Cádiz, 13 de abril de 2018

Caso clínico patológico

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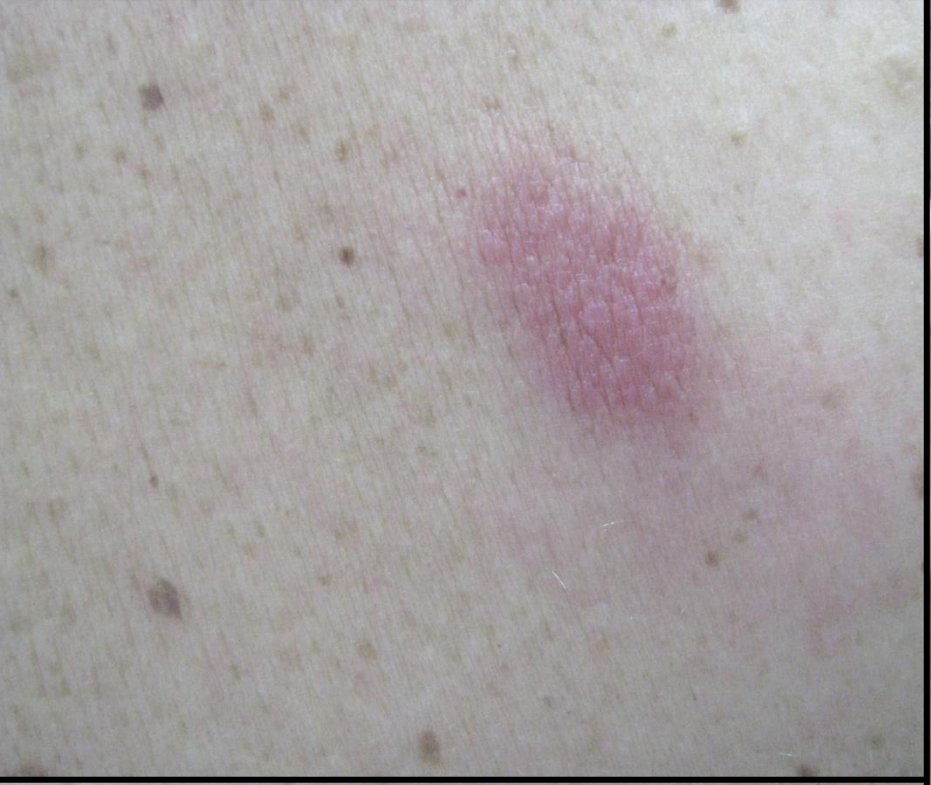
Sospecha clínica:
MASTOCITOSIS

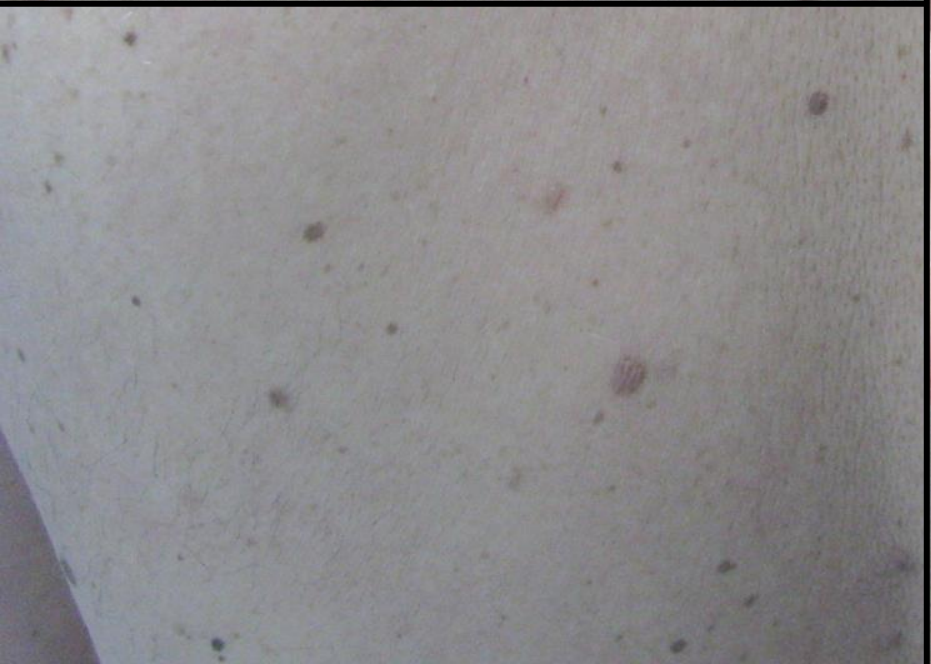
8 meses de evolución

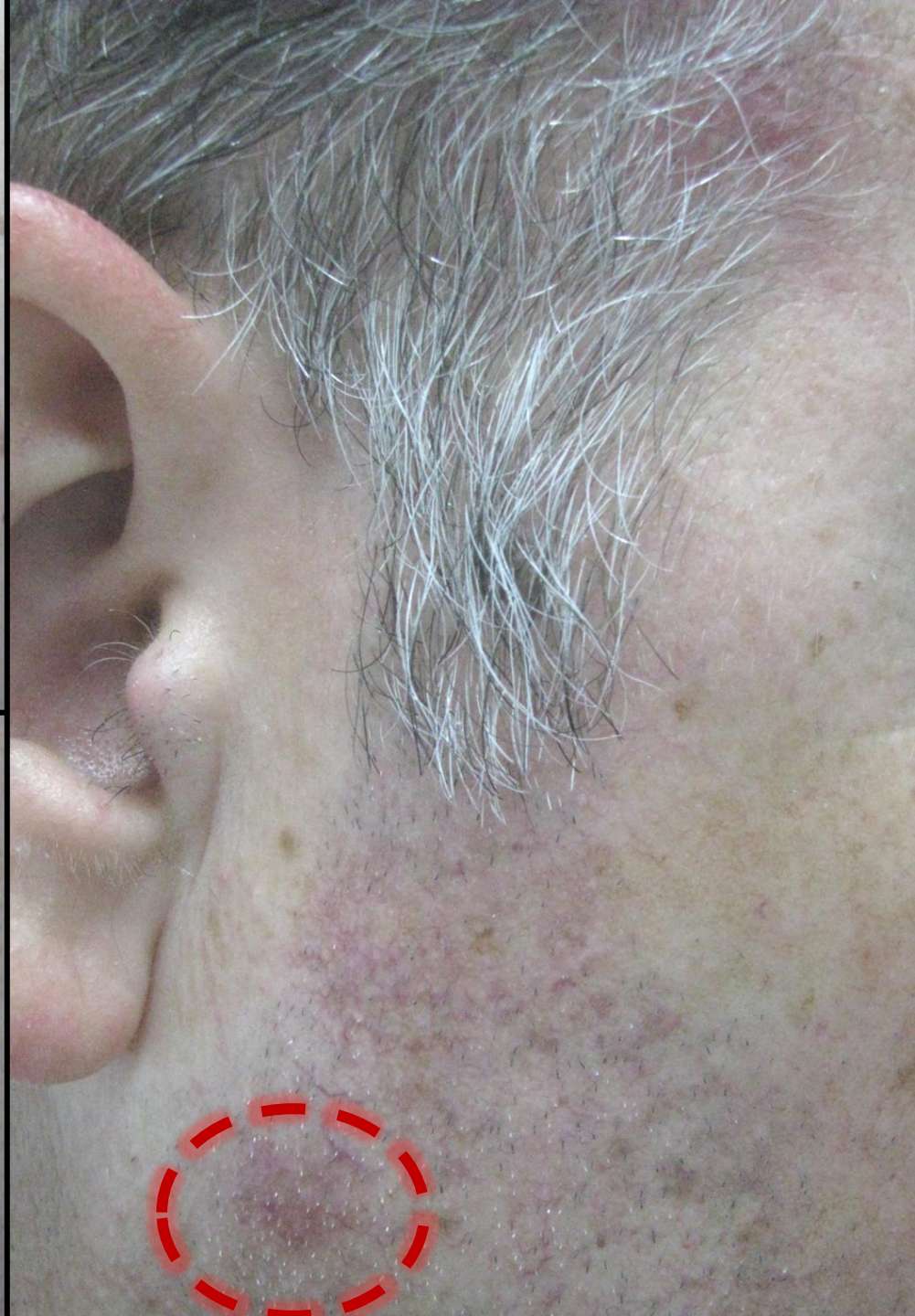
49 AÑOS

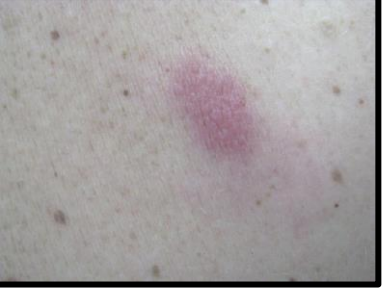
Prurito generalizado de 2 años de evolución

