



MASTOCITOSI E BPDCN: LE DIAGNOSI DIFFICILI

BOLOGNA, ROYAL HOTEL CARLTON
12 OTTOBRE 2026

BPDCN la diagnosi differenziale

Alessandro Pileri



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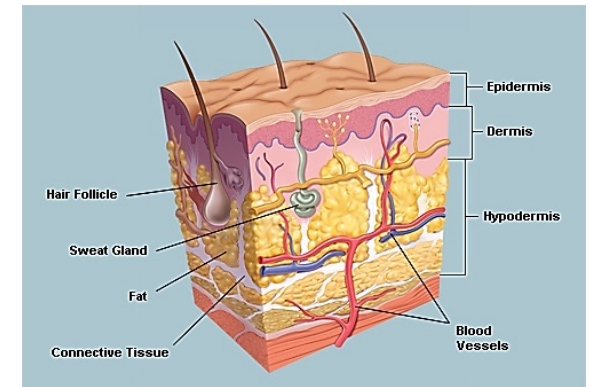
Disclosure: Alessandro Pileri

Company name	Research support	Employee	Consultant	Stockholder	Speakers bureau	Advisory board	Other
Kyowa Kirin			x				
Menarini Tsemline			x				
Recodsti Rare disease			x				

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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
- Awareness needed
- Lack of a defined pattern of clinical presentation



BPDCN: clinical features

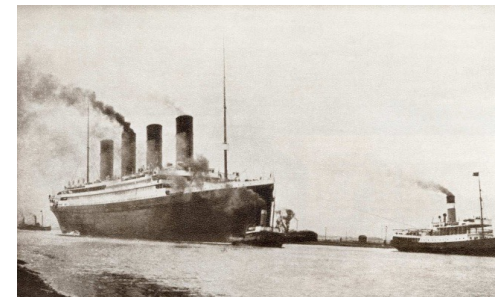
- Clinical presentation polymorphic
- Possible misdiagnosis
- Diagnostic delay
- NO anatomic preferred site
- Don't hit the rocks!!

GENERAL DERMATOLOGY

BJD
British Journal of Dermatology

The PROCLIFI international registry of early-stage mycosis fungoides identifies substantial diagnostic delay in most patients

J.J. Scarisbrick^{1,2,3,4}, P. Quaglino,^{2,3} H.M. Prince,³ E. Papadavid,^{2,3} E. Hodak,^{2,3} M. Bagot,^{2,3} O. Servitje,^{2,3} E. Berti,^{2,3} P. Ortiz-Romero,^{2,3} R. Stadler^{1,2,3}, A. Patsatsi,^{2,3} R. Knobler,^{2,3} E. Guenova,^{2,3} F. Child,^{2,4} S. Whittaker,^{2,3,4} V. Nikolaou^{1,2,3}, C. Tomasini,² I. Amitay,^{2,3} H. Prag Naveh,^{2,3} C. Ram-Wolff,² M. Battistella,^{2,3} S. Alberti-Violetti,² R. Stranzenbach,^{2,3} V. Gargallo,² C. Muniesa,² T. Koletska,² C. Jonak,^{2,3} S. Porkert,² C. Mitteldorf,² T. Estrach,² A. Combalia,² M. Marschalko,² J. Csomor,² A. Szepesi,² A. Cozzio,^{2,3} R. Dummer,² N. Pimpinelli,² V. Grandi,² M. Beylot-Barry,² A. Pham-Ledard,² M. Wobser,² E. Geissinger,² U. Wehkamp,^{2,3} M. Weichenethal,² R. Cowan,^{2,4} E. Parry,^{2,4} J. Harris,⁴ R. Wachsmuth,^{2,4} D. Turner,⁴ A. Bates,⁴ E. Healy,⁴ F. Trautinger,^{2,3} J. Latzka,² J. Yoo,^{1,2} B. Vydianath,¹ R. Amel-Kashipaz,¹ L. Marinos,² A. Oikonomidi,² A. Stratigos,² M.-D. Vignon-Pennamen,² M. Battistella,² F. Climent,² E. Gonzalez-Barca,² E. Georgiou,² R. Senetta,² P. Zinzani,² L. Vakeva,² A. Ranki,² A.-M. Busschots,² E. Hauben,² A. Bervoets,² F.J.S.H. Woei-A-Jin,² R. Matin,⁴ G. Collins,⁴ S. Weatherhead,⁴ J. Frew,⁴ M. Bayne,⁴ G. Dunnill,⁴ P. McKay,⁴ A. Arumainathan,⁴ R. Azurdia,⁴ K. Benstead,⁴ R. Twigger,³ K. Rieger,³ R. Brown,³ J.A. Sanches,³ D. Miyashiro,³ O. Akilov,³ S. McCann,³ H. Sahi,³ F.M. Damasco,³ C. Querfeld,³ A. Folkes,³ C. Bur,³ C.-D. Klemke,² P. Enz,³ R. Pujol,^{2,3} K. Quint,² L. Geskin,³ E. Hong,³ F. Evison,¹ M. Vermeer,^{2,3} L. Cerroni,² W. Kempf,² Y. Kim³ and R. Willemze²



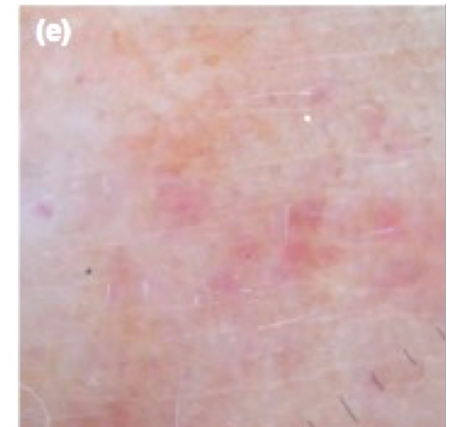
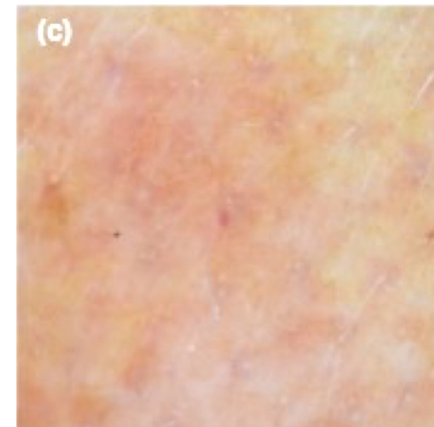
BPDCN



- Dermoscopy
- Two patients reported by Nicklas et al (CED 2021)
- Reddish and purplish non-blanchable structureless areas
- Surrounded by white halo alternated with homogeneous bluish-white areas
- No vessels seen

