

# MYCOSIS FUNGOIDES

## Cutaneous T-cell lymphoma

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Cutaneous T-cell lymphoma: clinical course, biology and pathology

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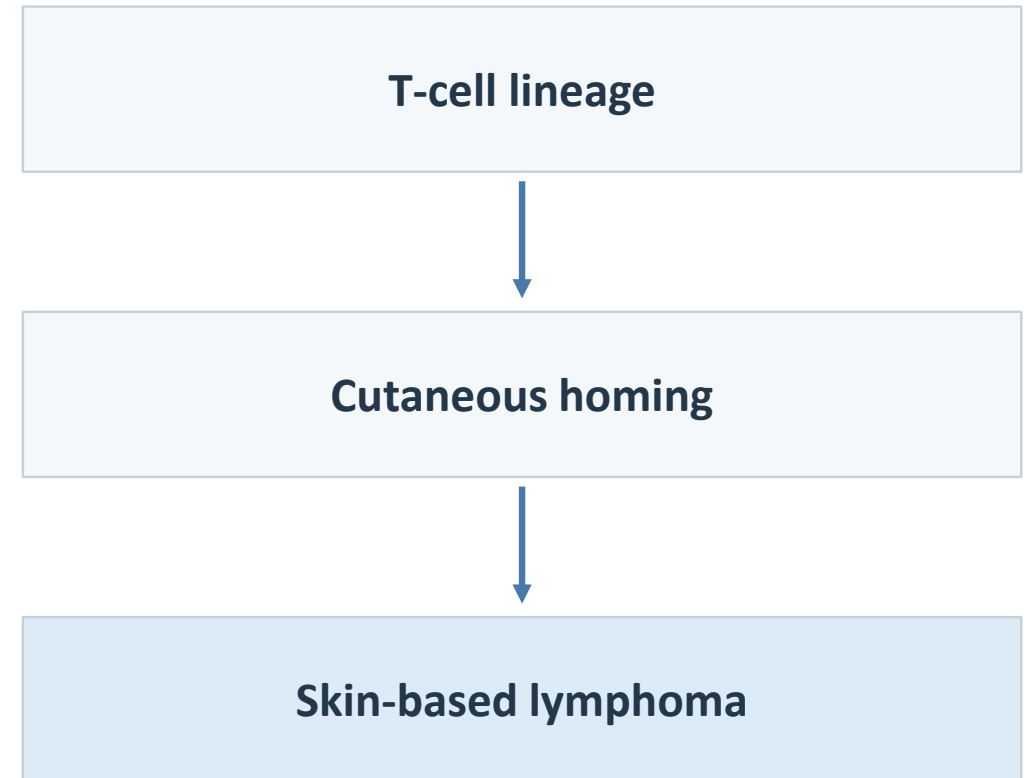
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# Learning objectives

- Define mycosis fungoides within the cutaneous T-cell lymphoma spectrum.
- Explain skin homing, clonality and the limits of molecular specificity.
- Correlate patch, plaque and tumor lesions with microscopic patterns.
- Interpret progression, Sézary syndrome and treatment context.

# A neoplasm of cells that migrate to skin

- Mycosis fungoides is discussed among tumors of cellular migrants to skin.
- The malignant lineage is lymphoid, not keratinocytic or melanocytic.
- Its clinical appearance reflects tissue localization by transformed T cells.



*Schematic (not histology)*

# Cutaneous T-cell lymphoma is a spectrum

- CTCL denotes multiple lymphoproliferative disorders involving the skin.
- Mycosis fungoides is a defined clinicopathologic member of that spectrum.
- Not every CTCL has the clinical evolution of mycosis fungoides.

| Umbrella category              | Mycosis fungoides                            |
|--------------------------------|----------------------------------------------|
| Several distinct presentations | Characteristic patches<br>→ plaques → tumors |

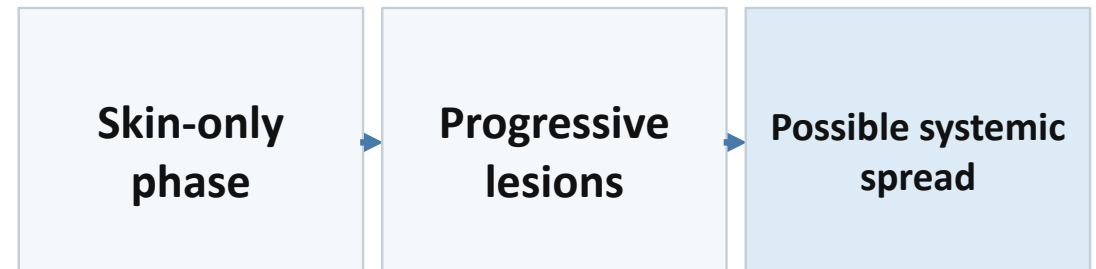
## Core definition: A skin-homing helper T-cell lymphoma

- The prototypic cells are CD4+ helper T lymphocytes with cutaneous tropism.
- Lesions generally begin in skin; Overt extracutaneous disease is late.
- Diagnosis rests on integration of clinical and pathologic patterns.

**CD4+ • skin-homing • clonal**

## An often prolonged cutaneous phase

- Most patients have skin-localized disease for years.
- This clinical indolence does not negate the neoplastic nature of the process.
- A subset eventually develops tumors or extracutaneous disease.



*Time and progression are variable*

## Age distribution does not exclude the diagnosis

- Mycosis fungoides may develop at any age.
- The disease is most common after 40 years of age.
- In compatible lesions, age alone should not substitute for morphology.

# The trunk is a frequent site of disease

- Early lesions commonly occur on truncal skin.
- With broader cutaneous involvement, extremities, face and scalp may be affected.
- Anatomic distribution is clinically useful but not independently diagnostic.

**Trunk**

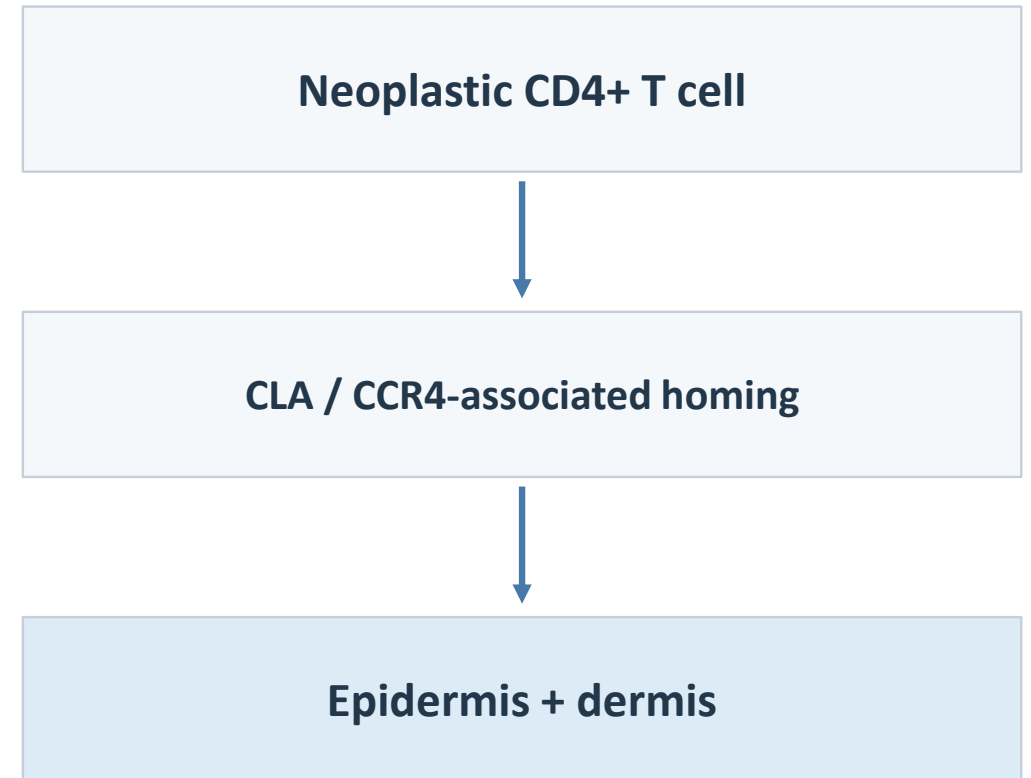
Extremities

Face and scalp

*Illustrative anatomic distribution*

# Why CD4+ tumor cells accumulate in skin

- Tumor cells express adhesion and chemokine-homing molecules.
- Cutaneous lymphocyte antigen contributes to skin localization.
- CCR4 is additionally highlighted in the lymphoid chapter.



*Schematic (not histology)*

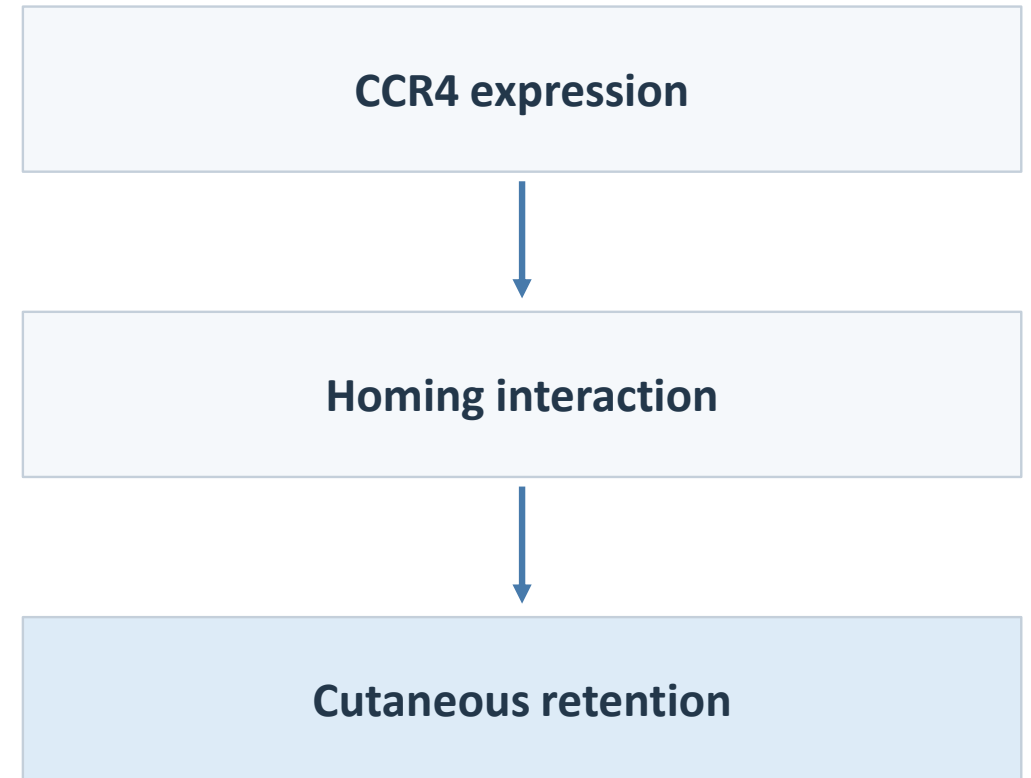
## Cutaneous lymphocyte antigen: Tissue targeting

- Cutaneous lymphocyte antigen is associated with T-cell trafficking to skin.
- Its expression provides a mechanistic link between lineage and site.
- Skin localization reflects homing biology, not epithelial origin.