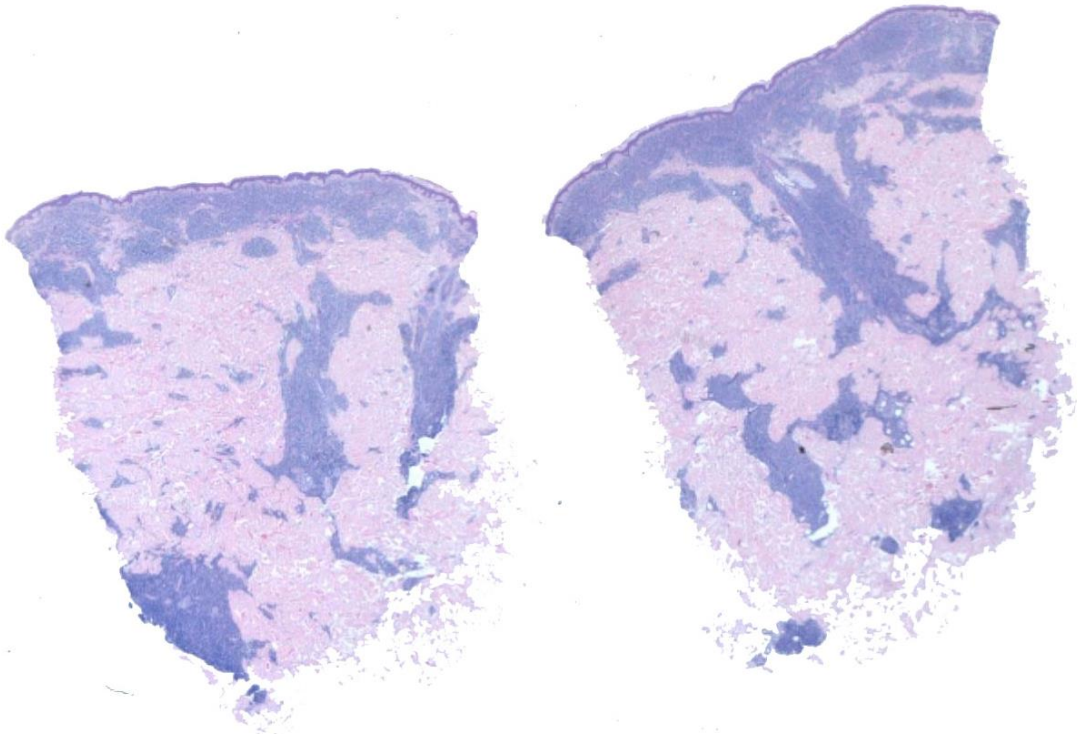
Prurito intensificado + placa eritematosa en espalda

