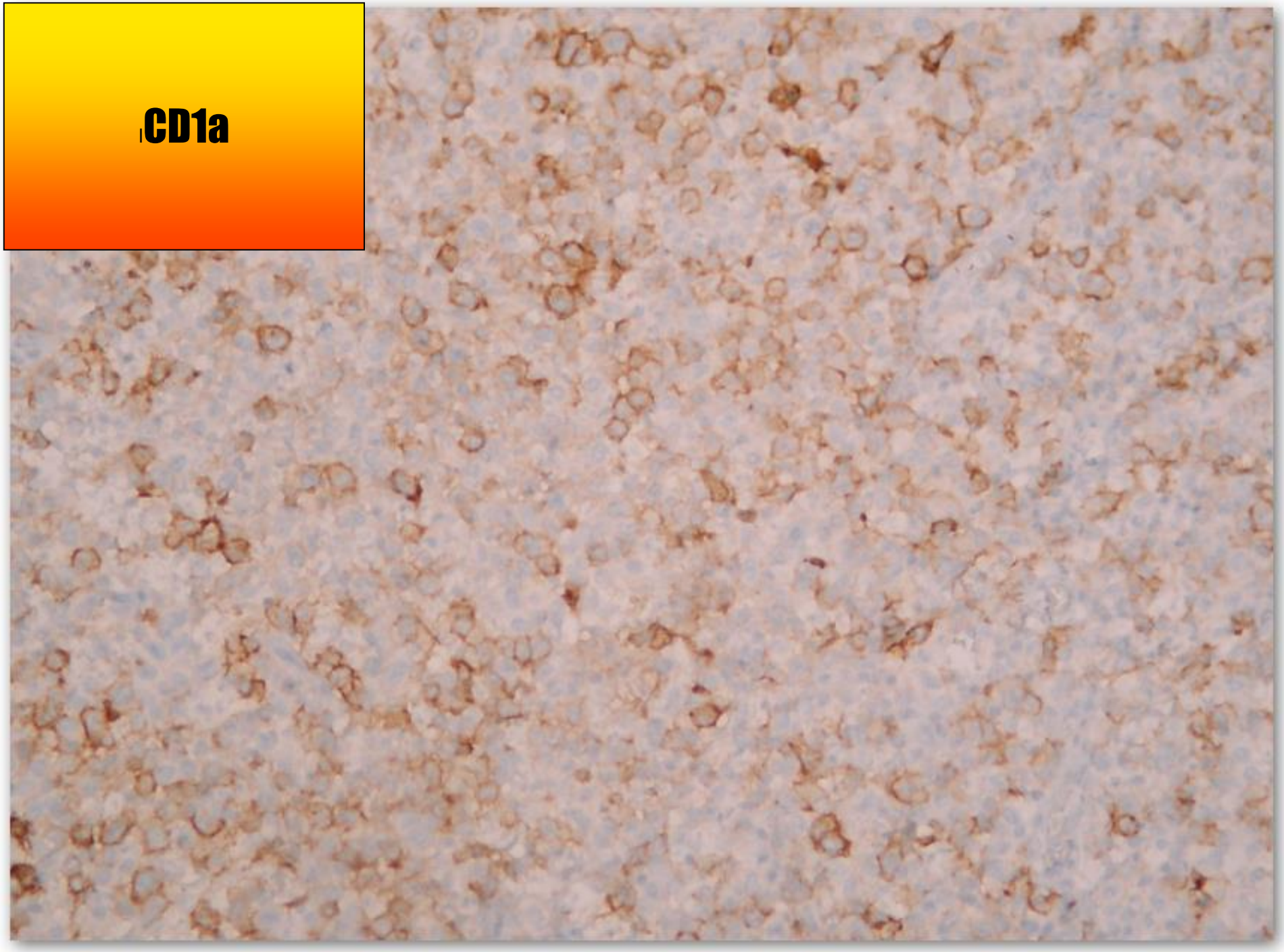
Základná imunonegativita:

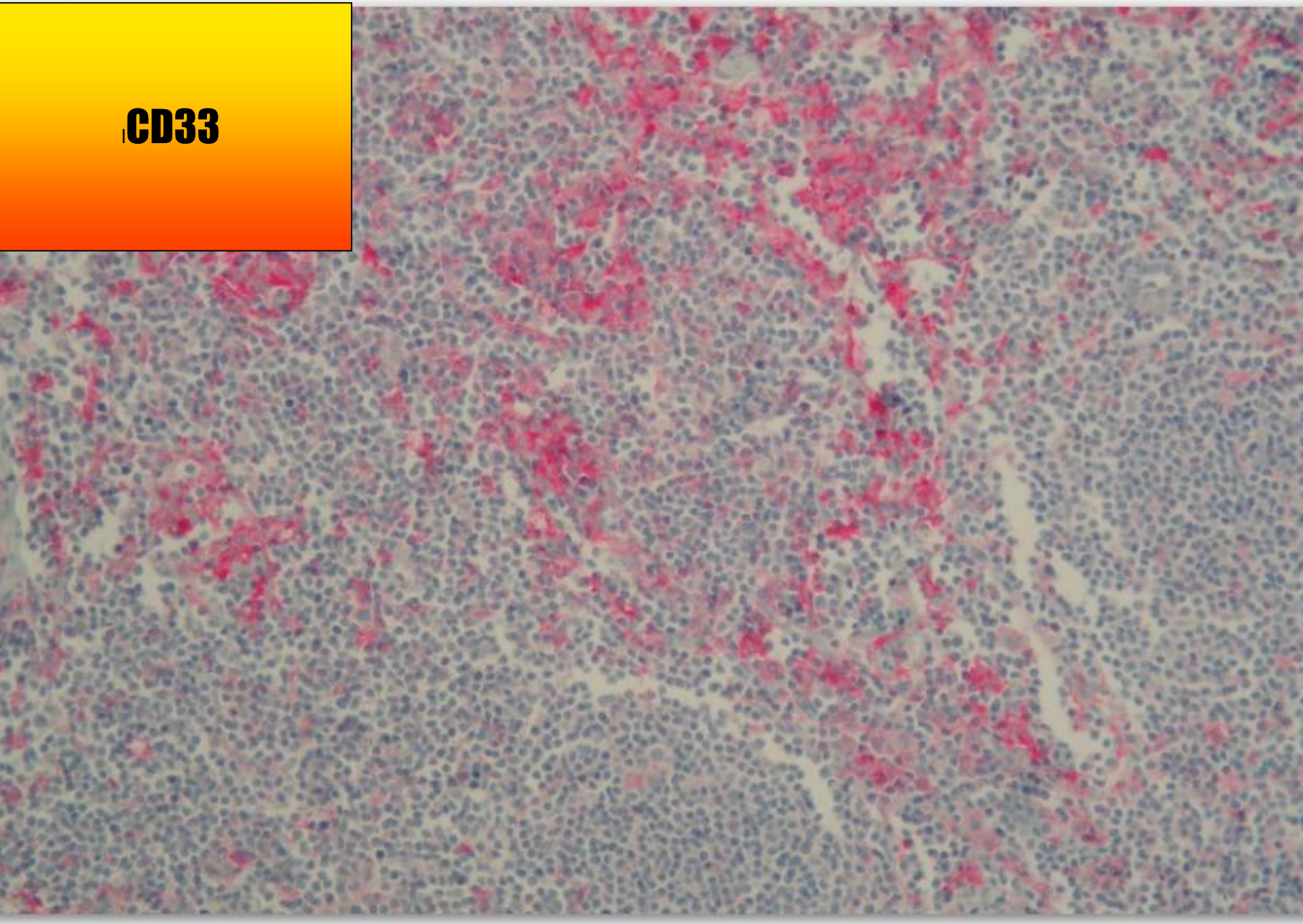
LCA, CD20, CD3, langerin, TdT, EBV, CD30, TIA, CD15, CD79a, CD7, CD23, CD21, CD123, granzyme B, CD117

