Il management del linfoma primitivo cutaneo di derivazione T-linfocitaria nell'ambito della regione Emilia-Romagna

Bologna, Royal Hotel Carlton
17 gennaio 2023

Collaborazione (Ematologia/Dermatologia) a Bologna

Alessandro Pileri

Professore Associato

UO Dermatologia

IRCCS Azienda Ospedaliero Universitaria – Policlinico di Sant' Orsola, Bologna.

Alma Mater Studiorum - Università di Bologna.

Dipartimento di Scienze Mediche Chirurgiche

Disclosures of Name Surname

Company name	Research support	Employee	Consultant	Stockholder	Speakers bureau	Advisory board	Other
Kyowa Kirin						X	
Recordati rare diseses			X				

Storia

- Ambulatorio Linfomi Cutanei
- Dal 2010
- Martedì e giovedì

Attualità

- 15 pazienti al giorno
- 4 posti riservati per pazienti interni (ricovero DSV-post dimissione)
- 1 posto riservato per pazienti esterni inviati direttamente dalla UO Ematologia
- 1 posto riservato a pazienti con diagnosi o sospetto di linfoma cutaneo inviati dal territorio (AUSL BO-Dermatologia)
- 1 posto per pazienti con diagnosi o sospetto di linfoma cutaneo inviati dalla UO Dermatologia
- 8 posti per pazienti ambulatoriali (controllo)
- Più di 1000 accessi/anno

Attualità

- Stadi precoci di MF (o altri linfomi T indolenti, es. lyp, CD4+ SMLPD)
- Valutazione esclusiva in Ambulatorio Linfomi della Dermatologia
- Il paziente con sospetto di malattia viene sottoposto a biopsia + es. istologico
- Il paziente in stadio iniziale viene sottoposto a staging secondo linee guida EORCT-CLTG
- Percorso terapeutico: vedi EORCT-CLTG
- In caso di non risposta invio c/o UO Ematologia, Ambulatorio Linfomi

Table 6. Recommended evaluation/initial staging of the patient with mycosis fungoides/Sézary syndrome

Complete physical examination including
Determination of type(s) of skin lesions
If only patch/plaque disease or erythroderma, then estimate percentage of body surface area involved and note any ulceration of lesions
If tumors are present, determine total number of lesions, aggregate volume, largest size lesion, and regions of the body involved
Identification of any palpable lymph node, especially those ≥ 1.5 cm in largest diameter or firm, irregular, clustered, or fixed
Identification of any organomegaly
Skin biopsy
Most indurated area if only one biopsy
Immunophenotyping to include at least the following markers: CD2, CD3, CD4, CD5, CD7, CD8, and a B-cell marker such as CD20. CD30 may also be indicated in cases where lymphomatoid papulosis, anaplastic lymphoma, or large-cell transformation is considered.
Evaluation for clonality of TCR gene rearrangement
Blood tests
CBC with manual differential, liver function tests, LDH, comprehensive chemistries
TCR gene rearrangement and relatedness to any clone in skin
Analysis for abnormal lymphocytes by either Sézary cell count with determination absolute number of Sézary cells and/or flow cytometry (including $CD4^+/CD7^-$ or $CD4^+/CD26^-$)
Radiologic tests
In patients with $T_1N_0B_0$ stage disease who are otherwise healthy and without complaints directed to a specific organ system, and in selected patients with $T_2N_0B_0$ disease with limited skin involvement, radiologic studies may be limited to a chest X-ray or ultrasound of the peripheral nodal groups to corroborate absence of adenopathy
In all patients with other than presumed stage IA disease, or selected patients with limited T_2 disease and the absence of adenopathy or blood involvement, CT scans of chest, abdomen, and pelvis alone ± FDG-PET scan are recommended to further evaluate any potential lymphadenopathy, visceral involvement, or abnormal laboratory tests. In patients unable to safely undergo CT scans, MRI may be substituted.
Lymph node biopsy
Excisional biopsy is indicated in those patients with a node that is either ≥ 1.5 cm in diameter and/or is firm, irregular, clustered, or fixed
Site of biopsy
Preference is given to the largest lymph node draining an involved area of the skin or if FDG-PET scan data are available, the node with highest standardized uptake value (SUV).
If there is no additional imaging information and multiple nodes are enlarged and otherwise equal in size or consistency, the order of preference is cervical, axillary, and inguinal areas.
Analysis: pathologic assessment by light microscopy, flow cytometry, and TCR gene rearrangement.