**CLA → cutaneous localization**

# CCR4 adds a chemokine-receptor component

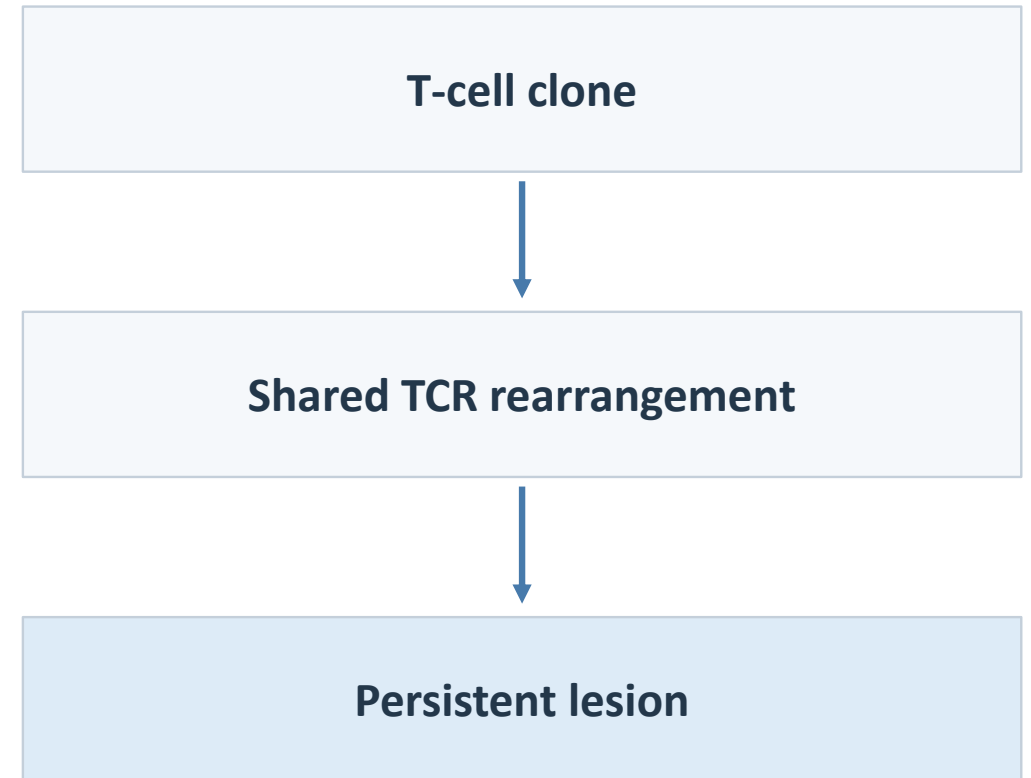
- The Chapter 13 discussion identifies CCR4 in skin-homing tumor cells.
- Chemokine receptors participate in positioning T cells within tissues.
- Cutaneous trafficking helps explain persistent skin lesions.



*Schematic (not histology)*

# Clonal T-cell receptor rearrangements

- Mycosis fungoides cells represent a clonal T-cell population.
- Clonal T-cell receptor gene rearrangements support shared lineage.
- Clonality must be interpreted with clinicopathologic findings.



*Schematic (not histology)*

# Genetic abnormalities are heterogeneous

- Changes involving oncogenes and tumor-suppressor genes occur in CTCL.
- Robbins does not identify a single mutation specific to this entity.
- No one mutation should be portrayed as its defining genetic test.

## Heterogeneous drivers

Multiple altered  
pathways

## No disease-specific driver

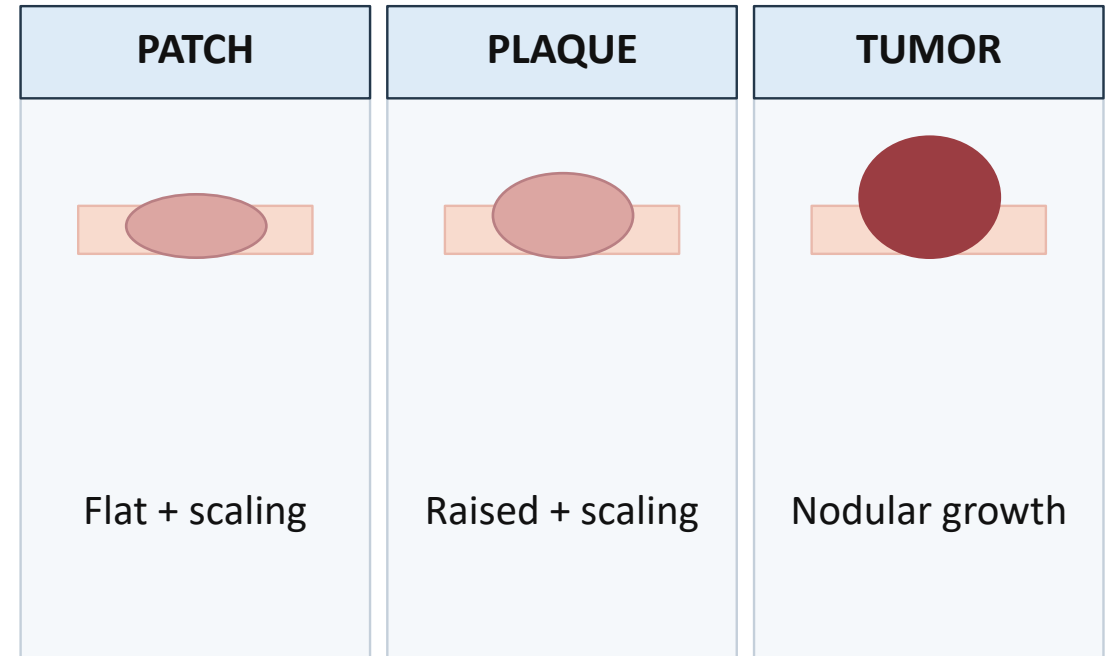
Diagnosis remains  
integrated

## A characteristic clinical-pathologic constellation

| Domain     | Typical observation                   | Interpretive role   |
|------------|---------------------------------------|---------------------|
| Clinical   | Persistent patches, plaques or tumors | Pattern recognition |
| Histologic | Dermal band + epidermotropism         | Tissue pattern      |
| Biologic   | CD4+ T cells; clonal rearrangement    | Lineage support     |

# Skin lesions form a clinical continuum

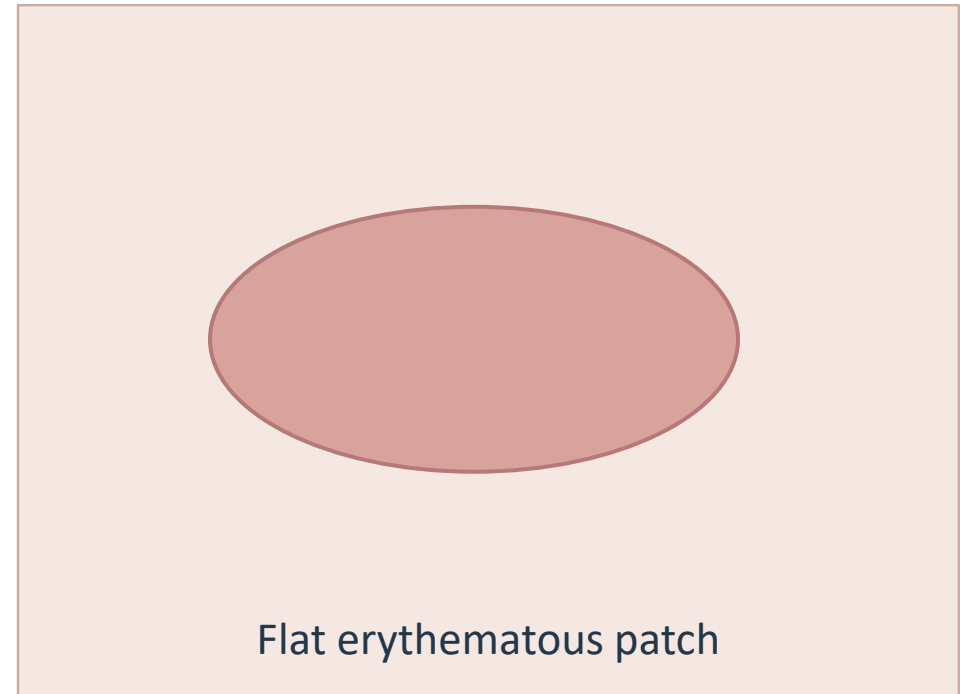
- The classic course is patch → plaque → tumor.
- The phases are useful teaching categories, not mandatory uniform steps.
- Different lesion morphologies may coexist in an individual.



*Schematic (not histology)*

## Patch lesions: Subtle cutaneous disease

- Flat erythematous or red-brown areas may show surface scale.
- Early lesions may resemble eczema.
- A persistent patch can precede more overt lymphoma for years.



*Schematic (not histology)*

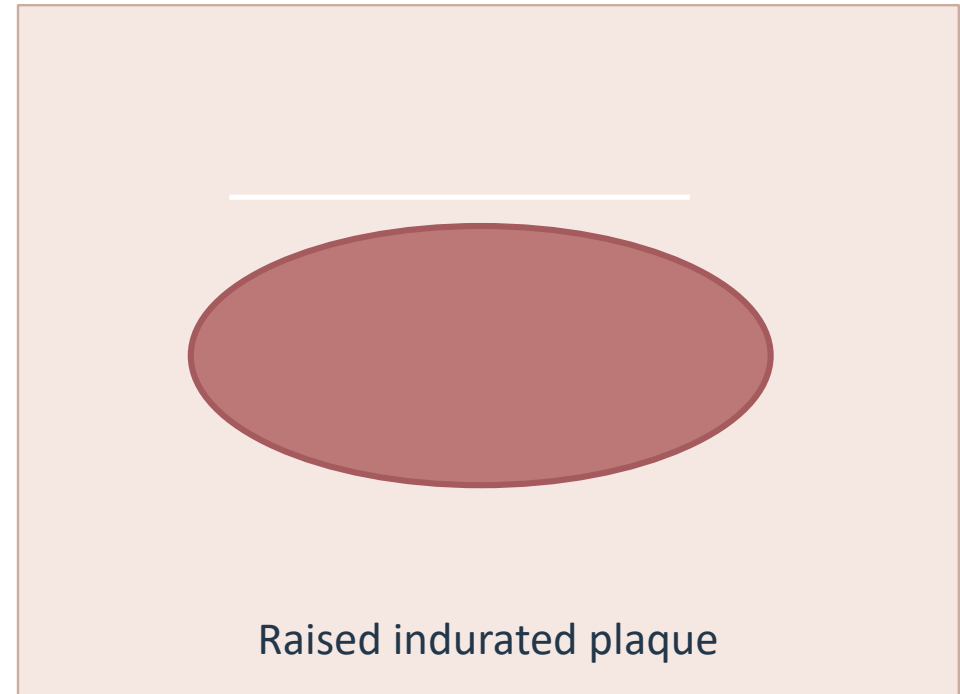
## The premycotic phase can appear inflammatory

- An eczematous presentation is typical of early mycosis fungoides.
- Obvious nodal or visceral dissemination is absent at this phase.
- Morphologic subtlety is a major reason for diagnostic uncertainty.

| Clinical surface  | Underlying process         |
|-------------------|----------------------------|
| Eczema-like patch | Neoplastic T cells in skin |

# Plaque lesions: Raised and indurated

- Plaques become elevated, scaling and erythematous.
- Irregular outlines and firmness may emerge.
- The appearance can be confused with psoriasis.



*Schematic (not histology)*

# Why plaque disease may mimic psoriasis

- Both processes can produce red, thickened, scaling plaques.
- The distinction therefore requires more than a descriptive skin examination.
- In mycosis fungoides, atypical T-cell epidermotropism is informative.

