Topic Area:
Nursing Care of the Patient with Cutaneous T-cell Lymphoma

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Nursing Care of the Patient with Cutaneous T-cell Lymphoma

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Objectives

- DEFINE AND DESCRIBE CUTANEOUS T-CELL LYMPHOMA (CTCL)
- DESCRIBE THE ETIOLOGY, PATHOPHYSIOLOGY, & DIAGNOSTIC CRITERIA OF CTCL
- DIFFERENTIATE BETWEEN INDOLENT AND AGGRESSIVE CTCL
- IDENTIFY PRIMARY NURSING CARE PRIORITIES WHEN CARING FOR CTCL PATIENTS

Cutaneous T-Cell Lymphoma

- Heterogeneous group of non-Hodgkin's T-lymphocytic diseases (3.9% of all NHL)
- Skin is primary site
- Variable presentations:
    - skin, tissue, blood
    - histopathology
    - immunophenotype
    - responsiveness to therapy
    - prognosis

Cutaneous T-cell Lymphomas

- Mycosis Fungoides & Variants
  - Folliculotropic MF
  - Granulomatous Slack Skin
  - Pagetoid Reticulosis
- Sezary Syndrome
- Adult t-cell leukemia/lymphoma
- Lymphomatoid papulosis
- Primary cutaneous large cell anaplastic lymphoma and extranodal natural killer-like (nasal type)
- Primary subcutaneous panniculitis-like
- Primary CD4 small/pleomorphic T-cell

WHO-EORTC Classification by Behavior

Indolent

- MF & variants
- Primary CD 30+
- Lymphomatoid papulosis (LyP)
- Subcutaneous panniculitis like
- Primary CD4+ small/medium pleomorphic

Aggressive

- Sezary Syndrome
- Primary cutaneous
  - NK nasal type
  - CD8 +
  - Gamma delta
  - Peripheral t-cell lymphoma unspecified

*Based on Willemze, R. et al., 2005 Blood. 105(10). 3768-3785

Clinical Presentation: Lesion Morphology

- Patch—flat, non-indurated
- Plaque—raised, indurated
- Tumor—nodular, raised ~1 cm
- Erythroderma- ≥80% redness; +/- scaling
- Sezary Syndrome (SzS)—B2 blood involvement + skin involvement