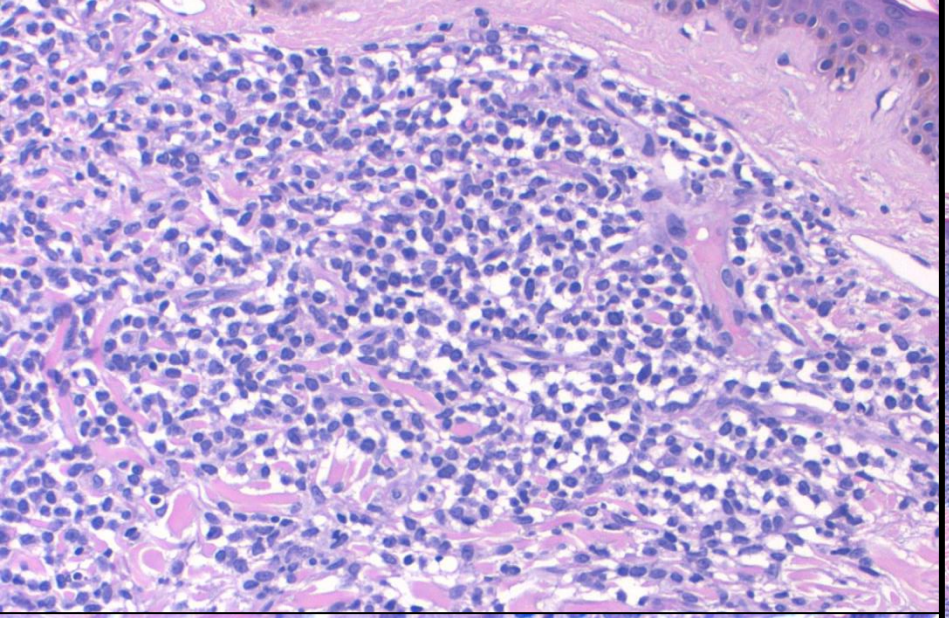


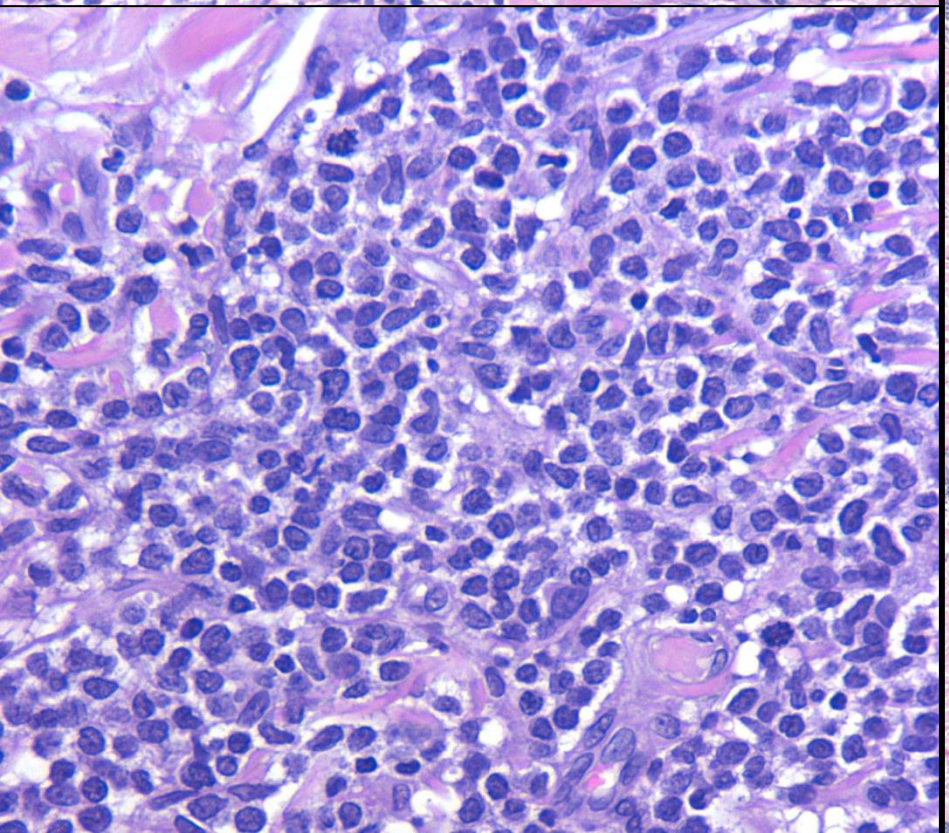


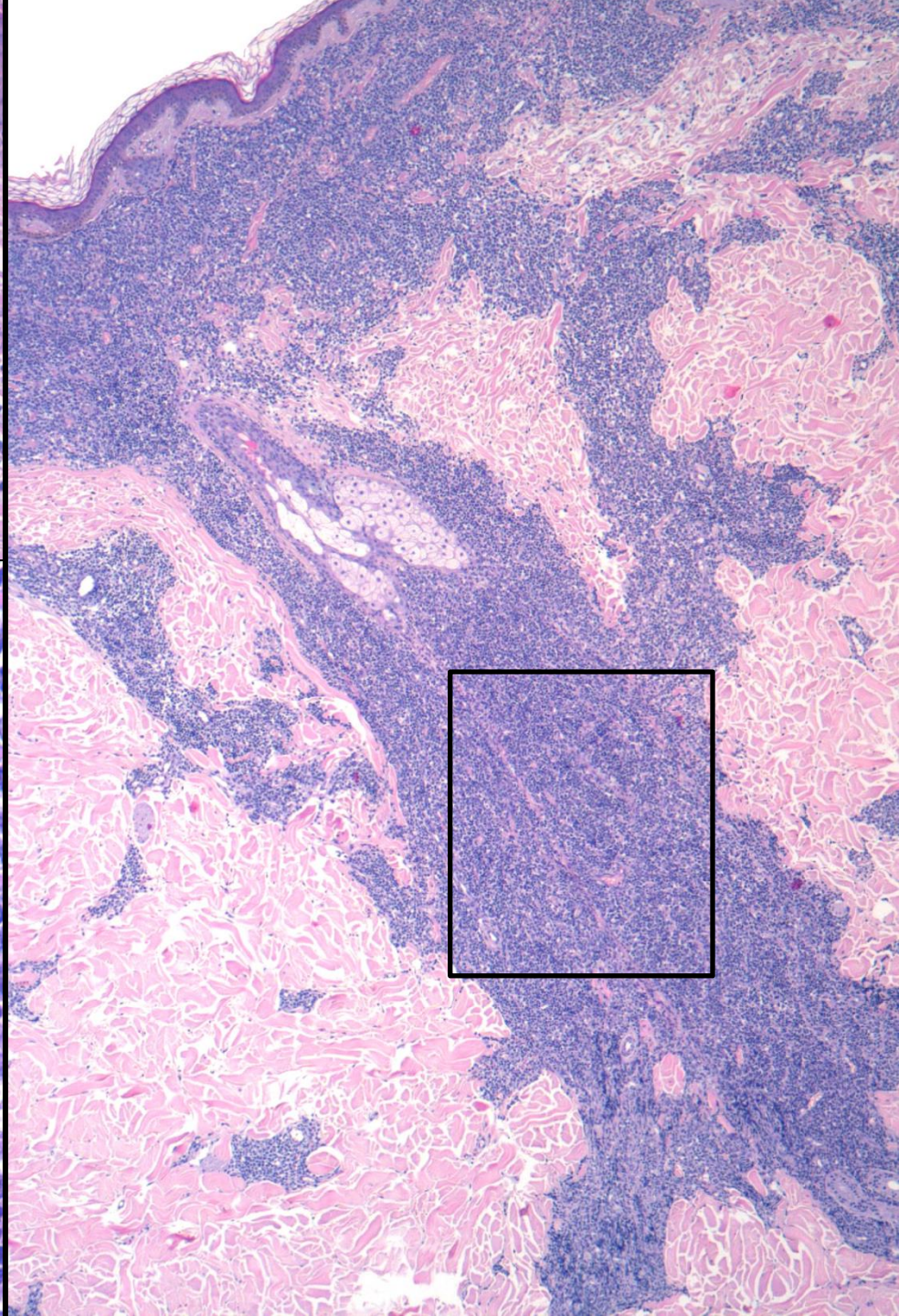


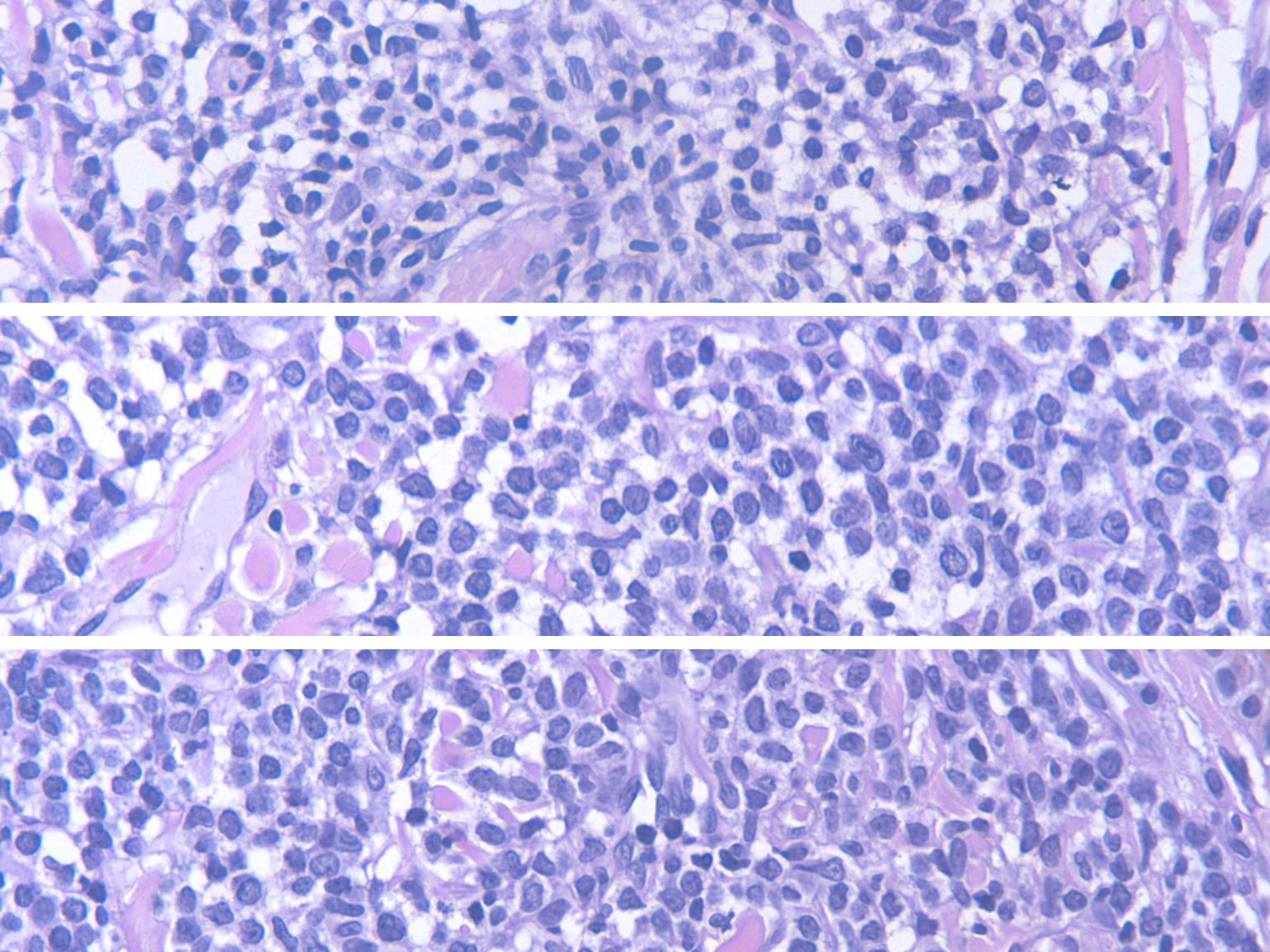