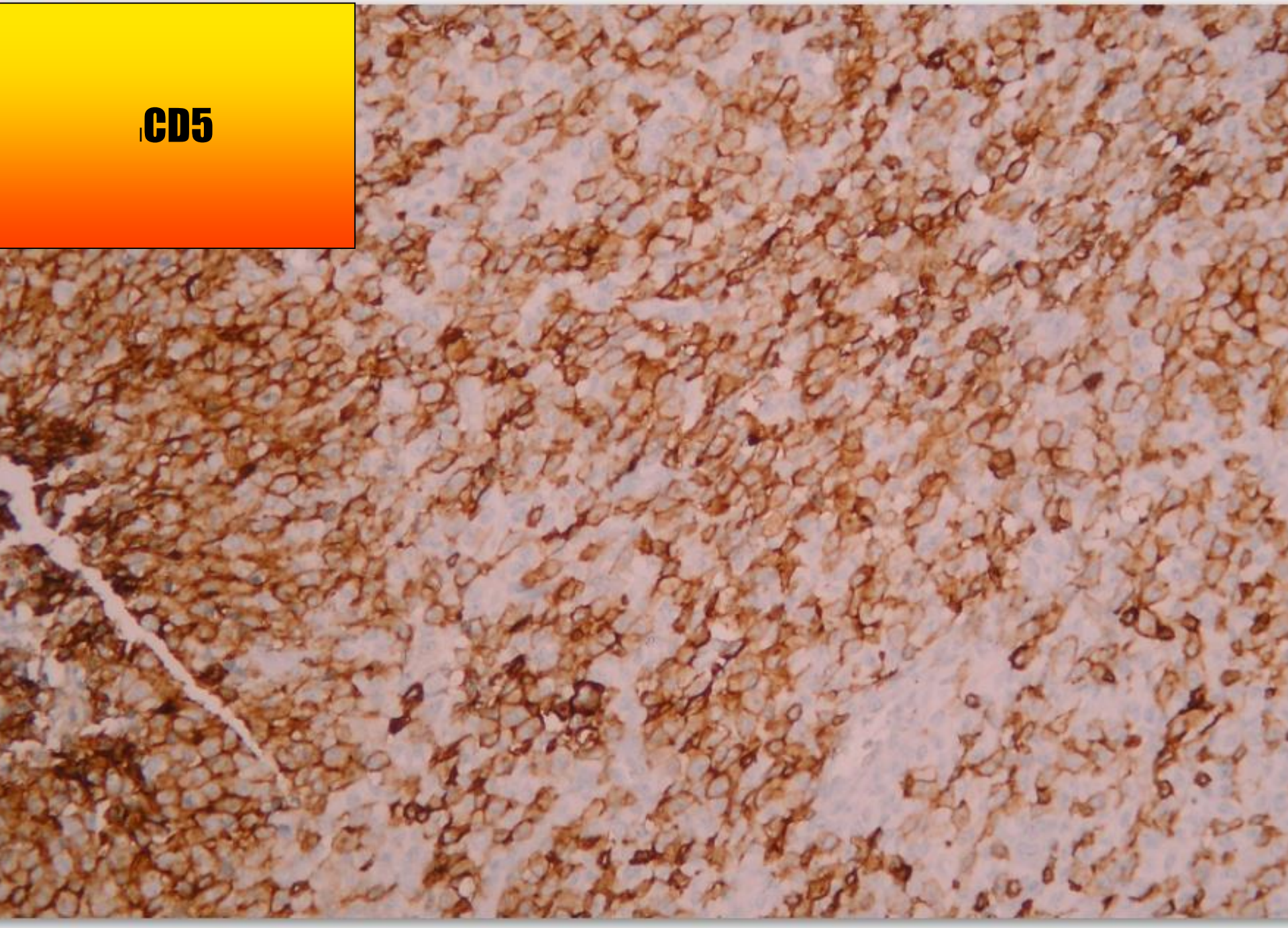
SÚHRN: Imunopozitivita.

