COURSE 8: Surgical Management of Visceral and Extremity Vascular Anomalies

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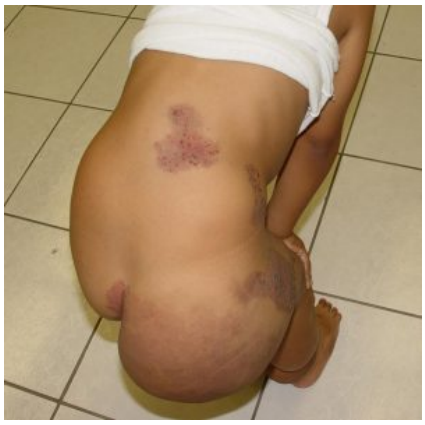
Introduction: Surgical management of visceral and extremity vascular anomalies



Most of the major vascular anomalies can occur in the viscera, in either isolation or in combination with other superficial malformations. These internal lesions, by their very nature, have historically garnered little attention. Vascular anomalies that are apparent and visually evident have been misunderstood for centuries. However, vascular anomalies that are visceral have received even less attention.

In the past, there was little focus on developing an understanding for, or a unique approach to, the diagnosis and surgical management of visceral vascular anomalies.

Today, experts who treat these rare and often complex and life-threatening anomalies are focused on developing evaluative and interventional techniques, including innovative operative procedures.

The utility of a modified endorectal pull-through technique for the treatment of colorectal venous malformations has been demonstrated. Resecting, debulking, and reconstructing visceral vascular malformations and their surrounding anatomies has recently become more available to the patient who previously had little, if any, option for treatment.

Understanding that there are limitations for surgical intervention in the treatment of visceral vascular anomalies, the latest focus is on combining knowledge of the biologic mechanisms involved in the development and progression of these lesions along with the surgical approach. This dual approach will give patients their best possible outcomes.

Objectives

Upon successful completion of this activity, participants should be able to:

- Identify and describe the types of vascular anomalies of the liver and gastrointestinal tract
- Define criteria to treat hepatic and intestinal vascular lesions
- Explain the options for treatment of visceral vascular anomalies
- Locate reliable resources to keep updated

Vascular anomalies of the liver

The term hemangioma is unfortunately often misapplied to many vascular lesions in the liver. It is crucial to distinguish true infantile hemangioma from other hepatic vascular tumors and malformations.

Infantile hemangioma of the liver occurs in several patterns, with differing presentations, risks, and treatments.

Adults do not have true hemangiomas. The vast majority of adult liver hemangiomas are actually venous malformations.

Angiosarcoma and epithelioid hemangioendothelioma are rare tumors of the liver, but primary hepatic kaposiform hemangioendothelioma does not occur.

Hepatic hemangioma: three patterns

Diffuse:

- Innumerable, packed in the liver with little normal hepatic parenchyma
- Massive hepatomegaly can cause fatal abdominal compartment syndrome
- No anemia or thrombocytopenia
- Always hypothyroid due to expression of type III iodothyronine deiodinase which inactivates thyroid hormone
- sually no high-flow cardiac state, but may be hypokinetic due to hypothyroidism
- These are infantile hemangiomas

Hepatic hemangioma: three patterns

Multifocal:

- Several to do ens
- Hypervascular with no central clot
- No anemia or thrombocytopenia
- May have macrovascular shunts causing high-output cardiac state
- These are infantile hemangiomas
- Can proliferate to become diffuse pattern

Hepatic hemangioma: three patterns

Focal:

- Singular, often large mass detected antenatally or in newborn
- Central clot after birth causes moderate, self-limited anemia and thrombocytopenia (usually not less than 20,000; this is NOT Kasabach-Merritt phenomenon)
- May have macrovascular shunts causing high-output cardiac state
- Not infantile hemangioma, but rather apidly Involuting Congenital Hemangioma (ICH)

Hepatic hemangioma: treatment

Focal

sually none; they will involute substantially over the first year. If uncontrollable CHF, shunts may be emboli ed. Supportive care. Hyperbilirubinemia is common and generally resolves. Hepatic failure is rare.

Multifocal

Sequential ultrasonagraphic evaluation if asymptomatic. Propranolol if CHF, enlarging substantially, or hypothyroid. Shunts can be emboli ed if severe CHF and insufficient time for involution.

Diffuse

Aggressive propranolol and thyroid replacement therapy. Add steroid if propranolol alone insufficient in extremis. Consider evaluation for hepatic transplantation if abdominal compartment syndrome nearing fatality.