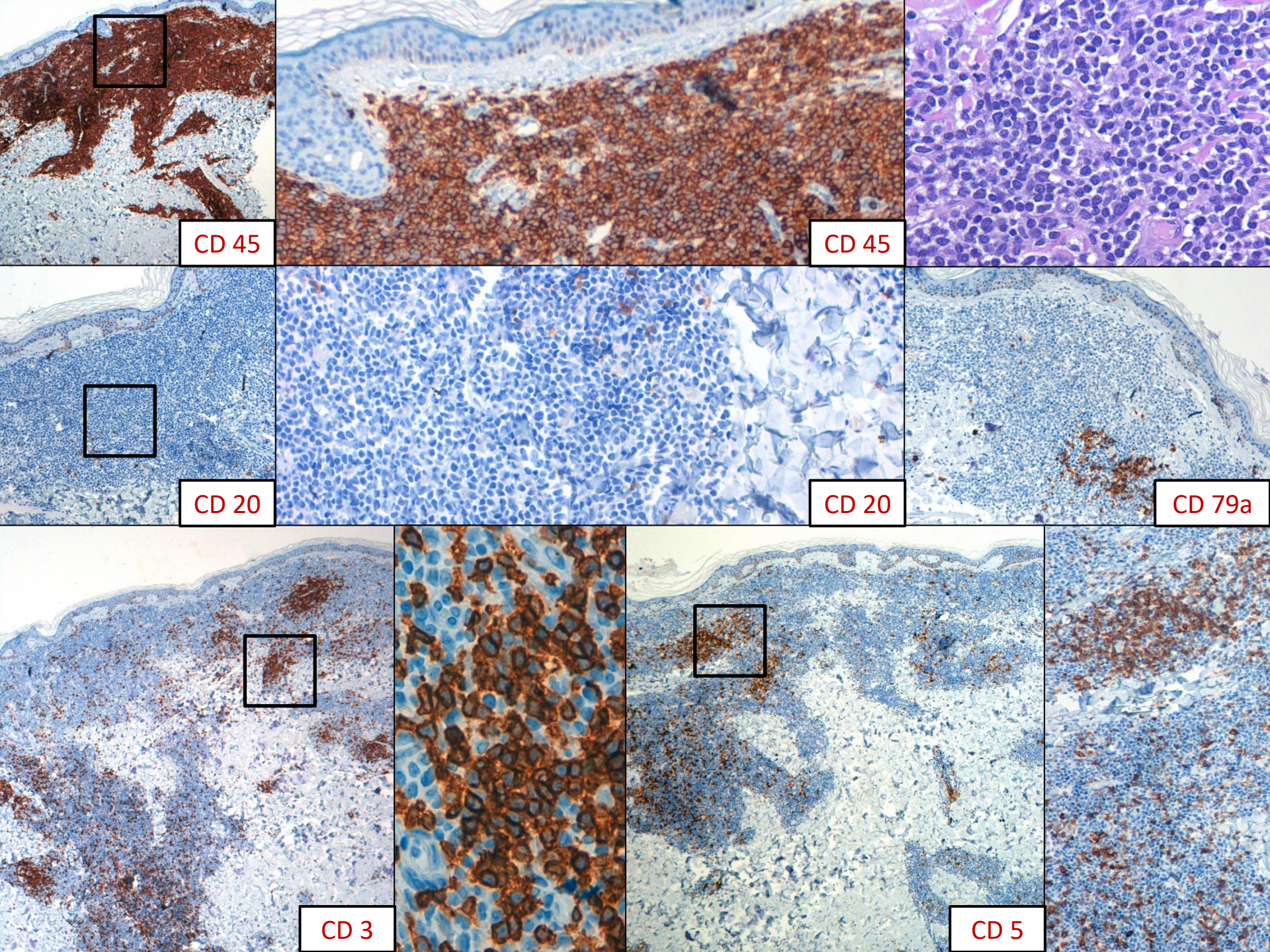








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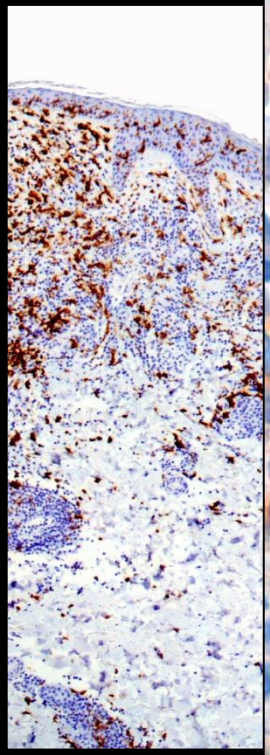
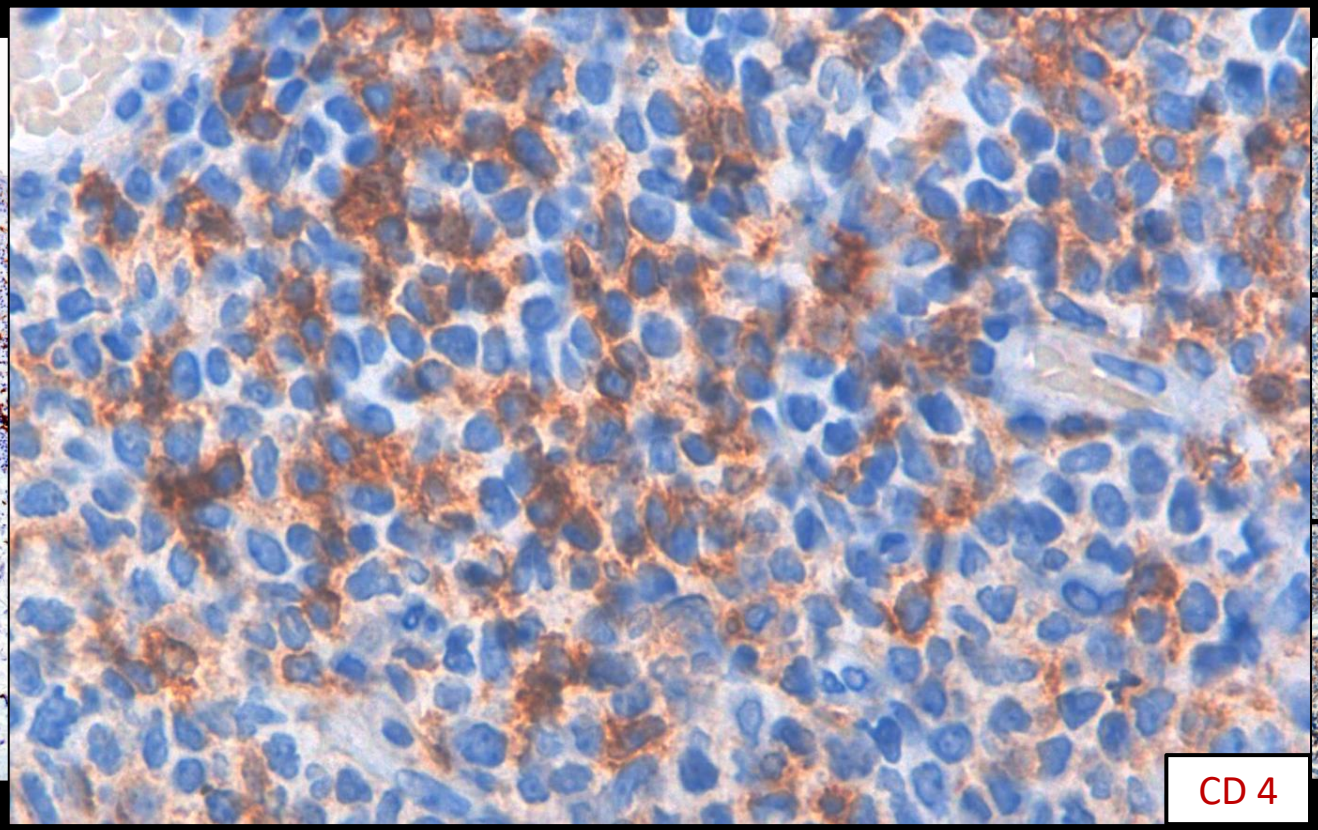
CD 5