C. Nicklas,¹ E. Bellolio,² E. Lopez,¹ I. Goldberg,³ S. Geller³
and C. Navarrete-Dechent⁴ 

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BPDCN

- Densely clus

Cureus
Part of SPRINGER NATURE

Review began 09/08/2024
Review ended 09/18/2024
Published 09/26/2024

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BPDCN: clinical features

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

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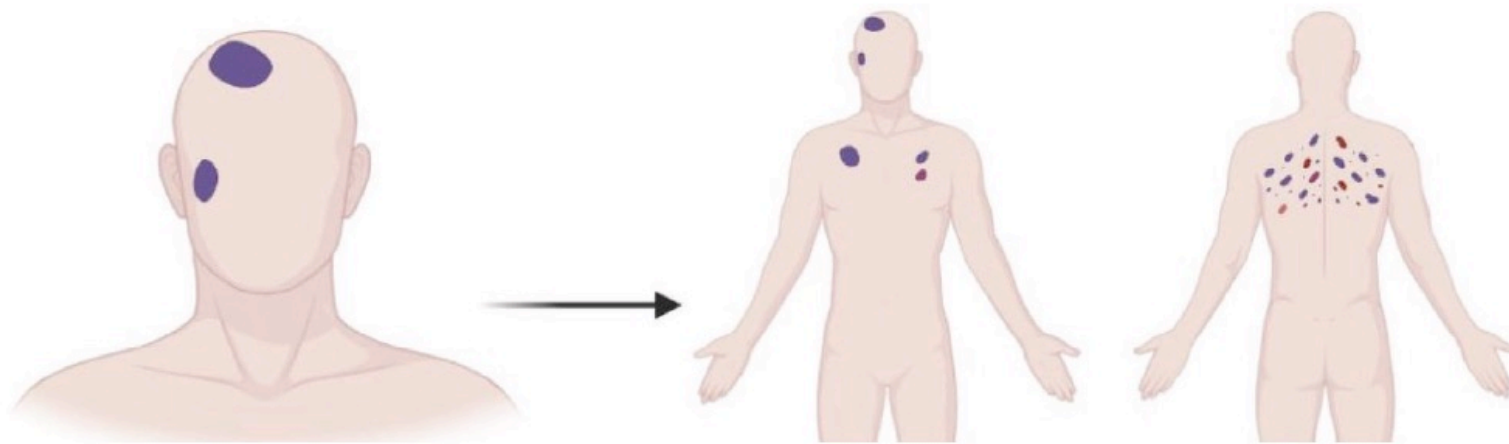
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MASTOCITOSI E BPDCN: LE DIAGNOSI DIFFICILI

A



B



BPDCN: clinical features

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

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

BPDCN: clinical features

- Isolated lesions
- Better prognosis?
- Multiple lesions
- Worse clinical outcome?

eruptive lesions) achieved a clinical response (2 CR and 3 PR). One patient initially experienced a SD, yet died of disease after a new progression (leukemization) 5 months later. All responding patients in both groups experienced relapse/progression, with a significantly different progression-free-survival (PFS): median 23 months (single-lesion-presentation) *versus* 9 months (multiple, eruptive lesion-presentation). Eight of 11 patients died, with a median overall survival of 18 months. One patient with a single-lesion-presentation is currently a long-lasting survivor (37 months). One with a multiple, eruptive lesion-presentation, after having achieved CR with 6-CHOP courses, has been treated with allogeneic stem cell transplantation from related donor (sister), and is now in stable CR since 10 months.

Blastic plasmacytoid dendritic cell neoplasm (BPDCN): the cutaneous sanctuary

A. PILERI¹, C. DELFINO², V. GRANDI², C. AGOSTINELLI³, S. A. PILERI³, N. PIMPINELLI²

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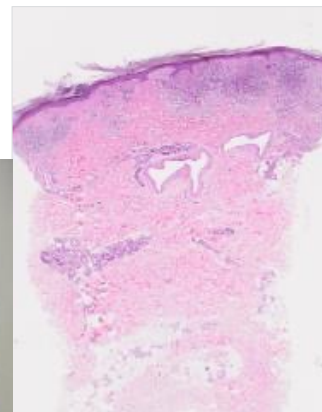
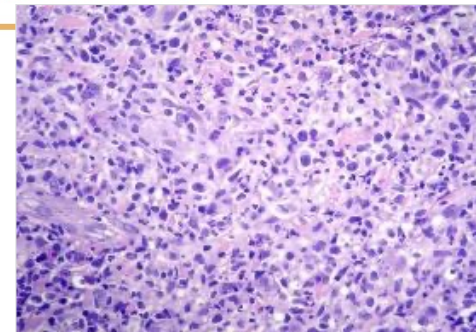


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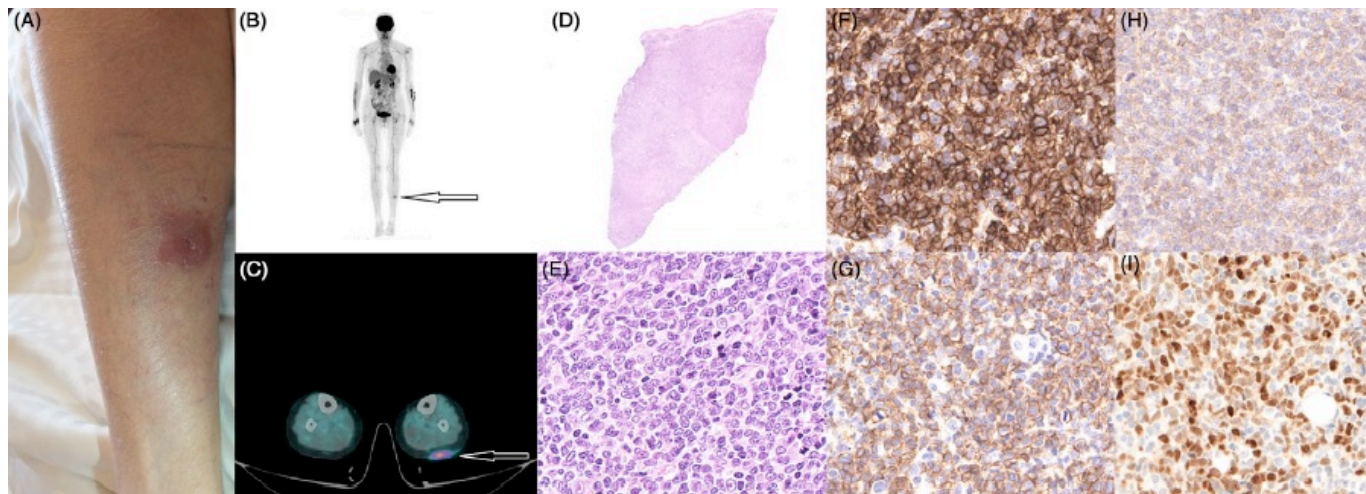


BOLOGNA, 12 OTTOBRE 2020

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DOI: 10.1002/jha2.370

HAEMATOLOGY IMAGES

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

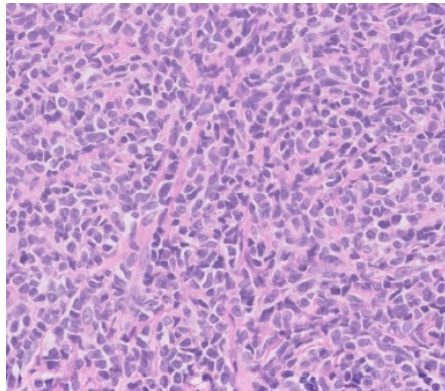
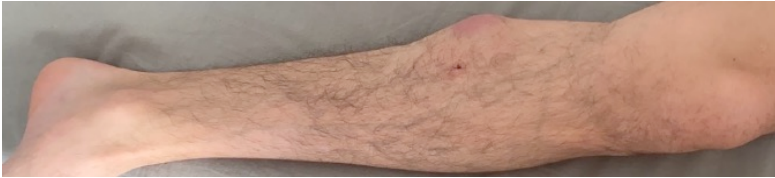
Isolated skin infiltration by a blastic plasmacytoid dendritic cell neoplasm

