EMBOLIZATION/SCLEROTHERAPY
of Intramuscular Venous Malformations

Robert J. Rosens, M.D.
Lenox Hill Heart and Vascular Institute
New York
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VEINOUS MALFORMATIONS
• LOW FLOW
• THE COMMONEST VASCULAR MALFORMATIONS ENCOUNTERED CLINICALLY BY A FACTOR OF 10:1 OVER AVM (HIGH FLOW)
• BY DEFINITION, ONLY VEINS INVOLVED
• TREATMENT RARELY ERADICATES LESION COMPLETELY BUT THE CLINICAL PROBLEM CAN OFTEN BE RESOLVED

VENOUS MALFORMATION TYPES
– CAVERNOUS
– INTRAMUSCULAR
– SUPERFICIAL
– KLIPPEL-TRENAUNAY
– MIXED

INFANTILE HEMANGIOMA – BENIGN ENDOTHELIAL TUMOR OF INFANCY
This is the only lesion you should refer to as “hemangioma”

VENOUS MALFORMATIONS
• CLINICAL PRESENTATION
  – MASS – SOFT, EMPTIES ON COMPRESSION OR WHEN ELEVATED
  – DISCOLORATION – DEPENDS ON DEPTH (e.g. “Port Wine Stain”)
  – PAIN – DEPENDENCY, EXERTION
  – BLEEDING
  – ACUTE THROMBOSIS

EVALUATION OF VENOUS MALFORMATIONS
• ULTRASOUND – EASY OFFICE STUDY – CONFIRMS LOW FLOW FLUID-FILLED SPACES OR CHANNELS
• PLAIN FILMS – PHLEBOLITHS, OCC. BONE CHANGES
• CT WITH CONTRAST
• MRI/MRA – BEST SINGLE EXAM
• VENOGRAPHY
• ARTERIOGRAPHY?
Angiography in apparently venous lesions?

Is it worth embolizing “microshunts” prior to sclero?

NO

Exception: Parkes Weber Syndrome

18 y o male with disabling pain, non-healing ulcer in wheelchair for 10 months

Unilaterality similar to KTS but with microvascular shunting
Typically diffuse malformation with limb swelling, venous hypertension, sometimes high output state

Pruning of small vessel shunting using microspheres

VENOUS MALFORMATIONS
TREATMENT OPTIONS

• SURGICAL RESECTION
• EMBOLIZATION – BY DEFINITION ARTERIAL SUPPLY GENERALLY MINIMAL
• SCLEROTHERAPY – ETHANOL, STS, BLEOMYCIN, DOXYCYCLINE
• LASER FOR SUPERFICIAL LESION (CUTANEOUS CAPILLARY VENOUS, E.G. PORT WINE STAIN)

6 weeks post embolization, ulcer healed, no pain
SURGERY FEASIBLE IN SELECTED CASES, BUT HIGH RISK OF RECURRENCE

BEST APPLICATION – LARGE BULKY LESIONS, SEVERELY AFFECTED OVERLYING SKIN

EARLY SURGERY (INFANCY, EARLY CHILDHOOD) OR OTHER INTERVENTION AMI HOLD PROMISE

INTRA-OPERATIVE EMBOLIZATION A VALUABLE ADJUNCT

DIRECT PUNCTURE VENOGRAPHY

PUNCTURE OF LESION DIRECTLY USING ANGIOCATH OR NEEDLE UNDER VISUAL, ULTRASOUND, OR FLUOROSCOPIC GUIDANCE. PRE-OP IMAGING ESSENTIAL FOR TREATMENT PLANNING

DIRECT EMBOLIZATION/ SCLEROTHERAPY

COLLAGEN SUSPENSION

SOTRADECOL (SODIUM TETRADECYL SULFATE)

- ALTERNATIVE SCLEROSANT TO ETHANOL (DETERGENT)
- HAS BEEN IN USE FOR VARICOSE VEIN TREATMENT FOR MANY YEARS
- AVAILABLE IN 1% AND 3% SOLUTIONS
- CAN BE USED AS FOAM (WITH SMALL AMT. CONTRAST AND AIR) – MORE POTENT
- LESS TISSUE/NEURO TOXICITY

Bleomycin

- In use as sclerosant for nearly 10 years
- Dose range much lower than that used in chemotherapy; lifetime cumulative dose is the limiting factor
- Primary concern has been pulmonary toxicity (fibrosis) which is risk when used as chemo agent
- There are scattered reports of both acute and longer term pulmonary toxicity when used as sclerosant but risk is generally considered low
- Precautions necessary to prevent hyperpigmentation when skin adhesives are used
- Should it be used as first line agent or reserved for cases when other agents with longer track record have failed?
Prevention of thromboembolic complications -
Automatic tourniquet, Pressurized IV heparin infusion

Cuff distends lesion for easier access and control of outflow
Continuous pressurized heparinized saline infusion via peripheral IV to prevent DVT

HEMOSTATIC MATRIX

Use to occlude tract on angiocath withdrawal
- Stops bleeding
- Reduces risk of sclerosant tracking to skin and causing ulceration

When treating Intramuscular lesions – use caution in calf and forearm

Staged procedures and steroids – risk of compartment syndrome

DIRECT STICK EMBOLIZATION – POST-OP

- EXPECT INCREASED SWELLING WHICH MAY LAST 2 - 3 WEEKS
- LESION GRADUALLY SHRINKS OVER 4 – 6 WEEKS
- MULTIPLE TREATMENTS MAY BE REQUIRED – LIMIT ON DRUG VOLUME AND ANATOMIC CONSIDERATIONS
- MAXIMUM DOSE OF STS AT ONE TREATMENT 0.5CC/KG
- POSSIBLE COMPLICATIONS INCLUDE DVT, ULCERATION, NERVE DAMAGE
- OFTEN POOR CORRELATION BETWEEN CLINICAL RESPONSE AND MRI FINDINGS (MECHANISM?)

