

What You Should Know About Klippel-Trenaunay Syndrome (KTS)

SESSION 68: SUPERFICIAL VEIN TREATMENT STRATEGIES III

51st VEITH Symposium 2024

November 19-23, 2024,
New York, NY

B. B. Lee, M.D., Ph.D., F.A.C.S.
Clinical Professor of Surgery, George Washington University, Washington, D.C. U.S.A.
Adjunct Professor of Surgery, Uniformed Services University of HS, Bethesda, MD, USA

DISCLOSURE OF CONFLICTS OF INTEREST

BYUNG-BOONG LEE, MD, PhD, FACS

Title:

What You Should Know About Klippel-Trenaunay Syndrome (KTS)

I do not have any relevant financial relationships with any commercial interests.

Klippel-Trenaunay Syndrome (KTS)

- Klippel-Trenaunay Syndrome (KTS) is one of the best-known terms to represent one unique 'syndromic' condition of congenital vascular malformations (CVMs) since French physicians, Maurice Klippel and Paul Trenaunay reported in 1900 (cf. Parkes-Weber Syndrome).
- Indeed, KTS represents a combined condition of venous malformation (VM) and lymphatic malformation (LM) together with capillary malformation (CM).
- But the 'marginal' vein (MV) is the most critical clinical issue among these many vascular malformation components.



B.B. Lee, MD
GWU

Klippel-Trenaunay Syndrome (KTS)

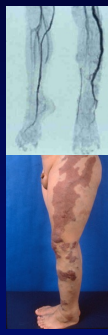
- Because, the MV is the most common VM component, mostly located superficially under the skin like common varicose veins deceiving the phlebologist.
- But, the MV is NOT a varicose vein; it is an 'embryonic' vein which failed to be involuted to remain after the birth so that many call it 'lateral embryonic' vein based on its unique location on the extremity, mostly along the lateral aspect of the lower limb.
- Indeed, MV is one of truncular VM lesions as the outcome of defective development during venous trunk formation/maturation period along the last stage of the embryogenesis.



B.B. Lee, MD
GWU

Klippel-Trenaunay Syndrome (KTS)

- Hence, the MV does not carry 'matured' vessel structure like ordinary varicose vein so that this defective vein wall, often missing the media, would result in the blood stagnation within this distorted vascular bed of the VM lesions.
- Such condition would precipitate continuous activation and subsequent consumption of clotting factors known as 'intraluminal coagulation process' to form the blood clots to result in the deep vein thrombosis (DVT) and further to pulmonary embolism (PE).



B.B. Lee, MD
GWU

Klippel-Trenaunay Syndrome (KTS)

- It further accompanies a unique condition of 'avalvulosis' to precipitate 'steadily progressing' chronic venous hypertension (CVI) as well, in addition to such high risk for the venous thromboembolism (VTE): DVT & PE.
- Besides, this MV often alarms for the further involvement by other VM conditions like 'aplasia of iliac vein; hypoplasia of femoral vein; intraluminal defects - web/stenosis/spur', resulting in defective condition of the venous system to cause potentially serious hemodynamic impact when mishandled.

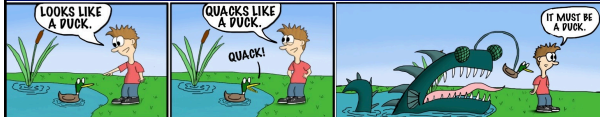


B.B. Lee, MD
GWU

Varicose Vein among KTS

B.B. Lee, MD
GWU

- Hence, whenever you should encounter the KTS with varicose vein, remember the MV as its VM component, mimicking the varicose veins but “the MV is actually an embryonic vein”.
- Therefore, such benign looking varicose veins among KTS patients would herald “hidden risk”, like a tip of the iceberg, of coexisting embryonic veins which is also mostly visible as well as palpable, like ordinary varicose veins!!!



Klippel-Trenaunay Syndrome (KTS)

B.B. Lee, MD
GWU

- Lastly, you should look for other infrequently coexisting VMs affecting the iliac-femoral vein system to make sure it is not overlooked, giving proper disposition *before* the commitment to MV management with phlebectomy/surgical excision and/or endovascular obliteration with laser & RF, etc.



Thank you for your attention!