## Visible similarity

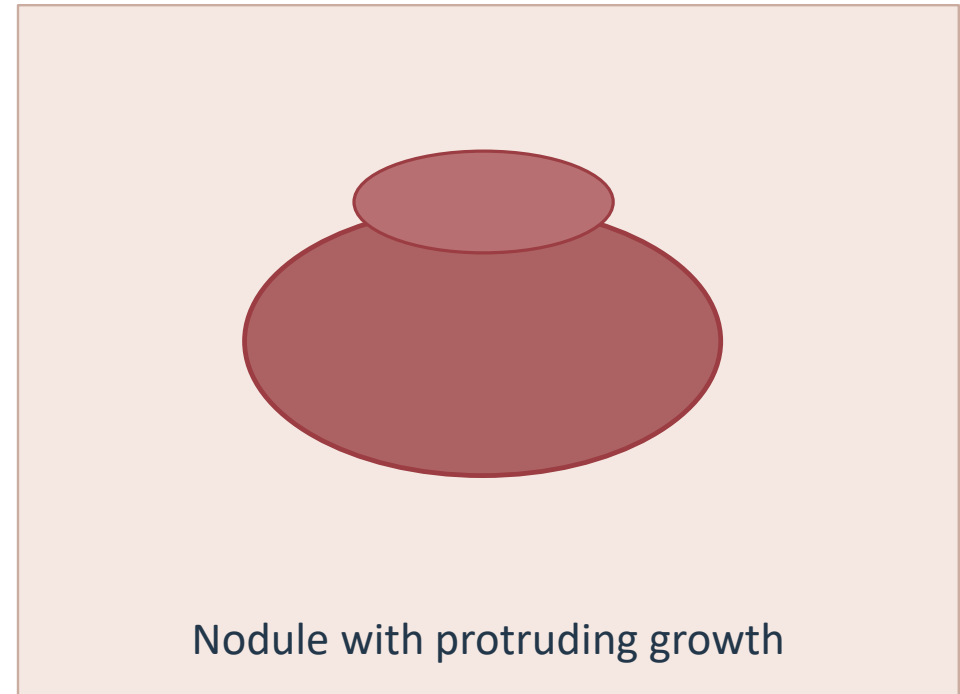
Erythema + scale +  
thickening

## Decisive difference

Neoplastic lymphoid  
tissue pattern

## Tumor-stage lesions are distinct from plaques

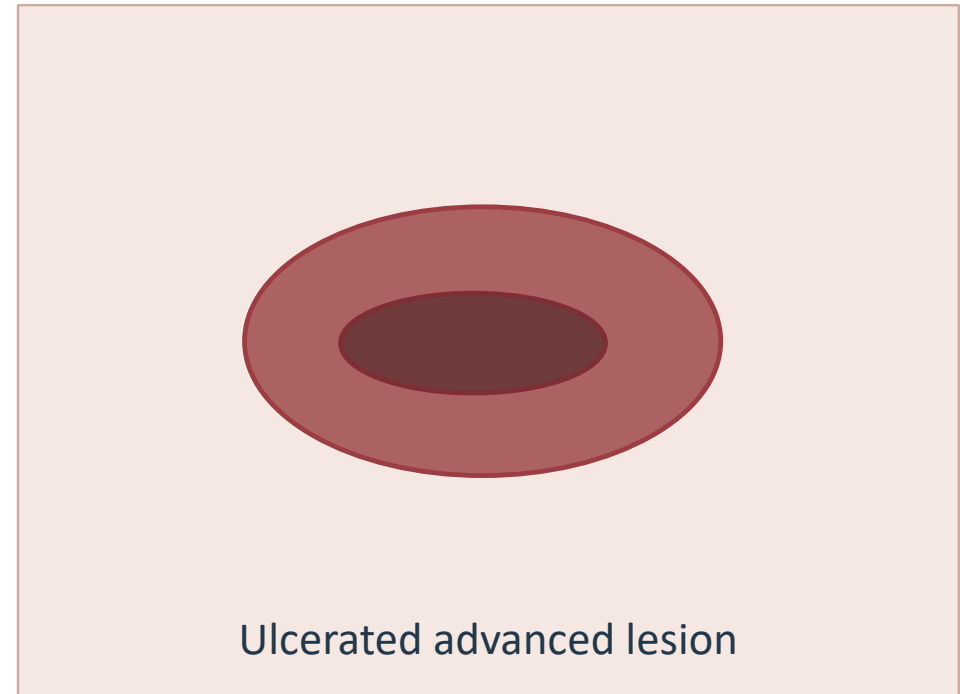
- Advanced lesions may form nodules with a fungating surface.
- Deep dermal growth contributes to nodule formation.
- Tumor development is associated with aggressive clinical behavior.



*Schematic (not histology)*

# Ulceration can complicate advanced lesions

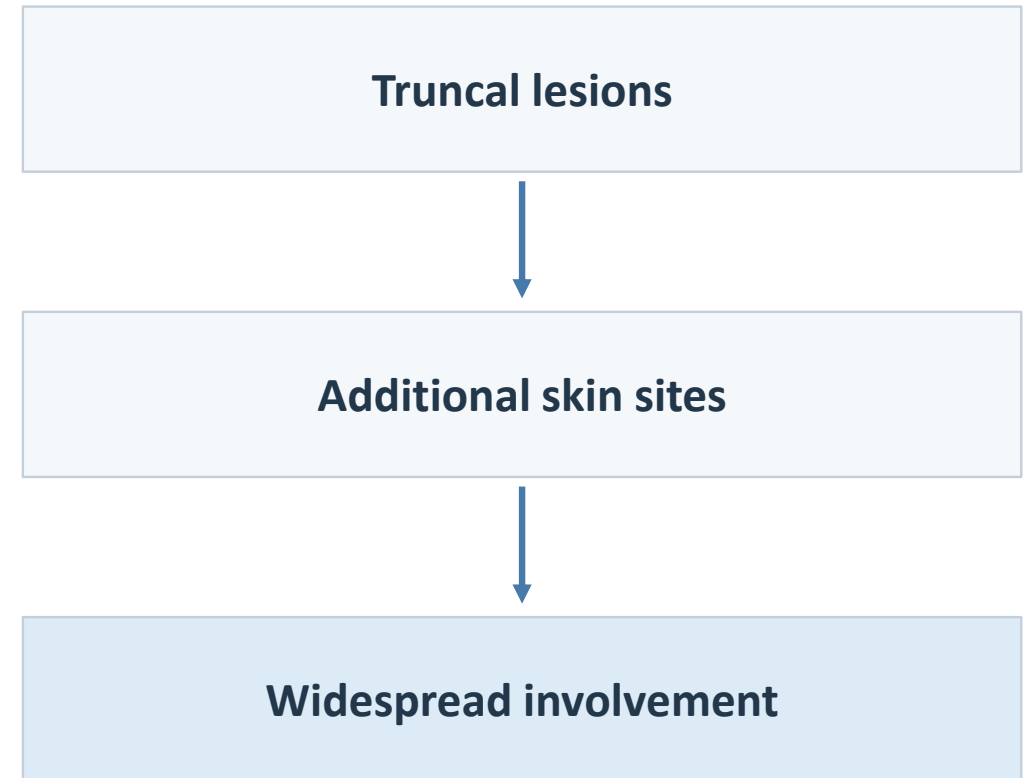
- Plaques or tumor nodules can ulcerate.
- Ulceration is a feature of some lesions, not a required diagnostic sign.
- The microscopic and clinical context must still establish the entity.



*Schematic (not histology)*

# Cutaneous distribution may become extensive

- Disease may expand beyond the trunk.
- Extremities, face and scalp can eventually be affected.
- Anatomic extension increases the need for systematic skin assessment.



*Schematic (not histology)*

## Patch, plaque and tumor are not synonyms

| Form   | Gross appearance                 | Broad significance            |
|--------|----------------------------------|-------------------------------|
| Patch  | Flat, scaly erythematous area    | Often early presentation      |
| Plaque | Raised, indurated scaling lesion | More established skin disease |
| Tumor  | Fungating or nodular mass        | Advanced cutaneous burden     |

# Clinical evolution is often slow

- Progression between forms may occur over prolonged intervals.
- Many patients remain localized to skin.
- Late nodal or visceral dissemination is a distinct clinical change.



*Time and progression are variable*

# Skin-surface burden influences prognosis

- Outcome to the percentage of body surface involved.
- Extent of cutaneous involvement is not merely cosmetic severity.
- This source does not provide numerical staging thresholds.