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



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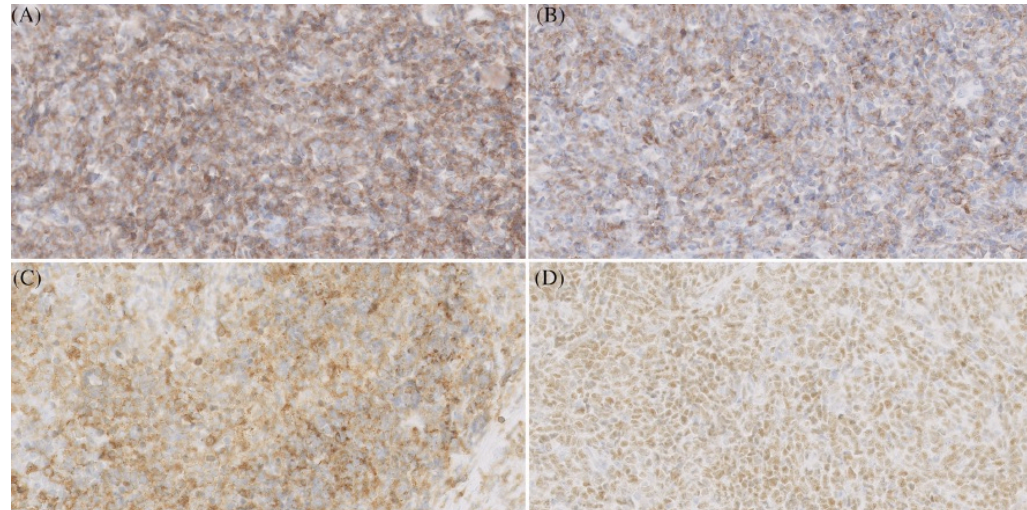
DOI: 10.1002/jha2.383

HAEMATOLOGY IMAGES

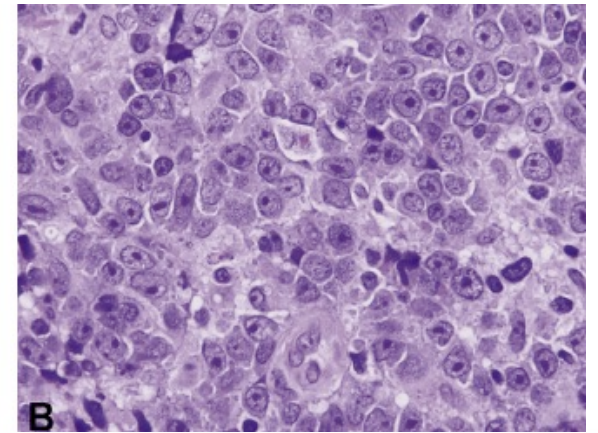
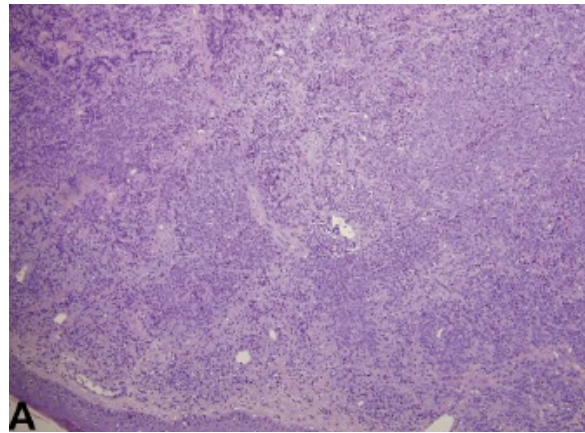
eJHaem

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



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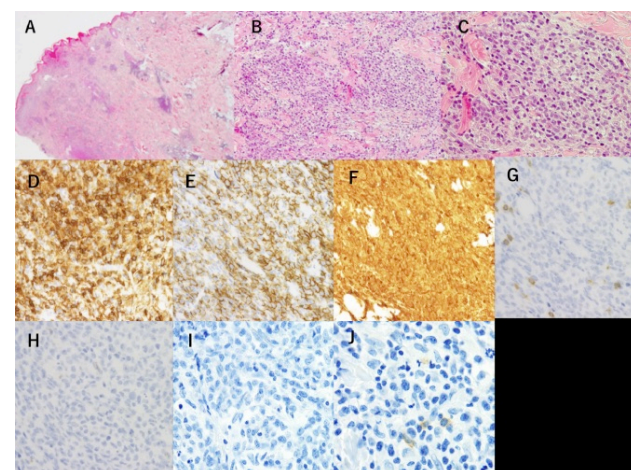
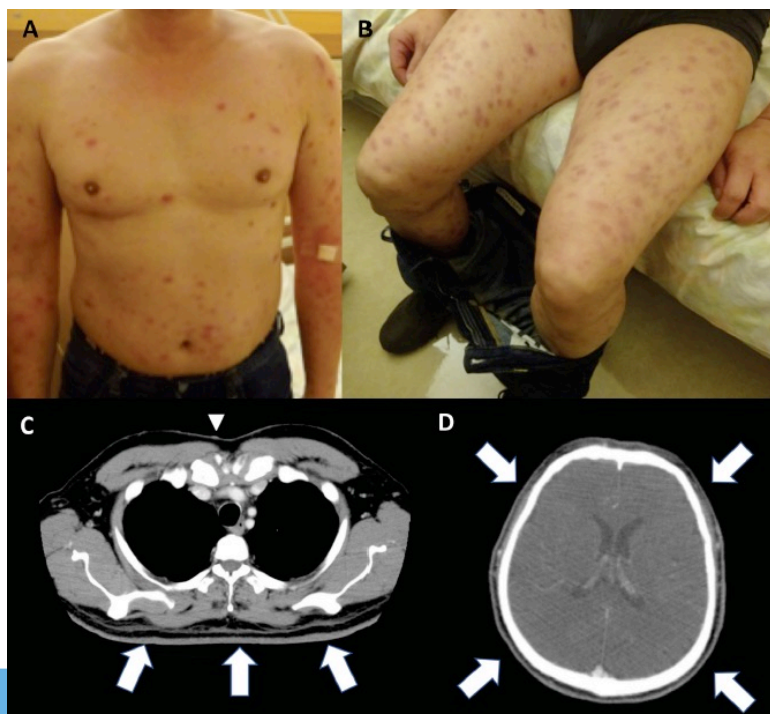
journal homepage: www.elsevier.com/locate/radcr