Difúzne:
CD4+/CD56+/CD43+/CD33+/CD5/
/S100+/CD1a+/lyzozým/CD163
Slabo fokálne pozit: CD13, CD14,
CD11c

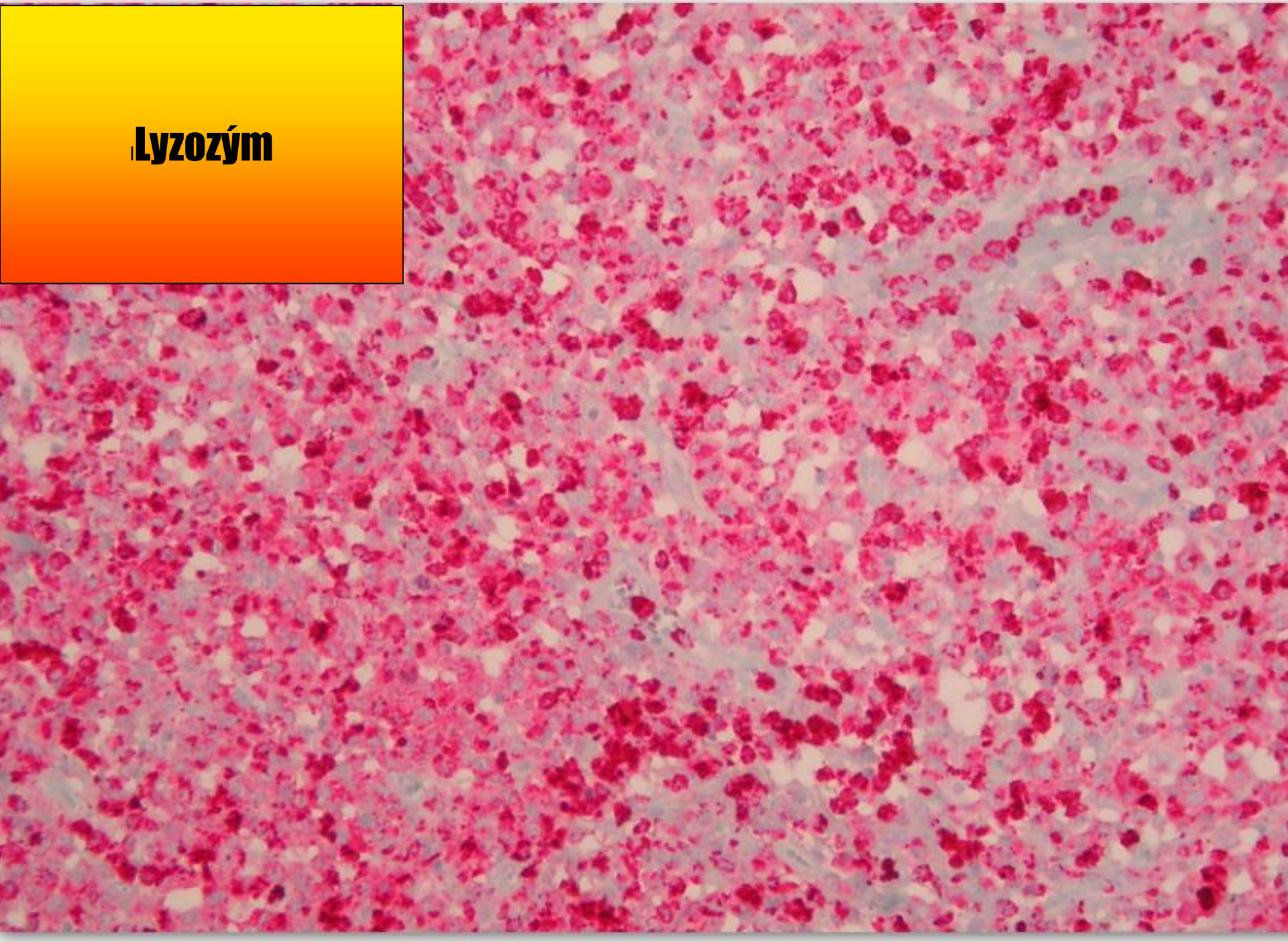
S100

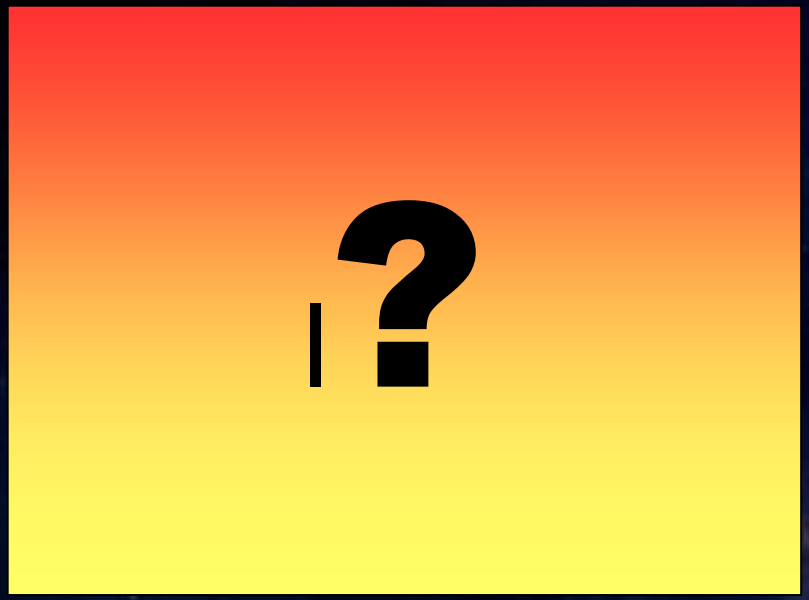

CD1a


CD33

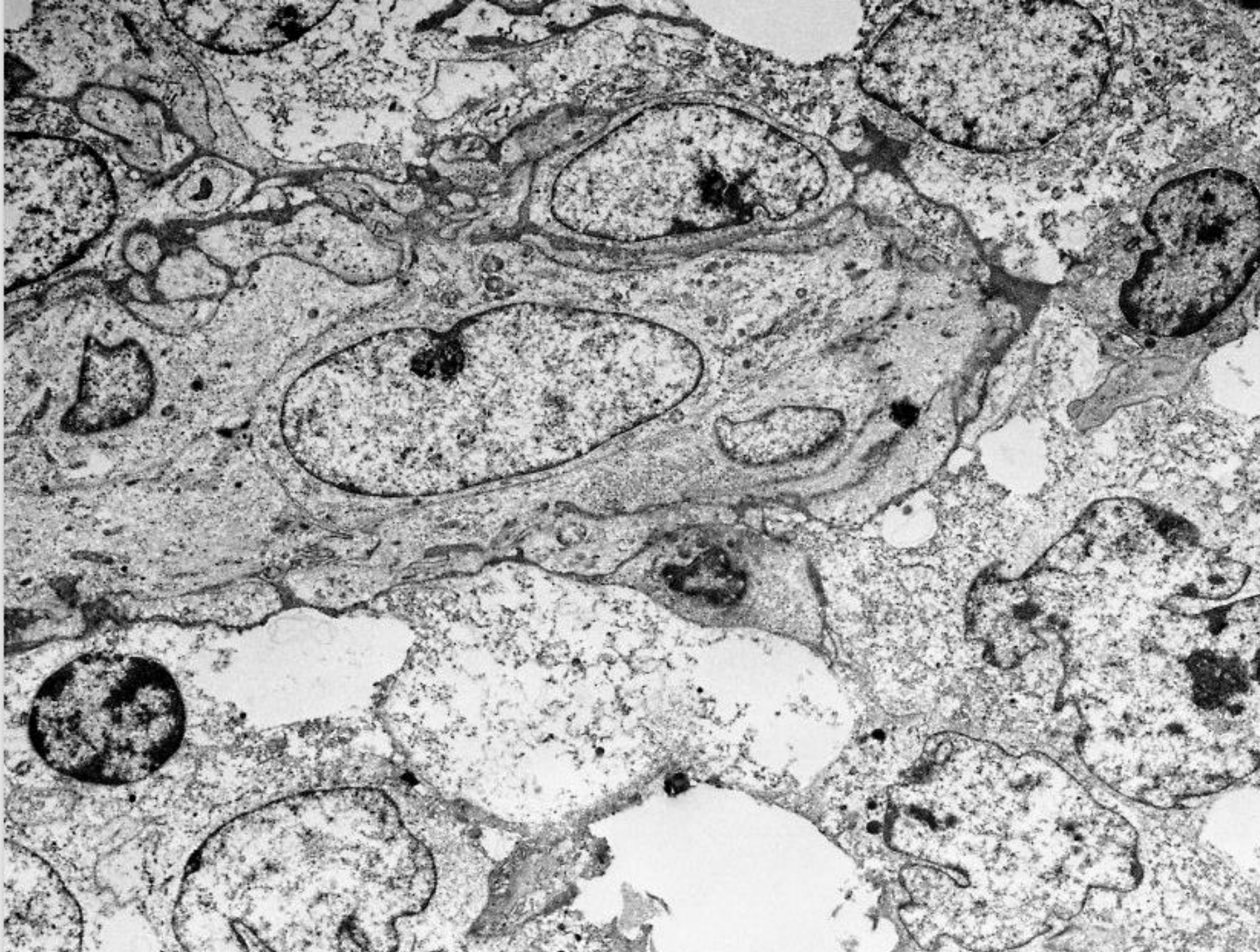

CD5

Lyzozým





BLASTIC INDETERMINATED DENDRITIC CELL TUMOR



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Review

Lymphoma classification update: T-cell lymphomas, Hodgkin lymphomas, and histiocytic/dendritic cell neoplasms

Manli Jiang, N. Nora Bennani & Andrew L. Feldman

Pages 239-249 | Received 15 Nov 2016, Accepted 09 Jan 2017, Published online: 29 Jan 2017

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ABSTRACT

