Phlegmasia Cerulea Dolens - A Limb Threatening Problem
Disclosures
Complications of DVT

• Early
  – Pulmonary Emboli
    • Life threatening “saddle embolus”
    • Incidence of PE is estimated to be approximately 60 to 70 per 100,000
    • PE is found in up to 50% of DVT patients (Silent PE)
Complications of DVT

- Early
  - What else??????

Phlegmasia Alba Dolens

Phlegmasia Cerulea Dolens

Venous Gangrene
Complications of DVT

- Late
  - PTS (Post Thrombotic Syndrome)
    - Leg swelling
    - Leg pain
    - Skin discoloration
    - Varicosities
    - Venous ulceration
Early Complications of DVT

1. Phlegmasia Alba Dolens
2. Phlegmasia Cerulea Dolens
3. Venous Gangrene
Phlegmasia Alba Dolens

- "Alba" means white (i.e. Albino)
- Referred to as "Milk Leg or White Leg"
- Historically seen in pregnant women (third trimester) or mothers who had just given birth
  - Compression of Lt iliac vein against the pelvic rim from an enlarged uterus
- Presently it is due to venous occlusion (DVT)
  - 40% of patients with phlegmasia alba dolens have an underlying malignancy
Pathophysiology

- **Phlegmasia “Alba” Dolens**
  - Thrombosis involving *ONLY* the major deep venous channels but *SPARING* the collateral veins
  - Venous drainage is decreased but still present via the superficial channels
  - At this stage the leg is painful, swollen and appears white (pale)
    - But no arterial compromise (either palpable or dopplerable pulses)
Pathophysiology

Phlegmasia Alba Dolens
- Venous Pressure
- Edema
- Tissue Pressure

Phlegmasia Cerulea Dolens
- Venous Pressure
- Edema
- Tissue Pressure

Venous Gangrene
- Ischemia
Phlegmasia Alba Dolens
Diagnosis

• Phlegmasia “Alba” Dolens
  – Diagnosis is made with venous duplex
    • Identify Acute DVT
    • Location of DVT (proximal vs distal)
    • Extent of DVT
  – Contrast enhanced CT scan can be helpful
    • To identify thrombus is pelvic veins or IVC
    • Check for underlying malignancy
Diagnosis
Treatment

• Phlegmasia “Alba” Dolens
  – Treatment
    • Admit to inpatient
    • Anticoagulation (IV heparin gtt)
    • Elevate affected limb
    • Vascular surgery consultation
      – Medical management vs. Endovascular venous lysis (CDT, mechanical thrombectomy, pharmacomechanical thrombectomy)
Phlegmasia Cerulea Dolens

- Phlegmasia “Cerulea” Dolens (PCD)
  - “Cerulea” means Blue
  - Referred to as “Painful Blue Leg”
- Rare condition that occurs in < 1% of DVT patients
- Up to 90% of patients with PCD have underlying malignancy (50% occult malignancy)
Phlegmasia Cerulea Dolens

- 50% of patients with PCD progress to Venous Gangrene
  - 30-50% Limb Amputation rate
  - Overall Mortality rate of 20-40%
Pathophysiology

- Phlegmasia “Cerulea” Dolens
  - Thrombosis causing COMPLETE occlusion of venous drainage (deep and superficial system)
  - Leads to increase in capillary pressure
  - Leads to exudation of fluid into the interstitial space
  - Leads to skin blistering

- Characteristics of PCD
  - Pain, swelling and most importantly cyanosis “blue appearance”
Pathophysiology
Pathophysiology
Diagnosis

- Phlegmasia “Cerulea” Dolens
  - Based on clinical signs and symptoms
    - Painful, blue, swollen leg
  - Venous duplex to confirm the diagnosis and localize the thrombus (purpose of intervention)
  - Contrast enhanced CT to identify centrally located and pelvic thrombus
Plegmasia Cerulea Dolens
Treatment

- Phlegmasia "Cerulea" Dolens
  - **THIS IS AN EMERGENCY!!**
  - **PATIENT IS GOING TO THE OR FOR SURGICAL VENOUS THROMBECTOMY**
  - Fluid resuscitation
    - Patients are hypotensive and sometimes in shock due to fluid extravasation and loss of intravascular fluid
- Anticoagulation
  - IV heparin bolus prior to OR
Treatment

- Phlegmasia “Cerulea” Dolens
  - Systemic tPA vs Catheter directed thrombolysis
    - Literature does not support either therapy as being successful
    - Time is tissue
  - Some surgeons recommend a brief (6 hours) of IV heparin gtt with profound leg elevation
    - If unsuccessful then will proceed with surgical venous thrombectomy
Surgical Treatment

• Phlegmasia “Cerulea” Dolens
  – Local, Regional or General anesthesia
  – Longitudinal groin incision to expose CFV, GSV and SFA
  – IV Heparin intra op if not given preop (Check ACT)
  – Venotomy to facilitate Fogarty catheter thrombectomy
    • American surgeons – place IVC filter (contralateral groin) before
    • European surgeons – no filter, perform thrombectomy with positive pressure ventilation or Valsalva maneuver
Surgical Treatment
Surgical Treatment
Surgical Treatment

