May - Thurner, Nutcracker and Paget - Schroetter Syndromes

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Disclosure Statement of Financial Interest

I, Robert Schainfeld, DO NOT have a financial interest / arrangement or affiliation with one or more organizations that could be perceived as a real or apparent conflict of interest in the context of the subject of this presentation.
May - Thurner (MTS) or Cockett’s Syndrome

- 1851 - Virchow sinistral (left-sided) DVT
- Dx in 2% - 5% of pts w/ venous dz
- Right CIA/compresses the left CIV against the lumbar vertebrae
- 25% of asymptomatic pts > 50% stenosis
- 3rd – 5th decade of life / women
May - Thurner Syndrome
Magenetic Resonance Venography
MRV of Abdomen / Pelvis
MRV of Abdomen / Pelvis
“Palacios - Schainfeld Syndrome?”
Diagnosis of May - Thurner Syndrome

- MRV (>95%)
- Venography (66%)
- IVUS (>95%)
- CTV

May - Thurner Syndrome
Clinical Presentation

Unilateral – L >> R

- Pain
- Edema
- Chronic venous insufficiency
- Venous ulceration
- Ilio-femoral DVT
- Phlegmasia cerulea dolens

Cryptogenic stroke ??
May-Thurner Syndrome in Patients With Cryptogenic Stroke and Patent Foramen Ovale
An Important Clinical Association

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Background and Purpose—We aimed to investigate the incidence of May-Thurner syndrome in patients with cryptogenic stroke with patent foramen ovale.

Methods—This was a retrospective study. All consecutive patients with cryptogenic stroke having undergone patent foramen ovale closure from January 1, 2002, to December 31, 2007, at our institute were included in this study. Pelvic magnetic resonance venography studies of all patients were reviewed to determine if features of May-Thurner syndrome were present. Medical records and invasive venography studies of all patients were reviewed when available. All patients with May-Thurner syndrome features on magnetic resonance venography were reviewed by a vascular medicine specialist to define any previous incidence of deep vein thrombosis or any signs of chronic venous insufficiency. All patients also had lower limb venous duplex performed to rule out lower limb venous thrombosis.

Results—A total of 470 patients from January 1, 2002, until December 31, 2007, with cryptogenic stroke underwent patent foramen ovale closure at our institute. Thirty patients (6.3%) had features consistent with May-Thurner syndrome on magnetic resonance venography. These patients were predominantly female (80%) with a mean age of 43.6±11.9 years. Twelve patients (40%) had abnormalities in their laboratory thrombophilia evaluation and 13 females (54.1%) were taking hormone-related birth control pills. Only 2 patients had a history and signs of chronic venous insufficiency. All patent foramen ovales demonstrated right-to-left shunting on tranesophageal echocardiography. Atrial septal aneurysms/hypermobile atrial septa were present in 70% of patients with May-Thurner syndrome.

Conclusion—May-Thurner syndrome has an important clinical association with cryptogenic stroke and patent foramen ovale. (Stroke. 2009;40:1502-1504.)

Key Words: PFO ■ May Thurner syndrome ■ cryptogenic stroke ■ iliac vein thrombosis
Wallstents

- Stainless steel
- Self expanding
- High radial force
- Flexible
- Wide range of sizes
Endovascular Treatment (EVT) of Chronic Iliac Vein Obstruction

**Objectives**

Compare stent-related and clinical outcomes, results and complications of EVT in limbs with non-thrombotic iliac vein lesions and post-thrombotic syndrome

NIVL = Non-thrombotic iliac vein lesion (NIVL / MTS)

PTS = Post-thrombotic syndrome

Neglen et al., J Vasc Surg, 2007: 979-990
EVT of Chronic Iliac Vein Obstruction

**Materials/Methods**

- 870 patients / 982 limbs
- Chronic symptoms (mean = 60 mos)
- NIVL = 518, PTS = 464 limbs
- Follow-up (mean = 22 mos), range 1-107 mos
- IVUS
- PTA / stenting

Neglen et al., J Vasc Surg, 2007: 979-990
EVT of Chronic Iliac Vein Obstruction

Technical Results

- Technical success = 97%
- Post-op stent thrombosis (<30 d) = 1.5%
- Primary, primary-assisted and secondary patency rates (6-years)
  - 79%, 100%, 100% (NIVL)
  - 57%, 80%, 86% (PTS)
  - ISR (>50%) = 5%
    - 10% (PTS), 1% (NIVL)