Am J Surg Pathol. 2012 Sep;36(9):1302-16.

Specific skin lesions in chronic myelomonocytic leukemia: a spectrum of myelomonocytic and dendritic cell proliferations: a study of 42 cases.

Vitte F[1], Fabiani B, Bénet C, Dalac S, Balme B, Delattre C, Verqier B, Beylot-Barry M, Vignon-Pennamen D, Ortonne N, Algros MP, Carlotti A, Samaleire D, Frouin E, Levy A, Laroche L, Theate I, Monnien F, Mugneret F, Petrella T.

and overall phenotypic expression, the 42 patients were classified into 4 clinicopathologic categories of MLC: (1) myelomonocytic cell tumors (MMCTs) (n = 18); (2) mature plasmacytoid dendritic cell proliferations (MPDCPs) (n = 16); (3) blastic plasmacytoid dendritic cell neoplasms (BPDCNs) (n = 4); and (4) blastic indeterminate dendritic cell tumors (BIDCTs) (n = 4). Considering all categories, the median age was 71.0 years with a male to female ratio of 5:1, showing a clear male predominance of 83.3%.

TABLE 6. Survival Results

	OS1				OS2			
Pattern	Meantime	CI−	CI+	*P*	Meantime	CI−	CI+	*P*
MMCT	49.4	24	74.8	0.60	13.9	5.9	21.9	0.77
MPDCP	53.2	33.5	72.8		23	10	36	
BPDCN	49.8	21.9	77.6		10.4	4.5	16.2	
BIDCT	124	29.9	218.2		6.8	3.5	10.1	

Results shown in months.

CI− indicates lower limit of the confidence interval; CI+, upper limit of the confidence interval.