**Limited skin burden**

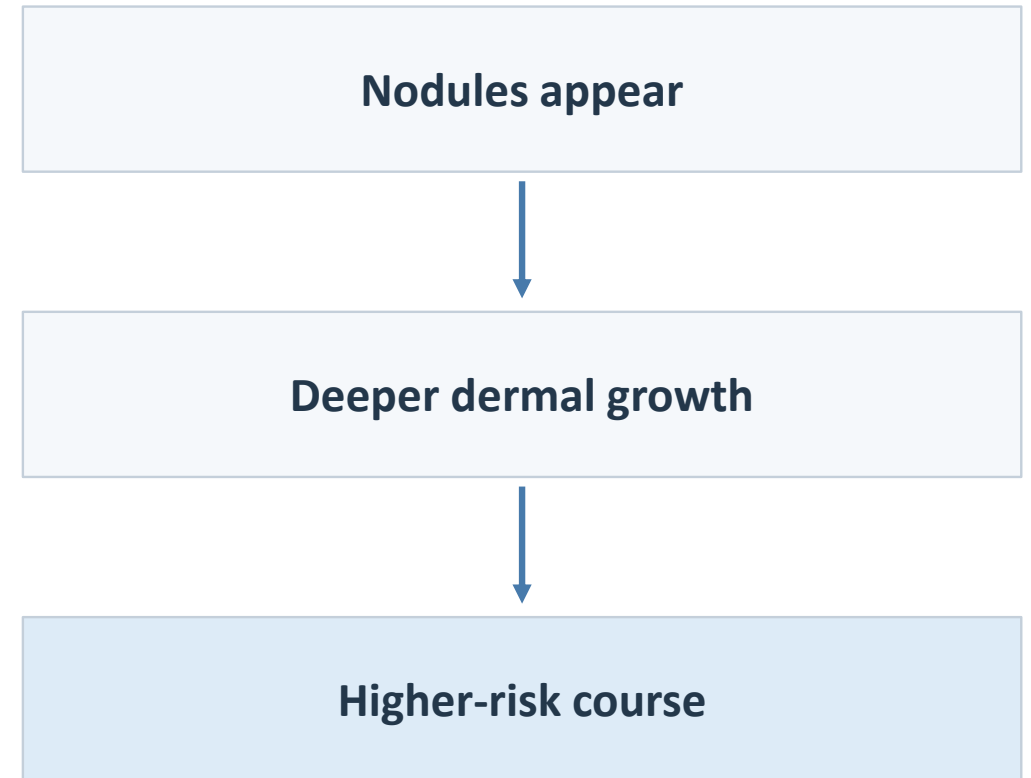
**More extensive skin burden**

**Tumors or systemic involvement**

*Qualitative risk relationships — no numeric cut-offs implied*

# Tumor nodules indicate greater risk

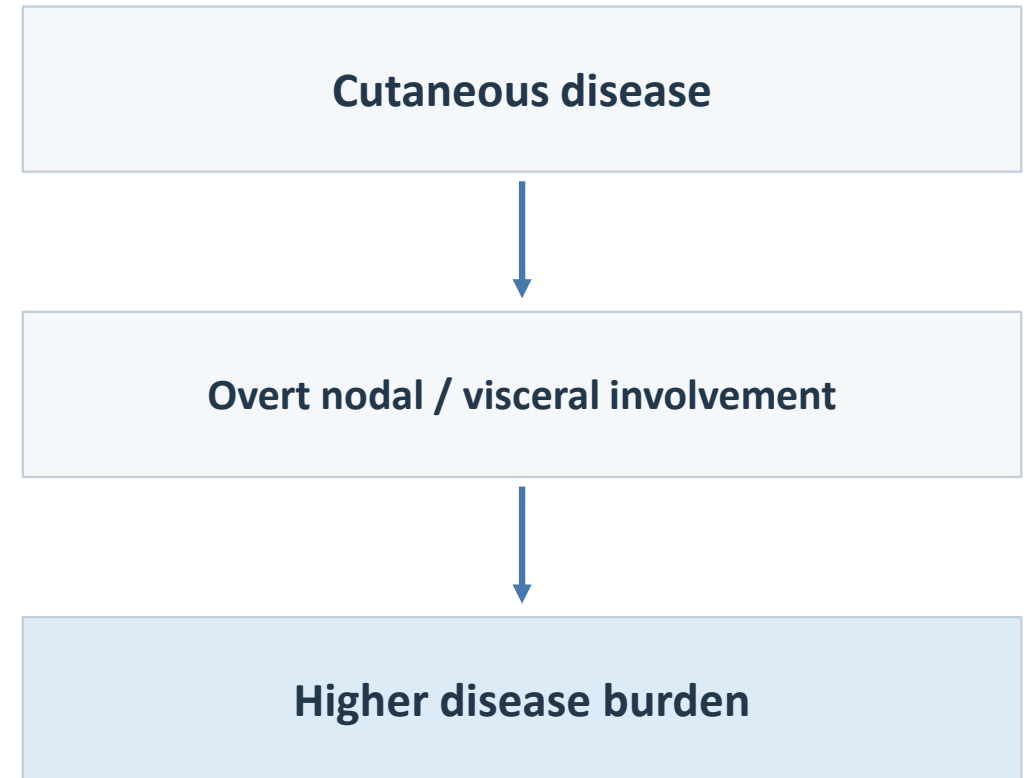
- The appearance of tumor nodules correlates with systemic spread.
- Nodular transformation reflects altered tissue growth behavior.
- Tumor presence is associated with more aggressive disease.



*Schematic (not histology)*

## Extracutaneous spread changes the interpretation

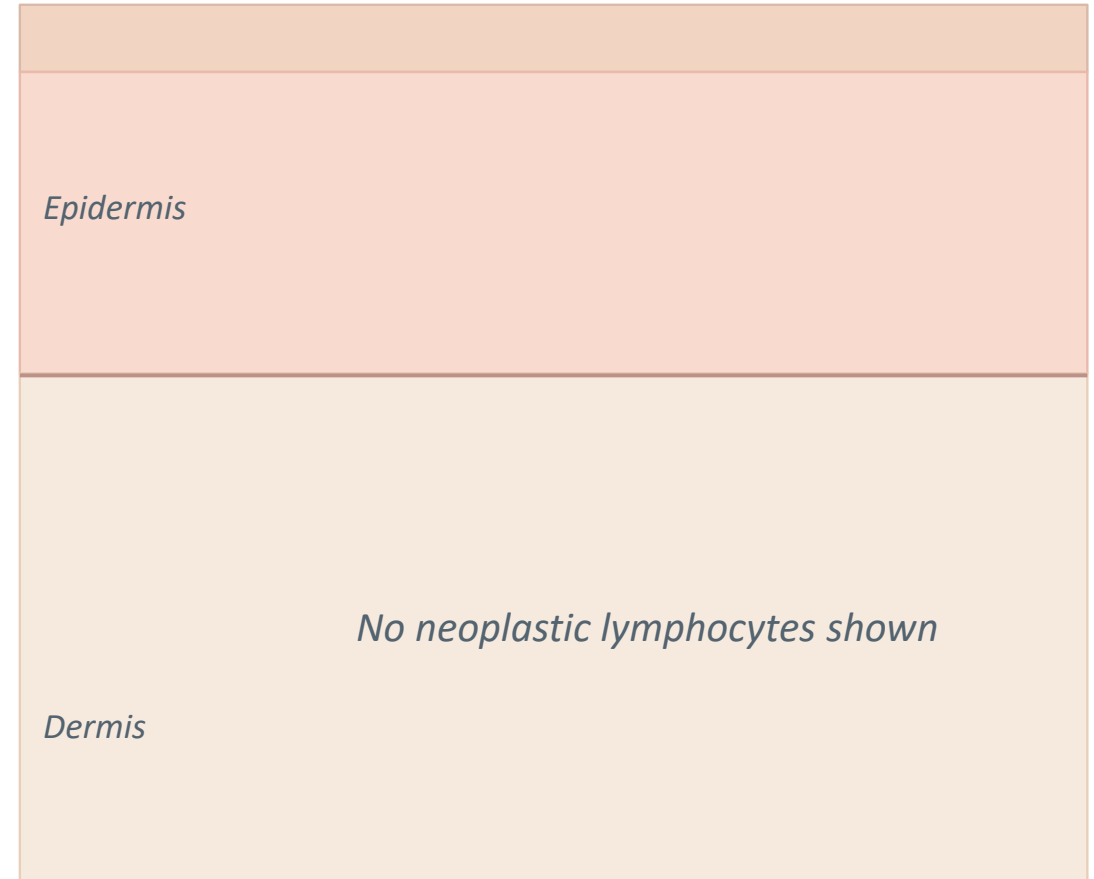
- Overt spread to lymph nodes or viscera is usually a late event.
- Presence of systemic disease is linked with a worse prognosis.
- Circulating cells alone are not equivalent to overt organ infiltration.



*Schematic (not histology)*

# Normal skin compartments orient the biopsy

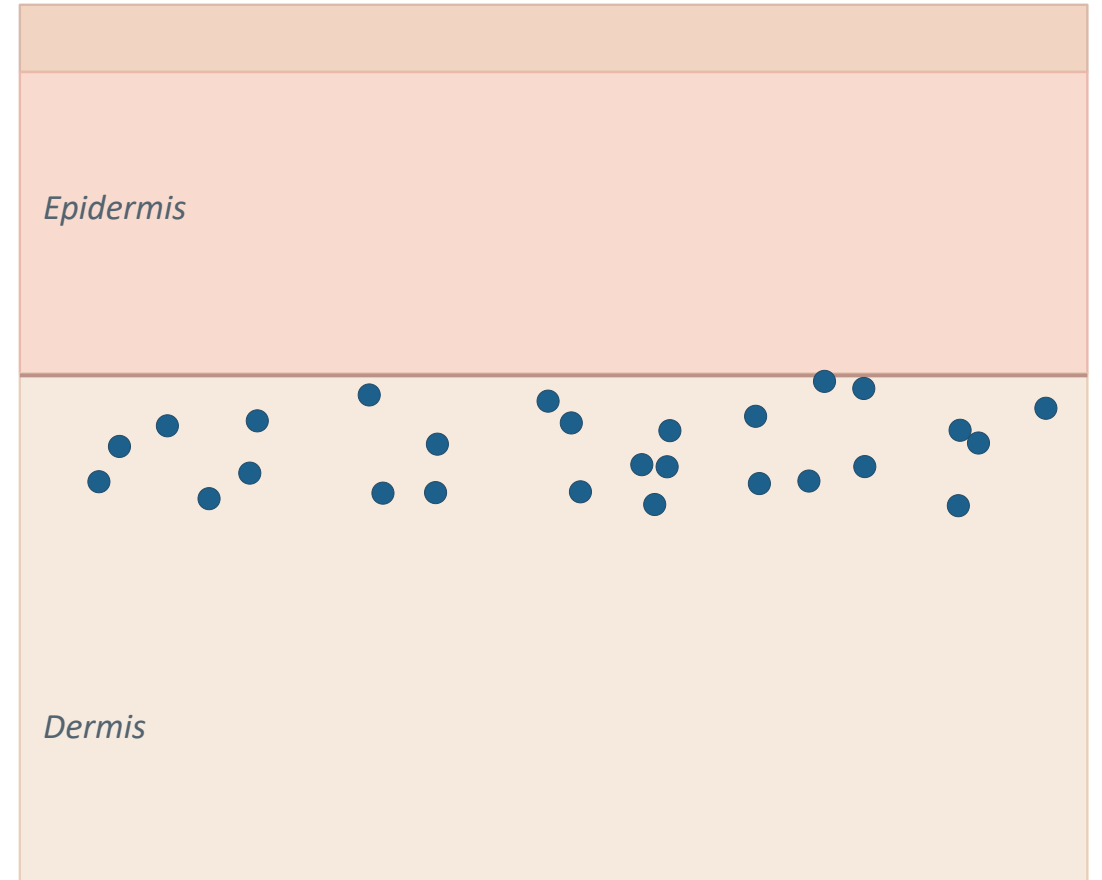
- Epidermis overlies the papillary and deeper dermis.
- Mycosis fungoides combines superficial dermal infiltration with epidermal involvement.
- Different clinical phases shift the depth and distribution of atypical cells.



*Schematic (not histology)*

# Superficial dermal band of atypical cells

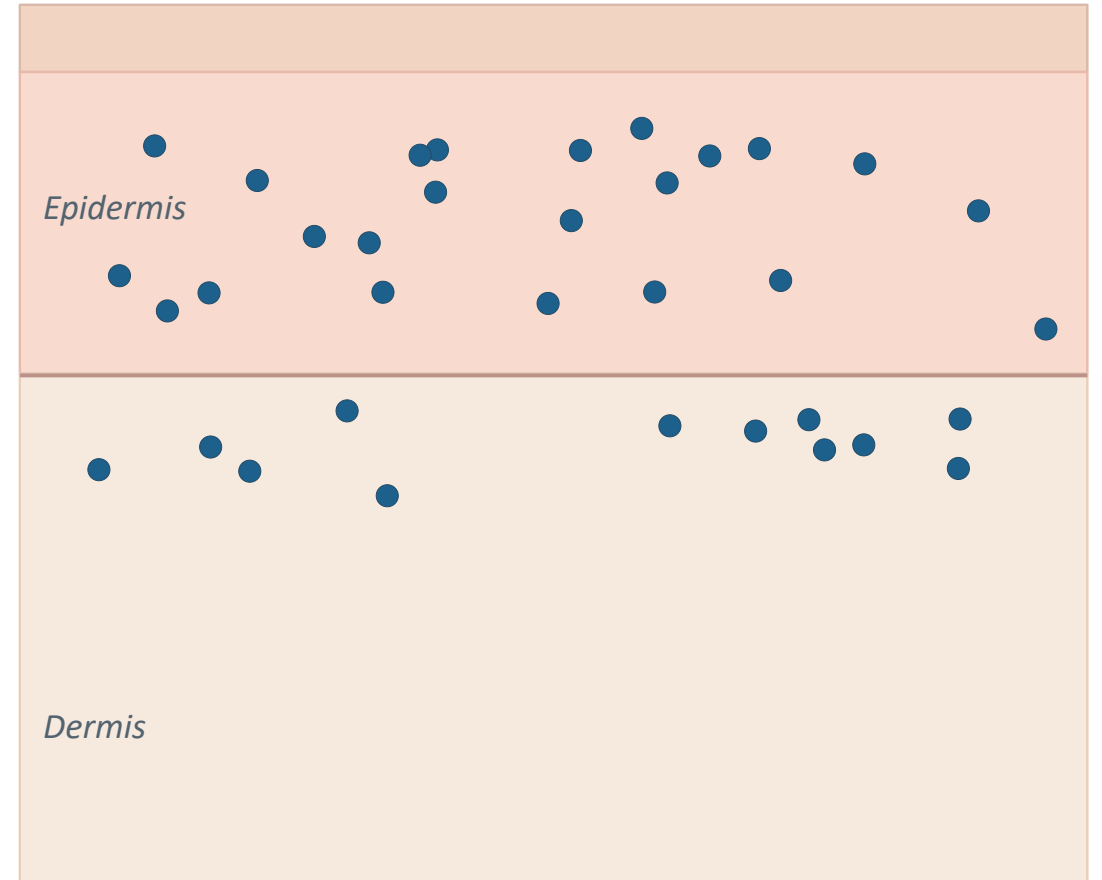
- A band-like population of atypical lymphocytes collects in superficial dermis.
- This localization is a characteristic morphologic clue.
- Band-like distribution must be assessed along with epidermotropism.



