

I quadri clinici osservati dal dermatologo

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Direttore: Prof.ssa Bianca Maria Piraccini

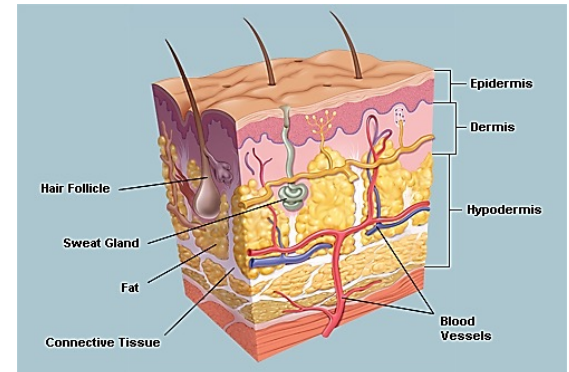


**NEOPLASIA A
CELLULE DENDRITICHE
PLASMATICI
BLASTICHE:**
NUOVE OPZIONI DIAGNOSTICHE
ED ALGORITMO TERAPEUTICO

Napoli
22 Febbraio 2024
Grand Hotel Santa Lucia

BPDCN: clinical features

- Anatomic involvement at the diagnosis
- **Skin** (60-100%)
- Bone marrow and peripheral blood (60-90%)
- Lymph node (40-50%)



BPDCN: clinical features

- Most cases have skin as VERY FIRST clinical manifestation (90%)
- To date case report or case series
- Lack of a defined pattern of clinical presentation
- Tricky and easy to misdiagnose

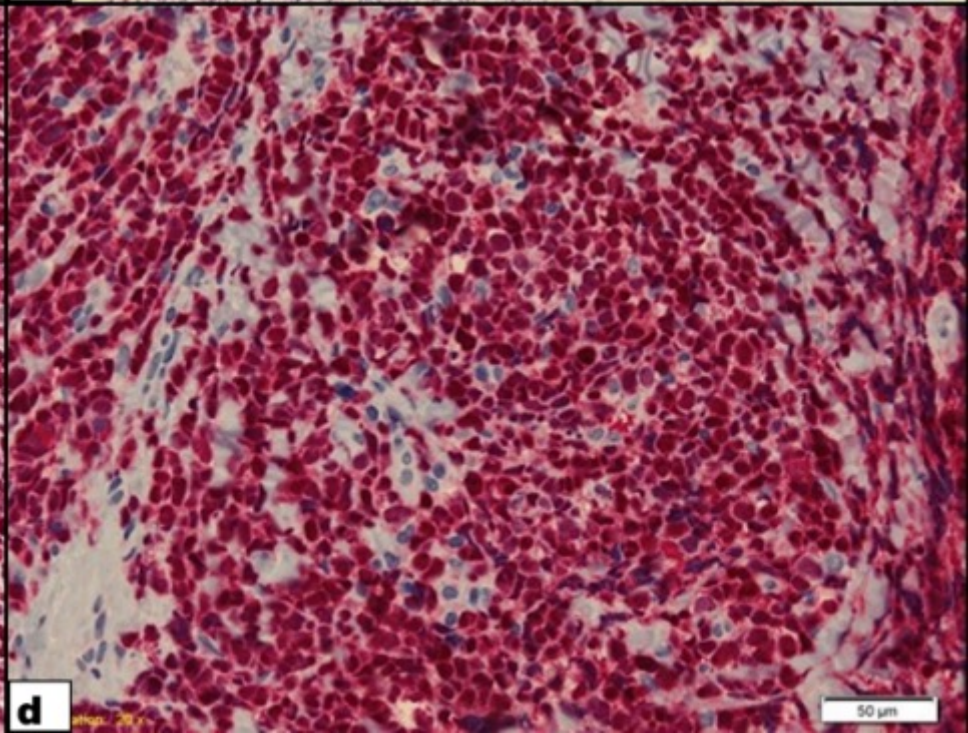
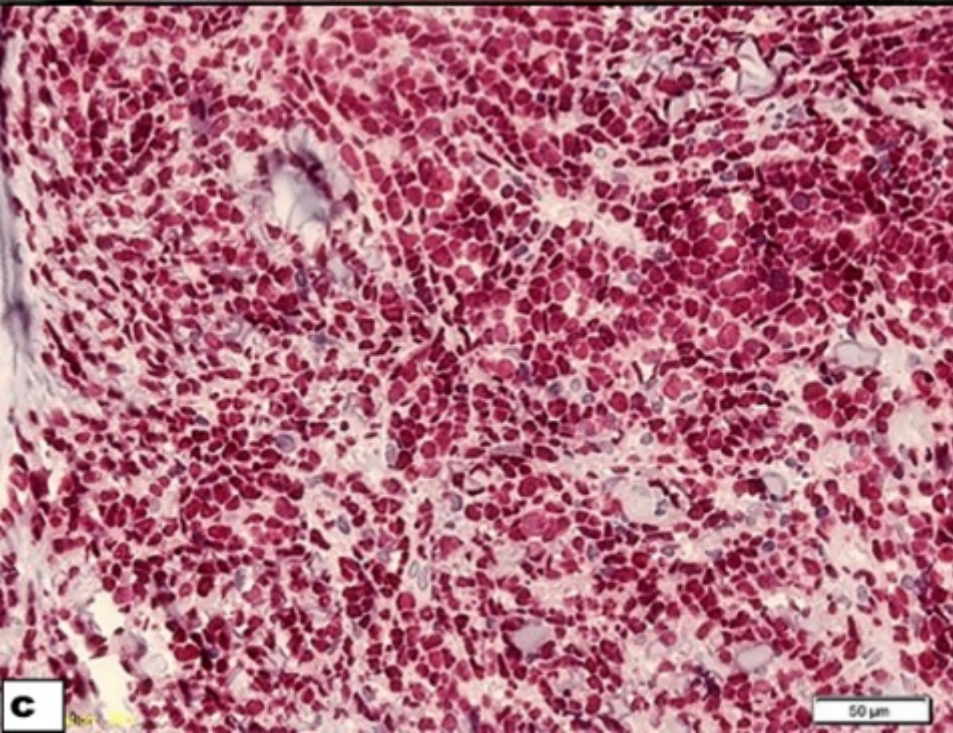
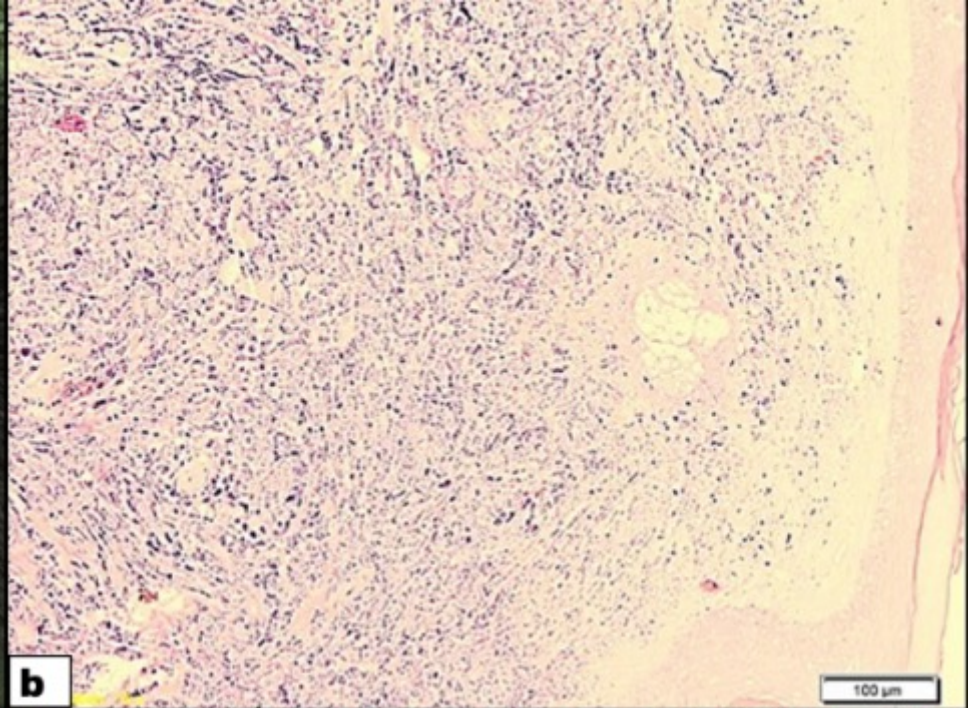
BPDCN: differential diagnosis

- Different clinical presentation
- Possible differential diagnosis
- **Possible misdiagnosis**
- **Rare disease**
- The eyes recognise what they know



BPDCN: clinical features

- NO anatomic preferred site
- Two types of clinical appearance
- Single-lesion
- Multiple-lesion (eruptive presentation)





BPDCN: clinical features

- Morphologically variable clinical presentation
- Macules
- Papules
- Papulo-nodular lesions
- Patches
- Plaques
- Nodules
- Size from some mm to 10 cm

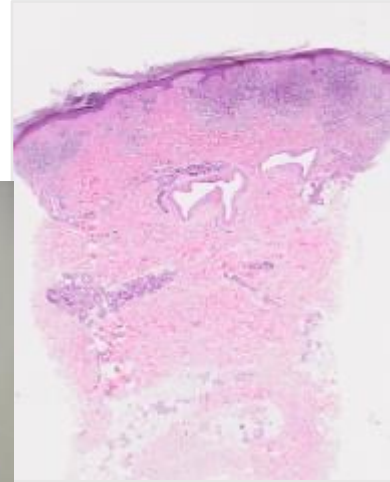
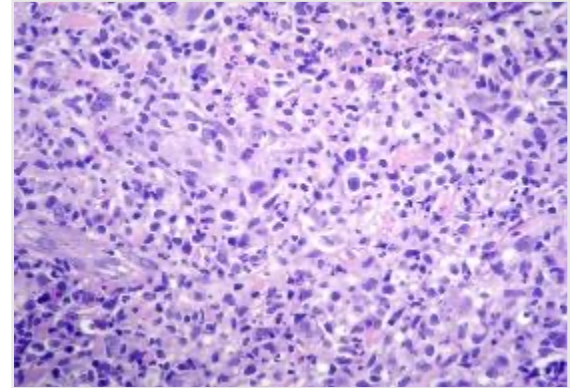
BPDCN: clinical features

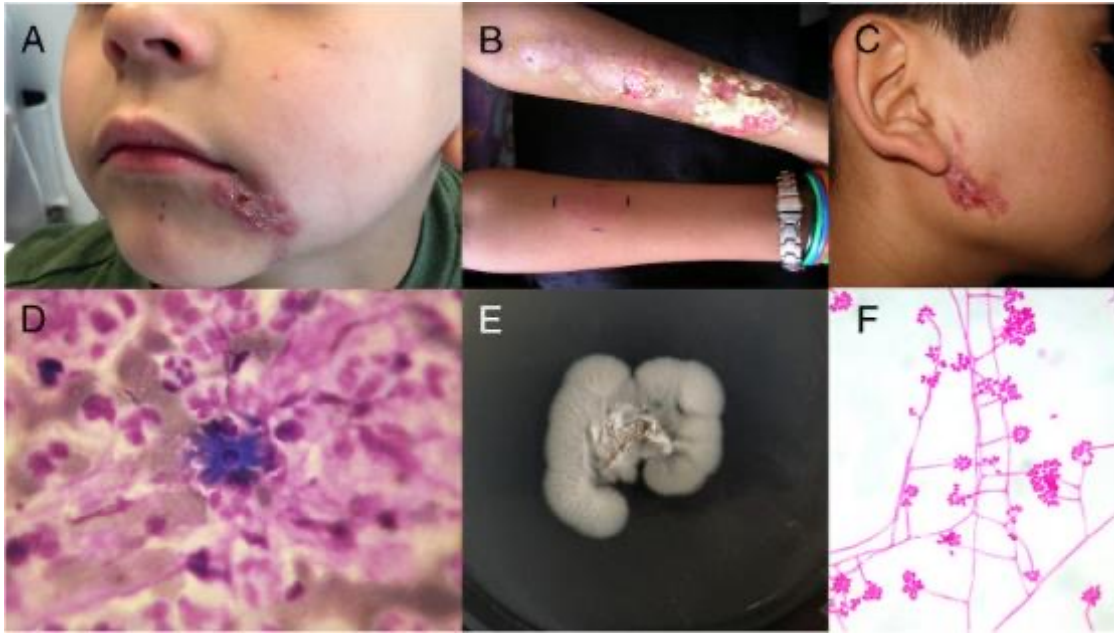
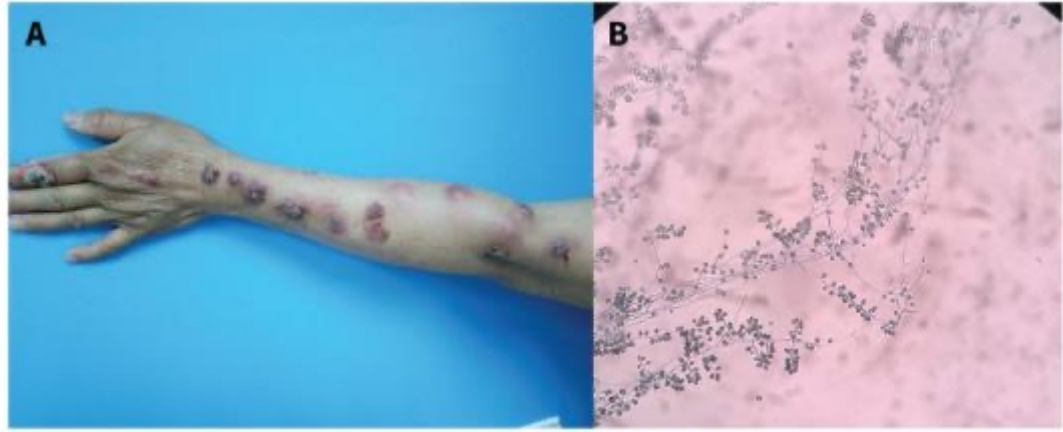
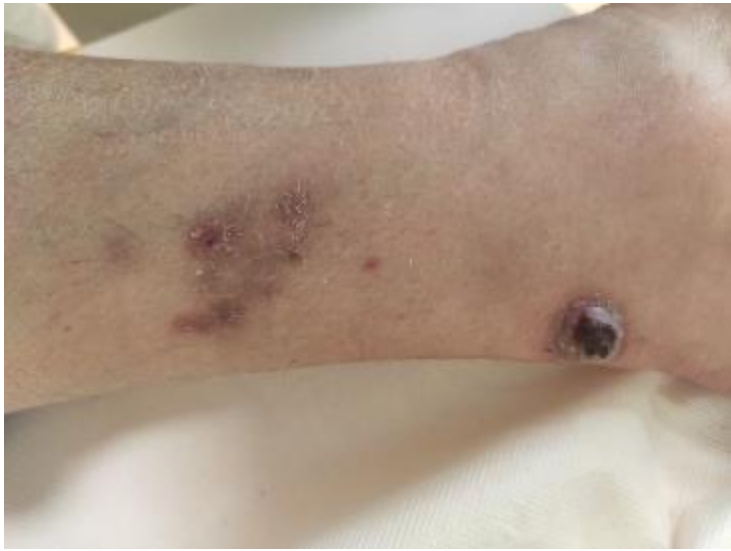
- **Isolated lesions**
- Violaceous or purplish-like
- Bruise-like