TABLE 2. Comparison of the Main Clinical Data for the 4 Categories

	All Patients	MMCT	MPDCP	BPDCN	BIDCT
No. patients (%)	42	18	16	4	4
Median age (y)	71	71.5	76.5	71.5	65.5
Sex					
Male	35 (83.3)	15 (83.3)	15 (93.7)	3 (75)	2 (50)
Female	7 (16.7)	3 (16.7)	1 (6.3	1 (25)	2 (50)
Presentation					
Unique lesion	6/42 (14.3)	2/18 (11.1)	1/16 (6.3)	1/4 (25)	2/4 (50)
Multiple lesion	35/42 (83.3)	15/18 (83.3)	15/16 (93.8)	3/4 (75)	2/4 (50)
Data not available	1/42 (2.4)	1/18 (5.6)	0/16 (0)	0/4 (0)	0/4 (0)
Chronology of skin lesions					
Before diagnosis of CMML	1/42 (2.4)	0/18 (0)	0/16 (0)	0/4 (0)	0/4 (0)
Concurrent to diagnosis of CMML	14/42 (33.3)	4/18 (22.2)	8/16 (50)	2/4 (50)	1/4 (25)
After diagnosis of CMML	27/42 (64.3)	14/18 (77.8)	8/16 (50)	2/4 (50)	3/4 (75)
Treatment					
None	4/42 (9.5)	1/18 (5.6)	1/16 (6.3)	1/4 (25)	1/4 (25)
Local corticoid	3/42 (7.1)	1/18 (5.6)	2/16 (12.5)	0/4 (0)	0/4 (0)
Cytoreductive	18/42 (42.9)	7/18 (38.9)	9/16 (56.3)	0/4 (0)	2/4 (50)
High-dose chemotherapy	7/42 (16.7)	4/18 (22.2)	0/16 (0)	2/4 (50)	1/4 (25)
Radiotherapy	2/42 (4.8)	0/18 (0)	1/16 (6.3)	1/4 (25)	0/4 (0)
Data not available	8/42 (19)	5/18 (27.8)	3/16 (18.8)	0/4 (0)	0/4 (0)
Treatment response					
None	8/42 (19)	3/18 (16.7)	2/16 (12.5)	1/4 (25)	2/4 (50)
Partial response	13/42 (31)	4/18 (22.2)	6/16 (37.5)	2/4 (50)	1/4 (25)
Complete response	11/42 (26.2)	4/18 (22.2)	5/16 (31.3)	1/4 (25)	1/4 (25)
Data not available	10/42 (23.8)	7/18 (38.9)	3/16 (18.8)	0/4 (0)	0/4 (0)
Outcome at the date of last follow-up					
Alive	22/42 (52.4)	9/18 (50)	11/16 (68.8)	1/4 (25)	1/4 (25)
Dead	19/42 (45.2)	8/18 (44.4)	5/16 (31.3)	3/4 (75)	3/4 (75)
Data not available	1/42 (2.4)	1/18 (5.6)	0/16 (0)	0/4 (0)	0/4 (0)

THE UPDATED WHO CLASSIFICATION OF HEMATOLOGICAL MALIGNANCIES

The 2016 revision to the World Health Organization classification of myeloid neoplasms and acute leukemia

Daniel A. Arber,[1] Attilio Orazi,[2] Robert Hasserjian,[3] Jürgen Thiele,[4] Michael J. Borowitz,[5] Michelle M. Le Beau,[6] Clara D. Bloomfield,[7] Mario Cazzola,[8] and James W. Vardiman[9]

[1]Department of Pathology, Stanford University, Stanford, CA; [2]Department of Pathology, Weill Cornell Medical College, New York, NY; [3]Department of Pathology, Massachusetts General Hospital, Boston, MA; [4]Institute of Pathology, University of Cologne, Cologne, Germany; [5]Department of Pathology, Johns Hopkins Medical Institutions, Baltimore, MD; [6]Section of Hematology/Oncology, University of Chicago, Chicago, IL; [7]Comprehensive Cancer Center, James Cancer Hospital and Solove Research Institute, The Ohio State University, Columbus, OH; [8]Department of Molecular Medicine, University of Pavia, and Department of Hematology Oncology, Fondazione IRCCS Policlinico San Matteo, Pavia, Italy; and [9]Department of Pathology, University of Chicago, Chicago, IL