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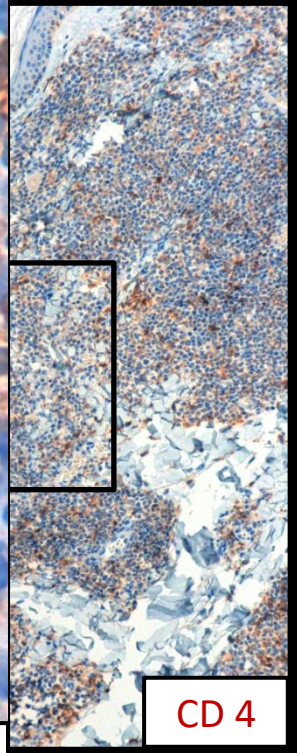
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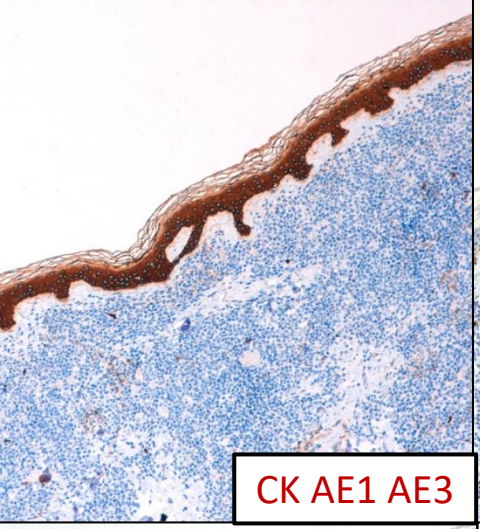
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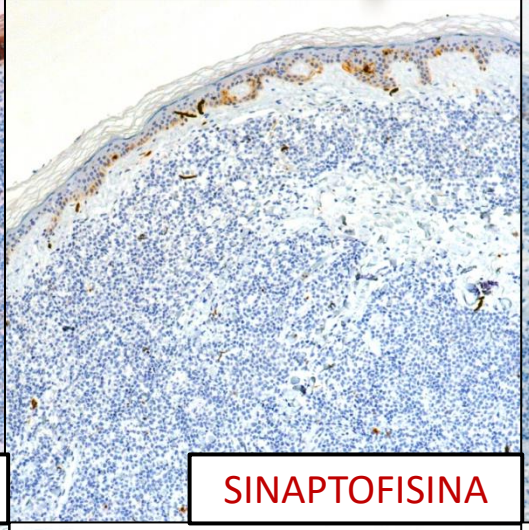
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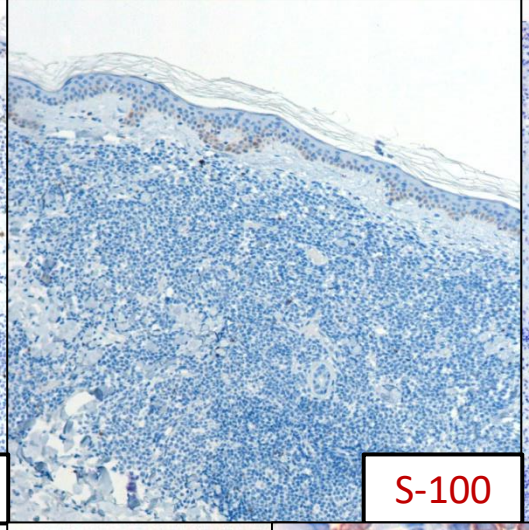
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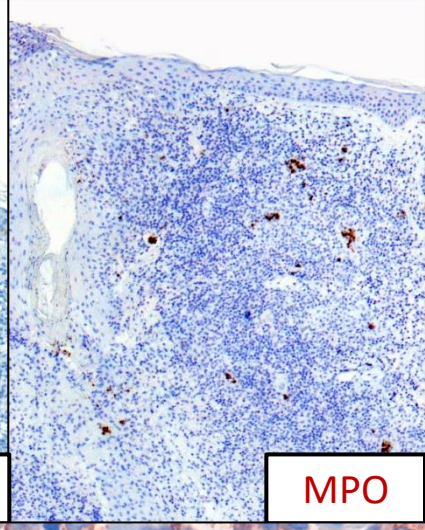
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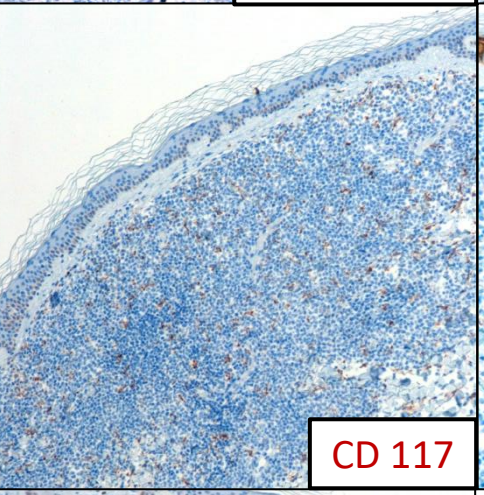
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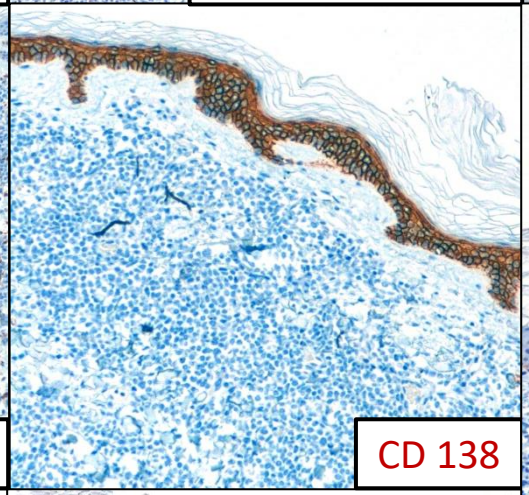
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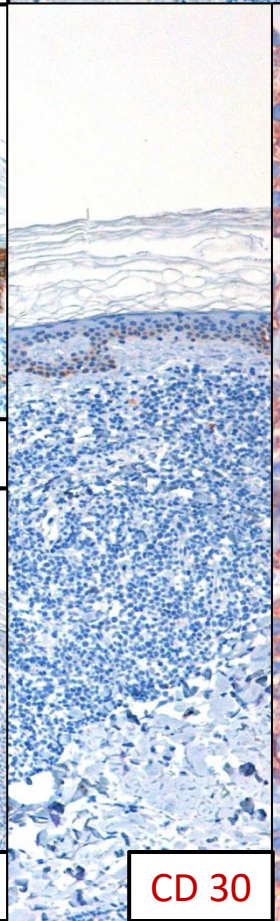
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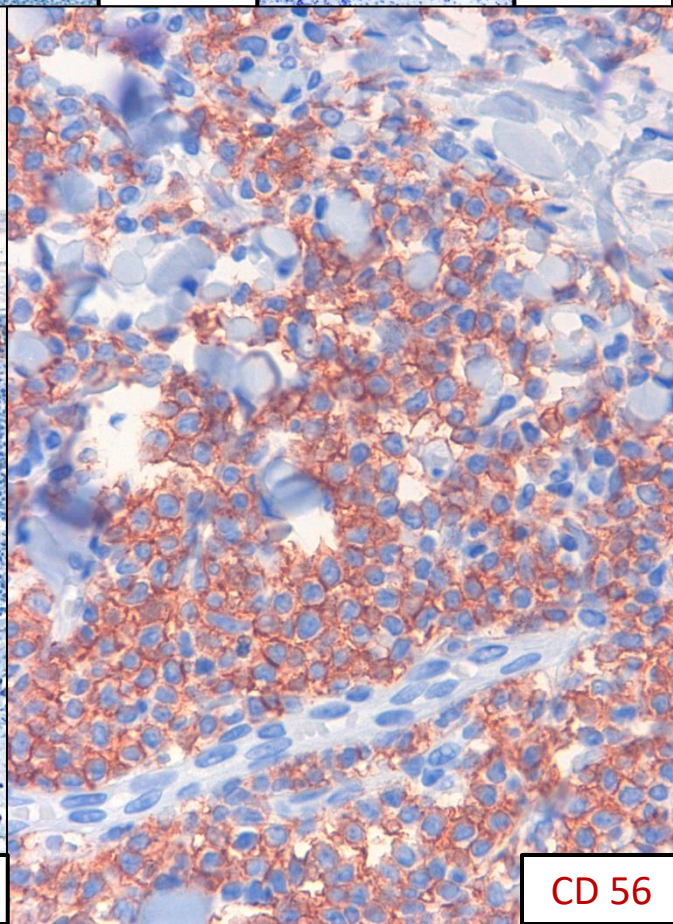
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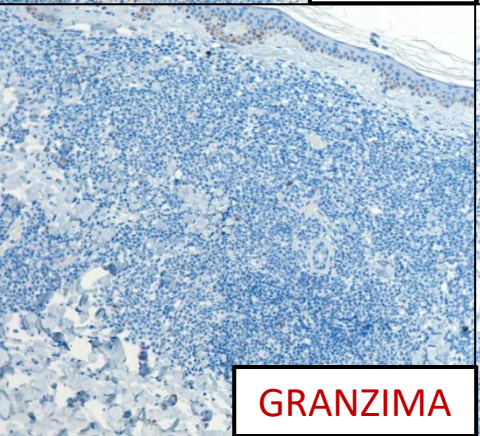
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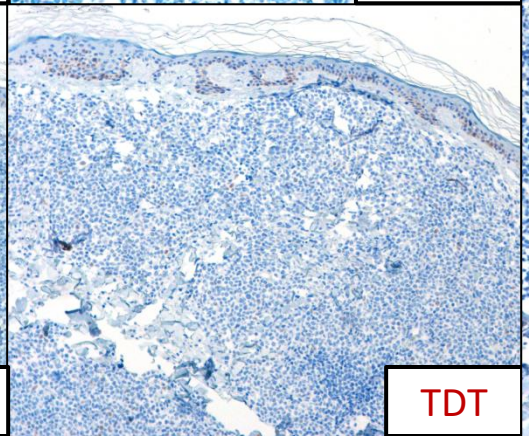
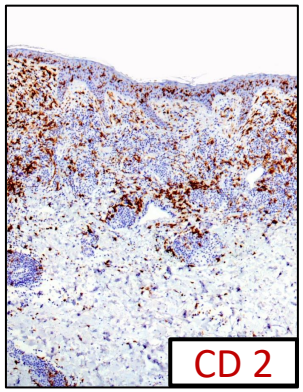
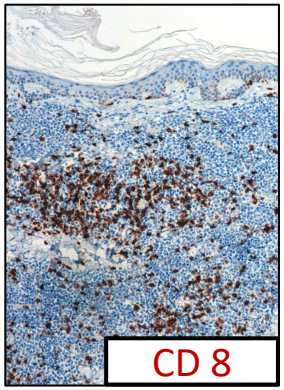
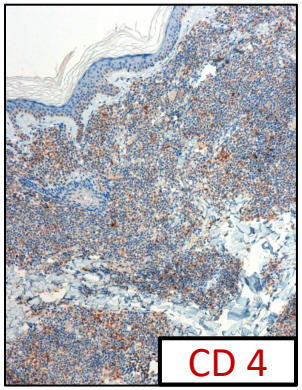

TDT

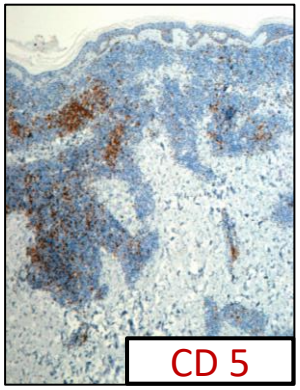
2 POBLACIONES LINFOIDES….

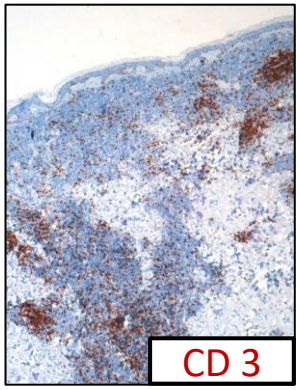
Población minoritaria: Células linfoides pequeñas

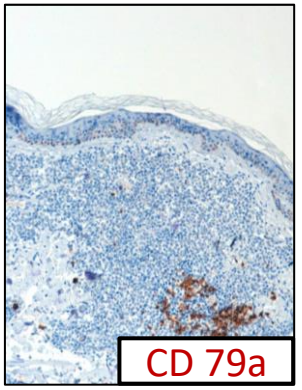




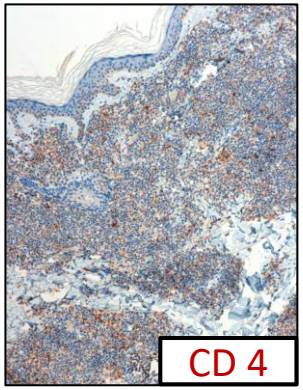


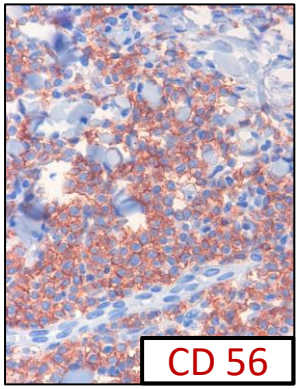






Población mayoritaria: Células hemato-linfoides grandes



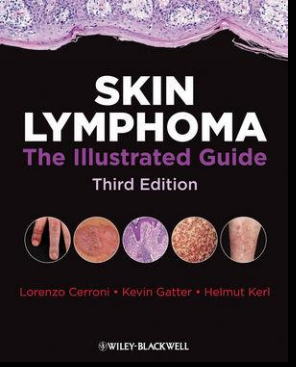


2 Poblaciones linfoides distintas….

¿ diagnóstico ?

¡Sabemos lo que no es!