DIFFICULT ACCESS

- NON-VISIBLE, NON-PALPABLE LESION
  – TOURNIQUET TO DISTEND LESION
  – US GUIDANCE
  – CT GUIDANCE
  – MR GUIDANCE – WOULD BE IDEAL IN MANY RESPECTS
  – MERGING PRIOR IMAGING STUDIES (CT, MR) WITH LIVE FLUOROSCOPY (FUSION IMAGING)

CT guided direct embolization
12 y.o. female with shoulder pain, lesion localized by prior MRI
MR Compatible Needles

Titanium alloy
Safe, relatively small artifact
Problems – very flexible, high surface friction

MR guided embolization

19 y.o. athlete with pain rt neck, only MRI showed lesion;
2 failed prior embolizations, one with US, one with CT

Using Titanium MR Needle, GadoLinum,
Sotradecol
Complete resolution of symptoms

KLIPPEL TRENAUNAY SYNDROME

- Commonest vascular malformation
- One or more extremities (usually leg)
- Venous anomalies & varicosities
- Some have hypoplastic deep veins
- Cutaneous lesion (port wine stain – actually capillary venous malformation)
- Hypertrophy of bone/soft tissues
- No AV shunting on angiogram

KTS – WHY DOES IT OCCUR?

- Abnormality in mesodermal fetal development vs segmental genetic mutation (mosaicism)
- Stereotypic clinical manifestations favor the latter
- Congenital but not genetically transmitted in vast majority

Receptor Tyrosine Kinases

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<th>Cell Membrane</th>
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<th>Akt/PKB</th>
<th>mTOR</th>
<th>Ras</th>
<th>Raf</th>
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**Identification of an angiogenic factor that when mutated causes susceptibility to Klippel-Trenaunay syndrome – isolated from affected tissues**

**WT**

**E133K**

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By A. Hammill, MD

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WT

E133K

**Receptor Tyrosine Kinases**

- EGFR, VEGFR
- AKT
- PI3K
- mTOR
- Raf
- Ras
- PTEN
- TSC2
- TSC1
- S6
- eIF-4E
- Green: activating mutations
- (all somatic, as germ line mutation would presumably be fatal - also known/potential oncogenes)
- Red: (stop signs): tumor suppression gene mutations
- (usually germinal inheritance)
K-T SYNDROME MANAGEMENT

- CONSERVATIVE – SUPPORT STOCKINGS, ELEVATION
- BASELINE US/VENOGRAM TO CONFIRM DX AND EVAL. DEEP SYSTEM
- ANGIO IF ANY ? OF ARTERIAL COMPONENT
- AVOID VEIN STRIPPING OR OTHER VENOUS SURGERY IF DEEP SYSTEM HYPOPLASTIC OR ABSENT
- EMBOLIZATION ONLY FOR ISOLATED CAVERNOUS COMPONENTS
- ROLE FOR ENDOVENOUS ABLATION TECHNIQUES

Deep Veins in KT Syndrome

- Contrast venography is NOT the gold standard in evaluating the deep veins of KT patients – the majority (>80%) DO have patent and intact deep veins even when venogram with tourniquets seems to indicate they are absent. Ultrasound is the best study to confirm this.
- This may mean that many KT patients have more treatment options than we thought in the past (EVLT, Sclero, etc).

VENOUS ABLATION IN KTS?

- LASER OR RADIOFREQUENCY

New Approach to KT Syndrome - re-route the plumbing

The Knee in KT Syndrome
Knee involvement should be treated more aggressively

- Knee involvement in KT may be associated with recurrent hemarthrosis (bleeding into joint) causing pain and eventual degenerative arthritis of joint (similar to hemophiliacs)
- Treat early and aggressively, including surgery with pre-op embolization

AT AGE 18....

Knee joint of 80 year old...

Often misdiagnosed

- Lack of physical findings, especially when not in KT setting may lead to misdiagnosis of JRA, PVNS, “growing pains”, etc. Only MRI confirms the diagnosis

Arthroscopic synovectomy

Goals are:
- Joint preservation
- Stopping recurrent hemarthrosis
- Delaying need for joint replacement
- Good working relationship with orthopedic surgeon essential

Study on hemophiliacs demonstrates that synovial hypertrophy occurs after only 2-3 episodes of hemarthrosis; hypertrophied synovia are the cause of cartilage destruction, joint space narrowing, bone erosions, subchondral cysts and other degenerative joint changes.
• So in theory, we should be arresting the bleeding episodes early in childhood using sclerotherapy rather than waiting for the synovial hypertrophy to occur, which would potentially require synovectomy after the joint damage has already occurred.

SUMMARY
• Intramuscular venous malformations are one of the commonest types
• Don’t call them hemangiomas
• Often delayed diagnosis as few physical findings unless part of a syndrome
• Dx made by MRI
• Most not amenable to resection
• Sclerotherapy will improve the sx in most pts
• Use caution in calf and forearm
• Knee involvement should be treated early and aggressively