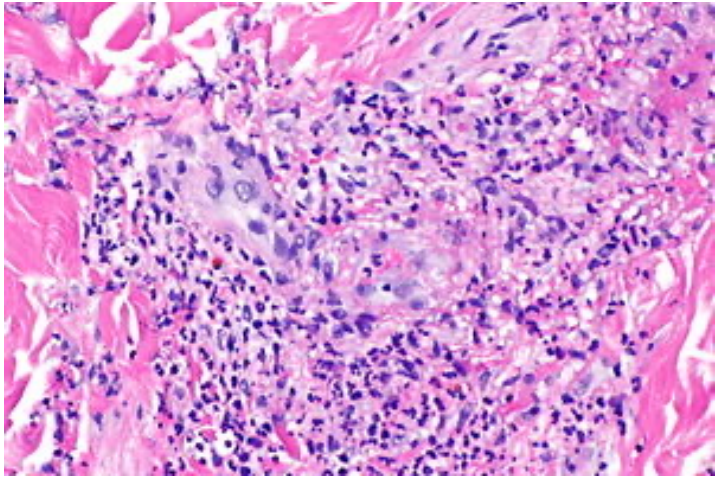
- **Multiple lesions**
- Eruptive appearance
- Lesions asymptomatic
- Rare ulceration



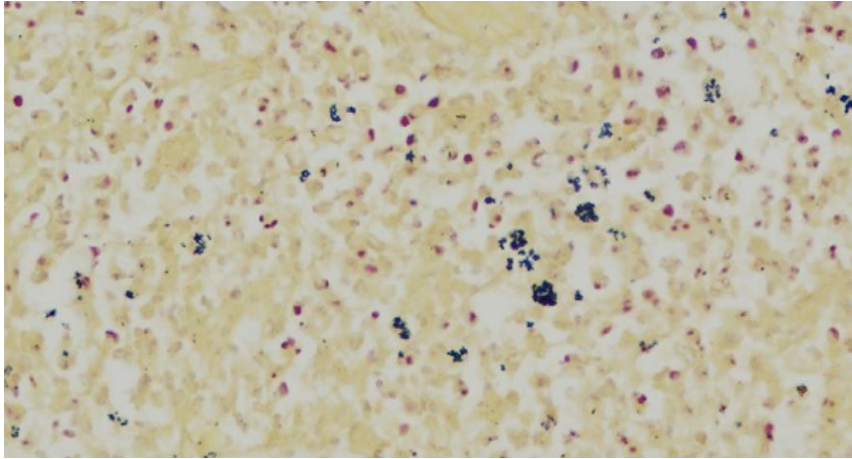












Actas Dermosifiliogr. 2011;102(2):142–145



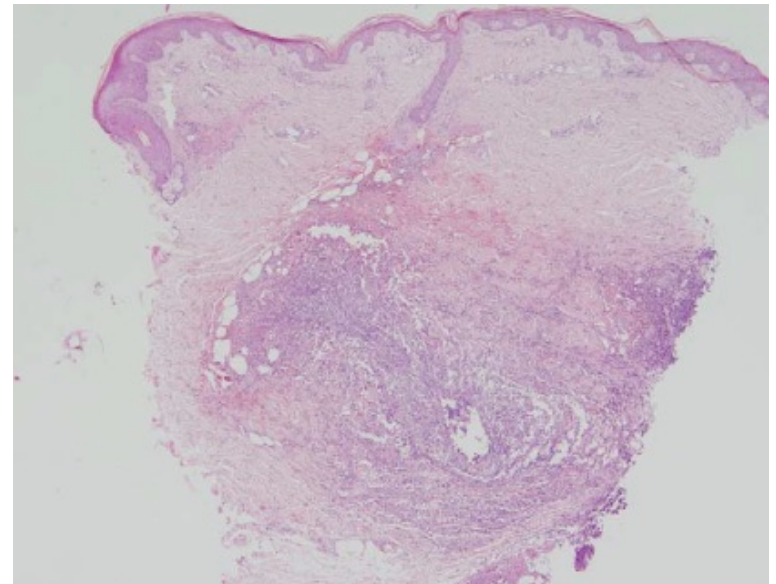
ACTAS Derma-Sifiliográficas

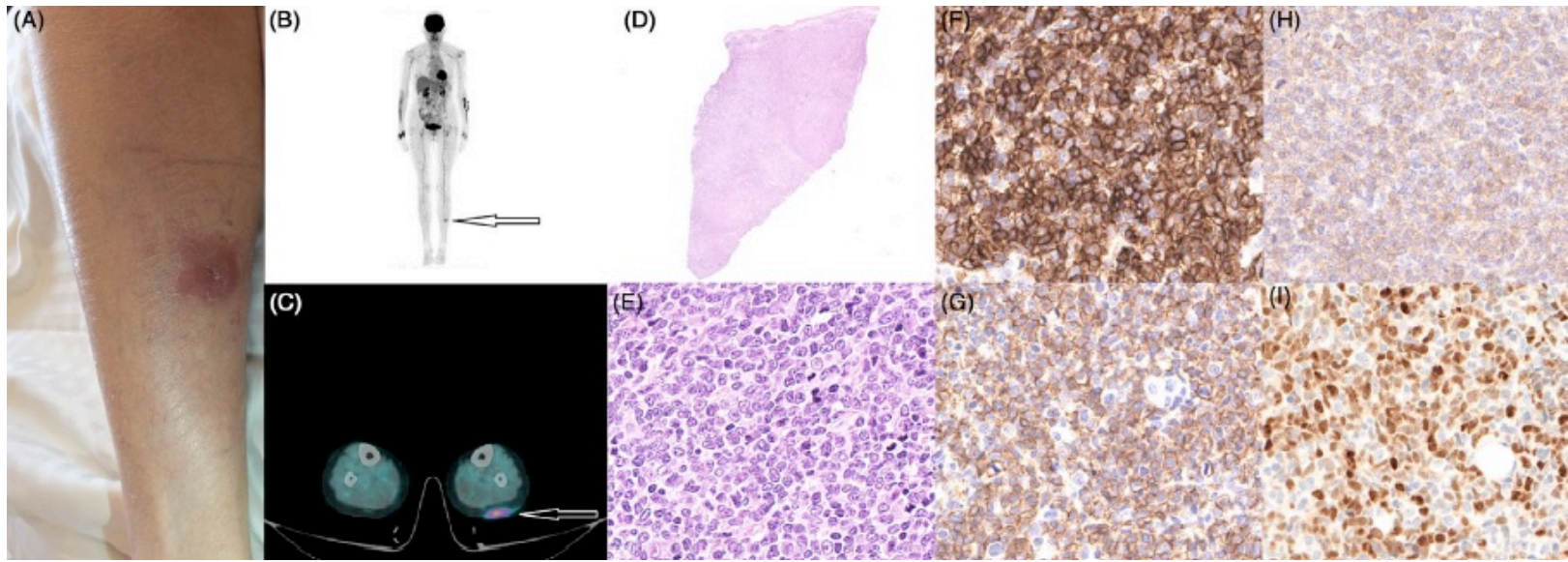
Full English text available at
www.elsevier.es/ad

CASO CLÍNICO

Embolismos sépticos cutáneos tras angioplastia

L. Sanz-Canalejas^{a,*}, U. Floristán-Muruzábal^a, M. Feito-Rodríguez^a,
E. Sendagorta-Cudós^a, M.J. Beato-Merino^b y P. Herranz-Pinto^a





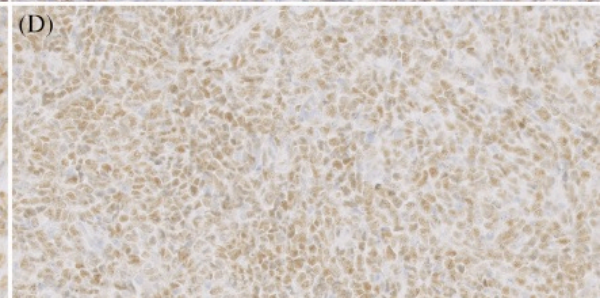
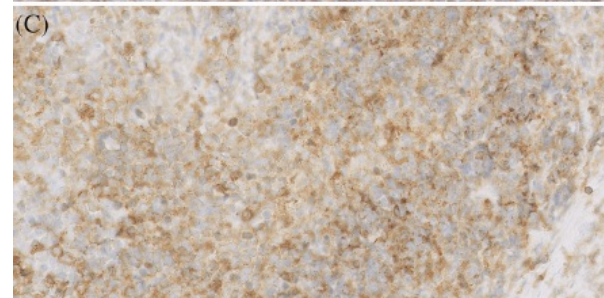
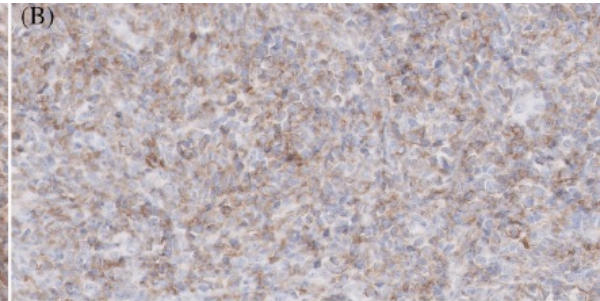
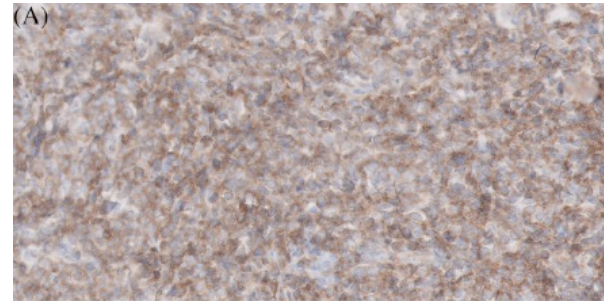
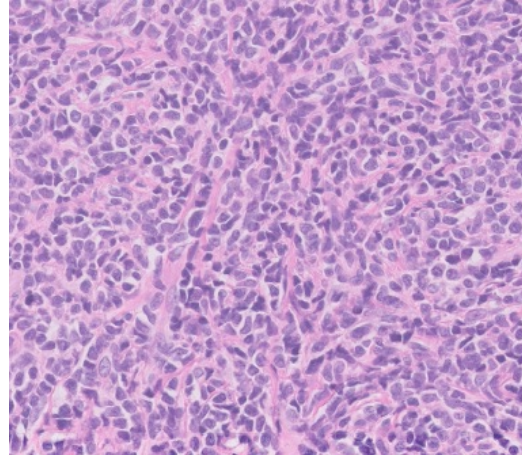
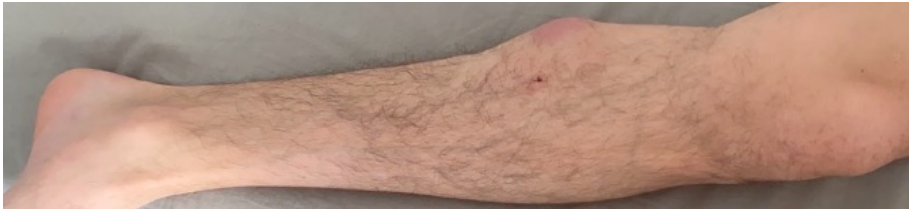
DOI: 10.1002/jha2.370