INFILTRACIÓN SUBCUTÁNEA POR PROCESO HEMATO-LINFOIDE NEOPLÁSICO



~~Neoplasia epitelial~~

Neoplasia hemato-linfoide

- Neoplasia de células plasmáticas
- ~~Neoplasia histiocitarias~~
- ~~Leucemia mielomonocítica crónica~~
- ~~Leucemia/Linfoma linfoblástica precursora~~
- ~~Linfoma B~~
- Linfoma T

Linfoma T

Linfoma NK/T extranodal de tipo nasal

Linfoma primario cutáneo T citotóxico y epidermotropo CD8+

Linfoma primario cutáneo T gamma - delta

CD56+ Lymphoma With Skin Involvement
Clinicopathologic Features and Classification

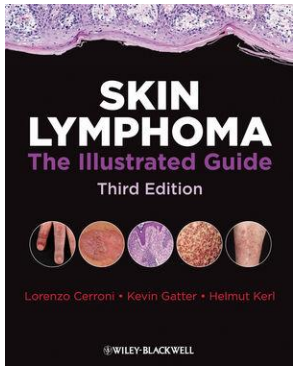
Robert Gniadecki, MD, DMS; Kristian Rossen, MD; Elisabeth Ralfkier, MD, DMS; et al

Table 4. Immunohistochemical and Molecular Characteristics of CD56+ Lymphoma*

Indicator	Blastic Lymphoma	Nasal-Type Natural Killer–Cell/T-Cell Lymphoma	Subcutaneous Panniculitislike Lymphoma
CD3+	15.7 (6.4-25.0)	63.6 (52.9-73.4)	81.3 (67.7-94.8)
CD4+	92.2 (85.6-98.7)	31.1 (17.6-44.6)	3.8 (0.1-11.2)
CD8+	1.6 (0.1-4.7)	2.1 (0.1-6.3)	7.4 (0.1-17.3)
CD30+	3.6 (0.1-10.4)	22.9 (11.0-34.8)	50.0 (22.5-74.5)
T-cell receptor gene rearrangement	4.8 (0.1-11.2)	22.2 (10.1-34.4)	32.1 (14.8-49.4)

*Data are percentage of patients (95% confidence interval). For CD3+, cytoplasmic or membrane expression was considered positive.

Linfoma T



Linfoma NK/T extranodal de tipo nasal

Linfoma primario cutáneo T citotóxico y epidermotropo CD8+

Linfoma primario cutáneo T gamma - delta

Epidemiología : Asia, sudamérica central. Adultos edad media

Presentación clínica

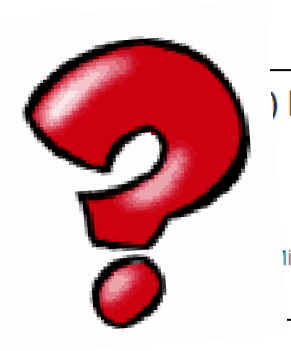
Etiología : EBV

CD 56 +
Granzima +
TIA 1 +

CD56-positive (na) lymphoma arising on the skin

Report of two cases and rev

Shin-ichi Ansai, Kunihiko Ma ikio Matsuda, Souichi Saitoh, Shinobu Suwa, Hiroki Saitoh, Mikio Ohtsuka



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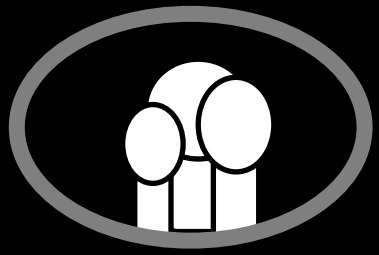
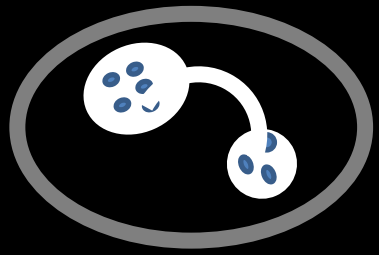




¿Qué hacemos con nuestro caso?

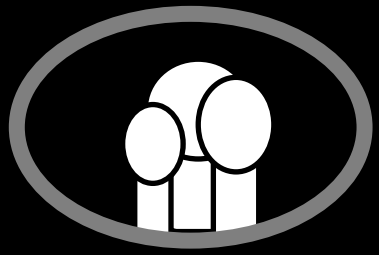
Lo informamos como entidad benigna

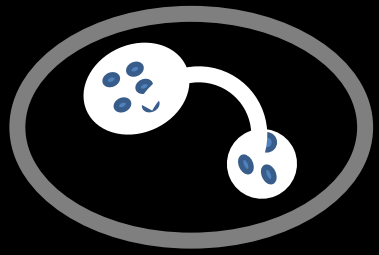
Dejamos el caso tal cual

Ampliamos panel IHQ y estudiamos

No es posible el diagnóstico

50:50





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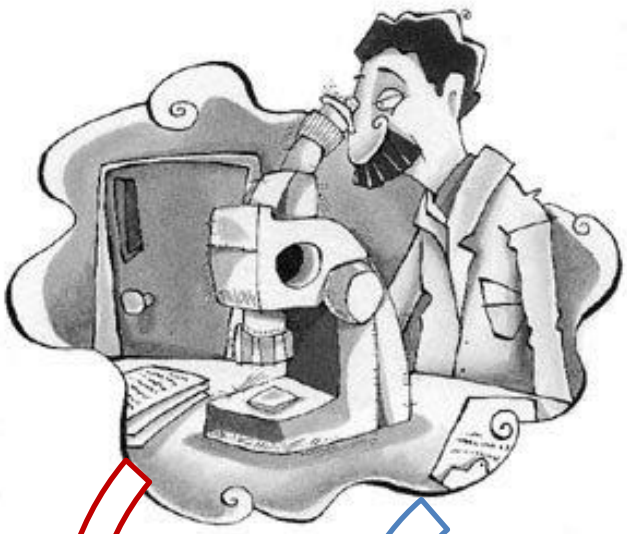

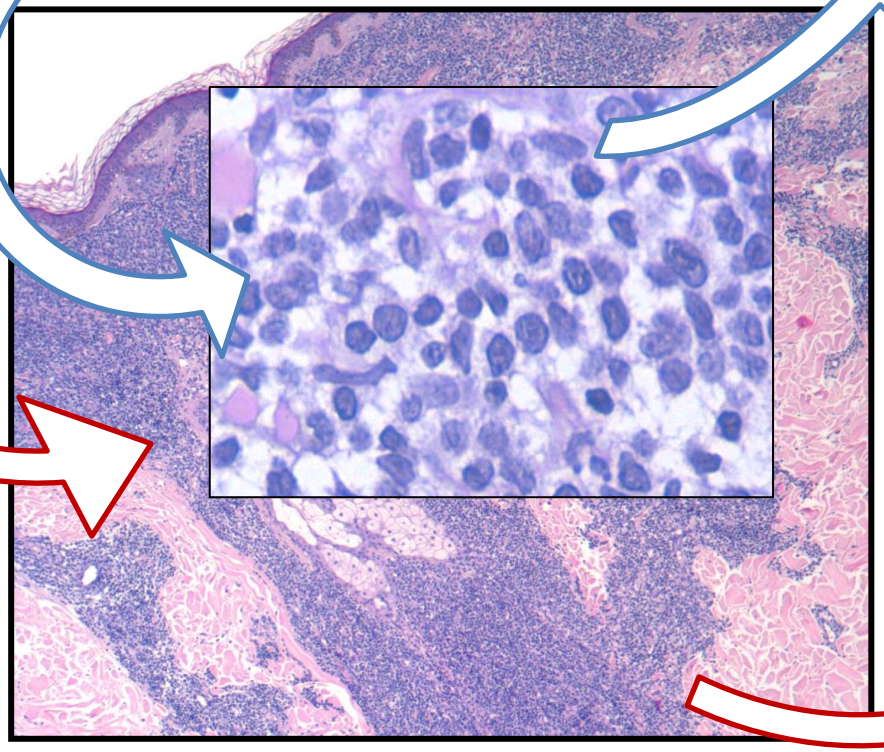


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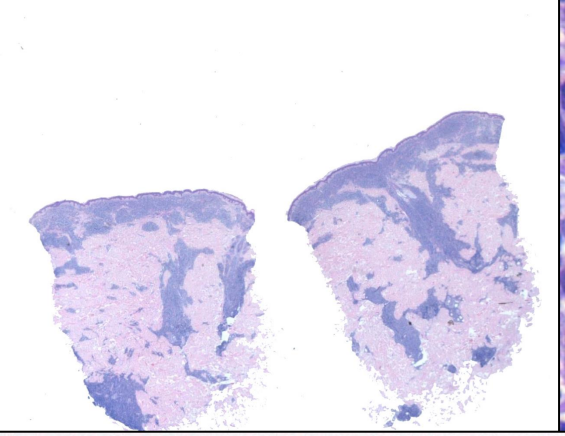