DIAGNOSI DIFFICILE

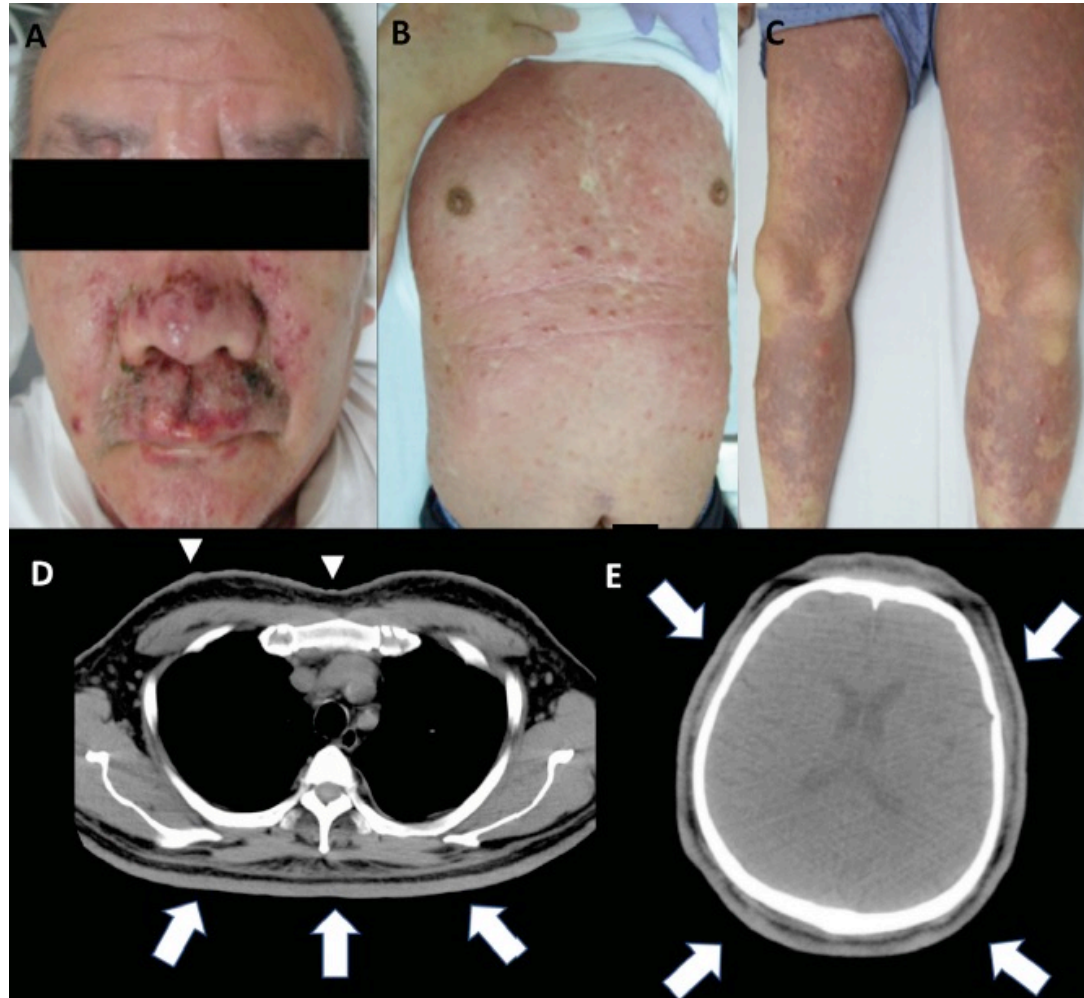
Case Report

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



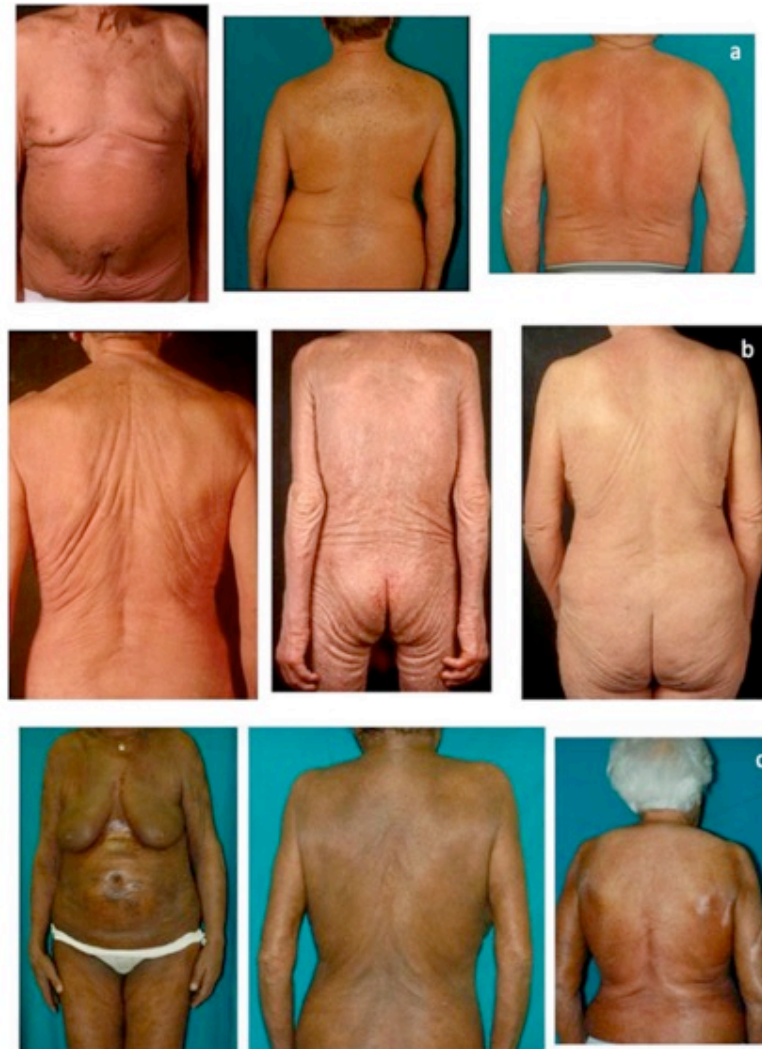
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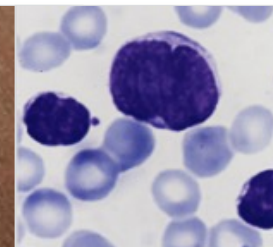
Article