Sezary Syndrome

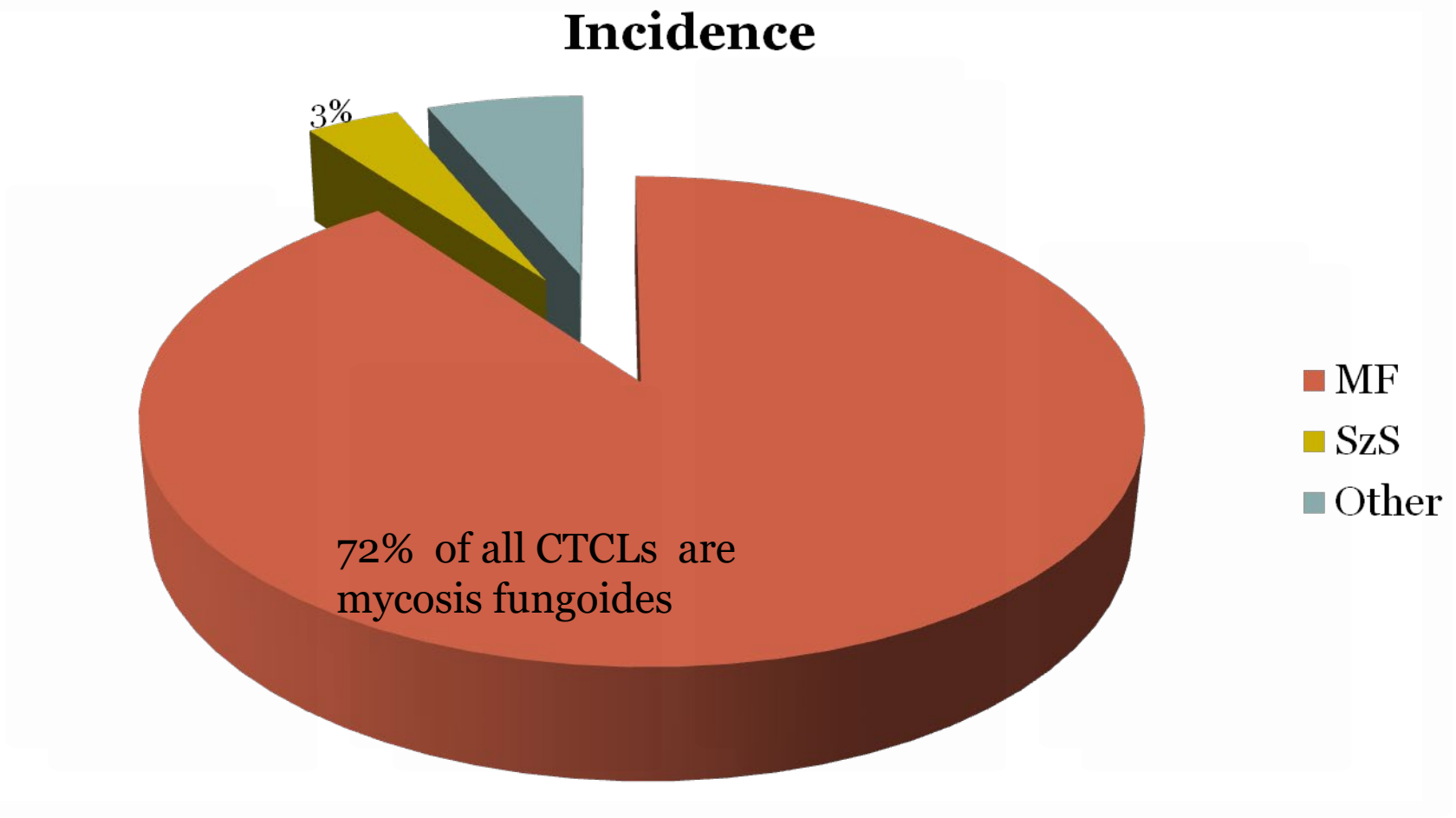
Historical Triad

- Diffuse erythema
- Lymphadenopathy
- Circulating Sezary cells

Signs and Symptoms

- Hyperkeratosis/fissuring
- Nail dystrophy
- Exfoliation/exudation
- Anemia
- Electrolyte imbalance
- albuminemia/edema
- Exfoliation/exudation
- Temperature dysregulation
- Pruritus
- Alopecia

Epidemiology of CTCL:2010



Criscione & Weinstock (2007) Incidence of CTCL in the U. S, 1973-2002. Archives of Dermatology 143(7), 854-859.

Epidemiology of CTCL

- Most common 5th to 6th decade
- 6.4 per million in U. S.
- Represents 2.9/million **increase** each decade since 1973
  - Geographic variances found increased incidence in areas with more physicians, increased income, higher education: related to better access to care???
- Male to female ratio is 2:1
- African American to caucasian is about 1.5:1

Criscione & Weinstock (2007) Incidence of CTCL in the U. S, 1973-2002. Archives of Dermatology 143(7), 854-859.