Attualità

- Stadi avanzati (progressione da stadio iniziale e/o diagnosi ex novo)
- Linfomi aggressivi (es. linfoma di Berti)
- Esecuzione biopsia
- Prescrizione di staging secondo linee guida EORCT-CLTG
- Invio c/o Ambulatorio Linfomi Ematologia
- Gestione eventi avversi alla cute
- Rivalutazione mSWAT score

REVIEW ARTICLE

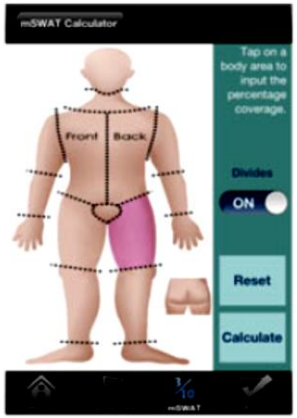
BJD
British Journal of Dermatology

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

J.J. Scarisbrick[1] and S. Morris[2]

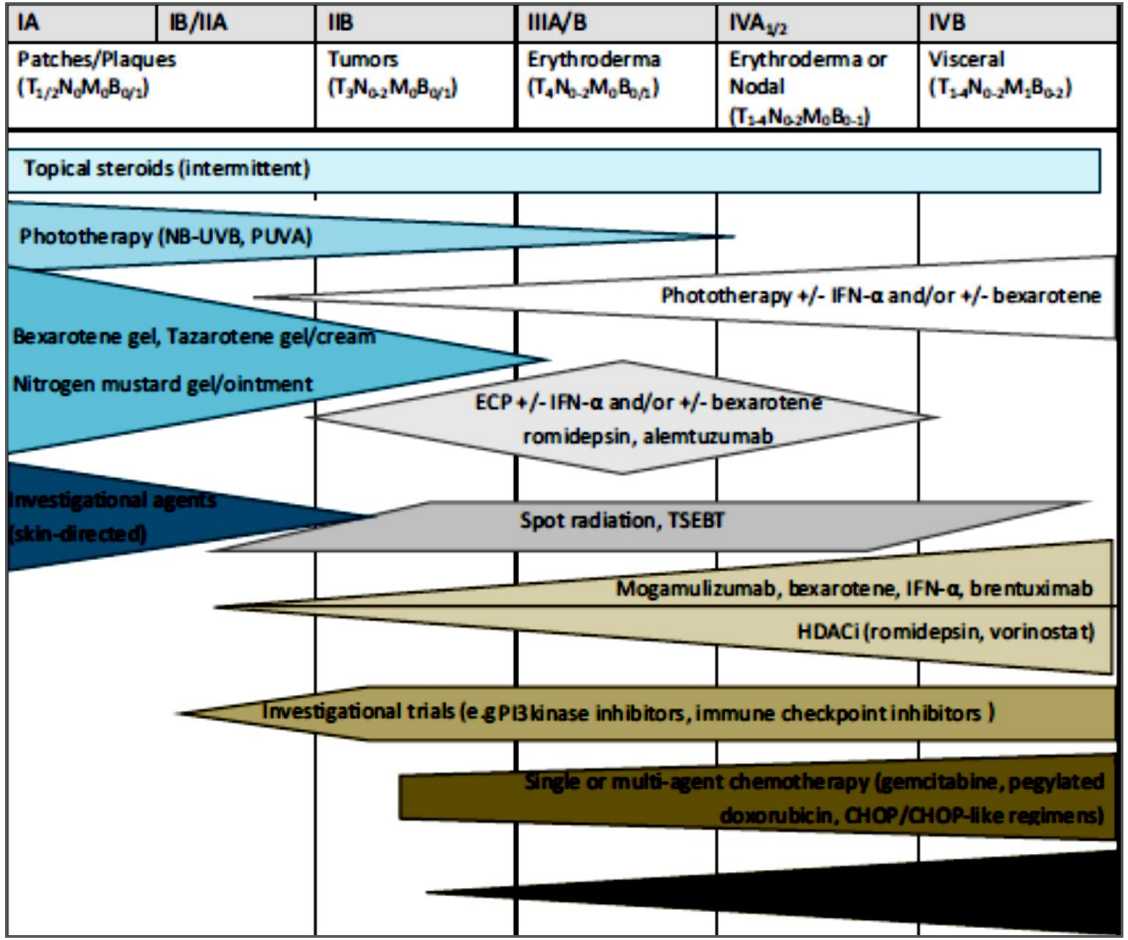
[1]Department of Dermatology, University Hospital Birmingham, Birmingham, B15 2TH, U.K.
[2]Guy's and St Thomas' NHS Trust, London, U.K.