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

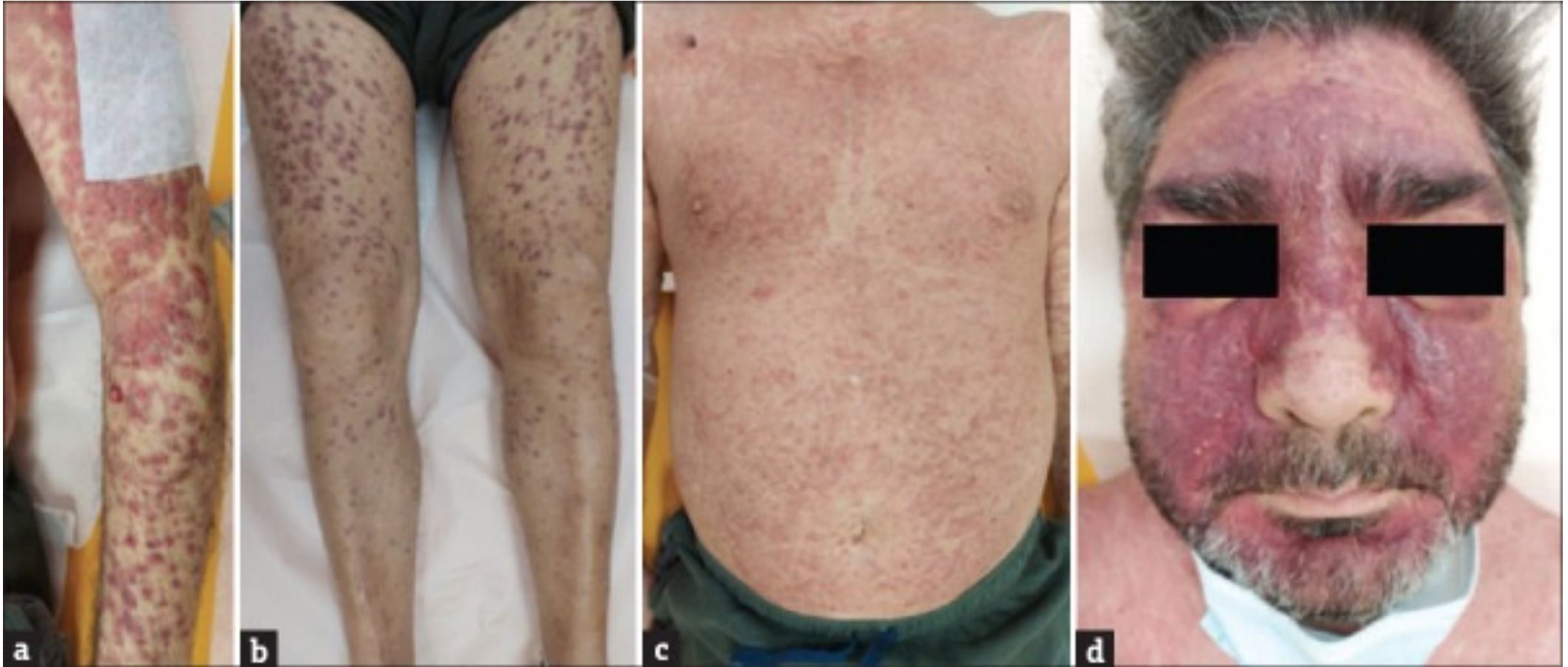
Gabriele Rocuzzo ^{1,*}, Silvia Giordano ^{1,†}, Gianluca Avallone ¹, Marco Rubatto ¹, Silvia Canonico ¹, Ada Funaro ², Erika Ortolan ², Rebecca Senetta ³, Paolo Fava ¹, Maria Teresa Fierro ¹, Simone Ribero ¹ and Pietro Quaglino ¹



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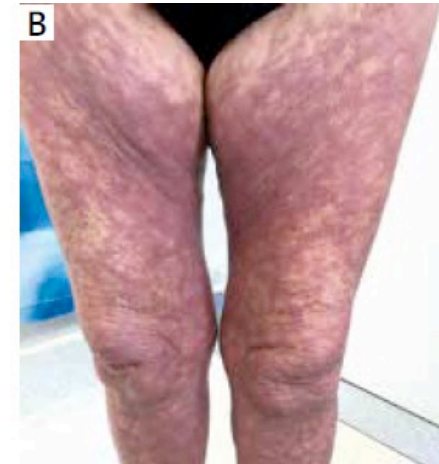
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Clinicopathologic retrospective analysis of blastic plasmacytoid dendritic cell neoplasms

Agnieszka Owczarczyk-Saczonek¹, Małgorzata Sokołowska-Wojdyło², Berenika Olszewska², Marta Malek³, Aleksandra Znajewska-Pander¹, Anna Kowalczyk⁴, Wojciech Biernat⁵, Grażyna Poniatowska-Broniek⁶, Wanda Knopińska-Posłuszny⁷, Zygmunt Koziol⁶, Roman Nowicki², Waldemar Placek¹

Adv Dermatol Allergol 2018; XXXV (2): 128–138
DOI: <https://doi.org/10.5114/ada.2017.72269>