HAFMATOLOGY IMAGES

Report of cutaneous loxoscelism caused by violin spider bite in Northern Italy

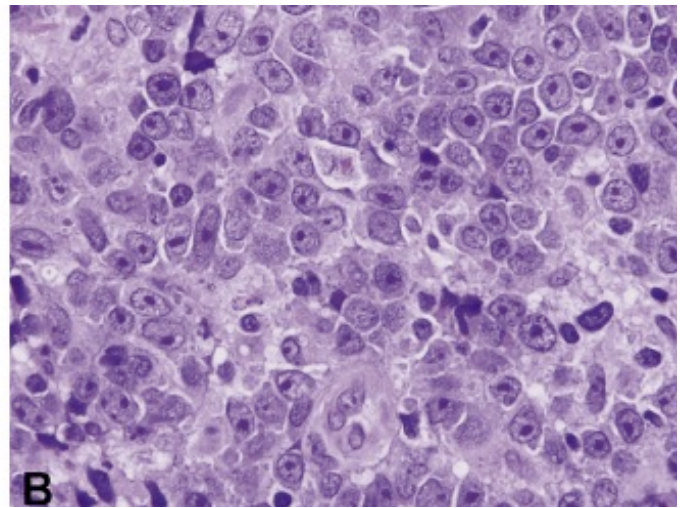
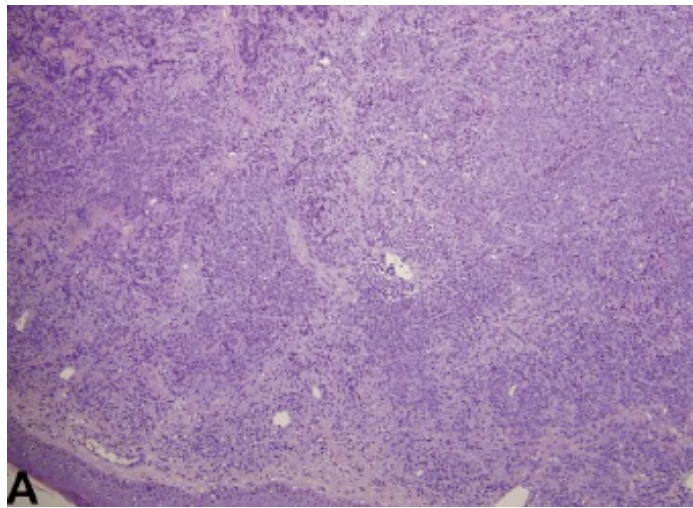
Isolated skin infiltration by a blastic plasmacytoid dendritic cell neoplasm

Grégoire Stalder^{1,#} | Dina Milowich^{2,#} | Sabine Blum¹ | Jacqueline Schoumans³ | Bettina Bisig⁴ | Olivier Spertini¹



Localized skin-limited blastic plasmacytoid dendritic cell neoplasm

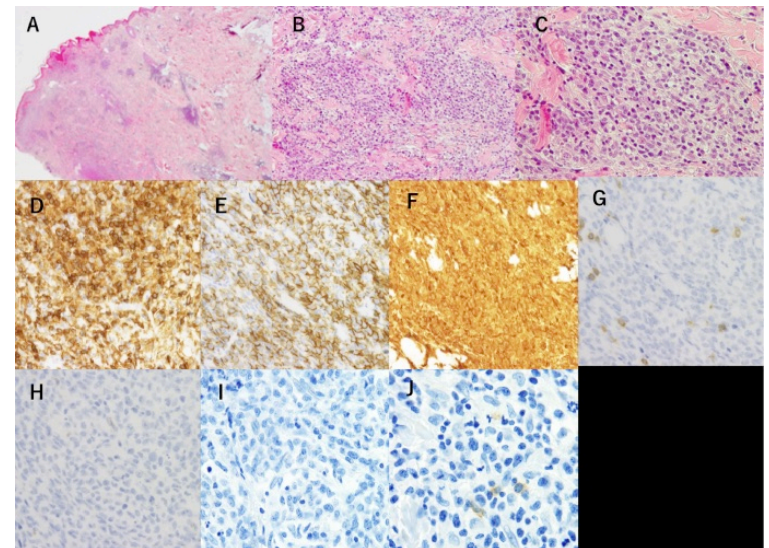
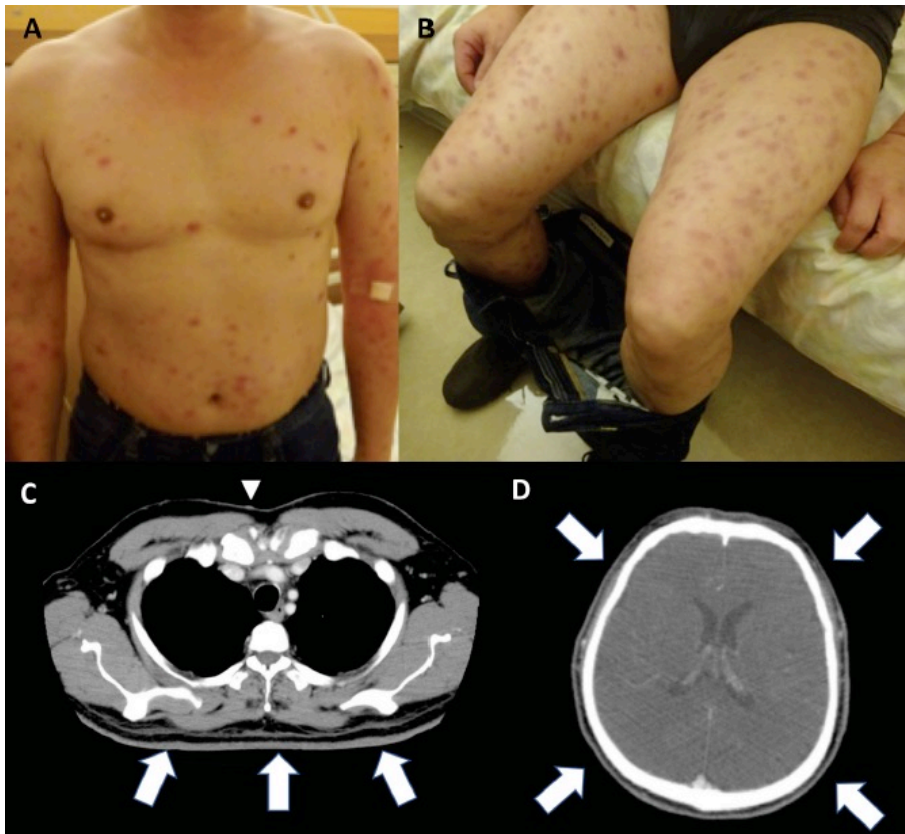
Edwin U. Suárez¹ | Javier Cornago¹ | Miguel Á. Piris² | Socorro M. Rodríguez Pinilla² | José L. López-Lorenzo¹ | Carlos Soto¹

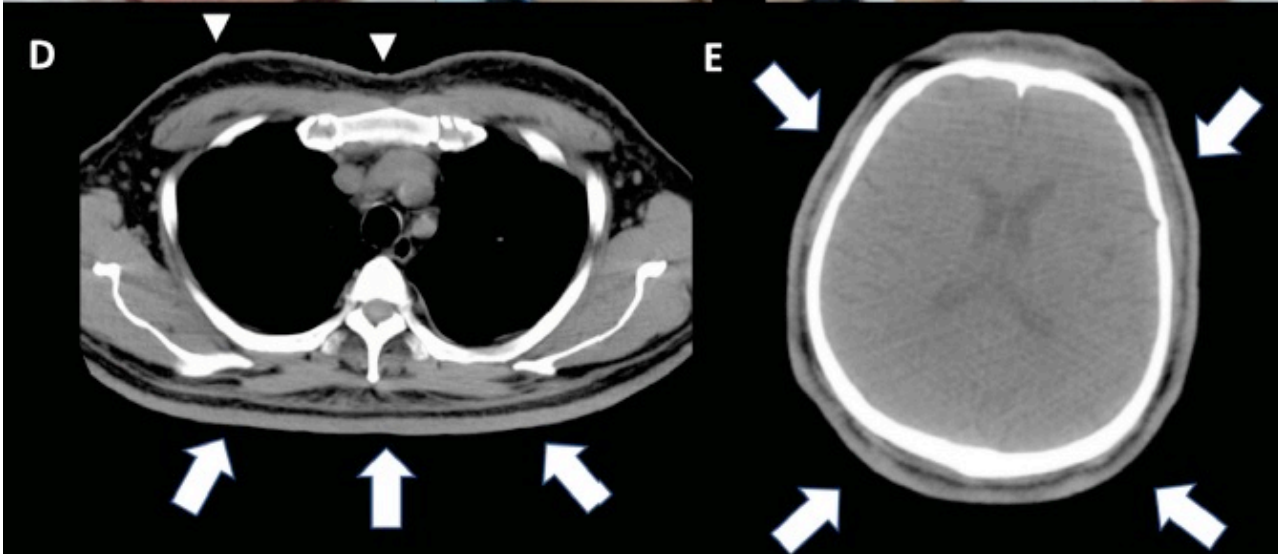
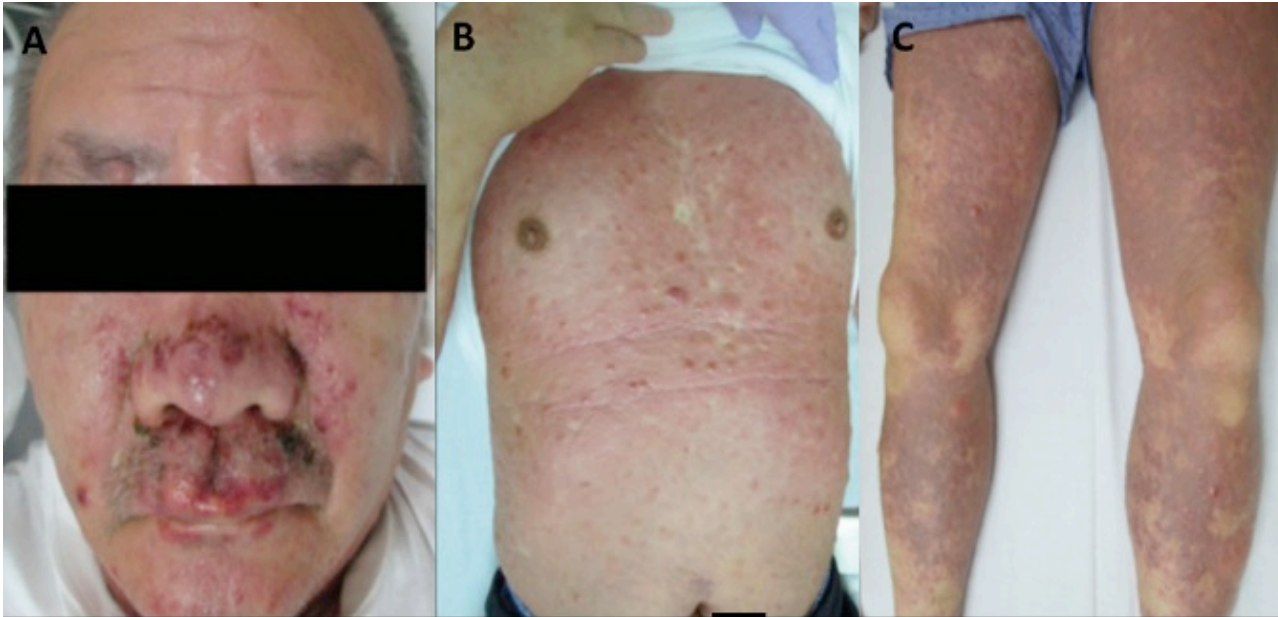


Case Report

Rapidly progressing blastic plasmacytoid dendritic cell neoplasm causing diffuse skin thickening: A case report with sequential computed tomography examinations

Kyohei Yoshioka^a, Ryo Kurokawa^{a,*}, Shiori Amemiya^a, Hiroaki Koyama^{a,b}, Kensuke Matsuda^b, Akira Honda^b, Mineo Kurokawa^b, Aya Shinozaki-Ushiku^c, Osamu Abe^a





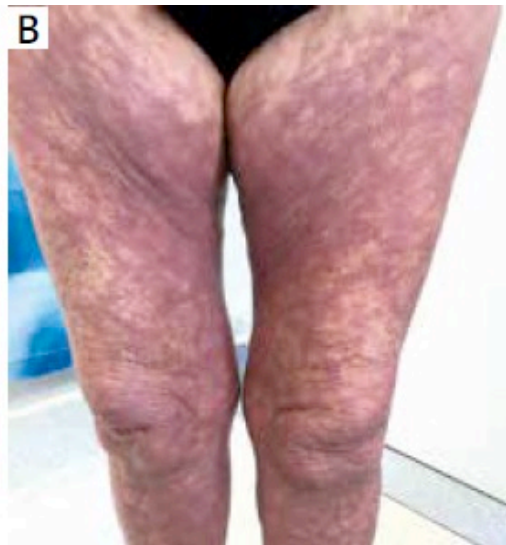


Article

Sézary Syndrome: Different Erythroderma Morphological Features with Proposal for a Clinical Score System

Gabriele Rocuzzo^{1,*†}, Silvia Giordano^{1,†}, Gianluca Avallone¹, Marco Rubatto¹, Silvia Canonico¹, Ada Funaro², Erika Ortolan², Rebecca Senetta³, Paolo Fava¹, Maria Teresa Fierro¹, Simone Ribero¹ and Pietro Quaglino¹