Etiology

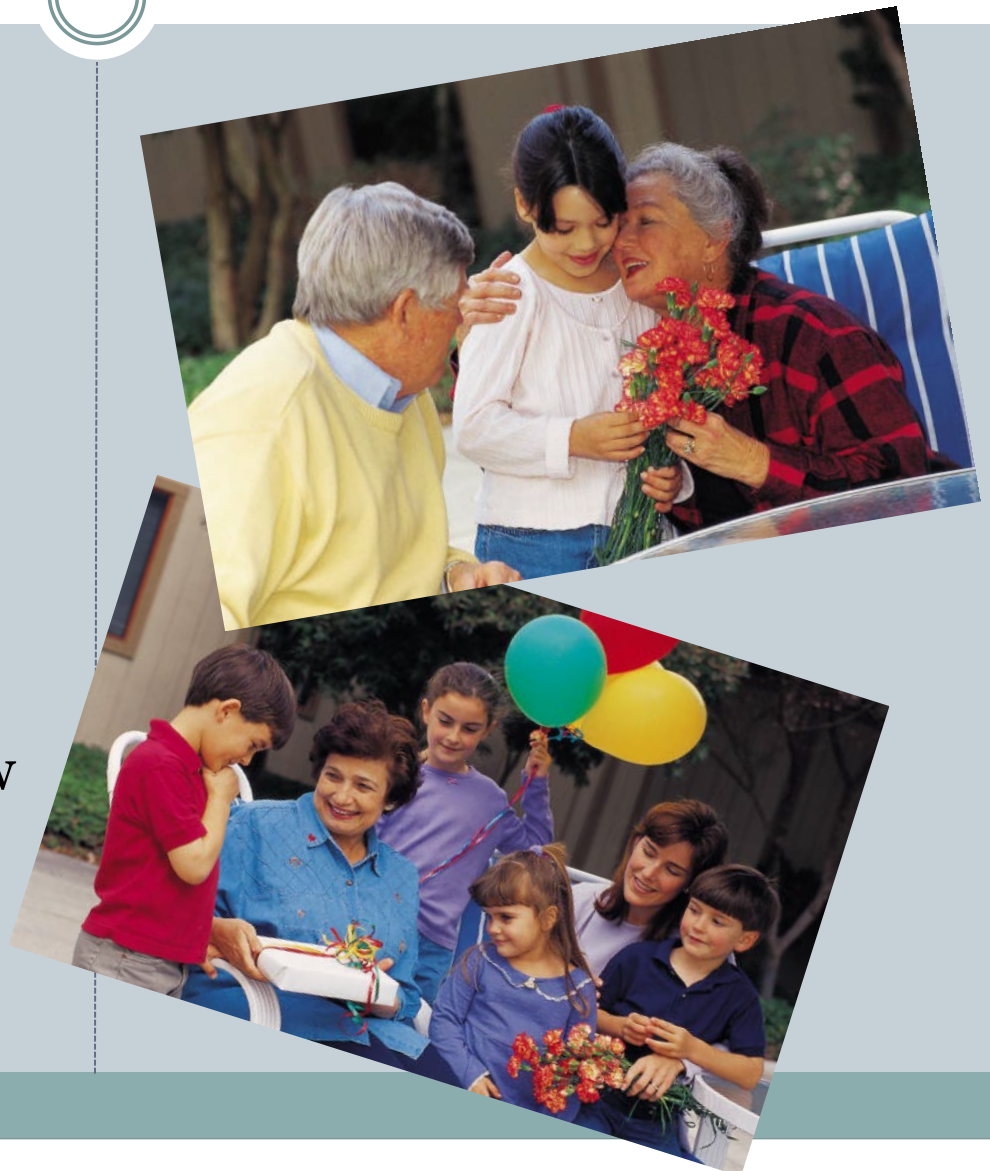
Cause is unknown

Theories:

- Environment
- Genetic
- Infections

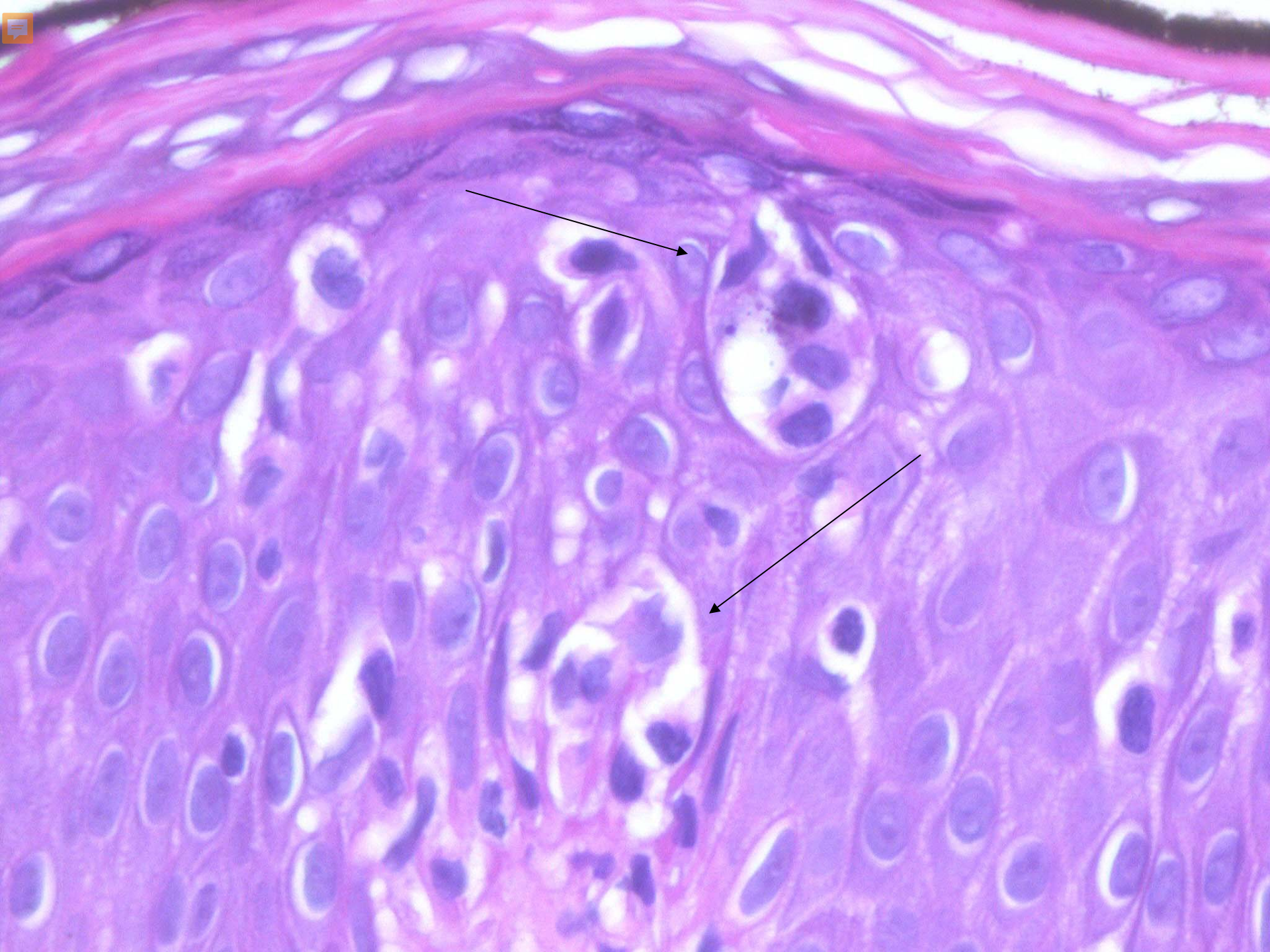
Etiology & Epidemiology: Questions & Concerns

- How did I get this disease?
- Did I do something to cause it?
- Does anyone else have it? No one I know has ever heard of it.
- Can I give it to anyone?
- Will my children or grandchildren inherit it?
- Will I die from it? If so, how will I die? (No really, HOW will I die?)



Pathophysiology

- Majority are malignancy of CD4+, CD45RO+ T-cells (memory cells)
- Clonal, mature T-cells home to epidermis and dermis, express cutaneous lymphoid antigen
- Pautrier's microabscess
  - pathogonomic collection of clonal malignant cells in epidermis
- Demonstrate loss of normal t-cell antigens: CD7, CD5, CD2, CD3, CD26



Pathophysiology

- Malignant cells release chemokines and cytokines such as IL4, IL5, IL6, IL10, IL13
- Results in clinical signs and symptoms i.e. scaling, induration, pruritus
- Inflammatory responses
- Proliferation of keratinocytes/skin lesions
- Immunosuppression

Diagnosis

- Clinical features and history
- Histopathology positive for superficial lymphoid infiltrate, atypia, epidermotropism
- Immunopathology demonstrating loss of pan t-cell markers (CD7, CD2, CD3, CD5)
- Molecular testing
  - + clonal gene rearrangement
- Hematology and chemistry lab work
- Flow cytometry
- Lymph node biopsy
- CT scan/PET scan
- Bone marrow biopsy

ISCL Diagnostic Criteria of Sezary Syndrome

- Absolute Sezary count 1000/mm3
- CD4:CD8 ratio > 10:1
- Positive clonality in blood by TCR PCR gene rearrangement
- Increased abnormal pan T-cell markers

  CD 32, CD3, CD4, CD5, CD26

Vonderheid E. et al. (2002). JAAD 46(1) 95-106.