Neglen et al., J Vasc Surg, 2007: 979-990
EVT of Chronic Iliac Vein Obstruction Clinical Results

- Pain - free @ (5 - yrs)
  - 6% - 60% (PTS)
  - 26% - 59% (NIVL)
- Absence of limb swelling
  - 3% - 42% (PTS)
  - 5% - 43% (NIVL)
- Ulcer healing = 58%

Neglan et al., J Vasc Surg, 2007:979-990
49 ♀ with:
• 2 day history of left thigh / calf swelling
• associated mild dyspnea on exertion
• 4 days prior had arthroscopic surgery of right knee

Past Medical History
- Gronblad – Strandberg syndrome
- subretinal hemorrhages of left eye
- PAD - intermittent claudication bilateral calves

Medications
- OCP
Physical Examination

Vital signs:
BP: 110 / 80 mmHg, P= 104, SaO2 = 94% (RA), T= 98.6 F

- **Cardiorespiratory exam:**
  - normal

- **Extremities:**
  - Left thigh and calf 2+ edema, non-tender, pedal pulses palpable bilaterally
Post power pulse spray
DVX catheter: 10mg tPA
Subsequent Plan

• Catheter - directed thrombolytic therapy
  – tPA @ 2 mg / hr via infusion catheter
  – Heparin @ 200 units per hour via sheath
  – Targeted PTT 40 - 50 seconds
Later that night …. (3 AM)

- Sudden visual field loss left eye
- Fundoscopy: New left sub-retinal bleed
- Heparin and TPA immediately discontinued

- ? IVC filter implantation (+/-)
- ? Mechanical thrombectomy without AC
- ? Surgical thrombectomy
- ? Compression stockings
Diagnosis ???
Pseudoxanthoma Elasticum

- Rare (1:70,000 - 160,000)
- Involves skin, eye and cardiovascular
- Retina – involvement in 85% of cases. Severe visual loss in ~ 5%
- Xanthomas (neck, groin, popliteal fossa)
- Abnormal GI bleeding and ? IC aneurysms
- Symptoms (CAD / ACS or PAD)
- Diffuse arterial disease with prominent collaterals
Angiography

Following left iliac vein stenting
Venogram @ 3 - months
(IVC retrieval)
Presentation of Upper Extremity Venous Thrombosis

- 2% - 4% of all venous thromboses
- US - incidence of 50,000 cases annually
- Involves brachial, axillary and subclavian
- Signs / Symptoms
  - Swelling / discoloration
  - Pain / discomfort in arm, shoulder, neck
  - Prominent superficial veins (Urschel’s sign)
Clinical Sequelae

- Pulmonary embolus ~ 12% symptomatic and up to ~ 36% may remain asymptomatic
- Venous hypertension ⇒ PTS (severe 13%)
- Loss of future vascular access or SVC syndrome
- Mortality 15% - 50% (underlying etiology)
- Recurrence after Tx ~ 2% - 8%
Etiologies of Upper Extremity Venous Thrombosis

• **Primary** axillo - SCV thrombosis
  (idiopathic or *Paget - Schroetter syndrome*)
  – No associated disease or trauma
  – Exertion - related

• **Secondary** axillo – SCV thrombosis
  – Recognized cause
  – 2o to central venous catheters (CVC), ICD, pacemakers
  – Systemic due to malignancy, thrombophilia, trauma
Paget - Schroëtter Syndrome

Leopold-von-Schroëtter, Vienna (1837-1908)

Sir James Paget, London (1814-1898)
Paget - Schroëtter Syndrome

• 2 - 4% of DVT involve upper limb
• Often secondary to repetitive upper extremity activity in the presence of a mechanical abnormality at the thoracic inlet
  – Arm abduction, cervical extension & shoulder depression
  – e.g. Weight lifting, baseball throwing, rowing, lobster fishing
• Repetitive compression results in fibrous tissue formation that permanently strangles the vein
• Most patients present after vigorous physical activity
  – ? Micro-trauma → activation of coagulation cascade
• Common anomalies
  - Young athlete with hypertrophied muscle
  - First or clavicular rib
  - Musculofascial bands
  - Cervical ribs
Ms SM

- 48 yo F presented with right upper extremity swelling
- HPI
  - Previously well & active
  - Woke up with pain & gross swelling of arm
  - Heavy lifting & mammogram few days prior
  - Multiple presentations to OSH
    - Rx as cellulitis
- PMH
  - HTN
  - Smoker
  - No VTE (DVT or PE)
Duplex Ultrasound
Orders to Dr. Charles Dotter !!!

