Special Diagnostic Considerations in CVM Patients

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- VEITH Symposium, November 18-22, 2015 -

Special Diagnostic Considerations:

- Localized Intravascular Coagulopathy (LIC)
- Aplasia or hypoplasia of deep venous trunks

Background & Significance:

- Coagulation disorders occur at a high frequency in patients with CVMs and are associated with potentially severe thrombo-embolic events and hemorrhagic complications

Disclosure:

- BTG Consultant

- Currently there are no defined guidelines around how patients with CVMs should be screened and/or treated for coagulopathy and when to introduce anticoagulation
Background & Significance:
• Thus, it is important to identify an accurate diagnostic algorithm for coagulopathies associated with VM to guide appropriate management of these potentially life threatening disorders
• IUP Consensus Update 2013 – Provides current guidelines for the management of coagulopathies associated with VMs

Pathophysiology:
• Blood stagnation within the lesion and altered hemodynamics lead to the activation of coagulation cascade with subsequent production of thrombin and the conversion of fibrinogen into fibrin

Pathophysiology:
• This process is followed by fibrinolysis, which is reflected by elevated levels of fibrin degradation products, including plasmin derived D-dimer epitopes

Pathophysiology:
• This simplified description of a complex hemopathologic pathway underlines pathogenesis of localized intravascular coagulopathy (LIC) and distinct coagulation profile that characterize this unique coagulopathy associated with VM

Terminology:
• In the literature, LIC is often erroneously labeled as Kasabach-Merritt Syndrome (KMS)
• KMS: a distinct clinical entity characterized by disseminated intravascular coagulation (DIC) and profound thrombocytopenia frequently associated with aggressive vascular tumors, classically hemangioendothelioma

Terminology:
• Clear distinction between KMS and LIC is important:
  – in contrast to patients with KMS, LIC can be treated with heparin based medications
• In addition pts with LIC have been inadequately treated (antiplatelet medications, interferon etc.)
Differential diagnosis LIC vs KMS:

- Platelet count in LIC is minimally diminished (in the 100-150 x 10^3/ml range)
- PT/aPTT, INR (can be elevated and can be wnl/non specific)

Pathophysiology:

- Newly-formed microthrombi in LIC bind to intravascular elementary Ca^{++} deposits and form pathognomonic stone-like structures called “phleboliths”

Diagnostic considerations:

- Phleboliths can be detected during physical examination by palpation in patients with superficial VM

X Ray
Ultrasonography
Therapeutic considerations:
- LIC is of important clinical concern due to the potential for leading to more serious thrombo-hemorrhagic events, including:
  - Deep vein thrombosis (DVT)
  - Pulmonary embolism (PE) with associated pulmonary hypertension
  - Disseminated intravascular coagulopathy (DIC)

Indications for treatment:
- Pain caused by LIC/Phleboliths
- Peri-procedure
  - in patients with extensive Malformations (surface area > 10cm²)
  - Elevated D-dimer and/or low fibrinogen
- KTS + elevated D-dimer and/or low fibrinogen
- Marginal Vein if risk of VTE is high

Treatment:
- Painful lesions with evidence of LIC
  - 7 to 14 days of weight adjusted LMWH w/compression therapy

Prophylaxis:
- Patients with extensive VMs and ++D-Dimer
  - Prophylactic treatment with weigh adjusted LMWH prior to the procedure AND diagnostic
    - ESPECIALLY IN PATIENTS WITH KTS
    - Marginal Vein = risk of VTE is high
Consider preoperative temporary IVC Filters in addition to anticoagulation in KTS patients and patients with truncular VM.

"Duke Protocol":

- Markovic J, Shortell CK, JVS 2014:

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Present in 8% of CVM patients (with venous predominance)

Diagnostic considerations:
• dceMRI affords the opportunity to use ONE imagining modality to obtain information with regard:
  – Flow dynamics (HFVM vs. LFVMs)
  – Relationship with surrounding and vital anatomic structures
  – Deep venous anomalies / anatomic variations
  – LIC / phlebolith detection

• Agenesis of the popliteal vein in a patient with Klippel-Trénaunay syndrome

• Prevalence of deep venous anomalies is even higher (18%) in patients with KTS
• Aplasia of the left iliofemoral vein segment

Time-Resolved MRA Acquisition
Conventional Dynamic MRA Acquisition
Lateral Draining Vein
Management of Anomalies

• Absent/abnormal deep system
  – Doesn’t exclude possibility of treating malformation
  – Evaluate each patient’s anatomy individually

• Lateral Draining Vein
  – Often too large to ablate
  – Consider stripping and ligation
  – Anticoagulation

Summary:

• Measurement of baseline D-dimer and fibrinogen levels:
  – strongly recommended as part of the initial laboratory evaluation in all CVMs patients

• Weight-adjusted LMWH treatment:
  – drug of choice in patients with elevated D-dimers and extensive, multifocal, infiltrating and/or painful VMs, KTS, MV for symptom relief and prevention of thromboembolic complications

Summary:

• Evaluation of patency and anatomic variations of the ENTIRE venous system (deep and superficial)

Summary:

• These assessment need to be included in the treatment planning in all patients with CVMs

Vascular Malformation Team Roster 2015/16:

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