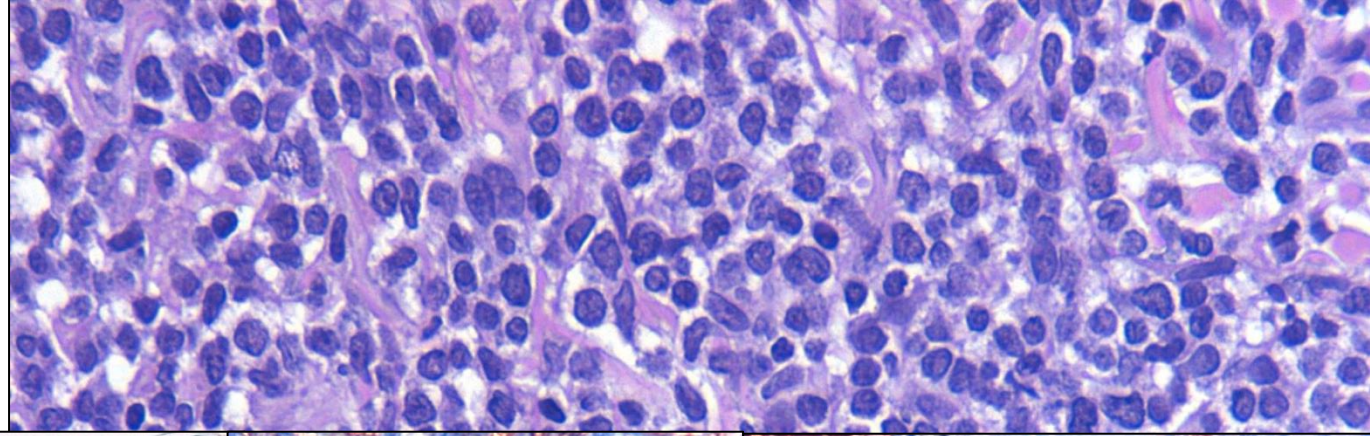

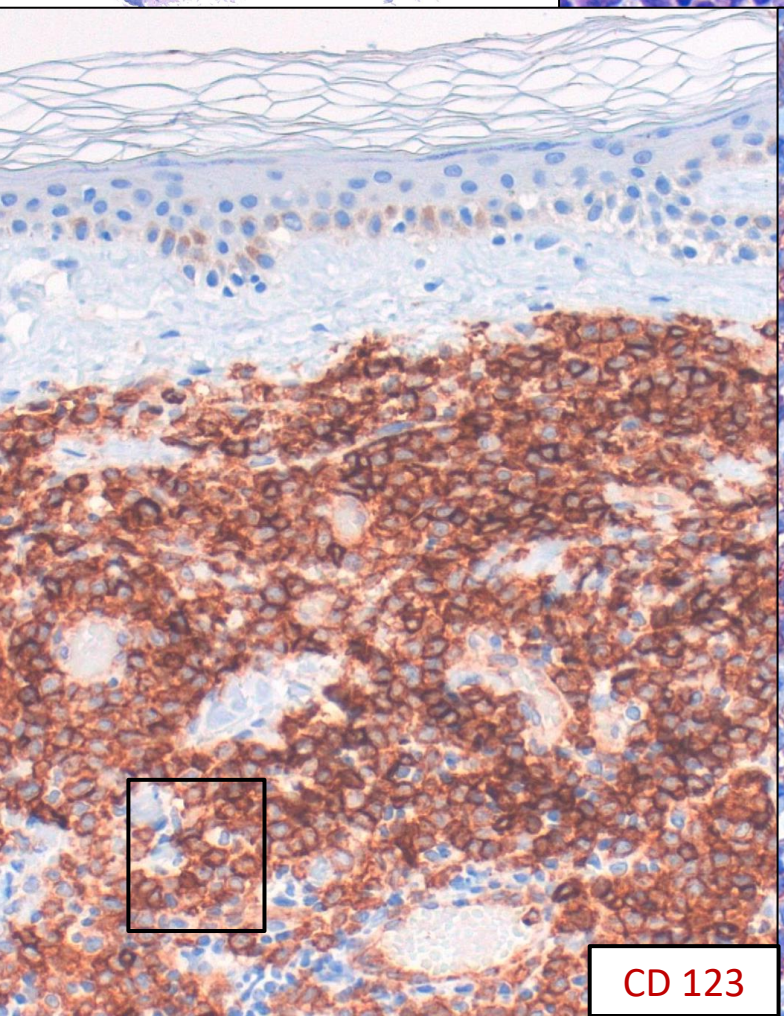

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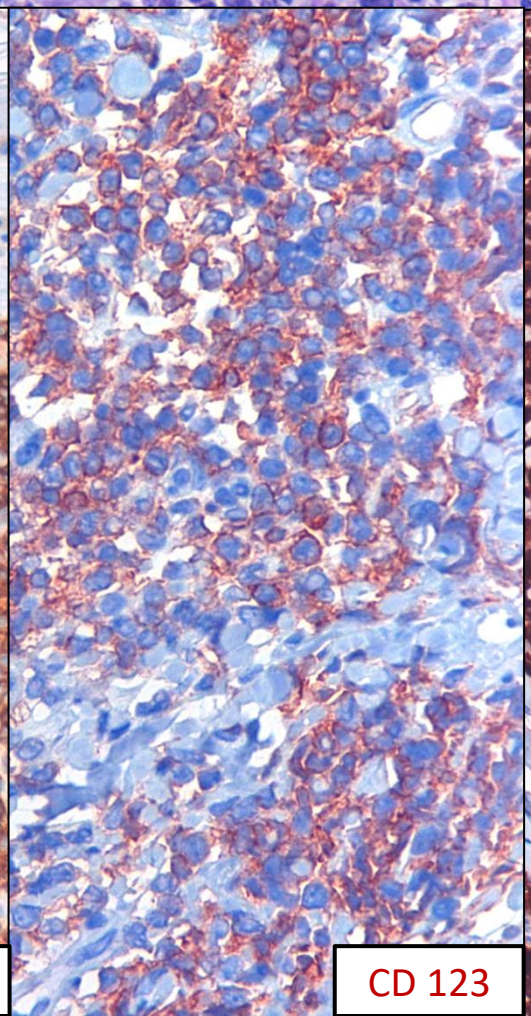

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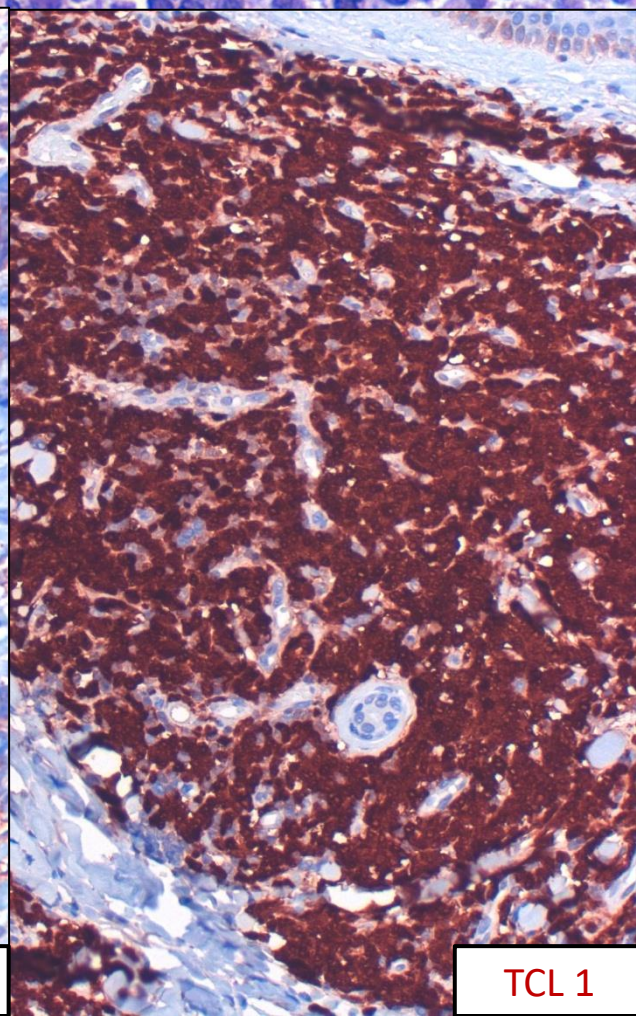
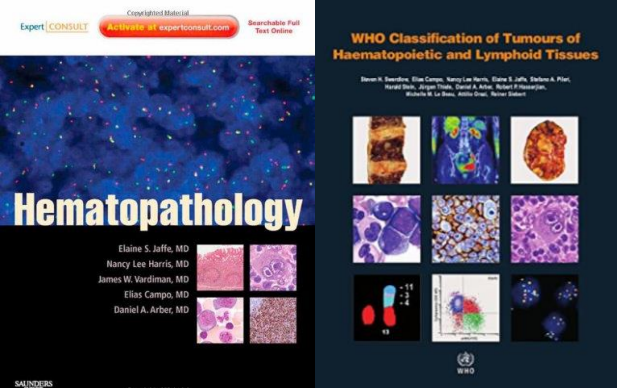