FOR ATOMIC PASS GRAFT TOMORROW

VISUALIZE BUT
DO NOT TRY TO FIX

Your signature on this request indicates your approval of the use of
and drugs appropriate to the examination.

Service
Surgery

2/13 Chia
Dr. Dotter’s “Rebuttal”
Venogram
Right Axillary - Subclavian Vein
EKOS Catheter
Ultrasound Accelerated Catheter-Directed Lysis

5F EKOS Peripheral System

- Multi Side Port Infusion Catheter
- Ultrasound Core Wire

• tPA @ 2mg/hr x 4 hrs
• Heparin (PTT 40-50)
Venogram (Post - tPA @ 4 hours)
Balloon Angioplasty (PTA)

- PTA 12 x 40 mm balloon
Final Venogram
Venogram @ 3 - weeks
(Post - 1st rib resection)
Venogram @ 3 - weeks
(Post - 1st rib resection)
Venogram
6 Month Follow-up
Fate of Contralateral Vein

- UCLA series: 61% with compression of contralateral vein on venography
- If normal in neutral position – stress (TOS)
- Role of surgery if Asx ???
  - elective repair if compression of vein in dominant arm and occupation exposes patient at increased risk for thrombosis
Treatment of Primary ASDVT

Results

- Largest retrospective series (50 - years)
  - 626 limbs / 608 patients
  - Best results in 511 / 548 patients < 6 weeks & prompt surgery
  - 24 / 42 limbs > 6 weeks all remained sx
  - 36 patients, no lysis
    - 10 - ASX
    - 25 (PTS) - despite first rib resection

Algorithm for the Management of 1° ASDVT

Venography

Thrombosis

Lysis

Anticoagulation

Evaluation

Symptomatic

Asymptomatic Abnormality

1st Rib Resection (early vs. delayed?)

Residual Stenosis

PTA +/- Stent

Normal Vein

Asymptomatic No Abnormality

Compression/Stricture

Urschel et al., Ann Thorac Surg, 2002:69
CT Abdomen
Nutcracker Syndrome
Nutcracker Syndrome (NCS)

• AKA, renal vein entrapment or mesoaortic compression of the left renal vein
• Results most commonly from compression of the left renal vein between the aorta and superior mesenteric artery, although other variants exist
Signs and Symptoms

• Hematuria
• Anemia
• Left flank and/or pelvic pain
• Orthostatic proteinuria and intolerance
• Left testicular pain (males)
• LLQ pain (females)
• Nausea and vomiting
• Varicocele and varicose veins
Demographic Features

- Childhood to 7\textsuperscript{th} decade of life
- Most symptomatic pts 2\textsuperscript{nd} to 3\textsuperscript{rd} decade
- Second peak in middle-aged women
- Not a hereditary phenomenon
Diagnosis of NCS

- Left renal venography
- CT
- MRI/MRA
- Abdominal ultrasonography
- DSA
Differential Diagnosis of NCS

- Pelvic Congestion Syndrome
- Renal stones
- Genitourinary malignancy
- Loin pain hematuria syndrome
Treatment of NCS

- Conservative Tx age < 18 YO
- ACE-I for orthostatic proteinuria
- Endovascular stenting
  - balloon-expandable or self-expanding
  - Cxs include migration, thrombosis, ISR, deformities and erosions
Surgical Treatment of NCS

• Medial nephropexy (excision of renal varices)
• Left renal vein (LRV) bypass
• LRV transposition between SMA and aorta
• Renal-to-IVC shunt
• Renal autotransplant
• Gonadocaval bypass
• Nephrectomy
• Coil embolization of ovarian vein with PCS
Compressive Disorders: Diagnosis or Bust