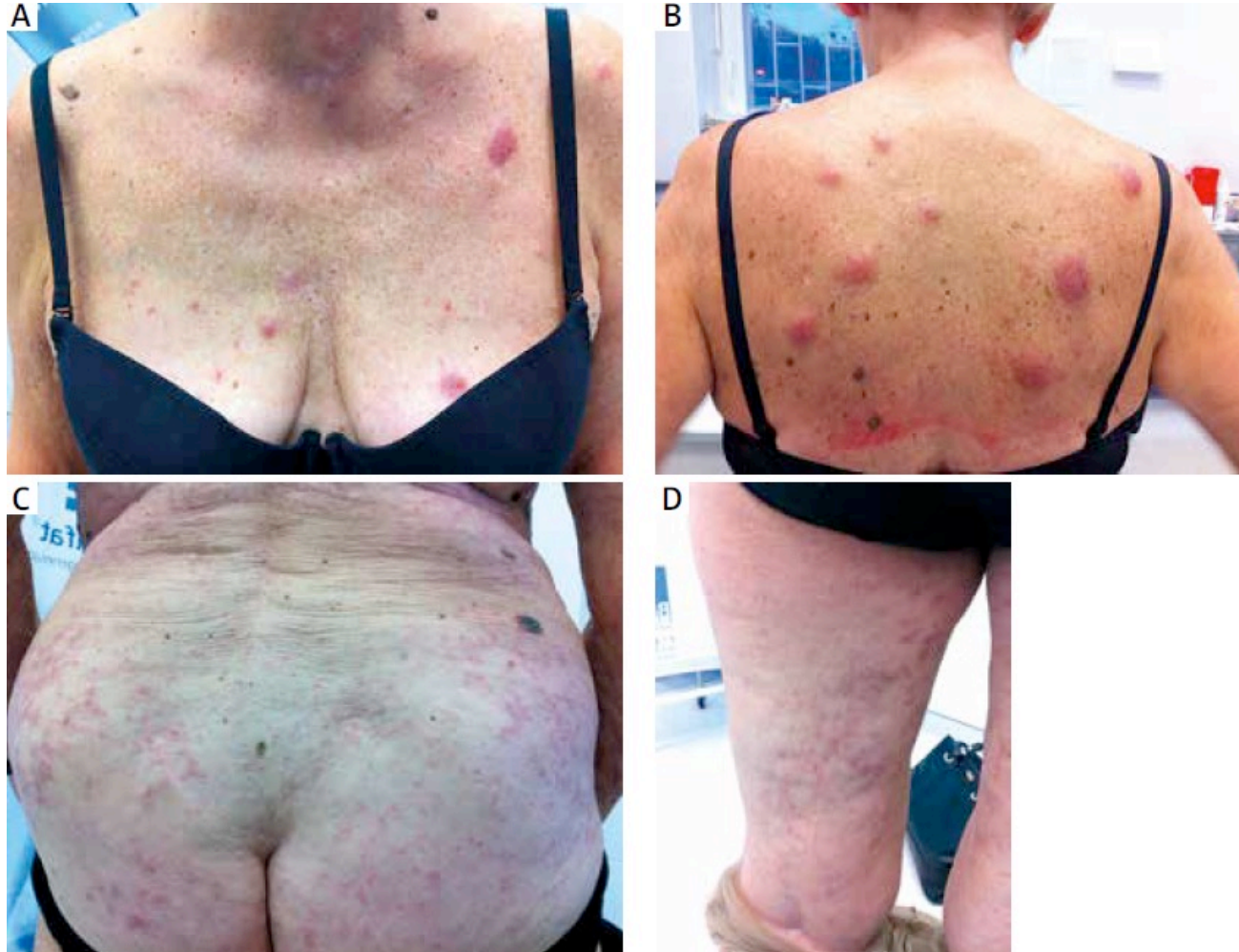
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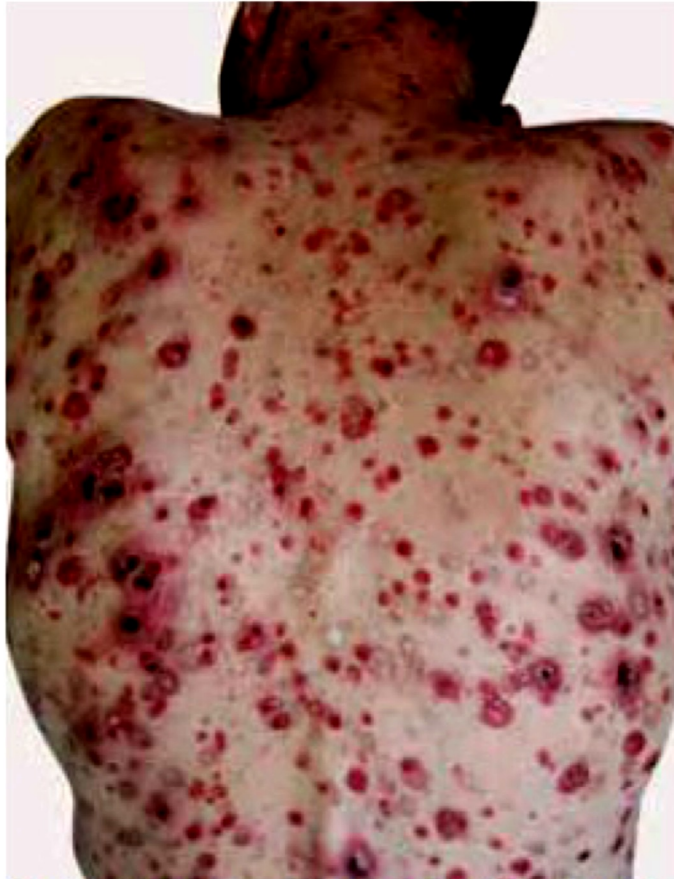


Fig. 6093 Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma. Lesions are often haemorrhagic; they are diffuse and associated with epidermal ulceration.

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

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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, X2.5) 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.

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488 Berti et al
AJP August 1999, Vol. 155, No. 2

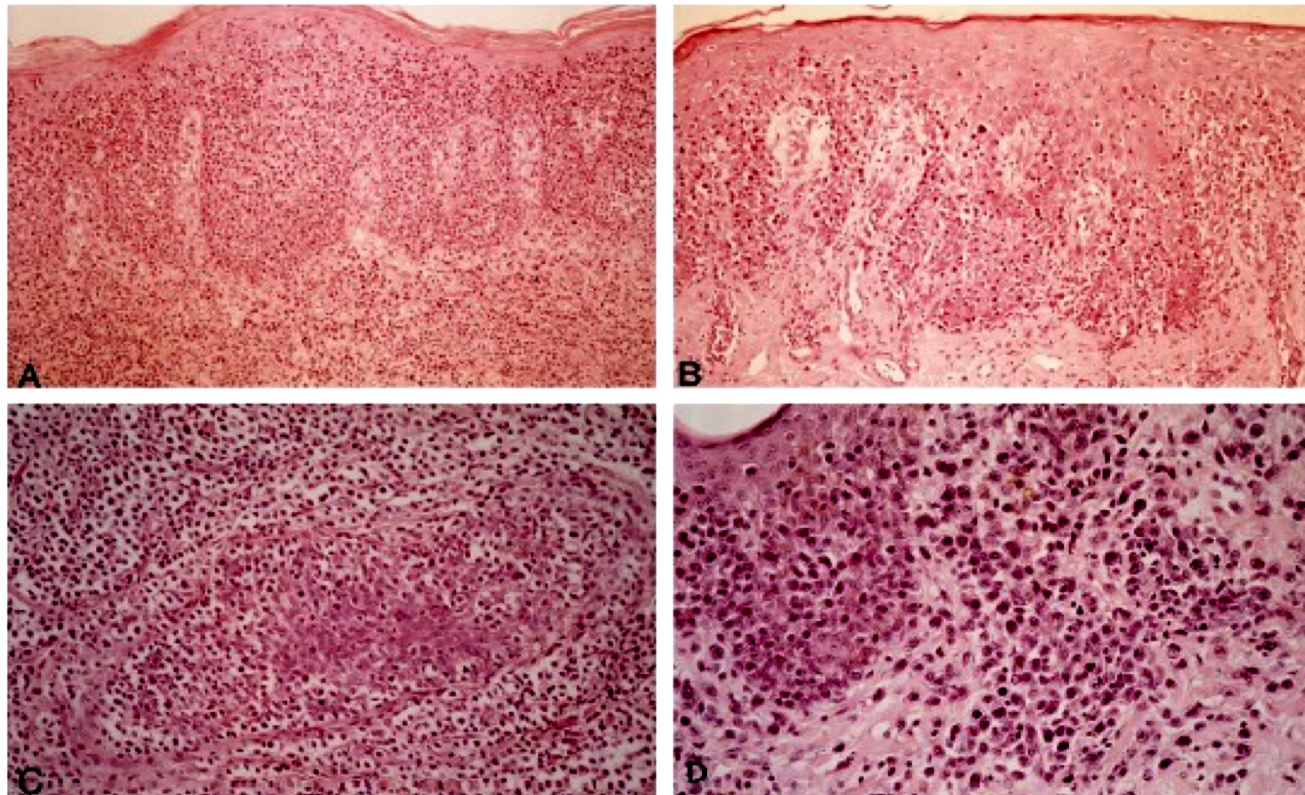


Figure 2. A: Histology (H&E; original magnification, $\times 100$) of case no. 4: well developed nodular lesion. Note the diffuse pleomorphic strongly epidermotropic T cell infiltrate. B: Histology (H&E; original magnification, $\times 100$) of case no. 1: early lesion. Note the intraepithelial pleomorphic 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.