Diagnostic Considerations

- Multiple mimickers of CTCL: psoriasis, eczema, large plaque parapsoriasis, drug eruptions, vitiligo, tinea, contact dermatitis
- Multiple biopsies may be required

- Variable lesion morphology:
  - Single or multiple lesions
  - Annular, arcuate, nummular, psoriasiform, eczematous, unilateral, regional (face, bathing suit area, generalized)
  - Coloration, hyper/hypopigmentation

Diagnosis: Patient Questions & Concerns

- Anger/denial/grief cycle
  - If only I were diagnosed sooner, I wouldn't be in this mess; I would be cured by now
  - Self-blame or anger at healthcare team
- Reassurance
  - Delayed diagnosis is a common occurrence
- Re-focus to current status and situation
- Help patient move toward acceptance of illness, current and future treatment plans

Staging: TNMB

- **T**umor burden in skin
- **N**odal involvement
- **M**etastatic involvement
- **B**lood involvement

T1	**< 10% BSA patch or plaque**
T2	> 10% BSA patch or plaque
T3	Tumor(s)
T4	Erythroderma

Staging: TNMB

- **T**umor burden in skin
- **N**odal involvement
- **M**etastatic involvement
- **B**lood involvement

No	No abnormal nodes
N1	Dermatopathic node
N2	Early involvement with MF
N3	Partial to complete effacement with atypical cerebriform cells

Staging: TNMB

- **T**umor burden in skin
- **N**odal involvement
- **M**etastatic involvement
- **B**lood involvement

Mo	**No systemic involvement**
M1 (stage IVb)	Systemic involvement with metastasis more commonly to lungs, liver, spleen, CNS

Staging: TNMB

- **T**umor burden in skin
- **N**odal involvement
- **M**etastatic involvement
- **B**lood involvement

B0	**< 5% Sezary cells**
B1	> 5% Sezary cells but < 1000 Sezary cells/mm3
B2	> 1000 Sezary cells/mm3, + clonality OR elevated CD4:CD8 ratio > 10:1

CTCL Staging System

Stage	Clinical picture	Skin	Nodes	Mets	Blood
IA	<10% patch or plaques	T1	No	Mo	0,1
IB	> 10% patch or plaques	T2	No	Mo	0,1
IIA	Up to 100% BSA	T1,2	N1,2	Mo	0,1
IIB	Tumors	T3	0-2	0	0,1
IIIA	Erythroderma	T4	0-2	0	0
IIIB					1
IVA1	Any	T 1-4	0-2	0	2
IVA2			3	0	0-2
IVB			0-3	1	0-2

Olsen, E. et. al. (2007) Blood

Nursing Care Issues

- Altered skin integrity
- Pruritus
- Homeostasis
- Pain
- Infection
- Psychosocial
- Activities of daily living
- Knowledge deficit (disease process, therapeutic options, chronic disease management)

Patient Care: Altered Skin Integrity

- Teach basic skin care
  - Self assessment for infection, fissures
  - Bathing techniques
    - Soak & seal
    - Antibacterial baths
  - Injury avoidance (finger nails, tape removal)
  - Topical therapy application
- Maintain nutrition and hydration
- Elevate edematous extremities

Patient Care: Pruritus Management

- Antihistamines
  - Sedation side effects & precautions
- Moisturization
  - Home humidification
- Interrupt scratch/itch cycle
- Oatmeal baths
- Wet wraps
- Mentholated/cooling topicals
- Non-irritating clothing
- Distraction techniques

Patient Care: Maintaining Homeostasis

- Cold intolerance
  - Warm, draft-free environment
    - Avoid exposure to extreme temperatures
      - Inability to sweat in some patients
  - Low grade fever may indicate serious infection
  - Layer clothing
- Metabolic/laboratory abnormalities
  - High protein, high quality carbohydrates
  - Nutritional supplements, iron, vitamins

Patient Care: Managing Pain

- Asses type and quality of pain
  - Tenderness, burning, throbbing, pressure sensitive (affects clothing choices)
- Treat for infection if present
- Pain relief measures
  - Medications
  - Moisturization
  - Special dressings, beds
  - Topical applications
  - Position change, distraction, massage, warm/cool compresses

Patient Care: Preventing Infection

- Super-infection with bacterial toxins
  - Erythrodermic and SzS problematic
  - Stimulates malignant T-cells—exacerbates symptoms
- Vigilant assessment and skin care
  - Topical antibiotics to open wounds
  - Chlorox/Vinegar baths
  - Phisohex showers
- Topical antibiotics: open wounds, test/treat bacterial carriers with mupirocin
- Oral antibiotics-culture skin as appropriate
- **Avoid central lines**

Patient Care: Psychosocial Issues

- Emotional distress: anger, denial, depression
  - Strategies to cope with chronic illness and cancer, referrals, support networks, medications
- Self image & sexuality
  - Feel trapped within own body, embarrassment, social stigma/misunderstanding with skin disease
  - Strategies to help
    - Take time to know the "real" person, positive reinforcement, discuss clothing choices, cosmetic camouflage, wigs, skin care to reduce symptoms
- Socioeconomic concerns

CLF List-Serve Quote

" After years and years of treatment, I am so tired of everything!! I mean I've been on PUVA, Targretin, Interferon….but nothing has ever put me in remission…..I'm sick of it!! All I want is a normal life. Do I demand too much?"

