Imaging and Management of Chylous Leak

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Chylous Leak
• Not a disease
• Sign of a more extensive disruption of central lymphatic channels
• “Lymphatic Hypertension”
• Crucial to differentiating various types of lymphatic anomalies

Lymphatic Anomalies
• Cystic “common” forms
• Central Conducting Lymphatic Anomalies (CCLA)
• Gorham-Stout Disease (GSD)
• Generalized Lymphatic Anomaly (GLA)
• Kaposiform Hemangioendothelioma (KHE)
• Kaposiform Lymphangiomatosis (KLA)
• Others: KTS, CLOVES, Noonan, lymphedema, etc.

Lymph vs. Chyle
• Chyle exists only in the “Chylous Circulation” – Lacteals, intestinal trunk, cisterna chyli, thoracic duct.
• Chyle leak = Disruption of central conducting lymphatics by injury, invasion or anomalies (e.g. Central Conducting Lymphatic Anomalies (CCLA)

Cystic “common” forms
• Macrocystic, Microcystic and Combined
• Contains lymph, not chyle.
• Old names: Cystic hygroma, lymphangioma

Lymphatic Malformation

Central Conducting Lymphatic Anomalies (CCLA)
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- Central channels
  - Perivascular
  - Lumbar trunks, cisterna chyli, thoracic duct
  - Their branches along axial skeleton
- "Truncular" types in Hamburg classification
- Often confused with other extensive types
- Malformations, tumors or injury

Central Conducting Lymphatic Anomalies (CCLA)

- Clinical hallmark: Chyle Reflux and Leak
  » Chylothorax, chylopericardium, chyloperitoneum, chyluria, etc.
  » Pulmonary Lymphangiectasia
  » Intestinal Lymphangiectasia:
    » Protein-losing enteropathy (PLE)
  » Bone involvement:
    » Dilated channels perforating the cortex, intact bone.
  » Nutritional, hematological and immune deficiencies
  » Lymphedema

Central Conducting Lymphatic Anomalies (CCLA)

- Pathologic hallmark: Lymphangiectasia, poor flow
  » Perivascular, retroperitoneal, mediastinal, neck
- Lymphatic dysfunction
  » Colloids and fluid leak
  » Cellular leak (lymphocytes)
  » Accumulation in interstitium, around airways, vessels
  » Loss of nutritional, hematological and immune factors
- Morbidity:
  » Recurrent infection
  » Fibrosis and respiratory failure

Central Conducting Lymphatic Anomalies (CCLA)

- Imaging findings:
  » Hallmark: Thick perivascular tissue
  » Replacement of retroper.-mediastinal fat
  » Interstitial and organ fluid
  » Chyle leak

Normal perivascular space: Fat

Perivascular tissue. Chyloous ascites
Central Conducting Lymphatic Anomalies (CCLA)

Gorham-Stout Disease (GSD)

Generalized Lymphatic Anomaly (GLA)
US of the Thoracic Duct

Conventional Lymphangiography

Intranodal Injection

Dynamic Intranodal Lymphangiography

Lipiodol

Node

Node
Dynamic Intranodal Lymphangiography

Abdominal Compression

Dynamic Intranodal Lymphangiography Augmentation with Saline

Congenital chylous leak

Courtesy of Ryan Arnold
Lymphatic duodenal reflux/fistula

CT Correlation

Non-conservative Treatment

- Embolization
  - Non-selective
  - Selective
- Surgical reimplantation of the TD
- Medical
  - Sirolimus: Limited benefit

Embolization of chylous leak

- Iatrogenic cause
- Target: TD or CC below leak
- Guided by lymphangiography
- Simple tools

**Embolization of chylous leak**

- Lymphangiography: identify leak
- Access cisterna chyli under fluoro (22-g Chiba) via anterior abdomen
- Guide wire (e.g. V18).
- Microcatheter (e.g. Rapid Transit, Renegade STC)
- Confirm leak
- Embolize TD below leak: coils or glue

**Embolization of chylous leak**

- General rules
  - Avoid embolization of TD in CCLA or congenital chylous leak
  - Promptly utilize the short window of lymphatic opacification
  - Only contraindication: Right to Left shunt
Thoracic Duct Embolization

Selective Embolization
- Iatrogenic, Congenital, secondary LH
- Target: Refluxing branch
  - Renal
  - Hepatic
  - Pulmonary: Plastic bronchitis*


Future of Lymphatic Imaging and Intervention: Unlimited

FOXC2 Mutation