INTRA-LIPOMATOUS CAPILLARY-VENOUS MALFORMATION

What is it??

• Intra-lipomatous Capillary-Venous Malformation.....???
• This is not described in the literature. The theory that is formulated by their evaluation is:
• Vascular malformations do exist in any and every tissue of the body. They often exist in a tissue without causing over time increased progressive growth of those tissues, merely infiltration of them.
• ILCVMs not only exist in fat, they cause a stimulation of the adipocytes with significant increased growth and increased vascularity resulting in pathology.

• On DSA studies, they are typified by an increased vascularity and dense contrast staining without AV shunts. Being that the capillaries are present, although malformed, they still are a check-valve that despite the increased in-flow vascularity, they prevent AV shunts. The malformed capillaries and malformed post-capillary veins stimulate their endothelial cells to produce angiogenesis factor that then causes increased in-flow neovascularity in the affected adipose tissues that are then stimulated and grow.

Financial Disclosure

• Nothing to disclose
A similar clinical model occurs that is described in the literature as the so-called “Intramuscular Hemangioma,” a misnomer in that they are histologically “Intramuscular Venous Malformations.” However, unlike “true” venous malformations, and their variant Glomovenous Malformations (“Glomangiomases”), their arterial vascularity is “normal” with late-phase weak contrast opacification (due to normal capillaries) of the malformed veins. The “intramuscular” VM form routinely demonstrates enlarged in-flow arteries with intense contrast staining WITHOUT AV shunts. Further, the infiltrated muscle routinely enlarges over time, just as in the “intra-lipomatous” form. These patients present as a growing muscle mass with or without pain. The primary age of presentation is in the first 3 decades. A growing muscle mass in this age group one must first consider some form of muscular sarcoma. In the work-up and eventual muscle biopsy, the VM is discovered and to everyone’s relief no sarcoma revealed.

Due to the significantly increased vascularity in ILCVMs, the veins become enlarged and engorged. This venous “hypertension” results in the pathologic changes noted clinically in the tissues affected as in this patient.

Abnormal Vascular and Lipomatous Cells
- Abnormally large (hypertrophied) fat cells
- There is hyper-endothelial cell proliferation around the vascular structures themselves.
- There is a dense matrix between the blood vessels and fat cells.
- Very large and proliferative vascular cells
- Close proximity to both vascular and fat cells.

Lateral View of the Tumor

Tumor from Above
11/22/2016

Excision of Tumor Mass

Successful Embolization of Veins
Dr. Yakes

Residual Defect After Flap Rotations

Six Weeks Later
What Went Wrong

- The original tumor was removed
- Margins when first looked at were free of vascular tissue at the periphery
- There remained an area of an open wound that did not heal
- The patient began to bleed from recurrent subcutaneous vascular malformation

Recanalizations of embolized vessels prior to 2nd surgery

Residual or Recurrent Lumbar Vascularity

RE-Operation Excision of all residual fat
Repeated ETOH injections

• The second surgery removed all visible fat cells by removing all fat and deep fascia.
• Blood vessels were followed deep into the paraspinal muscles, and ligated deep.
• Many repeated pre-op arterial embolizations using diluted ETOH, coils, & large particles to eradicate as much of the ILCVM arterial vascularity to minimize operative arterial bleeding.
• Direct puncture pre-op embo of the hypertensive massively enlarged post-capillary veins were thrombosed/occluded with nBCA & Onyx to prevent surgical bleeding from a vein source.
Where are we now

- 14 months of therapy
- 10 months after the last major surgery
- To the best of our current knowledge we have eradicated both the fat and viable vascular cells
- Have we cured Her? Only time will tell
- The patient on 11-21-15 returned to Cambodia and all phone and email contacts state she has no recurrence at this time and is clinically well.