PRIMARY CUTANEOUS B-CELL LYMPHOMAS: A PRACTICAL HIGH YIELD REVIEW

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T-CELL LYMPHOMAS

- **Mycosis fungoides**
- MF variants
- Folliculotropic MF
- Pagetoid reticulosis
- Granulomatous slack skin
- Sezary syndrome
- Adult T-Cell leukemia/lymphoma
- **Primary cutaneous CD30+ lymphoproliferative disorders**
  - Primary anaplastic large cell lymphoma
  - Lymphomatoid papulosis
T-CELL LYMPHOMAS

- Subcutaneous panniculitis like T-cell lymphoma
- Primary cutaneous gamma-delta T cell lymphoma
- Extranodal NK/T-cell lymphoma, nasal type
- Primary cutaneous T-cell lymphoma, unspecified
- Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma
- Hydra vacciniforme-like lymphoma
- Primary cutaneous acral CD8+ T-cell lymphoma
- Primary cutaneous CD4+ small/medium sized pleomorphic T cell lymphoma
- Angioimmunoblastic T-Cell Lymphoma
PRIMARY CUTANEOUS B-CELL LYMPHOMAS:

1. Primary Cutaneous Follicle Center B-Cell Lymphoma
2. Extranodal Marginal Zone Lymphoma of Mucosa Associated Lymphoid Tissue-MALT lymphoma (aka Primary Cutaneous Marginal Zone B-Cell Lymphoma)
3. Primary Cutaneous Diffuse Large B Cell Lymphoma, Leg type
4. Intravascular Diffuse Large B-cell Lymphoma
5. Precursor B lymphoblastic Lymphoma/Leukemia- Children/Young adults
6. Primary Cutaneous Large B-Cell Lymphoma, other
• **Primary cutaneous lymphomas** are malignant lymphomas confined to the skin at presentation only after complete staging procedures.

• Important because *6-10% of patients with systemic B cell NHL will develop cutaneous disease at some point in their illness*
DIAGNOSTIC WORK-UP

• CBC with differential and platelet count
• LDH
• Flow cytometry of peripheral blood mononuclear cells
• HIV
• CT chest/abdomen/pelvis or combined PET/CT. Include neck is palpable lymphadenopathy
• Unilateral bone marrow aspiration/biopsy if patient has cytopenia and for all patient prior to initiation of systemic therapy
A QUICK WORD ABOUT CLONALITY

• PCR commonly used to check for clonality
  • This can be performed on paraffin embedded tissue
• Monoclonality does not equal malignancy → have to take it in clinicopathologic context, also the absence of clonality does not exclude the presence of malignancy
• Monoclonal population can be identified when one or two dominant peaks substantially above that of the next highest background peak

Figure 3.1 The Immune System, Jed (© Garland Science 2009)
CASE #1

- A 75-year-old male presents with pink-orange nodule with telangiectasias located on forehead
Bcl-6+ neoplastic cells

+ monoclonal rearrangement in Ig heavy chain/ Ig kappa light chain

Primary Cutaneous Follicular Center lymphoma
PRIMARY CUTANEOUS FOLLICULAR CENTER LYMPHOMA

• Solitary or grouped plaques and tumors preferentially located on scalp/forehead or on the trunk (uncommon on legs)
• Tumor composed of neoplastic follicle center cells
• **Predominance of large centrocytes** (large cleaved cells) admixed with variable numbers of centroblasts (large, noncleaved cells with prominent nucleoli)
• **Histopathology patterns**: follicular, follicular and diffuse, or diffuse growth pattern
• B symptoms rare
• Lesions progress slowly → indolent clinical course → dissemination to extracutaneous sites uncommon (five-year survival rate at 95%)

**Tx:**
• Solitary lesions or lesions within one radiation field → radiation vs. surgical excision
• Extensive disease → rituximab
CASE #2

58-year-old male presenting with two well-circumscribed, erythematous nodules on the shoulder.
Primary Cutaneous Marginal Zone Lymphoma

Germinal centers highlighted with CD21 staining
PRIMARY CUTANEOUS MARGINAL ZONE-MALT LYMPHOMA (PCMZL)

- Primary cutaneous immunocytoma/primary cutaneous plasmacytoma are now grouped under PCMZL
- Look for recurrent pink-violet to red-brown papules, plaques, or nodules
- Lesions tend to favor upper extremity over lower extremity
- B symptoms not present
- Five-year survival at 98-100%

**Tx:**
- Solitary lesion/localized lesions → radiation therapy vs. surgical excision
- Asymptomatic multifocal disease → initial observation period
- Symptomatic multifocal disease → tx symptomatic lesions with IL triamcinolone, low dose radiation, or surgical excision rather than chemotherapy due to indolent nature of these tumors
- There is an association with Borrelia species infection European population → consider PCR testing and antibiotic therapy?
CASE #3

- A 78-year-old female with ulcerated red nodule on lower ankle presents to clinic
MUM1/IRF4

Primary Cutaneous
Diffuse Large B-Cell Lymphoma,
Leg type
PRIMARY CUTANEOUS DIFFUSE LARGE B-CELL LYMPHOMA, LEG TYPE

• Think about this when you see solitary or clustered, erythematous to red-brown or bluish nodules/tumors on distal aspect of leg in elderly women

• In 20% of patients tumors with similar morphologic and phenotypic features can arise in areas other than lower extremities, but still called Diffuse Large B-Cell Lymphoma, Leg type

• Unlike other cutaneous B cell lymphomas, these tumors commonly disseminate to extracutaneous sites

• 5-year survival of 40-50%

• **Tx:**
  - Rituximab + anthracycline based combination chemotherapy regimen (R-CHOP) followed by involved field radiation therapy
CASE #4

- 80 y/o male presents with indurated violaceous plaques on the thigh mimicking panniculitis
CD31+ staining of endothelial cells

Intravascular Diffuse Large B-Cell Lymphoma
INTRAVASCULAR DIFFUSE LARGE B-CELL LYMPHOMA

- Rare malignant proliferation of large B lymphocytes within blood vessels
- Most often systemic involvement (including CNS) from the onset and B symptoms common
- Look for indurated, erythematous or violaceous patches and plaques on trunk and thighs with prominent telangiectasias→ can look like panniculitis or vascular tumor
- Poor prognosis, aggressive course
- **Tx:**
  - R-CHOP systemic chemotherapy
Thank You!!!

That’s a Wrap!!!