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

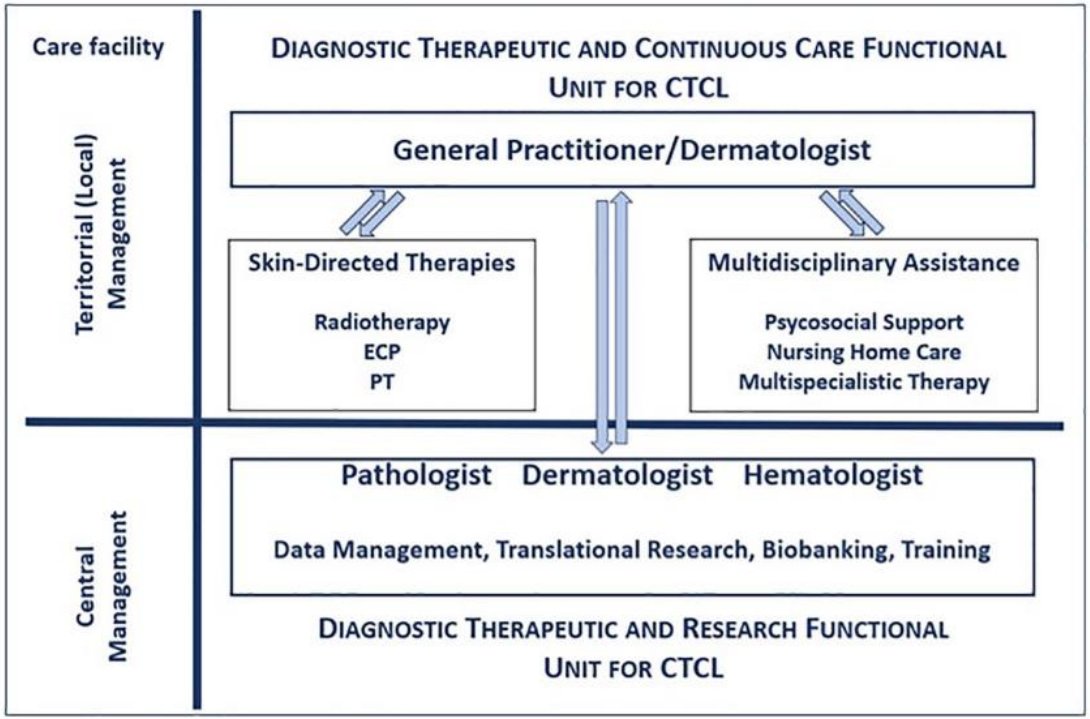
When scoring completed press calculate, which will weight scores and produce an mSWAT score/ 400


IA
IB/IIA
IIB
IIIA/B
IVA1/2
IVB
Patches/Plaques (T1/2N0M0B0/1)
Tumors (T3N0-2M0B0/1)
Erythroderma (T4N0-2M0B0/1)
Erythroderma or Nodal (T1-4N0-2M0B0-1)
Visceral (T1-4N0-2M1B0-2)
Topical steroids (intermittent)
Phototherapy (NB-UVB, PUVA)
Phototherapy +/- IFN-α and/or +/- bexarotene
Bexarotene gel, Tazarotene gel/cream
Nitrogen mustard gel/ointment
ECP +/- IFN-α and/or +/- bexarotene
romidepsin, alemtuzumab
Investigational agents (skin-directed)
Spot radiation, TSEBT
Mogamulizumab, bexarotene, IFN-α, brentuximab
HDACi (romidepsin, vorinostat)
Investigational trials (e.g PI3kinase inhibitors, immune checkpoint inhibitors)
Single or multi-agent chemotherapy (gemcitabine, pegylated doxorubicin, CHOP/CHOP-like regimens)