*Schematic (not histology)*

## Epidermotropism: Lymphocytes enter epidermis

- Atypical T cells infiltrate epidermis as individual cells.
- The finding distinguishes epidermal infiltration from a purely dermal band.
- Epidermotropism is prominent in many patch and plaque lesions.



*Schematic (not histology)*

## Pautrier microabscesses are intraepidermal clusters

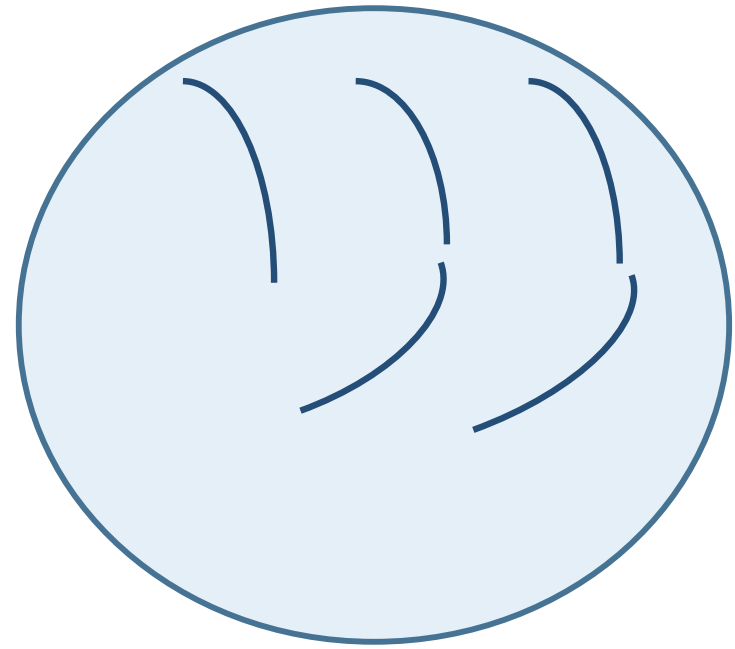
- Small groups of atypical lymphocytes can collect within the epidermis.
- These clusters are termed Pautrier microabscesses.
- They are lymphoid collections, not neutrophilic abscesses.



*Schematic (not histology)*

## Nuclear folding produces cerebriform contours

- Atypical lymphocytes may show markedly infolded nuclear membranes.
- The resulting outline appears convoluted or cerebriform.
- Nuclear contour complements, rather than replaces, assessment of architecture.

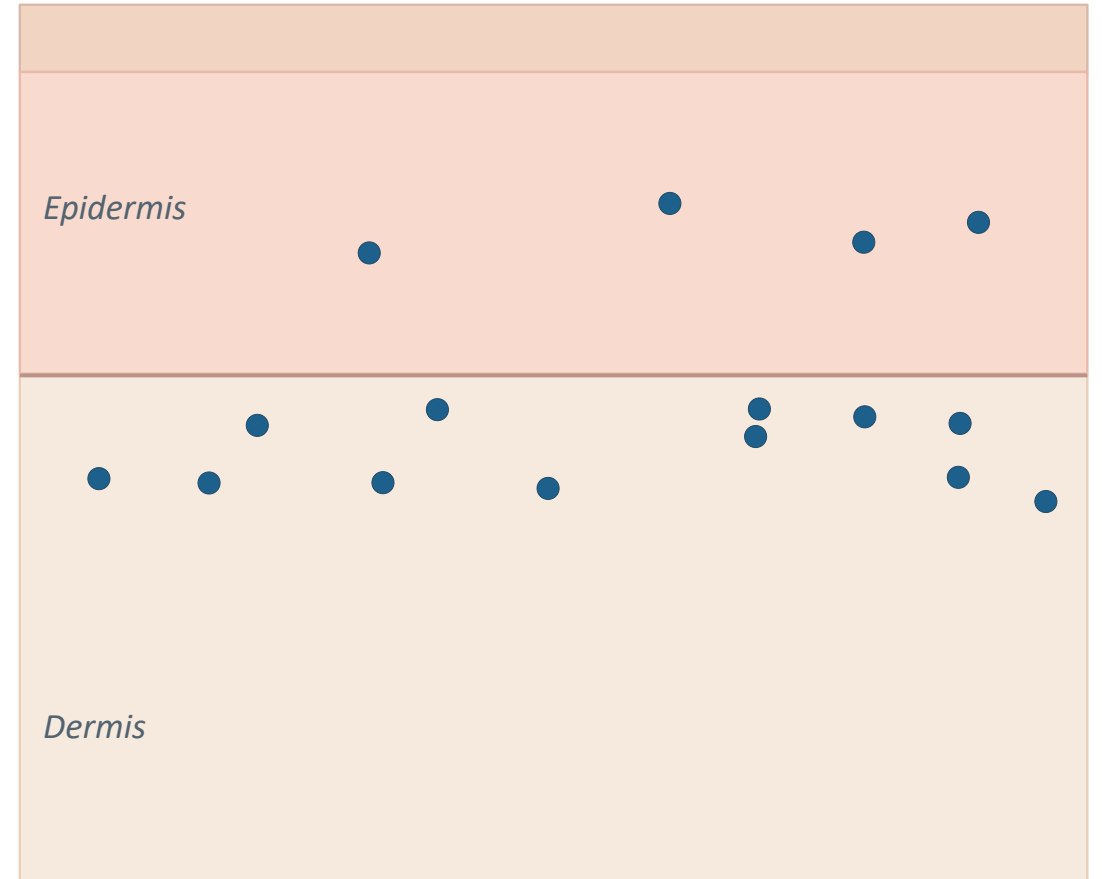


**Convoluted nuclear outline**

*Schematic (not histology)*

## Patch-stage histology may be subtle

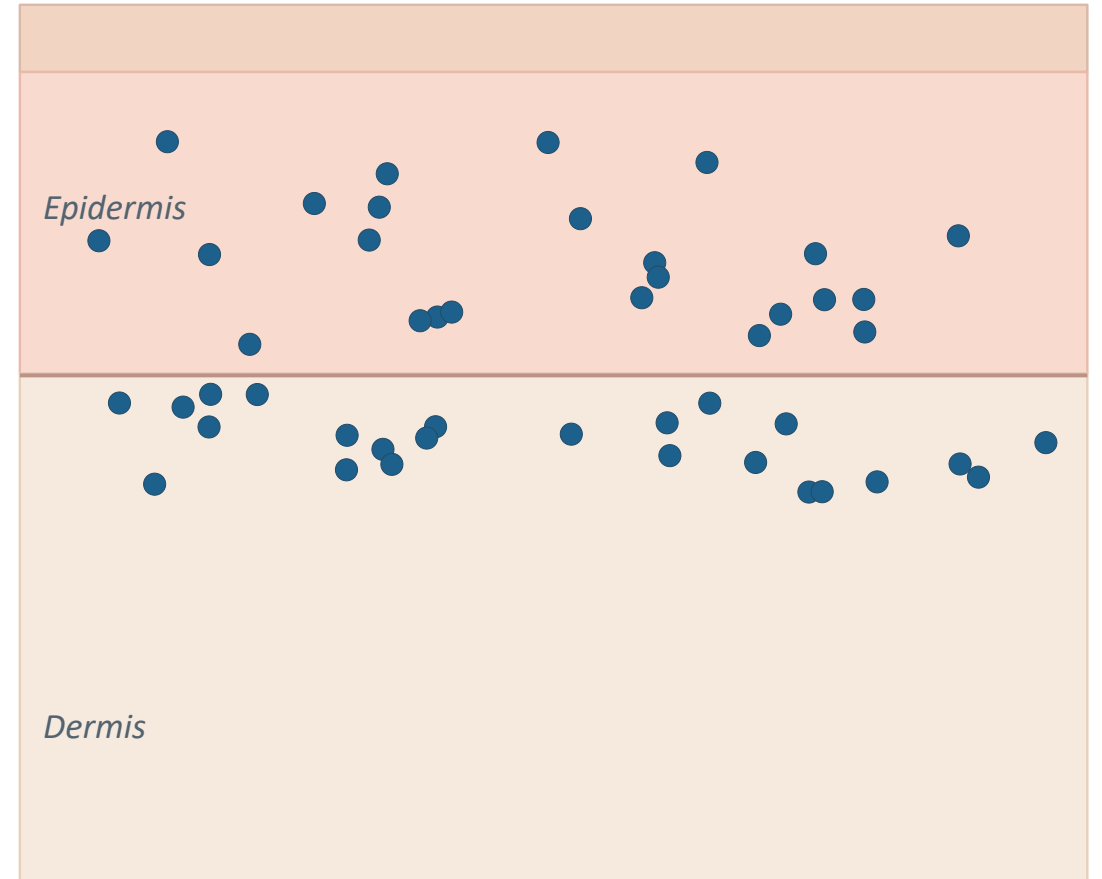
- The infiltrate may be superficial and limited in density.
- Epidermotropism can be present despite a clinically understated lesion.
- An inflammatory-appearing specimen does not automatically exclude MF.