• Phlegmasia “Cerulea” Dolens
  – Place Fogarty balloon in Common Iliac vein
  – Pass suction catheter parallel to Fogarty and try to suction out the internal iliac vein thrombus
  – Must confirm iliac vein flow
    • MANDATORY VENOGRAM
  – If Iliac vein stenosis/compression (MTS) is noted then may need balloon angioplasty + stenting
Surgical Treatment
Surgical Treatment

• Phlegmasia “Cerulea” Dolens
  – Thrombus in infrainguinal region is expressed manually using an Esmarch
  – Start wrapping at the base of the toes and proceed proximally to the groin incision
    • Passing fogarty distally will damage venous valves
Surgical Treatment
Surgical Treatment

• Phlegmasia “Cerulea” Dolens
  – Once the venous outflow is restored the venotomy is closed
  – An autogenous AVF is created in groin to increase iliac vein patency
    • GSV is divided and proximal end is anastomosed to SFA
  – Perform a 4 compartment lower leg fasciotomy
Surgical Treatment
Surgical Treatment

DOUBLE-INCISION LEG FASCIO TOMY

Used to adequately decompress all four compartments
Surgical Treatment

• Phlegmasia “Cerulea” Dolens
  – What if there is thrombus extending into the IVC?
    • Transperitoneal incision
    • Expose IVC below renal veins to bifurcation
    • IVC is opened and the thrombus is removed
Post Operative Treatment

- Phlegmasia “Cerulea” Dolens
  - Continue IV heparin drip x 5 days
  - Then transition to oral anticoagulation (coumadin, xarelto, eliquis) x 6 months
  - Gradient Compression Stockings
  - Once swelling decreased, return to OR to close fasciotomy sites or heal with secondary intention
  - Ligate groin AVF in 6-12 weeks
Venous Gangrene

- Venous Gangrene
  - Massive iliofemoral or IVC occlusion with extensive vascular congestion and venous ischemia
  - Thrombosis causing COMPLETE occlusion of venous drainage (deep and superficial system) AND ARTERIAL COMPROMISE
  - 50% of phlegmasia cerulea dolens progress to venous gangrene
Pathophysiology

- Phlegmasia Alba Dolens
  - Venous Pressure
  - Edema
  - Tissue Pressure

- Phlegmasia Cerulea Dolens
  - Venous Pressure
  - Edema
  - Tissue Pressure

- Venous Gangrene
  - Ischemia
Venous Gangrene

• Clinical Features
  – Excruciating Limb pain
  – Severe edema
  – Blistering with fluid extravasation
  – Superficial gangrene and necrosis
  – No motor or sensory to foot
  – IRREVERSIBLE (Phlegmasia Alba and Cerulea Dolens are reversible)
  – Treatment is AMPUTATION
Venous Gangrene
Case Study

- 75 year old male presents to ED with LLE pain and swelling x 1 day
- Denies CP or SOB
- Pt recently diagnosed with Stage IV Lung cancer and received 2 doses of chemotherapy via a mediport
- Vital signs are stable
- O/E – LLE is edematous but NO PHLEGMASIA, Pedal pulses are palpable
Case Study

• Blood work is normal
• LLE venous duplex to r/o DVT
• Venous duplex demonstrated acute femoropopliteal and tibial occlusive DVT, no extension into external iliac vein
Case Study
Case Study

- What do you do?
- Patient was discharged from ED with Xarelto (starter pack) x 21 days
- Instructed to follow up with vascular surgery in the next week
- Keep LLE elevated
Case Study

• Patient returns to ED 2 days later complaining of worsening pain and swelling
• A different ED physician evaluates patient
• O/E – LLE is much more swollen and appears white
• Repeat LLE venous duplex is ordered and now demonstrates Acute Left external iliac vein, femoral vein, popliteal vein and tibial vein DVT
• Vascular surgery consult obtained
Case Study

- Vascular surgery evaluates patient and feels that patient has developed Phlegmasia Alba Dolens
- Pt is admitted to ICU
- Initiate IV heparin gtt
- CT A/P with contrast to better evaluate IVC and pelvic veins
- Keep LLE elevated
Case Study

- No IVC clot
- Clot extends to the Lt CIV
Case Study

• Discuss options of medical management (IV Heparin vs. Endovascular thrombolysis/thrombectomy)
  – Risk of bleeding from venous lysis is high due to stage IV lung cancer

• Decision made to continue IV heparin and perform serial exams with the understanding that if this progress to phlegmasia cerulean dolens then patient will need surgery
Case Study

- Next morning the LLE pain is worse and now has spots of bluish purplish areas (new)
Case Study

- Pt taken to OR for surgical venous thrombectomy
  - Able to clear iliac vein clot
  - Venogram was normal
  - Able to express infrainguinal clot
  - AVF created in left groin
  - LLE prophylactic fasciotomy performed

- Post op patient continued on IV heparin gtt
Case Study

- Over the course of next few days, the LLE swelling, pain and color improved
- Fasciotomy sites closed on POD #4
- Pt given GCS
- Discharged on coumadin x 6 months
Take Home Points

• Phlegmasia “Alba” Dolens – “White” leg
  – Medical management (IV Heparin) vs Endovascular therapy
  – Check for Malignancy (40%)

• Phlegmasia “Cerulea” Dolens – “Blue” leg
  – Limb threatening emergency
  – Mandatory open intervention
  – Check for Malignancy (90%)
Take Home Points

- Venous Gangrene – “Game Over”
  - Amputation
Thank You