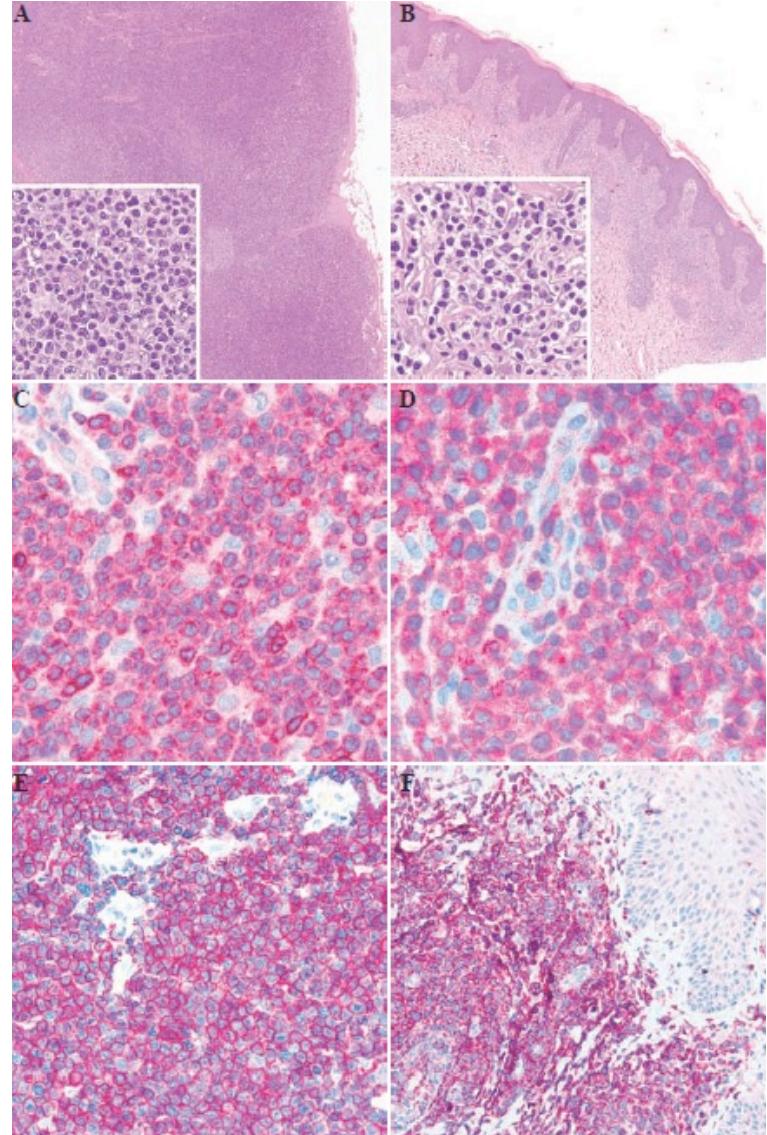
Clinicopathologic retrospective analysis of blastic plasmacytoid dendritic cell neoplasms

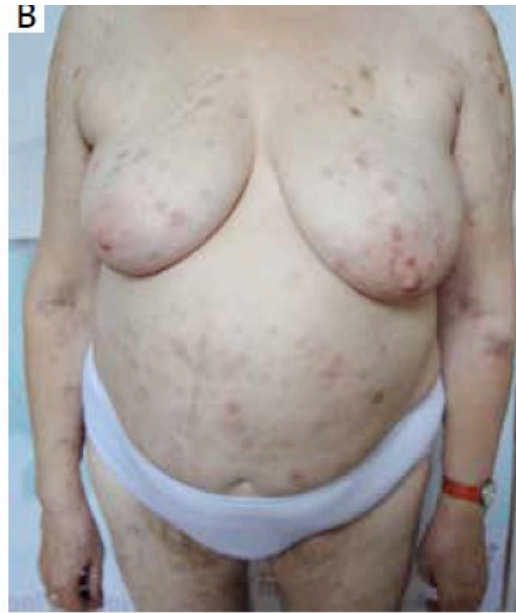
Agnieszka Owczarczyk-Saczonek¹, Małgorzata Sokołowska-Wojdyło², Berenika Olszewska², Marta Malek³, Aleksandra Znajewska-Pander¹, Anna Kowalczyk⁴, Wojciech Biernat⁵, Grażyna Poniatowska-Broniek⁶, Wanda Knopińska-Postuszny⁷, Zygmunt Koziolec⁶, Roman Nowicki², Waldemar Placek¹

Alessandro PILERI^{1,2}
Cinzia PELLEGRINI³
Claudio AGOSTINELLI⁴
Vieri GRANDI²
Annalisa PATRIZI¹
Pier Luigi ZINZANI³
Nicola PIMPINELLI²

Erythroderma and non-Hodgkin T-cell lymphoma: what else, apart from Mycosis Fungoides and Sézary syndrome?

Background: Peripherical T-cell lymphomas not otherwise specified







ICD-O code

9709/3

Epidemiology

This is a rare disease, accounting for < 1% of all CTCLs [26,228,2832]. It occurs mainly in adults. There are no predisposing factors. **Localization**



Fig. 6093 Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma. Lesions are often haemorrhagic; they are diffuse and associated with epidermal ulceration.



Fig. 5877 Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma. A large localized nodulotumoural lesion on the leg, showing ulceration and epidermal necrosis.

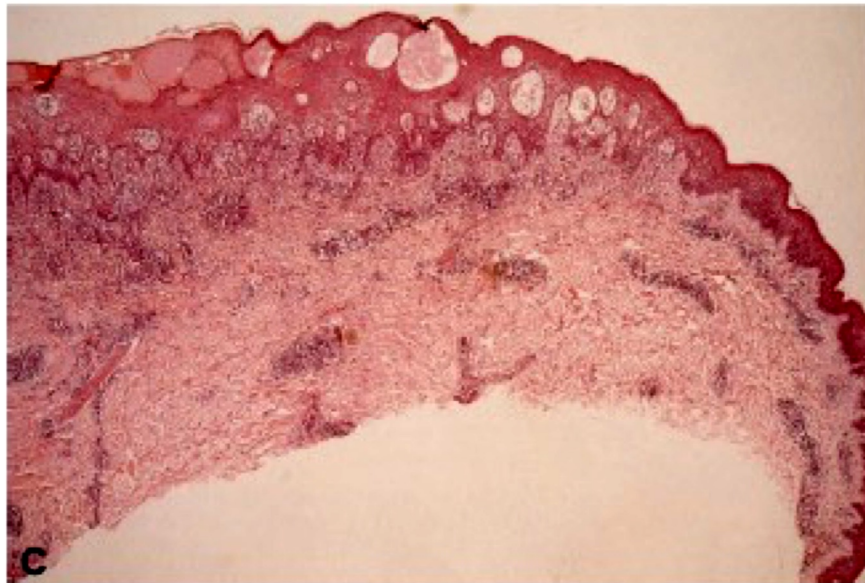


Figure 1. **A:** Clinical features of case no. 6: widespread eruption of patches, plaques, and papulonodular verrucous and hemorrhagic lesions. **B:** Clinical features of case no. 2: typical hemorrhagic and necrotic evolution of some lesions. **C:** Histology (H&E; original magnification, $\times 2.5$) of case no. 2: a perivascular and peridnexal, lichenoid, strongly epidermotropic infiltrate in an acanthotic and hyperplastic epidermis with spongiosis, blistering, and necrosis. **D:** Clinical features of case no. 5: particularly of the papulonodular and verrucous lesions. Note the central resolution of some lesions.

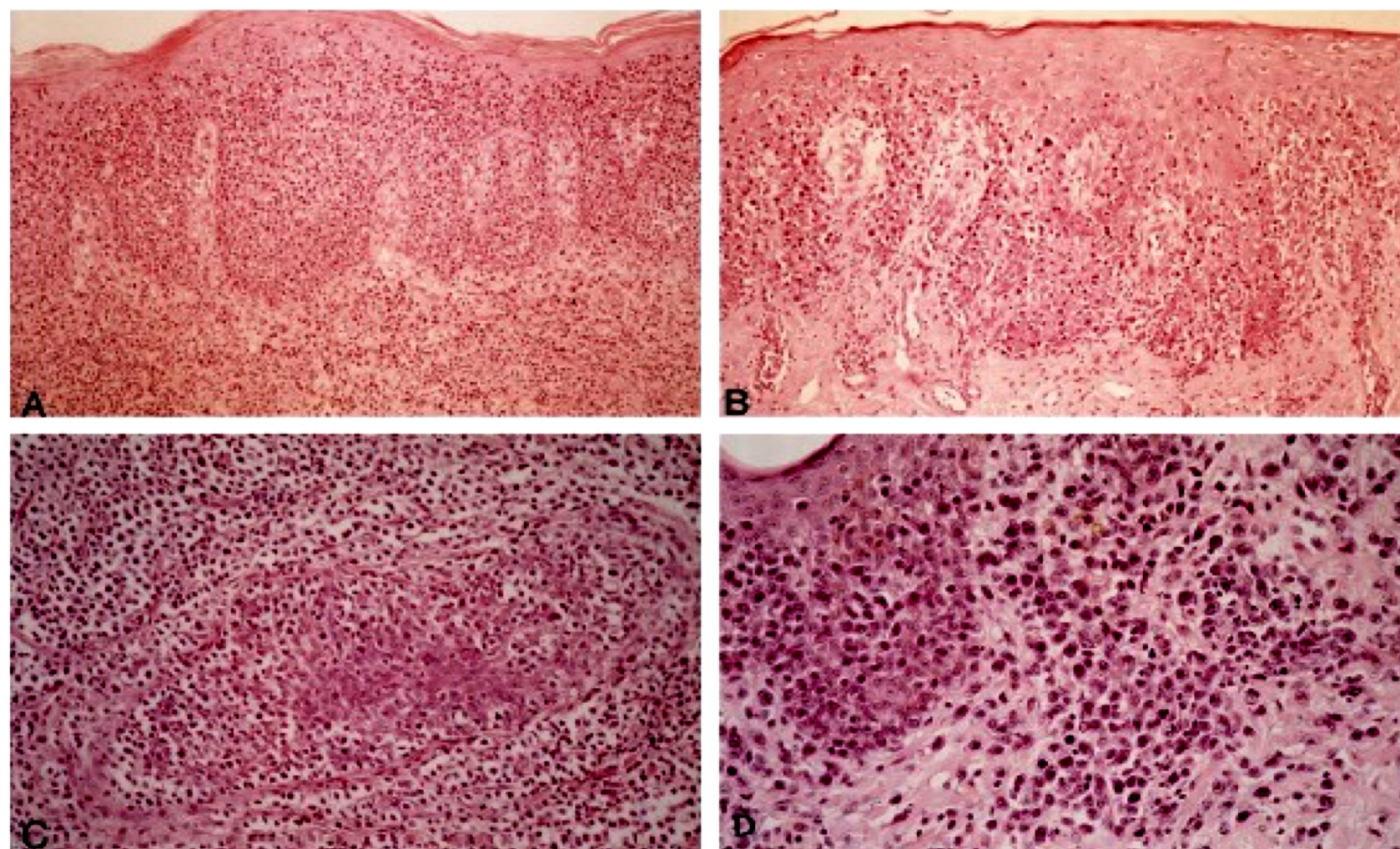


Figure 2. **A:** Histology (H&E; original magnification, $\times 100$) of case no. 4: well developed nodular lesion. Note the diffuse pleomorphic strongly epidermotropic T cell infiltrate. **B:** Histology (H&E; original magnification, $\times 100$) of case no. 1: early lesion. Note the intraepithelial pleomorphic atypical lymphoid infiltrate; the involved epidermis shows extensive keratinocyte necrosis. **C:** Histology (H&E; original magnification, $\times 200$) of case no. 3: the strongly adnexotropic small- to medium-size pleomorphic lymphocytes show a lymphoepithelioid pattern. **D:** Histology (H&E; original magnification, $\times 200$) of case no. 2: well developed tumoral lesion. High magnification of the infiltrate shows perivascular, strongly epidermotropic immunoblasts in the superficial dermis.

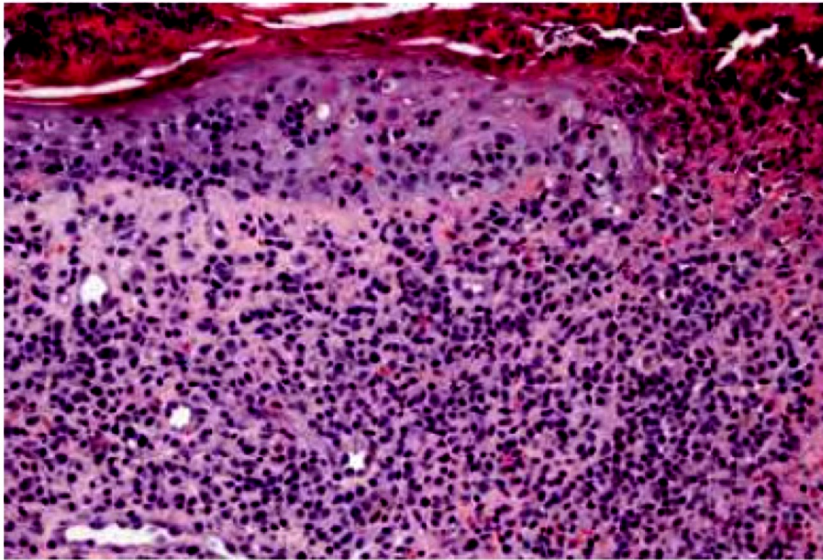


Fig. 6102 Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma. The atypical pleomorphic medium to large cell lymphoid infiltrate occupies the superficial dermis, extending to the epidermis in a pagetoid fashion and leading to epidermal necrosis.