*Schematic (not histology)*

## Plaque-stage histology emphasizes epidermotropism

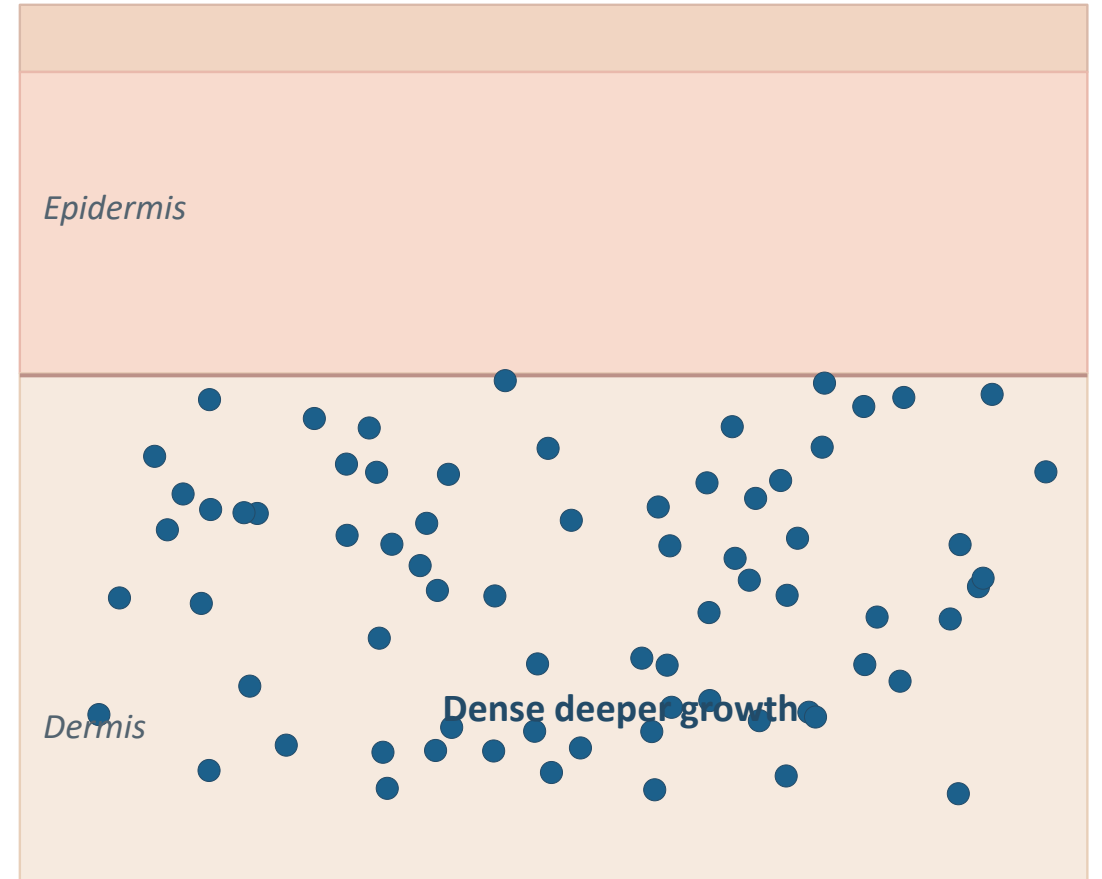
- Plaques frequently contain conspicuous epidermal atypical lymphocytes.
- An expanded superficial dermal infiltrate may coexist.
- Pautrier microabscesses can be seen in this setting.



*Schematic (not histology)*

# Tumor-stage histology becomes deeply dermal

- Malignant lymphocytes grow more extensively within dermis.
- The deep infiltrate produces clinically appreciable nodules.
- Tumor lesions may show less epidermotropism than earlier patches.



*Schematic (not histology)*

## Epidermotropism can diminish as tumors develop

- Patch/plaque disease typically involves the epidermis prominently.
- Advanced tumor-stage cells can grow deeply and lose epidermotropism.
- An absent epidermotropic pattern does not exclude advanced MF.

| Patch / plaque                        | Tumor                                              |
|---------------------------------------|----------------------------------------------------|
| Epidermal infiltration<br>conspicuous | Deeper growth; Often<br>reduced<br>epidermotropism |

## A single biopsy field is not the whole diagnosis

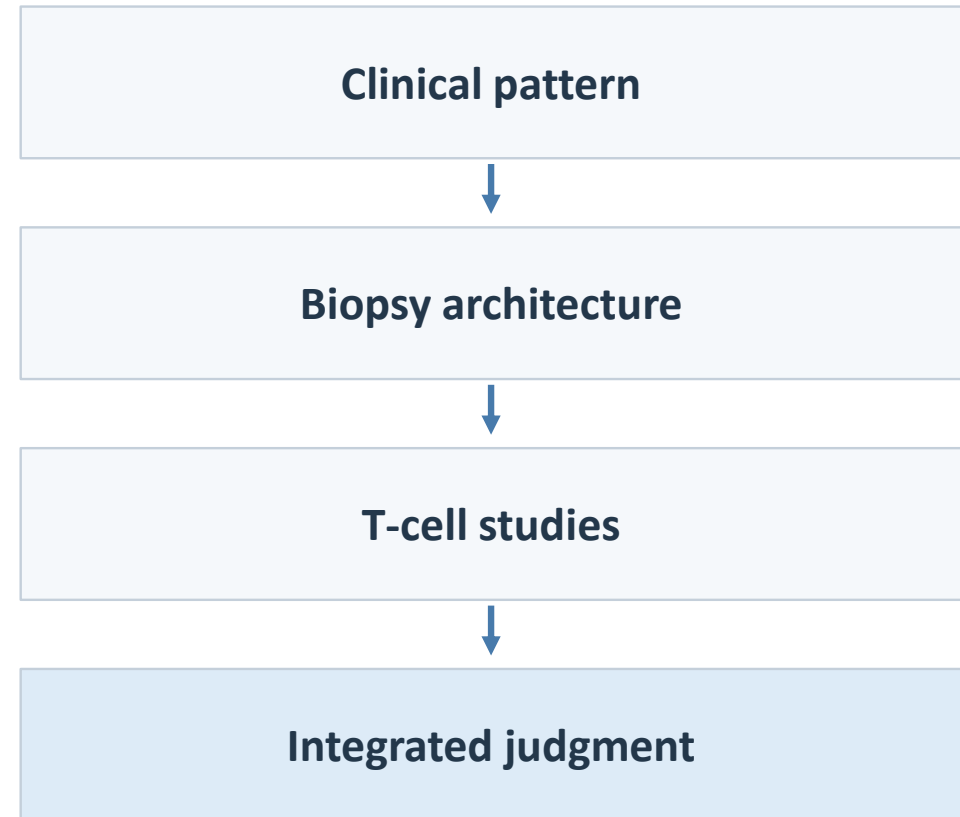
| Assess        | Question                      | Why it matters      |
|---------------|-------------------------------|---------------------|
| Distribution  | Where are cells concentrated? | Superficial vs deep |
| Epidermis     | Single cells or clusters?     | Epidermotropism     |
| Nuclei        | Convoluting contours?         | Cytologic support   |
| Clinical form | Patch, plaque or nodule?      | Stage correlation   |

## Morphology and clinical phase are linked

| Clinical form | Dominant microscopic emphasis                     | Visible correlate        |
|---------------|---------------------------------------------------|--------------------------|
| Patch         | Superficial sparse infiltrate;<br>epidermotropism | Flat scaly lesion        |
| Plaque        | Dermal band and epidermal atypical<br>cells       | Thickened scaling plaque |
| Tumor         | Dense deeper dermal infiltrate                    | Nodular mass             |

## Diagnosis integrates several lines of evidence

- A compatible lesion history prompts a representative skin biopsy.
- Morphology establishes the distribution of atypical lymphocytes.
- T-cell phenotype and clonality can provide corroboration.



*Schematic (not histology)*

## The T-cell phenotype is supportive

- MF is most often associated with a CD4+ helper T-cell population.
- Surface T-cell antigens may be aberrantly combined.
- Phenotype must be interpreted with morphology and clinical presentation.

| Expected lineage   | Possible aberrancy                  |
|--------------------|-------------------------------------|
| CD4+ helper T cell | Unusual T-cell antigen combinations |

## Clonality is evidence, not a complete diagnosis

- T-cell receptor rearrangements can demonstrate a shared clone.
- A molecular result does not reveal lesion architecture or clinical course.
- The final diagnosis is clinicopathologic, not a single-assay label.

| Molecular information      | Clinical/pathologic information |
|----------------------------|---------------------------------|
| Presence of a T-cell clone | Whether findings fit MF         |

## Early MF requires careful differential diagnosis

| Consideration          | Shared appearance        | Useful distinction                          |
|------------------------|--------------------------|---------------------------------------------|
| Eczema-like dermatitis | Erythematous scaly patch | Persistence + atypical epidermotropic cells |
| Psoriasis              | Scaling plaque           | Lymphoid infiltrate architecture            |
| Other CTCL             | Cutaneous T-cell lesion  | Entity-specific clinical presentation       |

## Eczema-like lesions are not always eczema

- MF may initially present as an inflammatory-appearing patch.
- Discordance between persistent lesions and a nonspecific label deserves review.
- The biopsy should be correlated with the lesion selected for sampling.

| Clinical mimic           | Pathologic concern                |
|--------------------------|-----------------------------------|
| Chronic eczematous patch | Atypical lymphocytes in epidermis |

## Psoriasis-like plaques require tissue correlation

- Scale and erythema occur in both disorders.
- Raised plaque morphology alone does not establish a neoplasm.
- In MF, lymphoid epidermotropism and cytologic atypia are key clues.

### Psoriasiform surface

Scale and thickening

### MF-specific concern

Atypical  
epidermotropic T-cell  
infiltrate

## Repeated evaluation may be necessary in subtle cases

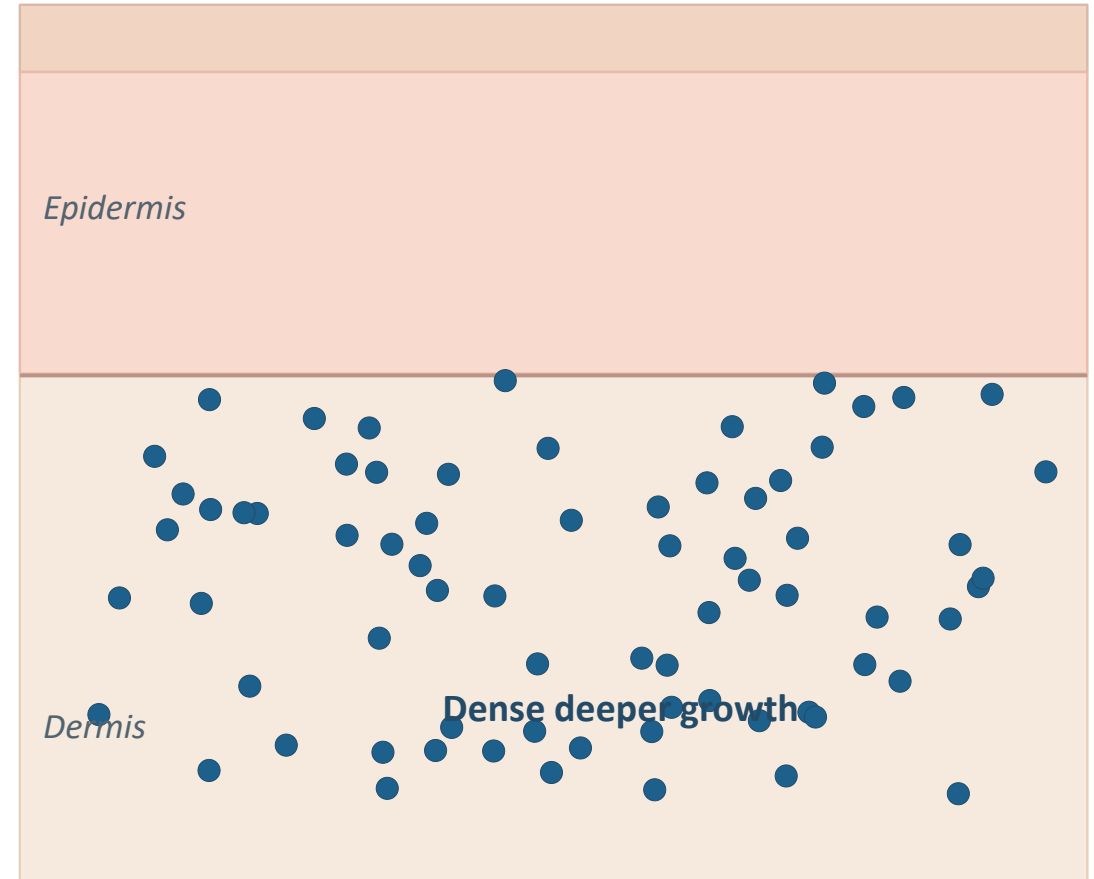
- Early patch-stage histology may be difficult to classify conclusively.
- Review of persistent lesions and additional biopsies can be necessary.
- An equivocal sample should not be forced into a definitive diagnosis.