Patient Care: Addressing Activities of Daily Living

ADL Impairment	Impact	Strategy	Comments
Hyperkeratosis palms and soles	Decreased dexterity, fine motor skills and mobility	Consult OT for adaptive equipment and clothing	
Deconditioning	Weakness, at risk for falls	Consult PT for strengthening program	Assess independence, fall risk, need for visiting nurses
Extremity edema	Decreased mobility, comfort, increased risk of falls	Elevate extremities Compression management Adaptive shoes	Monitor albumin and nutritional status
Ectropion & dry eyes	Diminished visual acuity	Consult ophthalmology, artificial tears	Surgical repair may be option

Treatments: Overview

- Early stage (IA-IIA): skin directed, clinical remission
- Later stages (IIB-IVB): systemic and combination Rx, palliation and control, multidisciplinary approach
- Skin directed, systemic, combination in setting of chronic disease state
- Previously failed therapies may be re-tried
- Patient's goals, socioeconomic issues, and QOL must be considered when choosing therapy

Patient Advocacy: What is IT?

- Facilitating communications with care team
- Assisting with navigation through health care system, treatment maze, insurance issues, and multidisciplinary care
- Ensuring educational needs are met
- Connecting to available resources
  - Financial
  - Social services
  - Patient groups: Cancer Care, ACS, Leukemia & Lymphoma Society, Cutaneous Lymphoma Foundation
- Connecting to support systems
- Advocating for patient rights and the right thing in all circumstances

Patient Advocacy: How To Do IT?

- Ask the question
- Make the suggestion
- Repeat the education
- Initiate the referral
- Facilitate a connection
- Permit the second opinion

Education, Financial, and Social Support

- Cutaneous Lymphoma Foundation
- Lymphoma Research Foundation
- Leukemia & Lymphoma Society
- American Cancer Society
- Pharma Patient Assistant Programs
- Triage Cancer

Patient Care: Quality of Life

QOL Aspect	% Indicating Importance
Worry about dying	80
Feel unattractive	62
Bothered by skin redness	94
Bothered by itching	88
Bothered by pain	41
Tired due to disease	66
Feel financially burdened	61
Sleep affected	66

- **Demierre, M. et al. Significant impact of CTCL on patient's quality of life: results of a 2005 national cutaneous lymphoma foundation survey. Cancer 2006 (930 surveyed with 68% response rate).**

Quality of Life: Patient Quote

"I have often joked that of all the cancers to have, this isn't a bad one since I should still die of old age. It's the constant stress of having the "sword of damacles" hanging over my head that is starting to wear…."

Quality of Life: Patient Quote

“And I too have felt the frustration of it all, still do get angry, sad and when I really feel like I am going to explode, I take my dogs out for a walk… and just let it out. I just yell as loud as I can to release the pain I feel inside…”.

Quality of Life: Patient Quote

"…I am sometimes jealous that she has a cancer (breast cancer) that can actually be cut out and cured. What a strange thing to be envious of!"

Prognosis: Patient Questions & Concerns

- Anxiety producing discussions
- QOL survey found 80% of patients worry about dying*
- Early stage: reinforce excellent age-matched longevity statistics
- Advanced stage: reinforce new therapies, clinical trials, assist with interpretation of statistics in literature

* Demiere, M. (2006). Significant impact of CTCL on patient's QOL. Cancer (107)10:2504-11

Patient Care: Nursing Implications

- Education: Listen to questions
  - Demonstrate & discuss skin care techniques
  - Discuss therapy instructions, side effects, follow up
- Review information and reassure often
- Patients need your advocacy and influence
- Connect patient to information sources and others with disease
- Tune in to your patient's QOL issues

Conclusions

- Chronic, uncommon, *complex* systemic disease of immune system with **variable** presentations
- Best practice = interdisciplinary care
- Experienced centers for complex cases
- Nursing care is vital to success with patients
- Multiple therapeutic options but no one best option for all
- Research invaluable and ongoing
- Quality of life MATTERS

THANK YOU