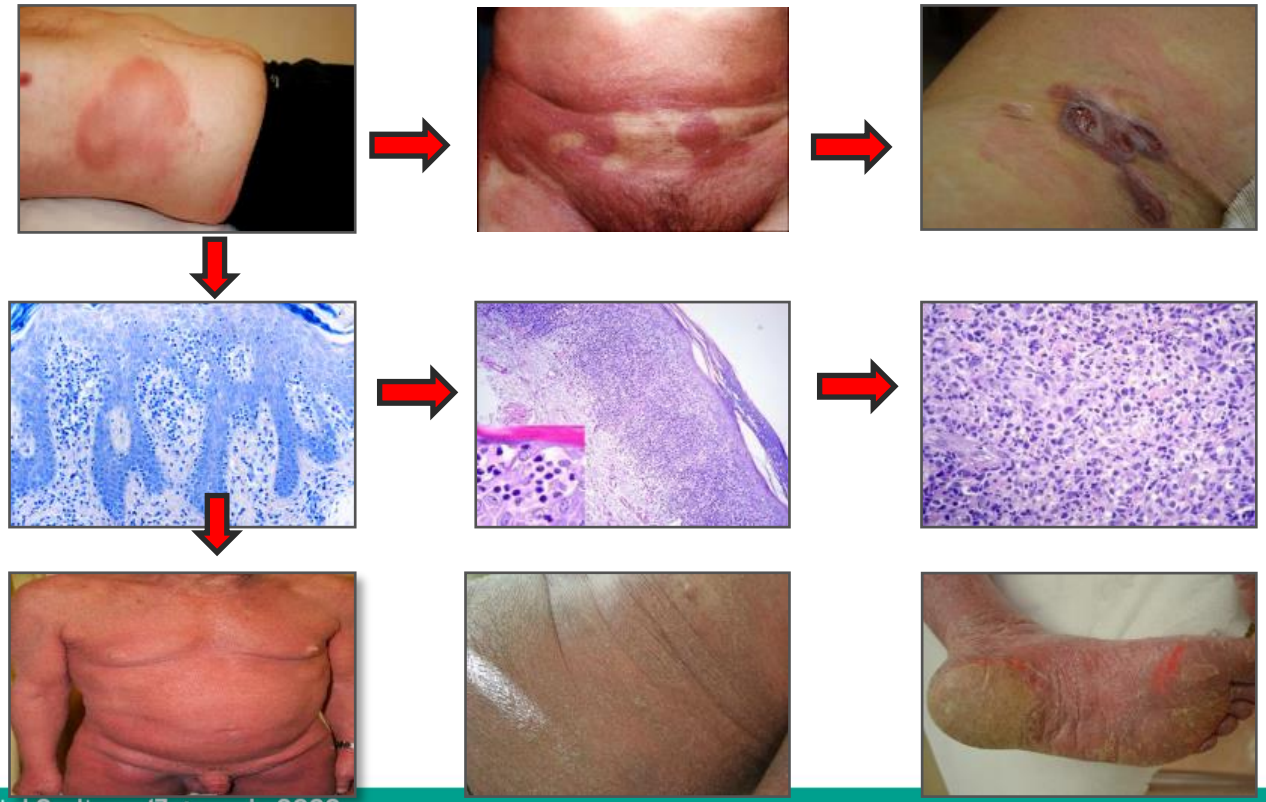
Attualità

- Discussione collegiale casi clinici
- Near future: organizzazione incontri multidisciplinari (dermatologo-ematologo-emolinfopatologo)
- Proiezione immagini cliniche raccolte

An integrated model for the management of patients with cutaneous T-cell lymphomas



Zinzani PL. *Hematol Oncol*, 2021; 39: 275-283




Grazie!!!