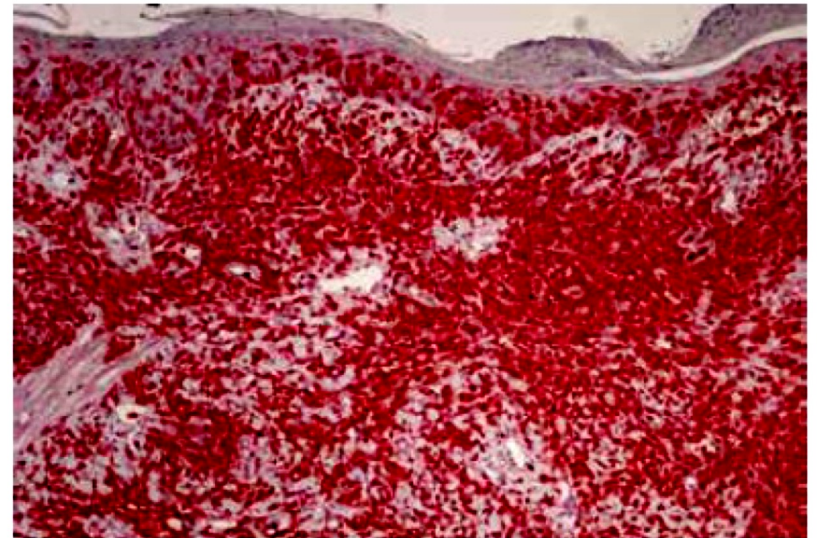
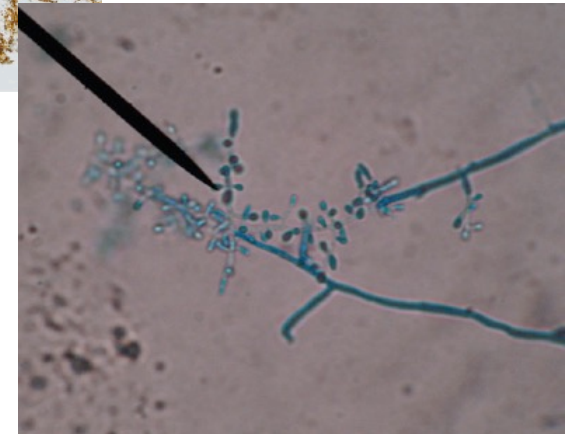


Fig. 6103 Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma. CD8 staining highlights the epidermotropic neoplastic cells in a skin section.

Juvenile Blastic Plasmacytoid Dendritic Cell Neoplasm

Kinan M. HAYANI¹, Gabriele ESCHERICH², Karoline KOCH³, Lars E. FRENCH¹ and Hans H. WOLFF¹

¹Department of Dermatology, University Hospital of Munich (LMU), Frauenlobstrasse 9-11, DE-80337, Munich, ²Clinic of Pediatric Hematology and Oncology, University Medical Centre, Hamburg-Eppendorf, Hamburg, and ³Department of Pathology, Hematopathology Section and Lymph Node Registry, University Hospital Schleswig-Holstein, Kiel, Germany. E-mail: Kinan.hayani@med.uni-muenchen.de
Accepted Jul 9, 2020; Epub ahead of print Jul 29, 2020



Survival outcomes in blastic plasmacytoid dendritic cell neoplasm by first-line treatment and stem cell transplant

Seongseok Yun,¹ Onyee Chan,¹ Daniel Kerr,¹ Nicole D. Vincelette,¹ Afshan Idrees,² Qianxing Mo,³ Kendra Sweet,¹ Jeffrey E. Lancet,¹ Mohamed A. Kharfan-Dabaja,⁴ Ling Zhang,^{2,*} and Lubomir Sokol^{1,*}

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare hematologic malignancy with dismal clinical outcomes. Conventional chemotherapies such cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) and hyperfractionated cyclophosphamide, vincristine, doxorubicin, dexamethasone alternating with high-dose cytarabine and methotrexate (CVAD) have been commonly used for the BPDCN treatment until a recent study showed promising outcomes in patients treated with SL-401 (Tagraxofusp). In this single-institution retrospective study, we identified a total of 49 consecutive BPDCN patients. Among 42 patients who received treatment, hyper-CVAD regimen was associated with higher complete response rate compared with CHOP-based regimens or SL-401 (91% vs 50% vs 50%), although the difference did not achieve statistical significance. Furthermore, there was no significant overall survival (OS) difference between patients treated with SL-401 vs other chemotherapies as their first-line treatment (hazard ratio = 1.597; 95% CI, 0.460-5.548; $P = .431$). Of note, patients who received allogeneic stem cell transplant (allo-SCT) had significantly longer OS (hazard ratio = 0.160, 95% CI, 0.045-0.56; $P = .041$). Extent of disease (skin vs bone marrow vs both) of younger age (<60 years old) did not have significant prognostic impact on OS. Collectively, our study confirmed the survival benefit of allo-SCT and suggests that conventional and intensive chemotherapies such as CHOP and hyper-CVAD as well as SL-401 would be comparable first-line choice for the BPDCN patients.

Clinical presentation

- Jan 2022
- Erythematous patch
- No symptoms
- Punch biopsy

Clinical presentation

- Case tricky
- Innocent patch..
- ..aggressive disease
- The differential diagnosis is seborroic dermatitis





JCMS Case Report

SAGE Open Medical Case Reports

Upadacitinib-induced paradoxical face and scalp dermatitis: A case report of a novel sequela

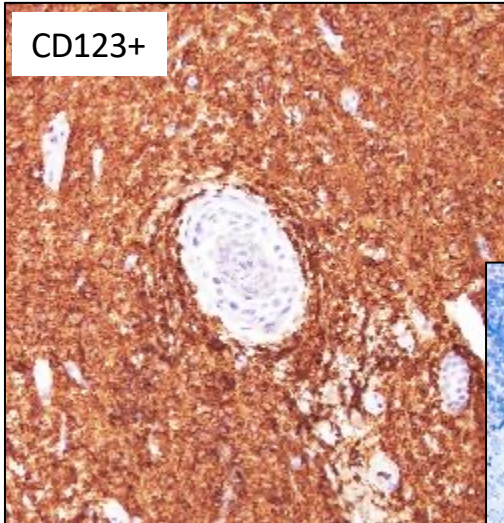
SAGE Open Medical Case Reports
JCMS Case Reports
Volume 11: 1-5
© The Author(s) 2023
DOI: 10.1177/2050319X231164271
journals.sagepub.com/home/sco



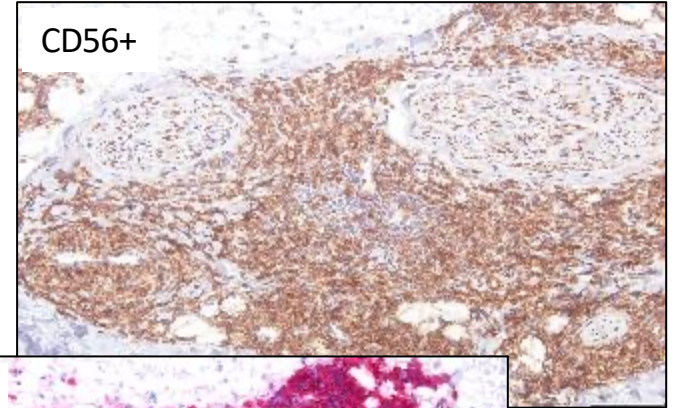
Elena Pastukhova¹, Alison Spurr^{1,2}, Quentin Nakonechny³
and Jennifer Lipson^{1,2}

The diagnosis of BPDCN relies on the demonstration of CD4, CD56, CD123, CD303, and TCL1 expression,

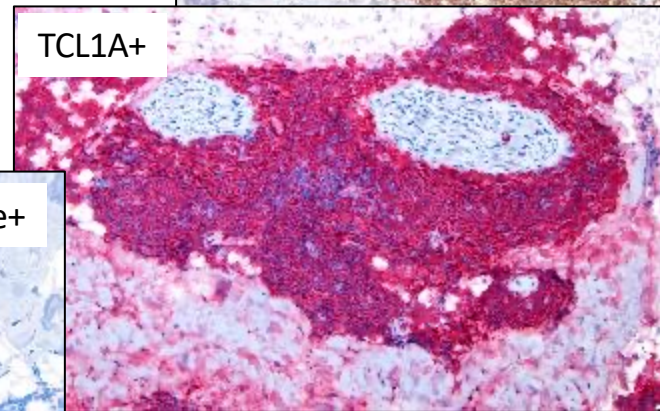
CD123+



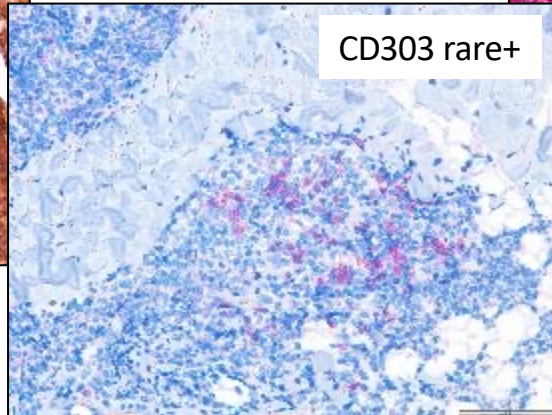
CD56+



TCL1A+



CD303 rare+



however the expression of CD4 and CD56, singly or in association, can be observed in several other hematological diseases

AML with monocytic differentiation

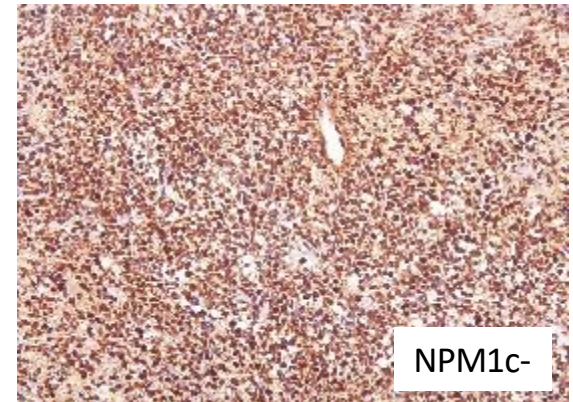
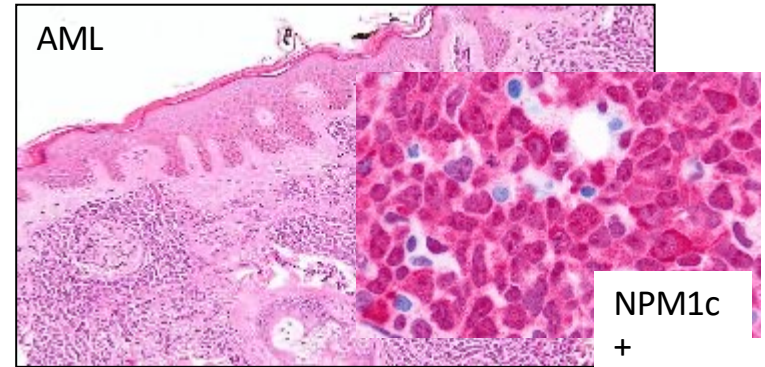
CD68
CD14
CD163