The World Health Organization (WHO) classification of tumors of the hematopoietic and lymphoid tissues was last updated in 2008. Since then, there have been numerous advances in the identification of unique biomarkers associated with some myeloid neoplasms and acute leukemias, largely derived from gene expression analysis and next-generation sequencing that can significantly improve the diagnostic criteria as well as the prognostic relevance of entities currently included in the WHO classification and that also suggest new entities that should be added. Therefore, there is a clear need for a revision to the current classification. The revisions to the categories of myeloid neoplasms and acute leukemia will be published in a monograph in 2016 and reflect a consensus of opinion of hematopathologists, hematologists, oncologists, and geneticists. The 2016 edition represents a revision of the prior classification rather than an entirely new classification and attempts to incorporate new clinical, prognostic, morphologic, immunophenotypic, and genetic data that have emerged since the last edition. The major changes in the classification and their rationale are presented here. (*Blood*. 2016; 127(20):2391-2405)

Table 1. (continued)

WHO myeloid neoplasm and acute leukemia classification
Blastic plasmacytoid dendritic cell neoplasm

Myeloid/lymphoid neoplasms with eosinophilia and rearrangement of *PDGFRA*, *PDGFRB*, or *FGFR1*, or with *PCM1-JAK2*
Myeloid/lymphoid neoplasms with *PDGFRA* rearrangement
Myeloid/lymphoid neoplasms with *PDGFRB* rearrangement
Myeloid/lymphoid neoplasms with *FGFR1* rearrangement
Provisional entity: Myeloid/lymphoid neoplasms with PCM1-JAK2
Myelodysplastic/myeloproliferative neoplasms (MDS/MPN)
Chronic myelomonocytic leukemia (CMML)
Atypical chronic myeloid leukemia (aCML), *BCR-ABL1*$^{-}$
Juvenile myelomonocytic leukemia (JMML)
MDS/MPN with ring sideroblasts and thrombocytosis (MDS/MPN-RS-T)
MDS/MPN, unclassifiable
Myelodysplastic syndromes (MDS)
MDS with single lineage dysplasia
MDS with ring sideroblasts (MDS-RS)
MDS-RS and single lineage dysplasia
MDS-RS and multilineage dysplasia
MDS with multilineage dysplasia
MDS with excess blasts
MDS with isolated del(5q)
MDS, unclassifiable
Provisional entity: Refractory cytopenia of childhood
Myeloid neoplasms with germ line predisposition

Myelodysplastic/myeloproliferative neoplasms

The myelodysplastic syndrome (MDS)/MPN category was introduced in the third edition to include myeloid neoplasms with clinical, laboratory, and morphologic features that overlap between MDS and MPN.[24] Based on accumulated scientific evidence, a provisional entity within the MDS/MPN unclassifiable group, refractory anemia with ring sideroblasts associated with marked thrombocytosis (RARS-T), has been accepted as a full entity, now termed MDS/MPN with ring sideroblasts and thrombocytosis in the 2016 revision. The 2016 revised criteria for diseases in this category are summarized in Tables 11-14.[25]

In MDS/MPN, the karyotype is often normal or shows abnormalities in common with MDS. Targeted sequencing of genes mutated in myeloid neoplasms detects mutations in a high proportion of cases of chronic myelomonocytic leukemia (CMML) as well as other MDS/MPN patients.[26] The most commonly mutated genes in CMML are *SRSF2*, *TET2*, and/or *ASXL1* (>80% of cases).[26,27] Other mutations which occur at lower frequency include *SETBP1*, *NRAS/KRAS*, *RUNX1*, *CBL*, and *EZH2*.[28,29] They can be helpful adjunct studies in difficult cases, particularly given the frequently normal karyotype of CMML, but should not be used alone as proof of neoplasia because some of these mutations occur in healthy older patients as so-called clonal hematopoiesis of indeterminate potential (CHIP)[30,31] (for further discussion, see "Myelodysplastic syndromes"). *ASXL1* is a predictor of aggressive disease behavior and has been incorporated into a prognostic scoring system for CMML alongside karyotype and clinicopathologic parameters.[27] Of note, *NPM1* mutation is seen in a rare subset of CMML (3%-5%) and appears also to herald a more aggressive clinical course.

Next generation sequencing solution or dissolution?

Take home message??

Don´t take any…..

….take a tripple mojito…

:)


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