*Time and progression are variable*

# Recognize a diagnostic pitfall in tumor disease

- Tumor-stage lesions can lose classic epidermotropism.
- Relying only on epidermal infiltrates risks overlooking advanced disease.
- Clinical evolution and dermal lymphoid morphology remain relevant.



*Schematic (not histology)*

## Sézary syndrome: A related but distinct entity

- Both diseases involve skin-homing CD4+ T-cell neoplasms.
- Sézary syndrome has generalized erythroderma, blood involvement and nodes.
- The clinical courses and patterns of tumefaction differ.

| <b>Mycosis fungoides</b>             | <b>Sézary syndrome</b>          |
|--------------------------------------|---------------------------------|
| Patches, plaques,<br>possible tumors | Erythroderma + blood<br>+ nodes |

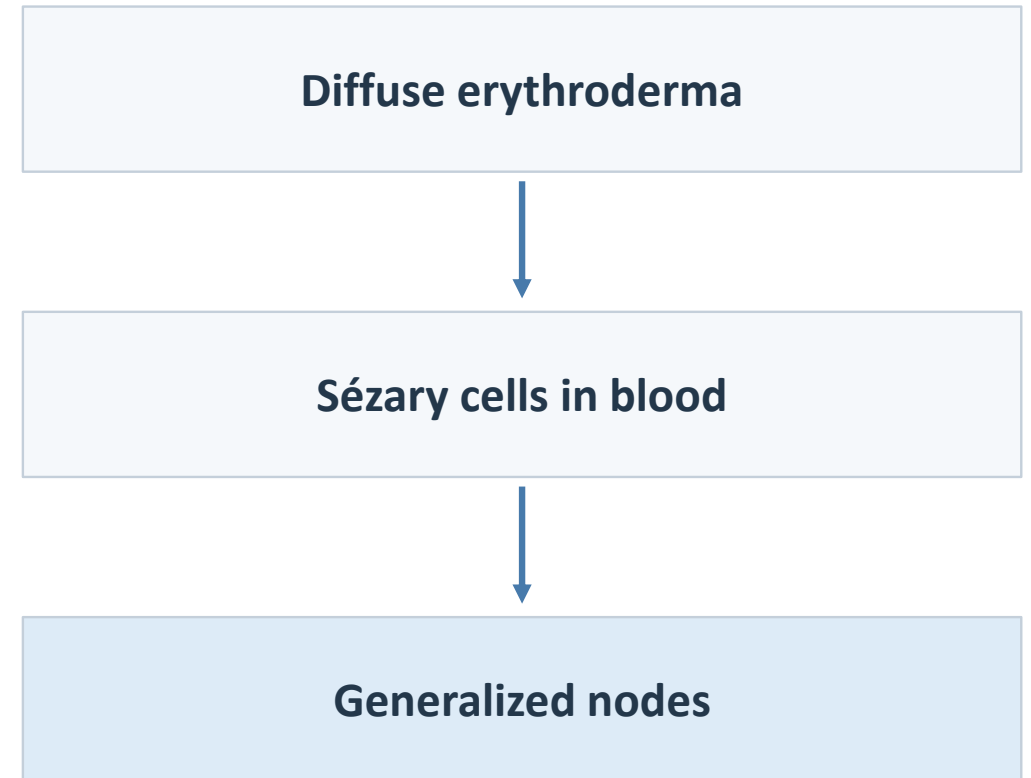
## Generalized erythroderma defines the skin picture in Sézary

- Sézary syndrome typically manifests with diffuse exfoliative erythroderma.
- This differs from the usual evolution of discrete MF lesions.
- Generalized lymphadenopathy commonly accompanies it.

**Erythroderma +  
lymphadenopathy**

## Blood involvement distinguishes Sézary syndrome

- Sézary syndrome includes leukemia of circulating neoplastic cells.
- These cells characteristically display cerebriform nuclear contours.
- Blood findings are interpreted together with skin and nodes.



*Schematic (not histology)*

# Mycosis fungoides versus Sézary syndrome

| Feature        | Mycosis fungoides                      | Sézary syndrome                  |
|----------------|----------------------------------------|----------------------------------|
| Skin           | Patch → plaque → tumor                 | Diffuse exfoliative erythroderma |
| Blood          | Sensitive tests may detect cells early | Overt leukemic involvement       |
| Tumors         | May develop                            | Skin tumefaction uncommon        |
| Overall course | Often indolent                         | Typically more aggressive        |

## Early cell circulation does not equal overt systemic disease

- Sensitive analyses can detect MF cells in blood, nodes or marrow early.
- Clinically evident extracutaneous disease usually develops much later.
- This distinction reconciles biological trafficking with cutaneous indolence.

### Sensitive detection

Tumor cells may  
circulate

### Overt dissemination

Usually a late clinical  
event

# Outcome in mycosis fungoides is variable

- MF often progresses indolently with prolonged skin localization.
- Chapter 13 reports a median survival of approximately 10 years.
- Extent, tumor nodules and systemic involvement modify prognosis.

**Limited skin burden**

**More extensive skin burden**

**Tumors or systemic involvement**

*Qualitative risk relationships — no numeric cut-offs implied*

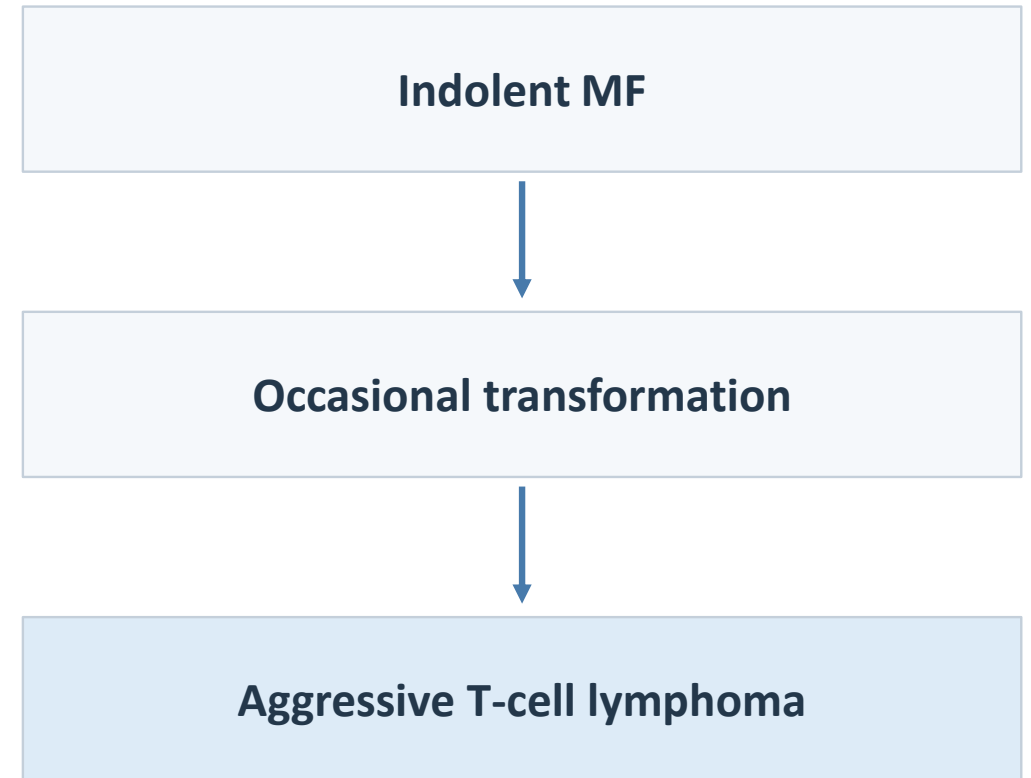
## Sézary syndrome generally has a worse course

- Chapter 13 characterizes Sézary syndrome as more aggressive than MF.
- It reports that only a minority remain alive five years after diagnosis.
- Population-level statements cannot predict an individual patient.

| MF                               | Sézary                            |
|----------------------------------|-----------------------------------|
| Often prolonged cutaneous course | More aggressive systemic behavior |

# Aggressive transformation can occur late

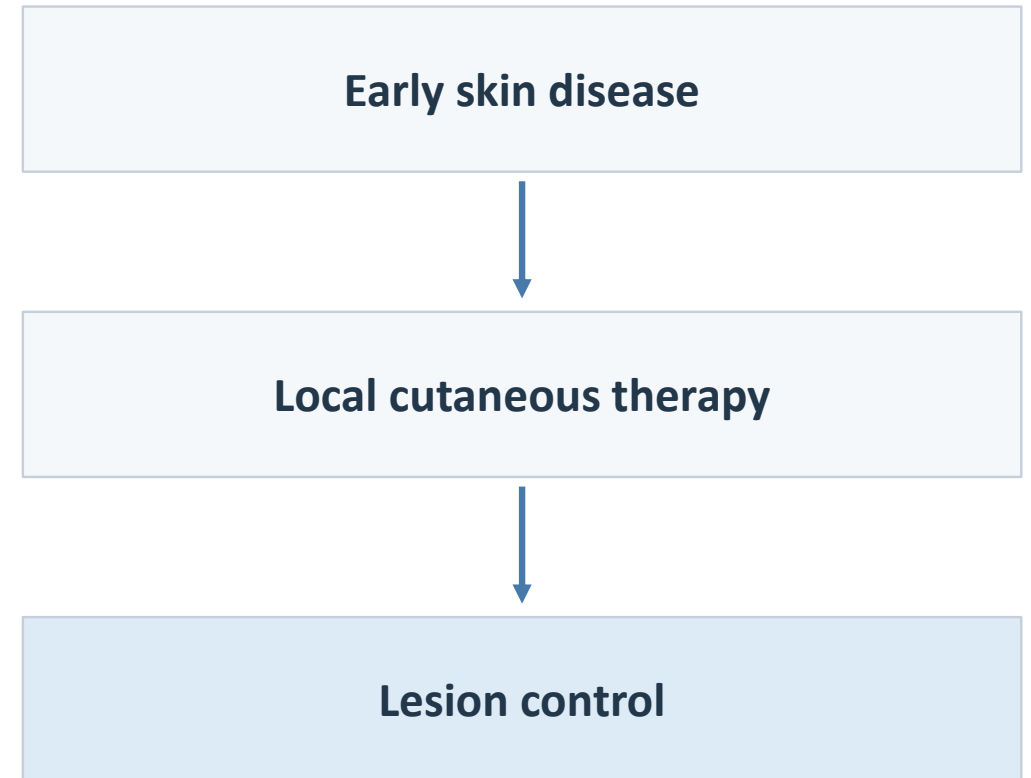
- MF occasionally evolves into aggressive T-cell lymphoma.
- This as a possible terminal event.
- New aggressive clinical changes warrant reconsideration of disease biology.



*Schematic (not histology)*

# Skin-directed therapy targets early lesions

- Localized patches and plaques are commonly managed with skin-directed measures.
- Topical steroids and ultraviolet light.
- The rationale is disease largely confined to cutaneous compartments.