TCL 1

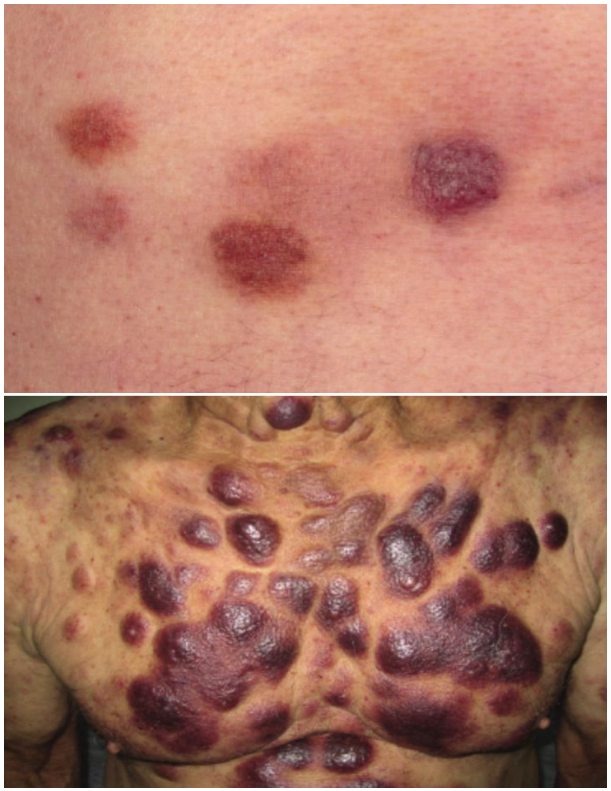
NEOPLASIA DE CÉLULAS DENTRÍTICAS PLASMOCITOIDES BLÁSTICA

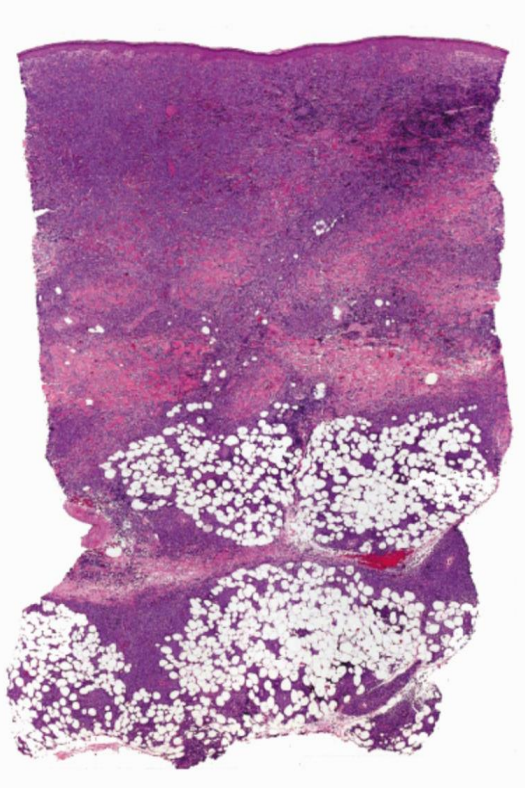


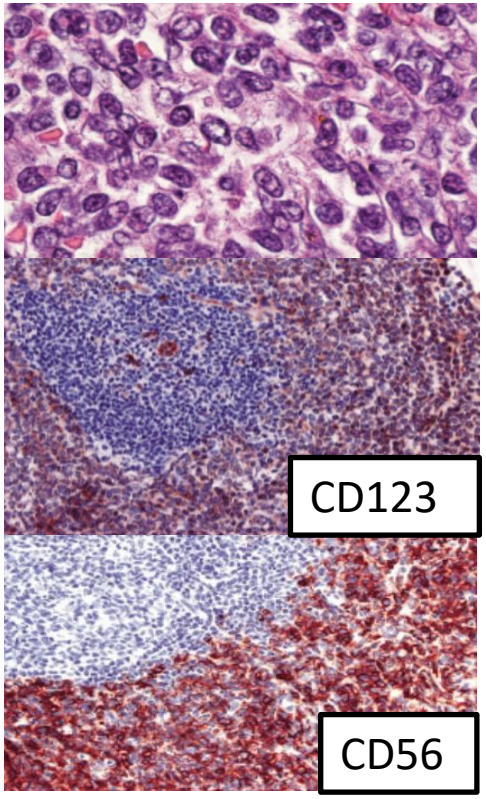
NEOPLASIA DE CÉLULAS DENTRÍTICAS PLASMOCITOIDES BLÁSTICA

0,25-0,7% de los linfomas. <1% de las leucemias

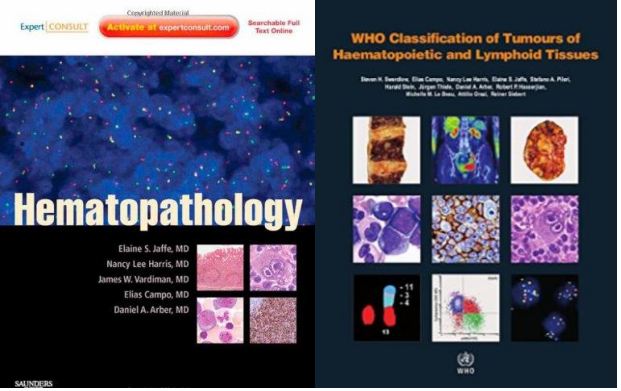
Clínica cutánea inicial, progresa rápido a cuadro sistémico







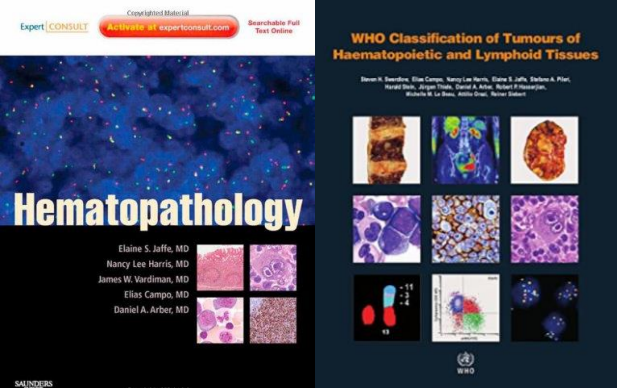
Cerroni L. Skin Lymphomas. 4th ed. Oxford, UK: Wiley-Blackwell; 2014.



NEOPLASIA DE CÉLULAS DENTRÍTICAS PLASMOCITOIDES BLÁSTICA

TABLA 1. Diagnóstico diferencial entre la neoplasia hematodérmica CD4+/CD56+ y otras neoplasias hematológicas con infiltración cutánea*

CARACTERÍSTICAS	NH	LC	LT/NK	LCT	LCB
Célula de origen	CDP / DC2	Mieloblasto, monoblasto	Célula NK, linfocito T citotóxico	Linfocito T	Linfocito B (diferentes etapas de maduración)
Morfología / histopatología	Infiltración de la dermis y TCS, zona de Grenz, células de aspecto monótono, tamaño mediano, núcleo redondo, nucleólos ocasionales y citoplasma basófilo escaso. En frotis, ocasionales prolongaciones citoplásmicas y zona clara de Golgi. Necrosis y angiocentricidad poco frecuente	Infiltración de la dermis y TCS, células de tamaño mediano a grande, núcleos redondos o plegados, cromatina muy fina, múltiples nucleólos prominentes, citoplasma abundante y en ocasiones prolongaciones citoplásmicas	Infiltración de la dermis y TCS, invasión y destrucción de vasos sanguíneos con necrosis extensa. Puede afectar epidermis. Células de tamaño variado, núcleo irregular u oval y citoplasma claro. Puede haber infiltrado inflamatorio asociado	Varios subtipos: MF, SS, ATLL, etc. Infiltración de la epidermis, dermis y TCS. Células pequeñas a medianas, núcleo cerebriforme (MF, SS) En ocasiones no hay afección epidérmica (SS) Células medianas a grandes con núcleo cerebriforme (ATLL)	Varios subtipos: PCMZL, PCFCL, PCLBCL, etc. Infiltración de la dermis y TCS. Predominan linfocitos pequeños, células B «de la zona marginal», células plasmáticas (PCMZL). Mezcla de centrocitos y centroblastos, patrón folicular o difuso (PCFCL). Predominan células grandes con morfología de inmunoblastos, patrón difuso (PCLBCL)



NEOPLASIA DE CÉLULAS DENTRÍTICAS PLASMOCITOIDES BLÁSTICA

CD123 NO ES UN MARCADOR ESPECÍFICO DE CÉLULAS DENDRÍTICAS PLASMOCITOIDES

LYMPHOID NEOPLASIA

SPIB, a novel immunohistochemical marker for human blastic plasmacytoid dendritic cell neoplasms: characterization of its expression in major hematolymphoid neoplasms

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Key Points

- SPIB is an Ets transcription factor involved in plasmacytoid dendritic cell differentiation.
- SPIB protein expression can be used as a marker for human blastic plasmacytoid dendritic cell neoplasms.

SPIB **is an Ets transcription factor that is expressed exclusively in mature B cells, T-cell progenitors, and plasmacytoid dendritic cells. In the present study, we developed a novel mAb against the SPIB protein and characterized its expression in major hematolymphoid neoplasms, including a series of 45 cases of blastic plasmacytoid dendritic cell (BPDC) neoplasms and their potential cutaneous mimics. We found that SPIB is expressed heterogeneously among B- and T-cell lymphoma types. Interestingly, SPIB is expressed in a large proportion of nongerminal center type DLBCLs. In cutaneous neoplasms, SPIB is overexpressed in all BPDC neoplasms, but none of its cutaneous mimics. SPIB remains overexpressed in all cases that lack 1 or 2 of the markers used for BPDC neoplasms (ie, CD4, CD56, TCL1, and CD123). We conclude that SPIB expression can be used as a tool for diagnosing BPDC neoplasms, but it needs to be tested in conjunction with the growing arsenal of markers for human plasmacytoid dendritic cells. (*Blood*. 2013;121(4):643-647)**

Evolución

Nuestro paciente: **Bx MO no afectada**.
En remisión completa a la espera **trasplante alogénico.**

Conclusiones

- Es rara, está infradiagnosticada.
- Presentación clínica indolente contrasta con la diseminación sistémica y agresiva de esta enfermedad.
- El diagnóstico diferencial se ha de considerar diferentes linfomas cutáneos, leucemias, y la histiocitosis de células de Langerhans, que se descartan por el perfil inmunohistoquímico.
- No existe una pauta terapéutica establecida. Trasplante alogénico de elección.

Muchas gracias por vuestra atención