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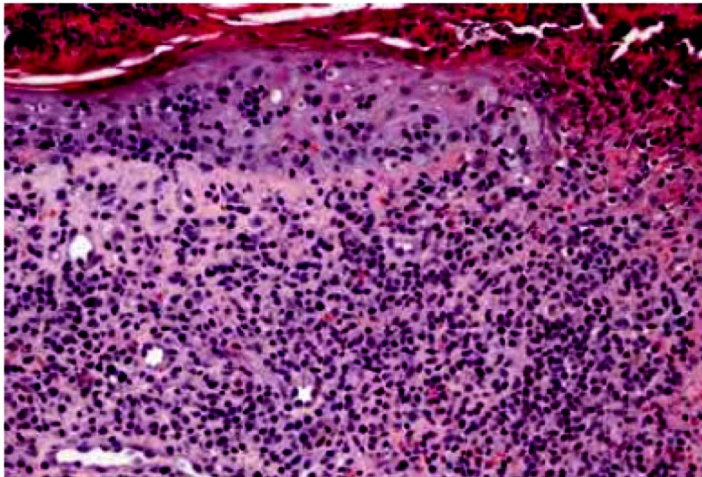


Fig. 6102 Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma. The atypical pleomorphic medium to large cell lymphoid infiltrate occupies the superficial dermis, extending to the epidermis in a pagetoid fashion and leading to epidermal necrosis.

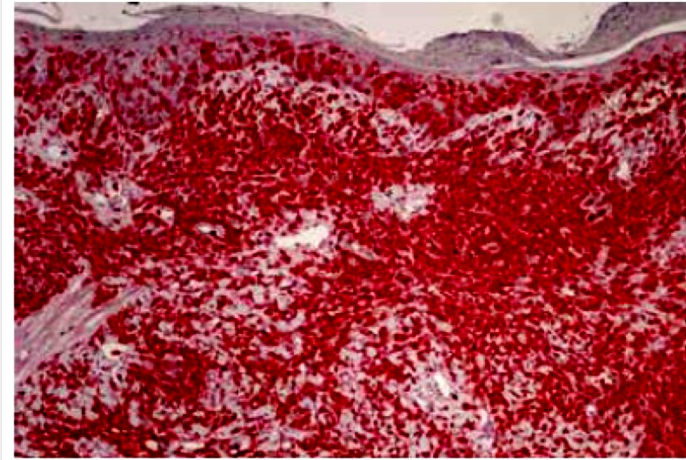


Fig. 6103 Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma. CD8 staining highlights the epidermotropic neoplastic cells in a skin section.

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Juvenile Blastic Plasmacytoid Dendritic Cell Neoplasm

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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



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



MASTOCITOSI E BPDCN: LE DIAGNOSI DIFFICILI

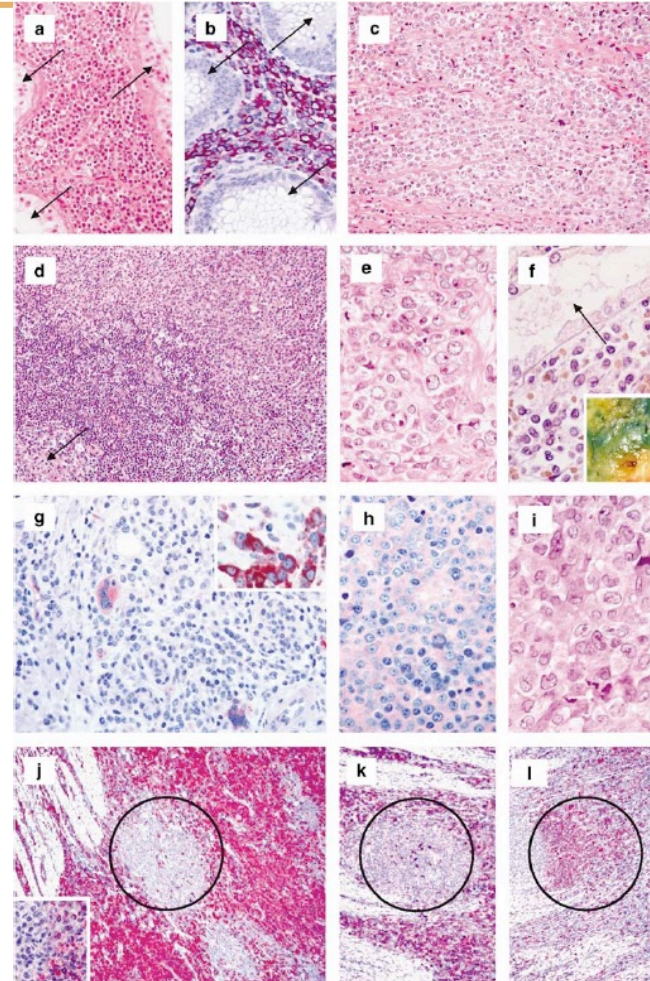
- Kaposi sarcoma
- No purpuric cherry-like nodules
- No history of immunosuppression
- No classical mucosal involvement



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- 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