*Schematic (not histology)*

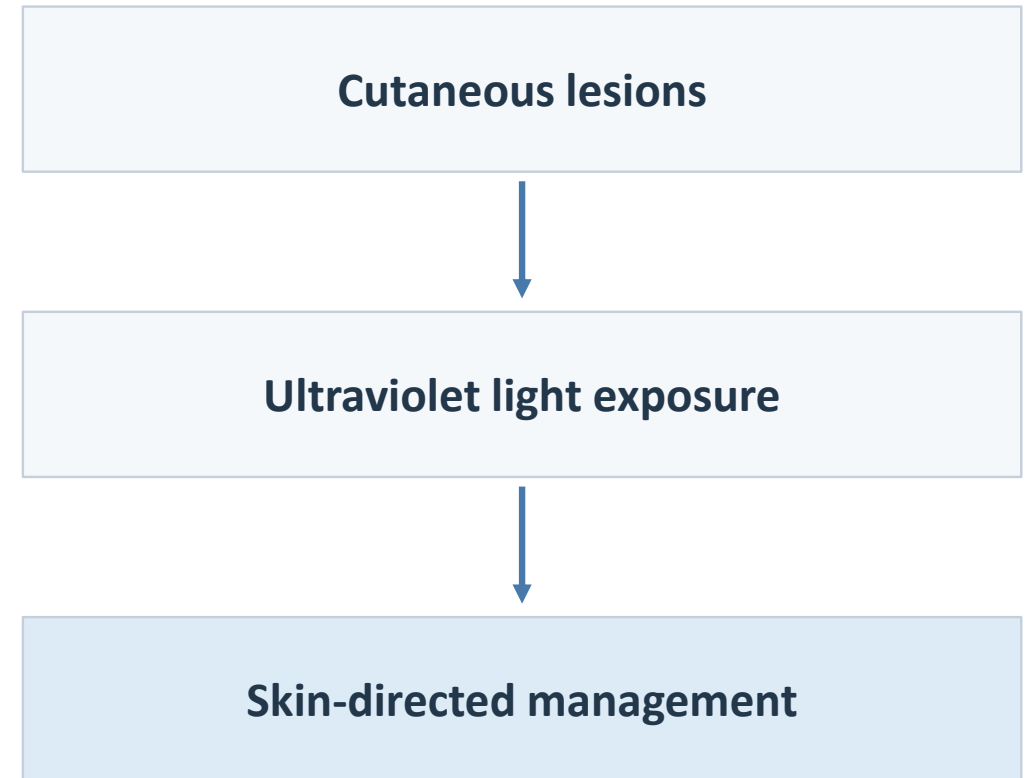
## Topical corticosteroids: Early-lesion approach

- Topical steroids as a treatment option for early MF.
- The treatment acts at the site of visible disease.
- The text does not specify regimens or response percentages.

| Setting            | Treatment modality      |
|--------------------|-------------------------|
| Early skin lesions | Topical steroid therapy |

## Ultraviolet therapy is another skin-directed option

- Ultraviolet light therapy is cited for early skin lesions.
- Phototherapy is distinct from systemic cytotoxic chemotherapy.
- Treatment choice must account for burden and stage.



*Schematic (not histology)*

## Advanced disease may require systemic treatment

- Systemic chemotherapy for more aggressive disease.
- Clinical tumor development or extracutaneous disease changes management.
- Therapeutic details beyond this principle require specialist guidance.

### Early skin disease

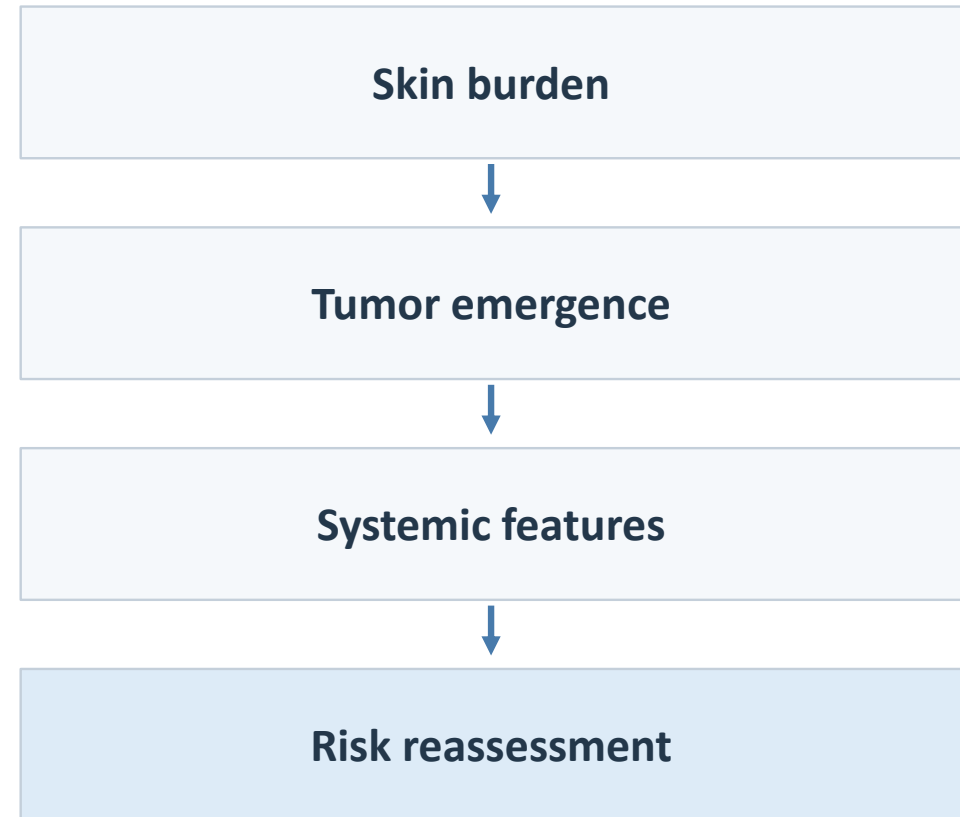
Topical / UV  
modalities

### Advanced aggressive disease

Systemic treatment

## Follow-up focuses on meaningful change

- Assess new plaque thickening, tumors, ulceration and broader skin involvement.
- Look for evidence of clinically overt extracutaneous disease.
- Clinical progression should trigger renewed clinicopathologic assessment.



*Schematic (not histology)*

# Case study: A persistent scaly patch

## Hypothetical teaching case

- A hypothetical patient aged 54 has a chronic red-brown, scaling truncal patch.
- The lesion has been described as eczema-like but persists.
- Question: Which tissue feature would most strengthen concern for MF?

## Case interpretation: Subtle clinical disease

### Reasoning

- Persistence and a truncal patch are compatible but not pathognomonic.
- Biopsy can disclose atypical epidermotropic T cells.
- Clonality and immunophenotype may support an integrated diagnosis.

**Clinical suspicion → representative biopsy → integrated interpretation.**

## Case study: Progression to a nodule

### Hypothetical teaching case

- A patient with long-standing scaling plaques develops a fungating nodule.
- The tumor contains a dense population of atypical cells deep in the dermis.
- Question: Must prominent epidermotropism still be present?

## Case interpretation: A changed tissue pattern

### Reasoning

- Nodule formation corresponds to greater dermal tumor burden.
- Epidermotropism may diminish in advanced nodular lesions.
- New tumors also alter assessment of prognosis and systemic spread.

**Deep dermal growth can dominate tumor-stage disease.**

# Morphology challenge: Identify the pattern

- Superficial dermal band plus intraepidermal atypical cells:
- What is the name for individual-cell epidermal migration?
- What is the name for small intraepidermal clusters?



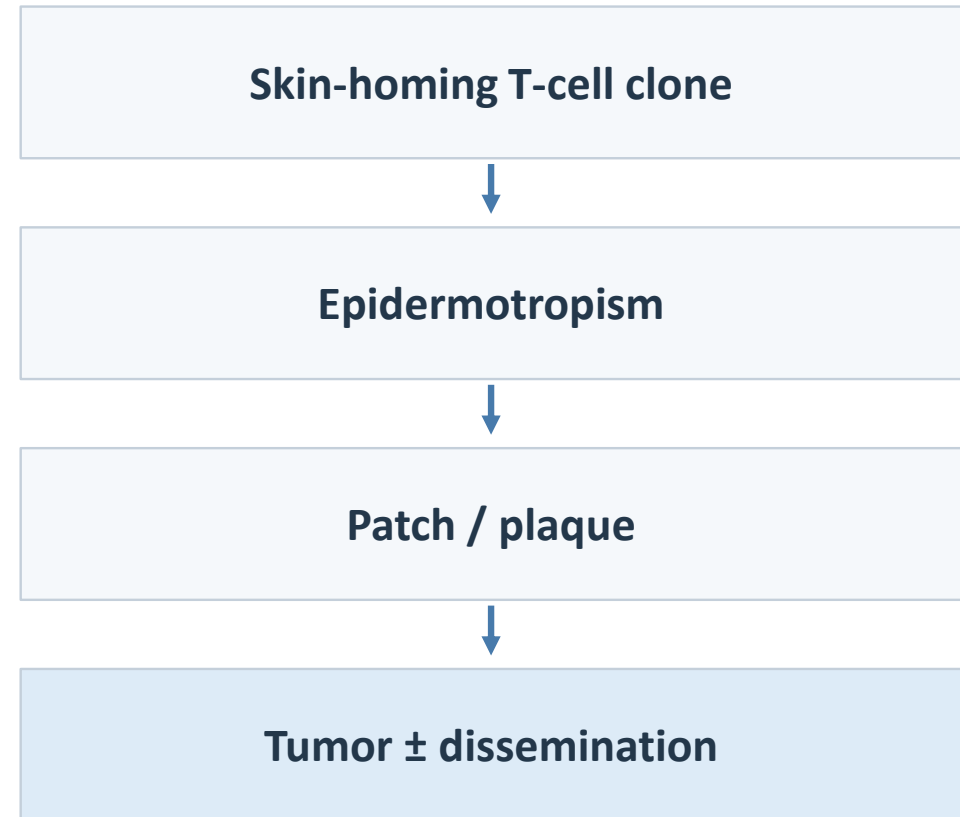
*Schematic (not histology)*

## Morphology challenge: Terms and interpretation

| Observation                     | Term                    | Interpretation                         |
|---------------------------------|-------------------------|----------------------------------------|
| Atypical cells within epidermis | Epidermotropism         | Neoplastic cells cross tissue boundary |
| Small intraepidermal clusters   | Pautrier microabscesses | Focal lymphoid aggregates              |
| Convolutated lymphoid nuclei    | Cerebriform contour     | Atypical cell morphology               |

# Integrating the disease in one mechanism

- Clonal, skin-homing CD4+ T cells underlie the lymphoma.
- Superficial dermal and epidermal infiltration produces early lesions.
- Progressive dermal growth and eventual spread explain advanced disease.



*Schematic (not histology)*

# High-yield discriminators

| If you see...                             | Think about...               | Do not forget...          |
|-------------------------------------------|------------------------------|---------------------------|
| Scaly chronic truncal patches             | Possible early MF            | Inflammatory mimics       |
| Superficial band + epidermotropism        | Characteristic MF morphology | Clinical correlation      |
| Deep dermal tumor; little epidermotropism | Advanced tumor-stage MF      | Epidermotropism can wane  |
| Erythroderma + blood disease + nodes      | Sézary syndrome              | Different clinical entity |

## Core teaching conclusions

- MF is a skin-homing, usually CD4+ clonal T-cell lymphoma.
- Clinical lesions classically evolve through patch, plaque and tumor patterns.
- Band-like dermal infiltrates, epidermotropism and Pautrier clusters are key clues.
- Tumor nodules and overt systemic spread indicate higher-risk disease.

## Source references

Robbins, Cotran & Kumar. Pathologic Basis of Disease. 11th ed. 2026. Chapter 25, pp. 1044–1046.

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