CD4
CD56
CD123

CD303
TCL1A
TCF4

BPDCN

Cytoplasmic expression
of NPM1

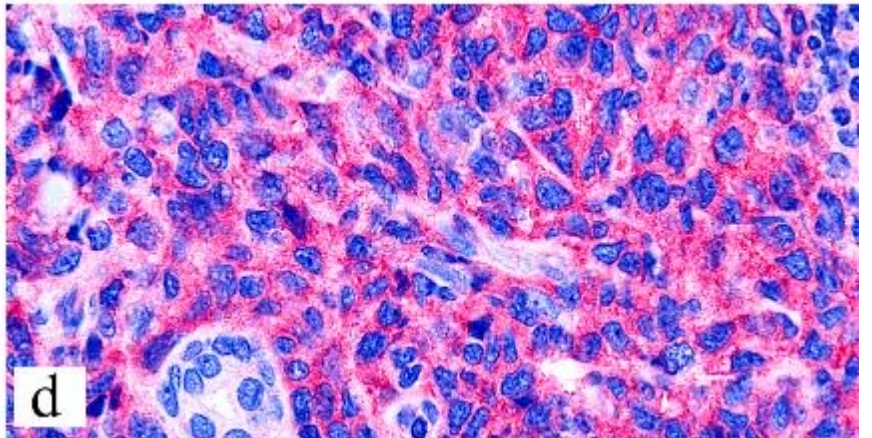
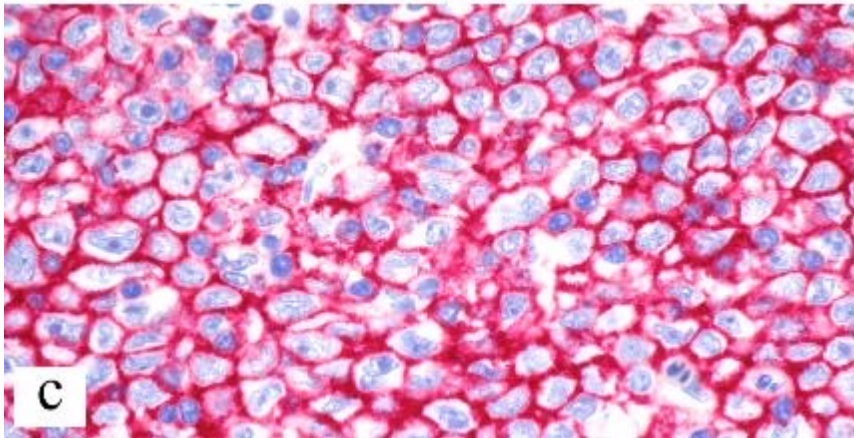
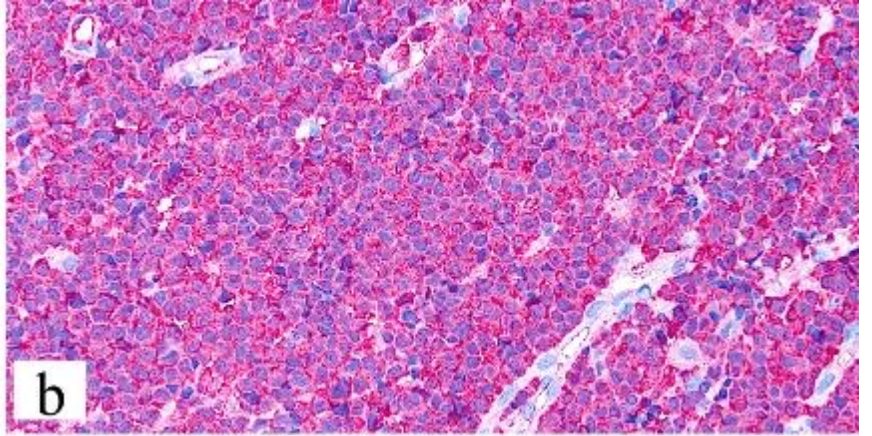
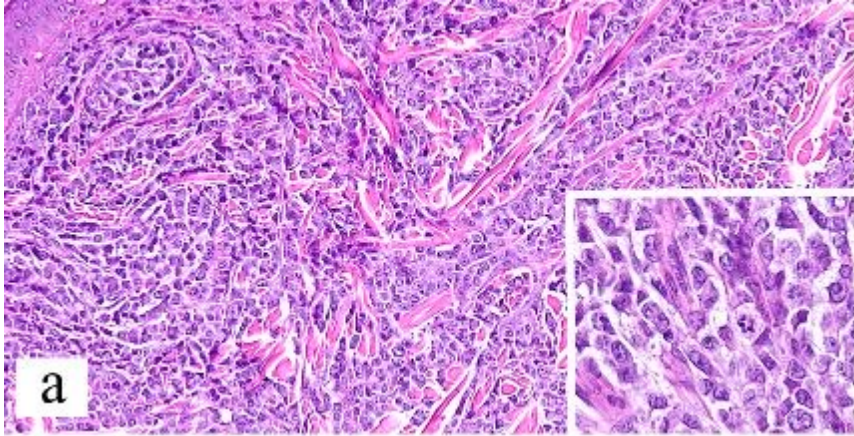


Lions for lambs

- Paziente di 86 anni
- Da sei mesi medicazioni sul territorio per un'ulcera
- Inviata per non risposta alle medicazioni
- Eseguita biopsia







- Diagnosi di BPDCN
- ...nel mente disseminazione leucemica
- Exitus
- Diagnosi post-mortem



Blastic Plasmacytoid Dendritic Cell Neoplasm with Pulmonary Involvement and Atypical Skin Lesion





Blastic plasmacytoid dendritic cell neoplasm with leukemic presentation, lacking cutaneous involvement: Case series and literature review

Michael J. Rauh^a, Fazlur Rahman^b, David Good^c, Jeffrey Silverman^d, Michael K. Brennan^e, Nikolay Dimov^f, Jane Liesveld^g, Daniel H. Ryan^h, W. Richard Burack^h, John M. Bennett^{g,h,*}

^a Department of Pathology and Molecular Medicine, Richardson Laboratory, Queen's University, Kingston, ON, Canada

^b Section of Oncology/Hematology, West Texas Medical Associates, San Angelo, TX, USA

^c Department of Clinical Pathology, Division of Hematopathology, Sunnybrook Health Sciences Centre, Toronto, ON, Canada

^d Department of Internal Medicine, North York General Hospital, Toronto, ON, Canada

^e Department of Pathology, North York General Hospital, Toronto, ON, Canada

^f BioReference Laboratories Inc., Elmwood Park, NJ, USA

^g Department of Medicine, James P. Wilmot Cancer Center, Hematology/Oncology, University of Rochester Medical Center, Rochester, NY, USA

^h Department of Pathology, Hematopathology Section, University of Rochester Medical Center, Rochester, NY, USA

M.J. Rauh et al. / *Leukemia Research* 36 (2012) 81–86

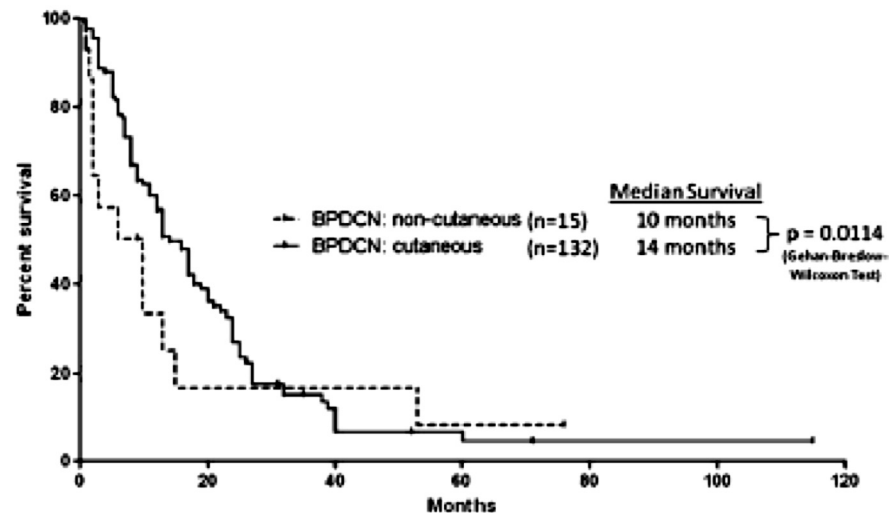
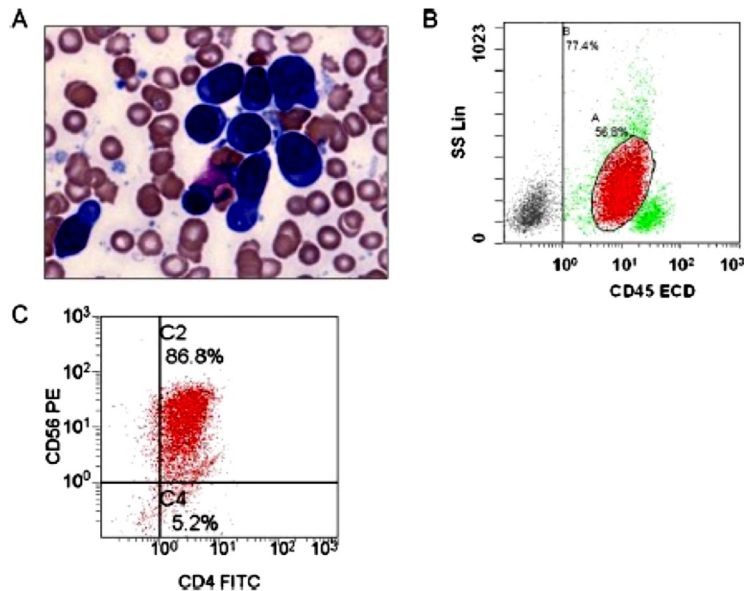
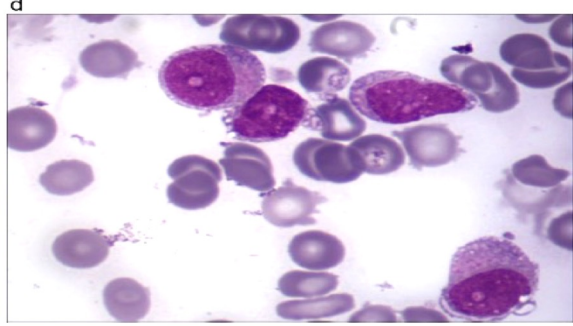
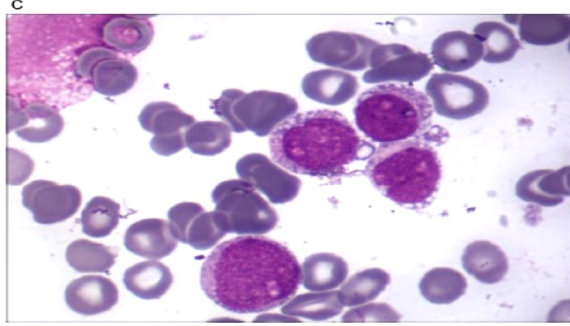
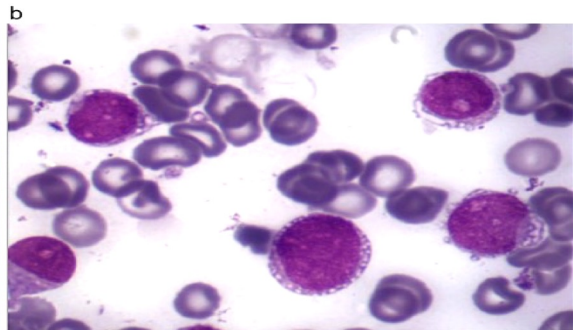
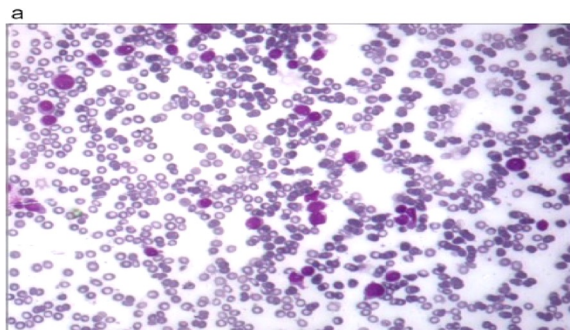
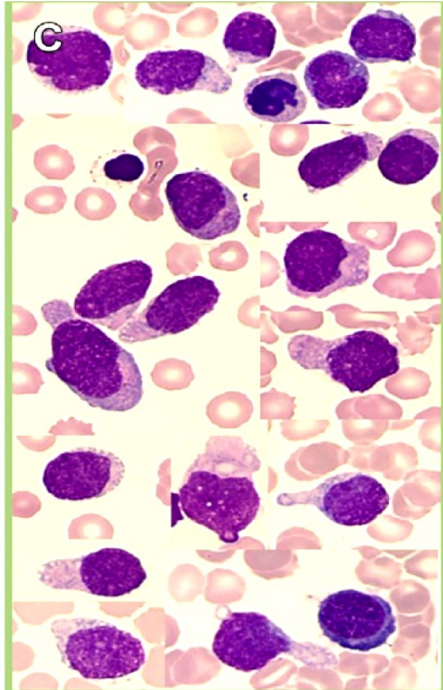
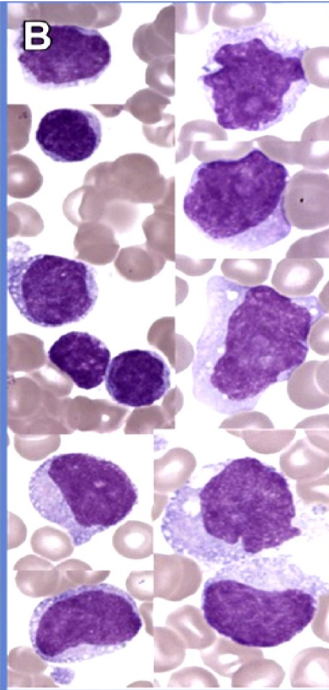
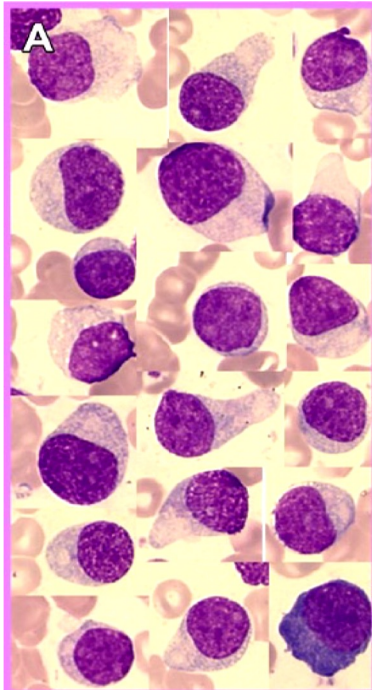
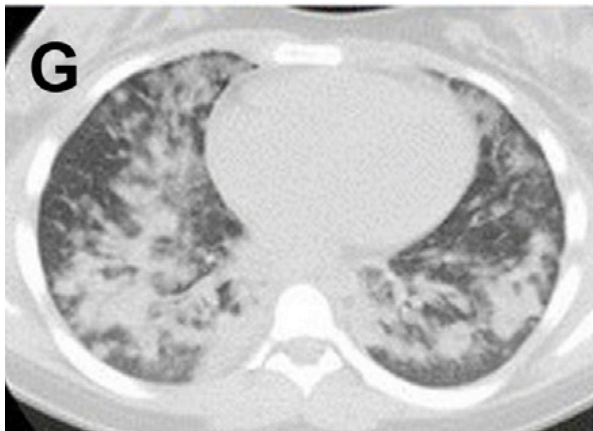
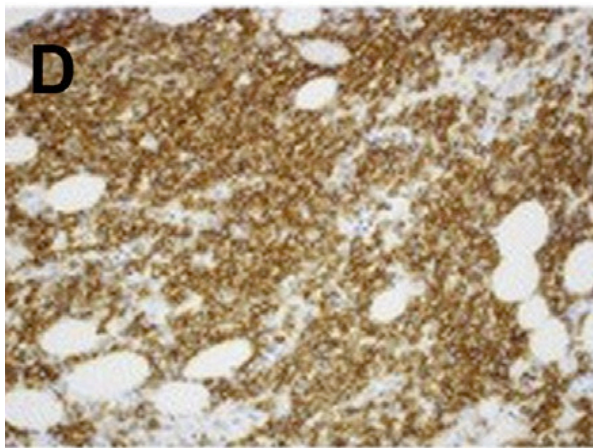


Fig. 2. Kaplan-Meier survival curves for BPDCCN cases with and without cutaneous presentation. Percent survival is plotted over time (months) for non-cutaneous (i.e. leukemic presentation) BPDCCN cases (n= 15; dashed line) and those with skin involvement (n= 132; solid line), using survival data (where available) for the cases presented in Tables 1 and 2.