MASTOCITOSI E BPDCN: LE DIAGNOSI DIFFICILI



MASTOCITOSI E BPDCN: LE DIAGNOSI DIFFICILI

- 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

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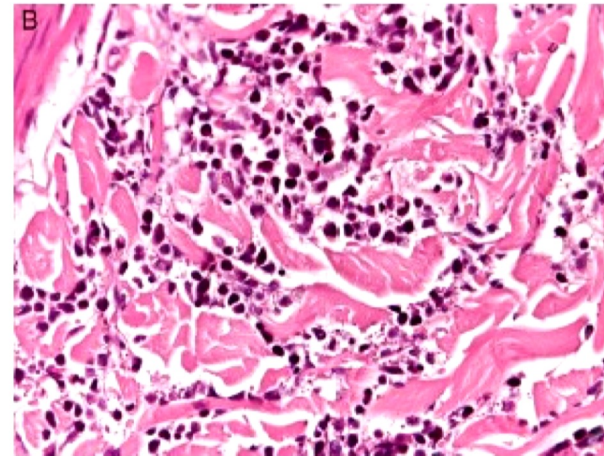
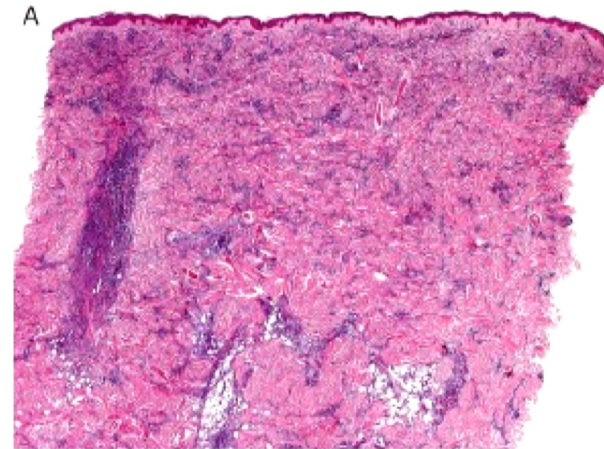
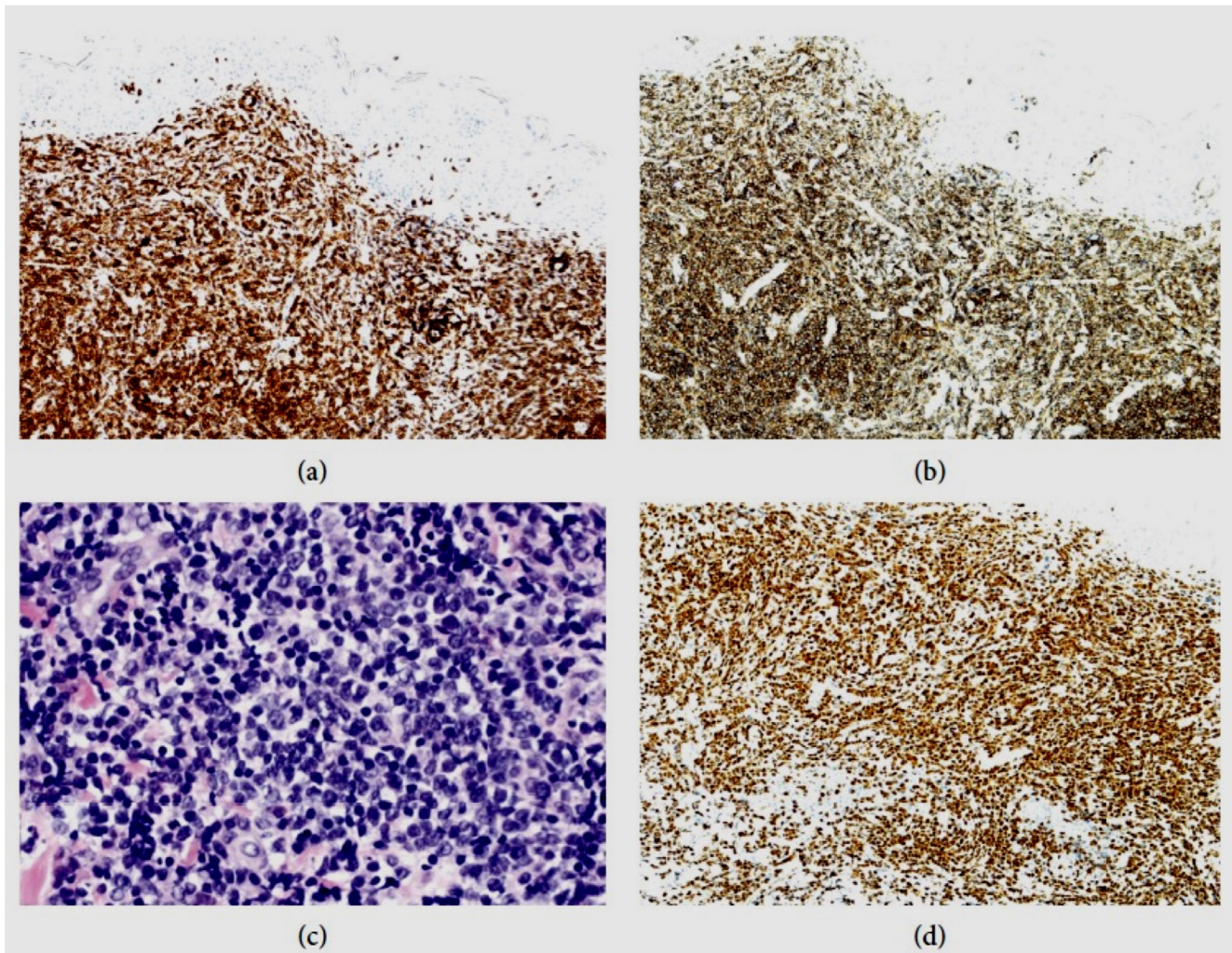


FIGURE 5. Histological patterns of blastic plasmacytoid dendritic cell neoplasm. A, Interstitial pattern resembling myeloid leukemia; (B) note indian filing.

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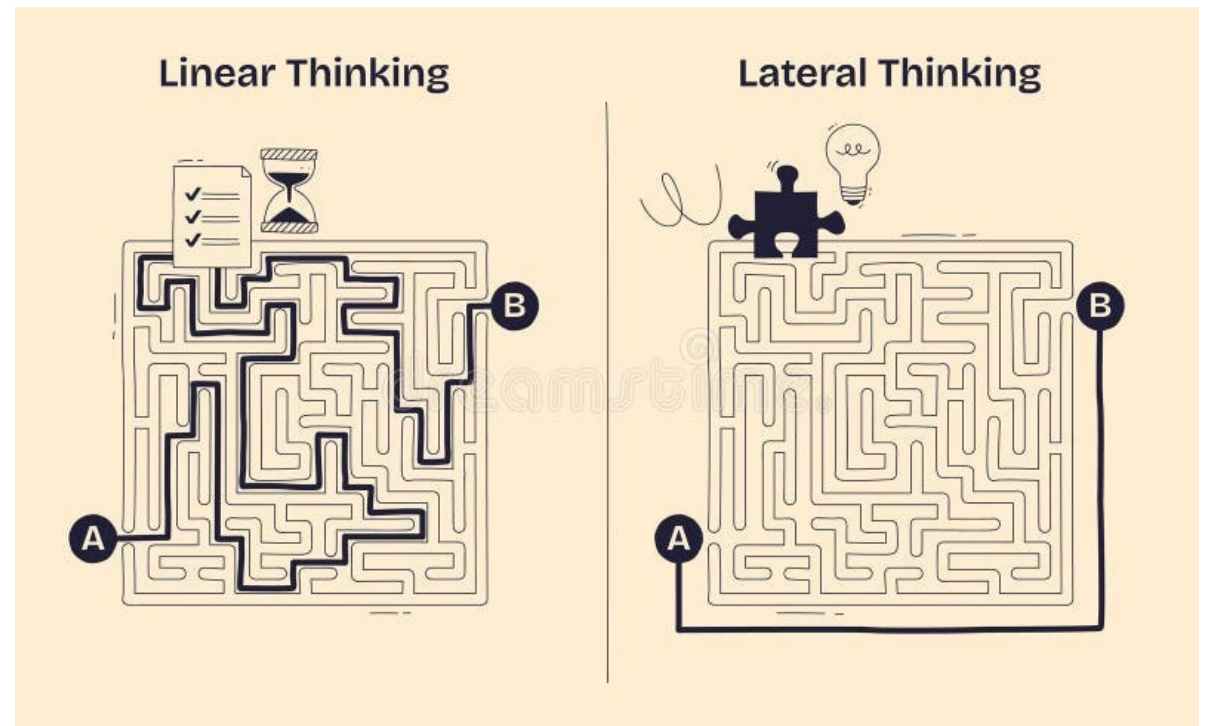
- Positivity for T-cell antigens and CD56.
- Angiocentric and angio-destructive growth
- Positive for CD3 and cytotoxic markers
- EBER+ and EBV at in situ hybridization

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- MDT warranted
- Communication with other specialist
- Awareness



Thank you!!!



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