Hepatic hemangioma: surgery

No role for surgery in treatment of hepatic hemangioma, except:

- iopsy if presentation/imaging not classic; needle biopsy usually sufficient
- Transplantation for abdominal compartment syndrome despite aggressive pharmacotherapy

Intestinal hemangioma

- Extremely rare
- Presents with I bleeding in early infancy
- Visible as mesenteric/bowel hypervascular thickening on ultrasonography
- Visible on CT with contrast and M I, but may not be necessary if high index of suspicion due to cutaneous lesions and ultrasonographic findings
- sually diffuse
- Treat with propranolol, bowel rest, transfusions as necessary
- Avoid resection, unless very focal (rare)

Pulmonary hemangioma

- Exceedingly rare
- Infants who present with airway hemorrhage and multiple vascular nodules in lung should be suspected of having cutaneovisceral angiomatosis with thrombocytopenia/multifocal lymphangioendotheliomatosis with thrombocytopenia (CAT/MLT) rather than hemangioma

Reasons to consider debulking of extremity/ torso vascular malformation

- Appearance
- Weight
- Function
- Infection
- leeding
- Prevention of progressive deformity

Risks of debulking

- Bleeding
- Pain
- Difficult wounds
- Prolonged drainage
- Infection
- Scarring
- Clots
- In ury to important structures
- Expansion of ad acent areas
- Rarely achieves perfection

Surgical debulking: Tackle one defined area at a time

- Focal lesions may be extirpated nearly entirely with closure by ad acent tissue advancement
- Avoid tissue expanders (infection risk, particularly with LM)
- Most lesions are diffuse: dissect one anatomic area extensively and stage further areas
- Unresected areas will likely expand:
  - It s not cancer – complete excision unnecessary, T
  - Aim for near-total removal or re-expansion will be disappointing

Debulking pearls

- Girth debulking for subcutaneous bulk is satisfying
- Intramuscular debulking is generally unrewarding (except for focal painful mass)
- Don't elevate flaps more than 180 degrees of extremity (post-debulking circumference); can do much more than 180 pre-debulking circumference
- Always place closed suction drains under advancement flap closure
- Ectatic veins with central communication should be obliterated by endovascular techniques before debulking to prevent pulmonary embolism or clot formation in stagnant vein segments
- Pre-operative low molecular weight heparin advisable with consumptive coagulopathy and/or ectatic veins
- Tourniquet is your friend (but be mindful of duration)
- Do not injure major nerves/vessels
- Anomalous veins in the subcutaneous tissues can generally be removed, regardless of si e

Post-op management

- Elastic compression – but not too tight
- Elevation
- Avoid body weight on flaps
- Leave drains as long as necessary until taper off (could be several months for lymphatic lesions; usually days for pure venous lesions)
- Counsel families to EXPECT wound separation, breakdown, necrosis; pleasant surprise if no problems.
- Infection common with lymphatic anomalies; slow to clear (at least 14 days); antibiotics if occur

Course 08: Lesson Quiz

1. Hepatic infantile hemangioma patterns are:

A. Singular large mass
B. Focal, diffused, multifocal
C. Mosaic
D. Deep and focal

. A focal hepatic infantile hemangioma will typically require:

A. Frequent ultrasounds
B. High dose steroids for up to three years
C. No treatment
D. Sequential resections

. Infantile hemangioma of the liver is never present in:

A. Adults
B. Diabetics
C. Caucasians
D. Pregnant women

4. The current treatment protocol for diffuse hepatic infantile hemangioma includes:

A. Thyroid replacement therapy with alpha interferon
B. Aggressive propranolol and thyroid replacement therapy
C. Observation only
D. Low dose steroids

. Pulmonary hemangiomas (airway hemorrhage and multi-vascular nodules) are often confused with:

A. LVM (lymphatic venous malformation)
B. CAT/MLT (cutaneousvisceral angiomatosis with thrombocytopenia/Multifocal lymphangioendotheliomatosis with thrombocytopenia)
C. KHE (kaposiform hemangioendothelioma) with MLT (Multifocal lymphangioendotheliomatosis with thrombocytopenia)
D. CVAVM (capillary venous arteriovenous malformation)

6. Hepatic infantile hemangiomas are usually not treated surgically, except for:

A. They are never treated surgically
B. Biopsies only
Biopsies and abdominal compartment
C. syndrome
D. Reduction of multiple small infantile hemangiomas of liver

7. Diffuse hepatic infantile hemangiomas:
   A. Frequently present with high-flow cardiopathy
   B. Are always hypothyroid
   C. Present with frequent anemia
   D. Never require treatment

8. Which of the following is NOT a reason for debulking an extremity/torso vascular malformation?
   A. Infection and/or bleeding
   B. Appearance
   C. Function
   D. Thinning of malformation

9. The primary treatment regimen for intestinal hemangiomas include:
   A. Frequent transfusions
   B. Observation only
   C. Serial resections
   D. Propranolol, bowel rest, and transfusions as needed

AUTHOR
PROFILES

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Dr. Fishman is Board-certified in Surgery and Pediatric Surgery. He is the Stuart and Jane Weitzman Family Chair in Surgery, Vice-Chair of Surgery for Clinical Operations, Co-Director of the Vascular Anomalies Center, President of the Physicians' Organization, and Senior Vice-President for Access and Business Services at Boston Children's Hospital. He is Professor of Surgery at Harvard Medical School.

Dr. Fishman is past president of the International Society for the Study of Vascular Anomalies. He has authored over 200 articles and chapters. He co-authored the comprehensive reference text Mulliken and Young's Vascular Anomalies, 2nd Edition.

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Course 08: Lesson Quiz Answer Key

1. B
2. C
3. A
4. B
5. B
6. C
7. B
8. D
9. D