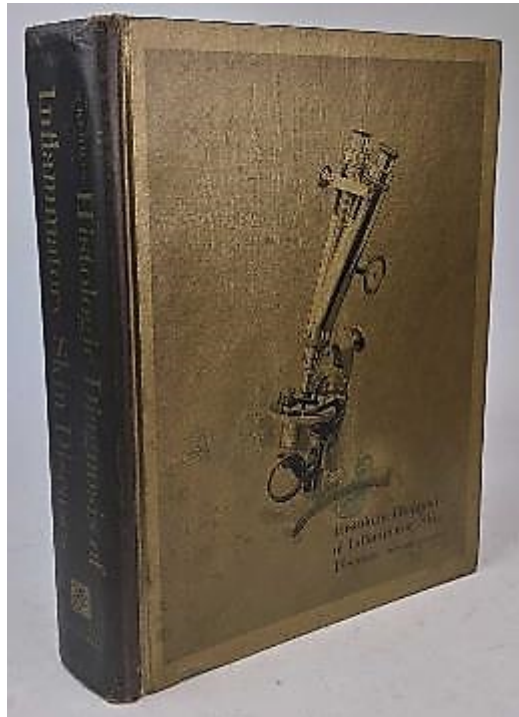
Cutaneous Manifestations of Blastic Plasmacytoid Dendritic Cell Neoplasm—Morphologic and Phenotypic Variability in a Series of 33 Patients

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Abstract: Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a neoplasm derived from precursors of plasmacytoid dendritic cells. Cutaneous involvement represents often the first manifesta-

Key Words: Blastic plasmacytoid dendritic cell neoplasm, CD4⁺/CD56, hematodermic neoplasm, phenotype

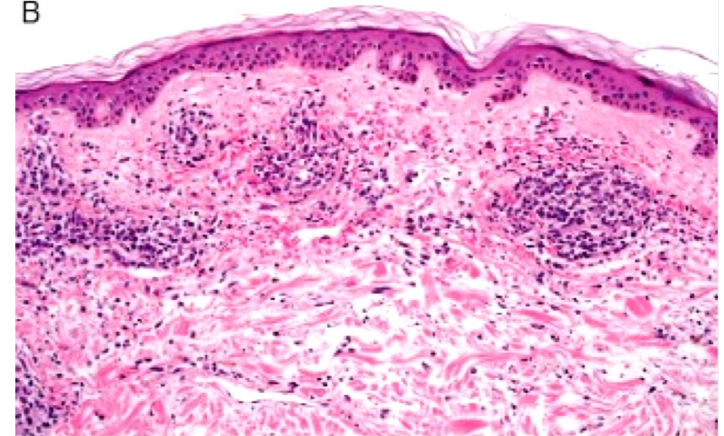
(*Am J Surg Pathol* 2010;34:75-87)



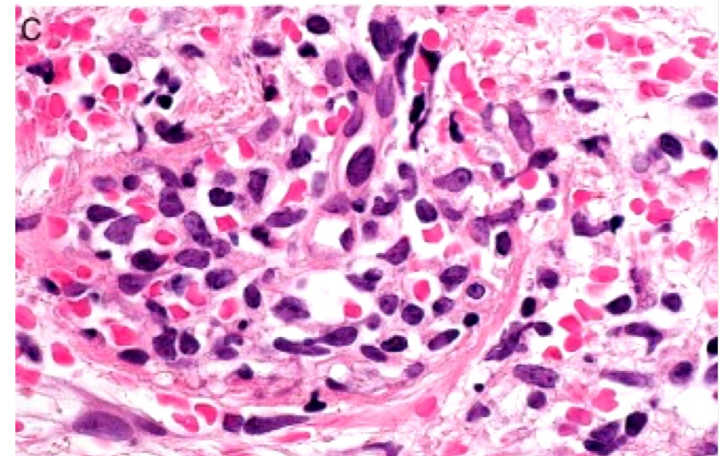
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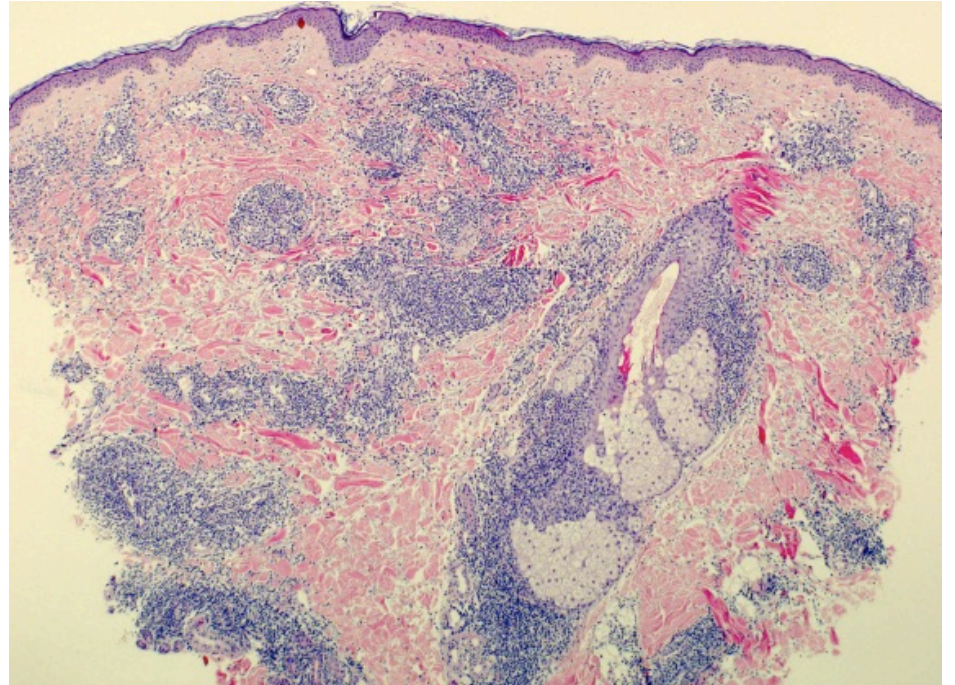


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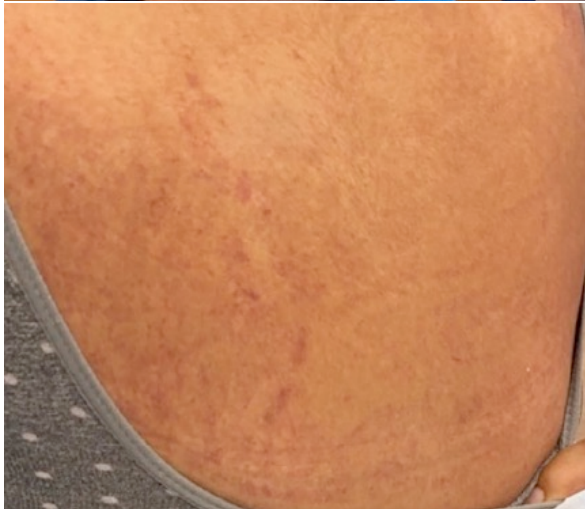
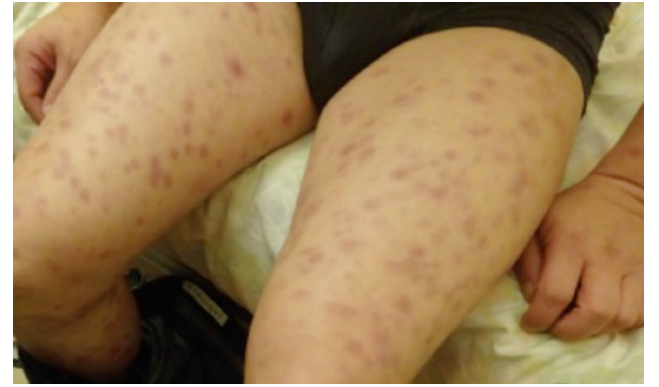
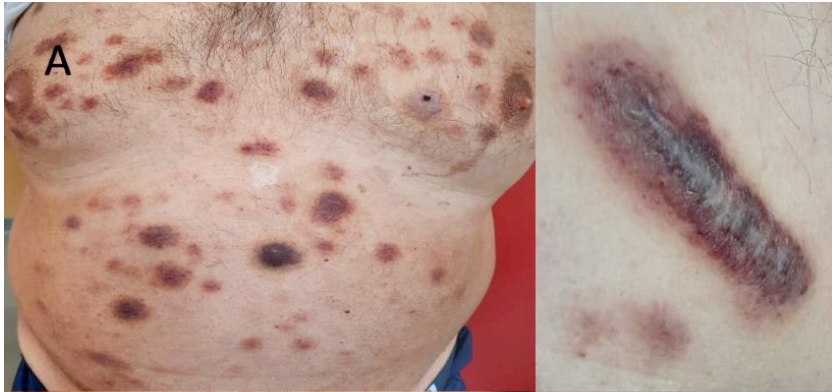
C





BPDCN: DD

- Petechiae
- Insect bites
- Kaposi sarcoma
- Myeloid sarcoma
- T-ALL/LBL
- Extra nodal NK-lymphoma



BPDCN: DD

- Petechiae
- Insect bites
- Easy to differentiate...no resolution within some days



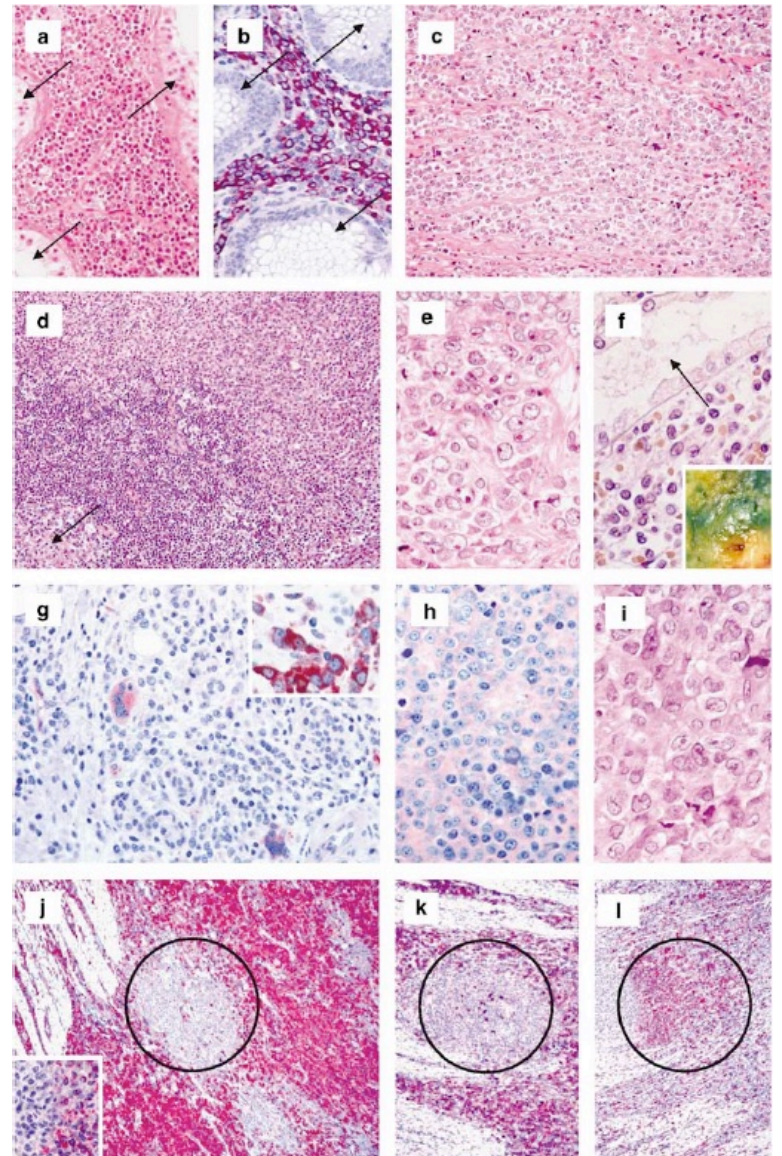
BPDCN: DD

- Kaposi sarcoma
- No purpuric cherry-like nodules
- No history of immunosuppression
- No classical mucosal involvement



