Phlebology Board Review 2015

Lymphedema and Lipedema

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Disclosures

• None
Lower extremity swelling

1. Lymphedema

1. Lipedema

2. Hybrid swelling
Lymphedema
Physiology

- 90% interstitial fluid transported via veins
- 10% via lymphatics
  - High molecular weight protein
Lymph 101

- Afferent lymph vessels
- Subcapsular sinus
- Lymphoid follicle
- Medullary cords
- Efferent lymph vessel
- Capsule

Regional lymph nodes:
- Cervical nodes
- Tracheobronchial nodes
- Axillary nodes
- Aortic nodes
- Iliac nodes
- Inguinal nodes

Right lymphatic duct
- Internal jugular vein
- Jugular trunk
- Subclavian trunk
- Subclavian vein
- Bronchomediastinal trunk

Thoracic duct
- Aorta
- Cisterna chyli
- Intestinal trunk
- Lumbar trunk

Lymphatic collecting vessels
Lymphedema

- Lymph stagnation occurs when lymph flow decreases by 80%
- High protein edema: 1.1 to 5 g/ML
  - Lack lymph channel
    - Congenital absence
    - Surgical resection
  - Lymph channel obstruction
    - Malignancy
    - Fibrosis
  - Phlebolymphtedema and similar phenomenon:
    - Abnormal venous systems overwhelms a normal lymphatic system
Inflammation

- Macromolecules
- RBC extravasation
- RBC degradation fibrin+hemosiderin
- Activated Mast cells
- Activated Macrophages
- Leukocyte adhesion and margination and diapedesis
- Endothelial –Leukocyte activation E-selectins L-selectins
- EC activation ICAM-1, VCAM-1, MCP-1, MIP-1β expression
- TGF-β1
- Fibroblast
- Provisional matrix Collagen-ECM TGF-β1
- Proteolytic activity Collagen-ECM degradation
- IL-1, TNF-α, MMPs
ISL Stages of Lymphedema

1. Pitting
   Elevation of limb reduces swelling
   <20% increase in limb volume

2. Elevation of limb does not reduce swelling
   Pitting is present in early Stage II
   Pitting is absent in late Stage II due to tissue fibrosis
   20% to 40% increase in limb volume

3. Lymphostatic elephantiasis
   Pitting is absent
   Trophic skin changes present
   >40% increase in limb volume

Exaggerated skin creases

Hyperkeratosis

Squaring

Hypoplastic nails
“Ski Jump Nails” = Primary Lymphedema
• Inability or failure to pinch or pick up a fold of skin at the base of the 2nd toe
• Positive = lymphedema
• Negative = lipedema
Peau d’orange

11.06.2012

Courtesy of Steve Dean DO
ISL Stage III Lymphedema: Elephantiasis

Cobblestoned Appearance
ISL Stage III Lymphedema: Elephantiasis (Nostras Verrucosa)

- Severe, non pitting edema
- Hyperkeratosis
- Fibrosis, induration, nodules
- "Cobblestoning"
- Papillomas/tumors
- Ulcerations

Massive Localized Lymphedema [MLL]

- “Pseudotumors”
- “Pseudosarcomas”

Brewer 2011 *Ann Plast Surg* - 41 pts; average weight 421 lb [160-619]
Diagnosis

• Physical and clinical diagnosis
• Imaging exams may be performed
  – Venous duplex
  – MRI or CT of abdomen and pelvis
  – Nuclear lymphoscintigraphy
  – Near-infrared fluorescence (NIRF) molecular imaging
  – Indocyanine green dye
Decongestion: Manual lymphatic drainage
Lipedema
Lipedema

- Grossly enlarged buttocks, thighs, calves
- Spares the foot; stops at the ankle- "ankle cut-off" sign
- Bilateral & symmetrical
- Torso relatively normal; disproportionate lower extremity involvement
- Non-pitting; tender; soft
- Easy bruising
- Frequently misdiagnosed as lymphedema
## Stages of Lipedema

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>Stage I</td>
<td>Skin surface normal, nodular (small) fatty tissue structure</td>
</tr>
<tr>
<td>Stage II</td>
<td>Skin surface uneven (peau d’orange), nodular (big) fatty tissue structure</td>
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<tr>
<td>Stage III</td>
<td>Lobular deformation due to increased fatty tissue</td>
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</tbody>
</table>
Lipedema: Stage I

Smooth
Lipedema: Stage II

Nodular
Stage III Lipedema

Lobular
“Hybrid” or Mixed Lower extremity Swelling
Phlebo-lymphedema
Phlebolymphedema

Dean SM, Ann Vasc Surg. 2013 Dec 11
Lipo-lymphedema
Lipo-phlebedema
What is the cause of the patient’s swelling?