BPDCN: DD

- Myeloid sarcoma
- RARE skin appearance
- AGGRESSIVE disease
- At histology MS can be CD43+ and CD68+
- Myelomonoblastic differentiation with expression of CD4, CD56, and CD123
- In MS morphologic evidence of mature or maturing granulocytes
- Expression of CD117, CD34, MPO (myeloperoxidase) and lysozyme



BPDCN: DD

- Cutaneous involvement by T-acute lymphoblastic leukemia/lymphoma (T-ALL/LBL)
- T-ALL/LBL and BPDCN are CD4+ and TdT+
- T-ALL/LBL is positive for T-cell–specific ag (CD3, CD5, CD2, and CD7)
- TCR positive in T-ALL/LBL

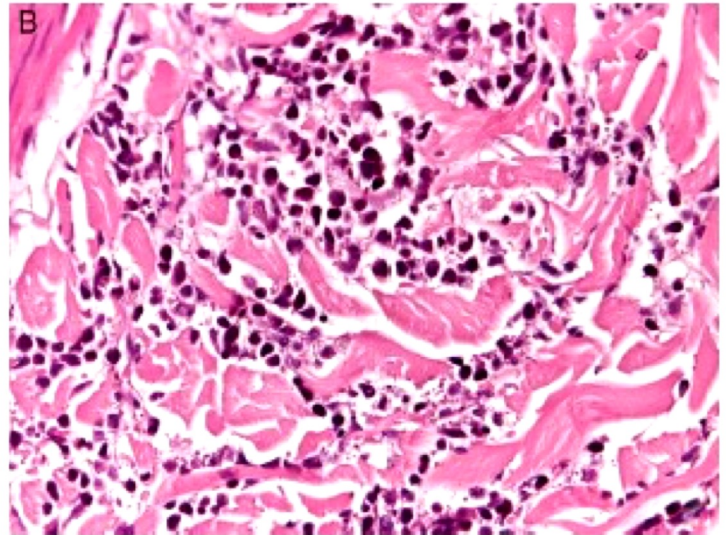
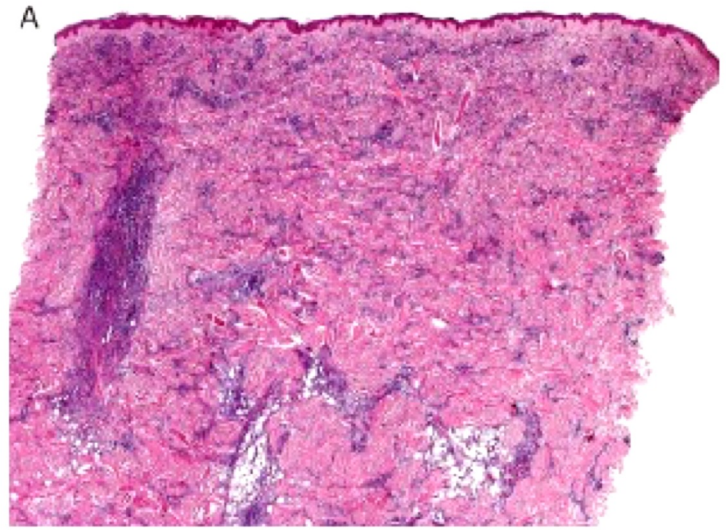
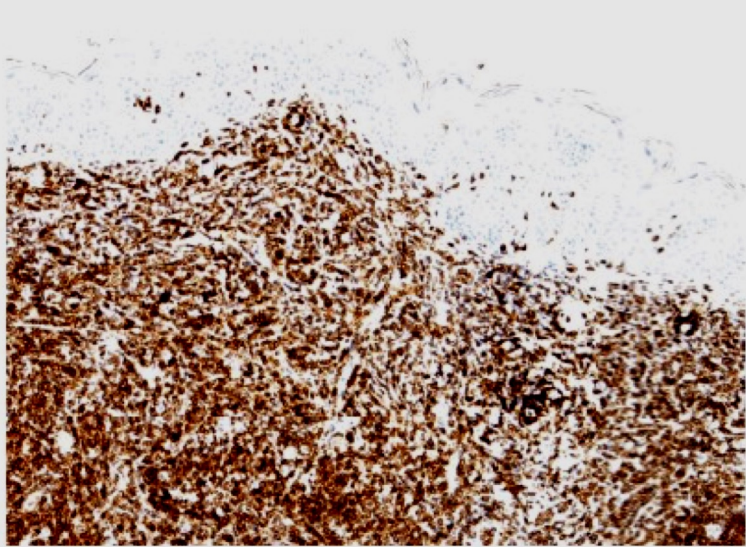


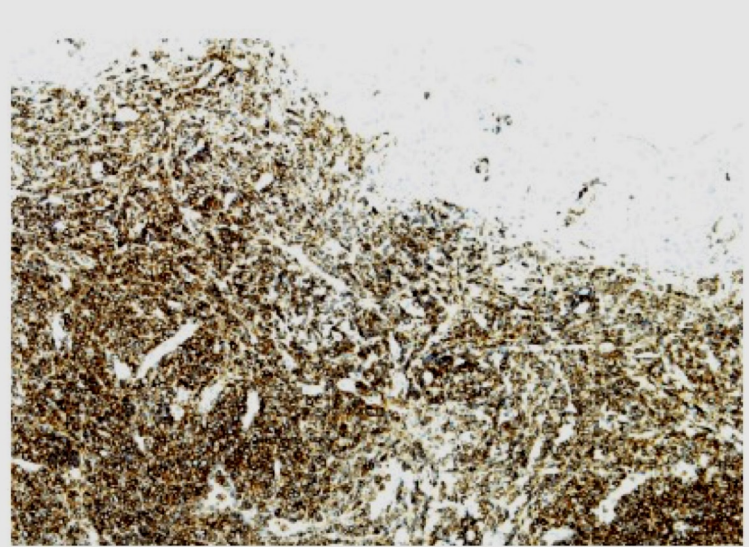
FIGURE 5. Histological patterns of blastic plasmacytoid dendritic cell neoplasm. A, Interstitial pattern resembling myeloid leukemia; (B) note indian filing.

BPDCN: DD

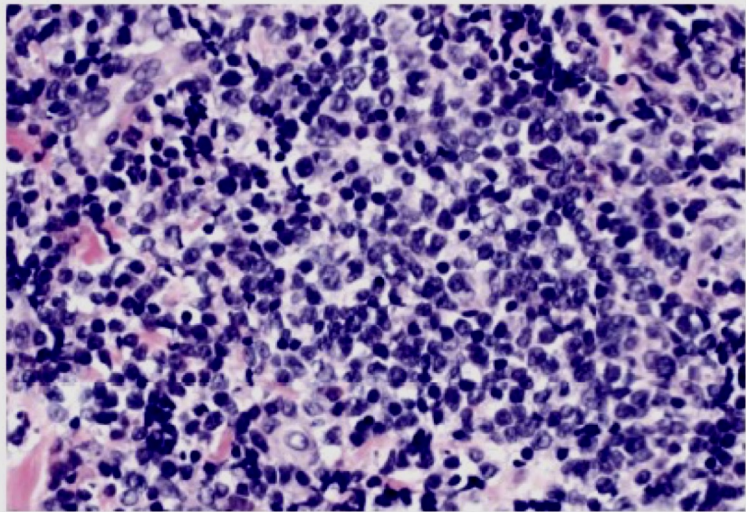
- Positivity for T-cell antigens and CD56.
- Angiocentric and angio-destructive growth
- Positive for CD3 and cytotoxic markers
- EBER+ and EBV at in situ hybridization



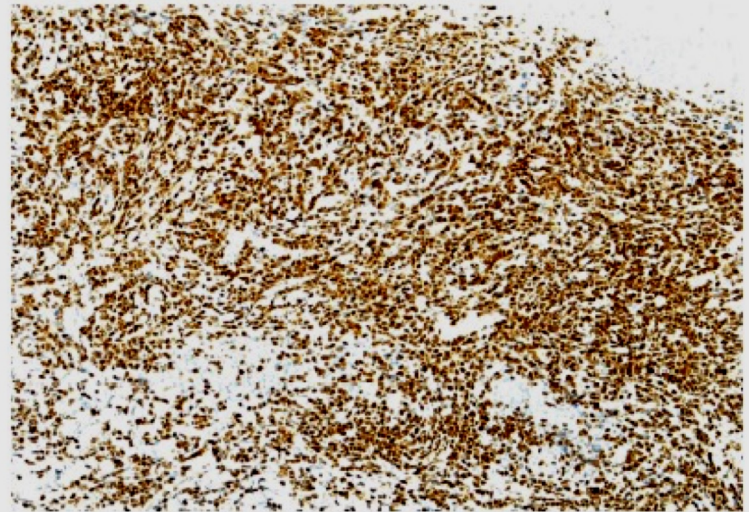
(a)



(b)



(c)



(d)

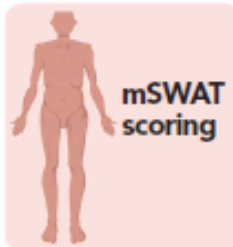
Outline of diagnostic evaluation for BPDCN

Cutaneous and systemic evaluation

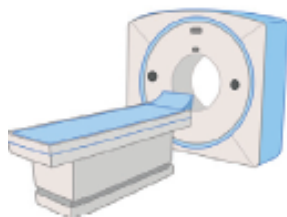


Violaceous skin lesions on inspection

- Skin biopsy and baseline photography



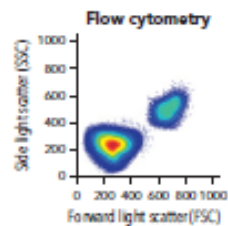
- PET-CT or CT scan for assessment of lymph nodes and/or other extra-medullary disease



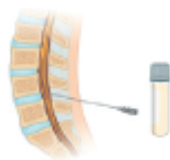
Cytomorphological evaluation



- Bone marrow aspirate and biopsy

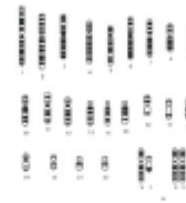


- Flow cytometry evaluation
1. CD123, CD4, D56
 2. CD303
 3. TCL1
 4. TCF4



- CSF evaluation for assessment of CNS disease

Adjunct tests



- Cytogenetics



- NGS for myeloid mutation assessment

Prognostic factors, prognostic indices and staging in mycosis fungoides and Sézary syndrome: where are we now?

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What does this study add?

- This is a review of clinical, haematological, pathological and genotypic changes affecting survival in MF/SS.
- The development of an international prognostic index to be adopted alongside staging may aid the management of patients.

Skin scoring

Skin tumour burden is not accounted for within TNMB staging. Several studies have found that multiple skin tumours have a worse disease-specific survival and overall survival than solitary lesions.^{10,12,18} A tumour burden index (TBI) was developed as a prognostic tool.²⁶ TBI was defined as equal to $1 + (\text{patches} \times 2) + (\text{plaques} \times 2) + (\text{tumour} \times 1.3)$, where patches $> 30\%$ of BSA were equal to 1 and presence of plaques or tumours was equal to 1. In a validation involving 116 patients both TNM and TBI provided prognostic information but discrimination of survival curves was better for TBI. The TBI has not been further validated in CTCL.

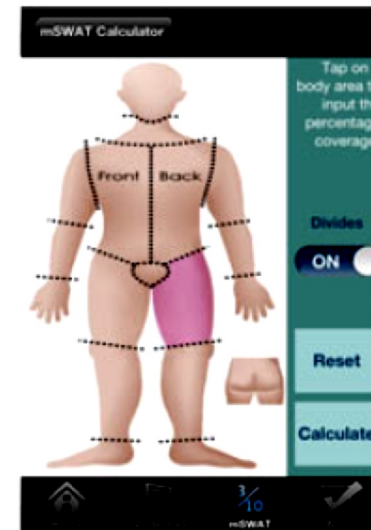
How big is your hand and should you use it to score skin in cutaneous T-cell lymphoma?

J.J. Scarisbrick¹ and S. Morris²

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So, when considering 'How big is your hand and should you use it to score skin?', we suggest that a palmar surface of 0.5% BSA should be used as the measurement tool to score skin, as this is relatively constant with age, stature and race. Our training day highlighted that even with a constant training method there is significant interuser variability. Where possible the same scorer should score any individual.



The scored area is highlighted in pink and further areas may be scored similarly

Non-erythrodermic:				
Total score : 13/0				
Involvement in Patient's skin				
Body region	BSA	Patch	Plaque	Tumour
Head	7	0/0	0/0	0/0
Neck	2	0/0	0/0	0/0
Anterior trunk	13	0/0	0/0	0/0
Arms	8	0/0	0/0	0/0
Forearms	6	0/0	0/0	0/0
Hands	5	0/0	0/0	0/0

When scoring completed press calculate, which will weight scores and produce an mSWAT score/400

	Clinics	Stage	T
EARLY-STAGE		IA	T1
		IB	T2
		IIA	T1-T2
/ANCED-STAGE		IIB	T3
		IIIA	T4
		IIIB	T4

- X 1

- X 2

- X 4

- Score 0 → 400

BPDCN

- Aggressive disease
- Median survival 14 months
- Prompt and accurate diagnosis required
- Start staging procedures
- Proper treatment of patients

BPDCN

- Role of dermatologist is important
- Guide pathologist
- Send patient to hematologist
- AVOID to waste time with unproper treatment (i.e. steroids → transient vanishing of lesions)



Thank you!!!

