Phlebolymphedema: Hall Mark Of Combined Insufficiency Of Venous-Lymphatic System

45th Annual VEITHsymposium- 2018
SESSION 110: EXAMINING THE EVIDENCE
November 13-17, 2018         New York, New York

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Phlebolymphedema

The venous and lymphatic systems are mutually dependent dual outflow system of the circulation as an integral part of a single 'phlebolymphatic unit' complementing to each other.

Under normal physiological conditions, the venous capillary can handle 80 to 90% of liquid portion only, ultrafiltered through its 'closed' system, while the lymphatic capillary reabsorbs the rest 10 to 20 % of total liquid filtered out from arterial capillary together with all the protein micromolecules through its 'open' system.

Hence, Phlebolymphedema (PLE) represents a ‘combined’ condition of chronic venous insufficiency (CVI) and chronic lymphatic insufficiency (CLI) as the outcome of simultaneous failure of dual outflow ‘veno-lymphatic’ system.

When one of these two mutually interdependent systems should fail, it gives additional burdening to the other system. When this additional loading should exceeds its limit, such condition would precipitate the failure of other system as well, resulting in a total failure of these two ‘inseparable’ systems together, resulting in ‘Phlebolymphedema’.

PLE is therefore, an inevitable outcome of inseparable venous and lymphatic system when both systems should fail together.

‘Primary’ PLE represents a combined condition of CVI caused by the venous malformation (VM) and CLI by the lymphatic malformation (LM) simultaneously.

The most common VM to cause CVI is ‘marginal vein’ with venous reflux/hypertension, followed by deep vein dysplasia (e.g. iliac vein agenesis, hypoplastic femoral vein) or defective vein (e.g. web, stenosis, aneurysm, ectasia) with venous outflow obstruction/hypertension.

CLI is mostly due to ‘primary lymphedema by truncular LM lesion (e.g. lymphatic dysplasia: aplasia, hypoplasia, or hyperplasia). Extratruncular LM (lymphangioma) seldom involved to the CLI.
These two vascular malformations, VM and LM together, to cause primary PLE is the most common vascular disorder of Klippel-Trenaunay Syndrome (KTS), remaining as an ultimate challenge due to delicate interrelationship between venous and lymphatic systems with mutual interdependency. Clinical manifestation of the PLE is therefore, extremely variable depending upon the etiology (primary and secondary) and the degrees/extent of the CVI and CLI, and clinically more distinctive along the lower extremity.

**Primary Phlebolymphedema**
- CVI of secondary PLE, on contrary to primary one, is mostly due to the sequelae of post-thrombotic syndrome (PTS) following the deep vein thrombosis (e.g. iliac vein thrombosis).
- CLI of secondary PLE is generally secondary outcome of regional/local lymphedema following steady progress of the local tissue damage (e.g. ulcer) by recurrent infections.
- However, occasionally, overlooked subclinical condition of primary lymphedema accelerates the deterioration of the underlying benign primary venous disorder (e.g. reflux) to cause CLI.

**Secondary Phlebolymphedema**
- Secondary PLE developed along the end stage of CVI cause the local condition more complicated with local/regional lymphedema often as newly added condition. It becomes an additional burdening to CVI to make the clinical management much more difficult.
- Secondary PLE also shows visibly strained lymphatic system infrequently as a victim of abnormal venous condition, especially when the lymphatic system is in marginally compensated condition.

**Phlebolymphedema**
- The assessment of the PLE should start with proper diagnosis of its etiology to differentiate two different types: primary and/or secondary PLE.
- The assessment of two separate condition of CVI and CLI should include the accurate estimate on its severity, extent, and secondary outcome/damage.
- And simultaneous evaluations of venous and lymphatic systems together are essential.

**Diagnosis in general**
- Non-invasive tests (e.g. Duplex ultrasonography) alone is generally sufficient for the basic assessment of the extent/severity of the CVI. But for the secondary PLE, ascending/descending phlebography is infrequently indicated as additional diagnostic evaluations.
- The lymphoscintigraphy on affected limb is essential for the CLI to assess the functional status of the lymphatic system.
Baseline therapy for the PLE is the compression therapy reinforced with decongestive lymphatic therapy to control the CVI and CLI together.

Marginal vein with the reflux as the cause of CVI can be treated with the resection or embolo-sclerotherapy as long as the deep vein system is fully developed to be able to handle diverted blood influx.

Deep vein reconstruction to relieve the CVI caused by deep vein dysplasia (e.g. aplastic/ hypoplastic iliac-femoral vein) can be beneficial when there is a clear evidence for the hemodynamic gain to improve indolent ulcers.

Secondary PLE with the CVI by PTS should be treated more aggressively to relieve the cause of obstruction/ reflux with various forms of open surgical (e.g. bypass) and endovascular therapy (e.g. angioplasty and stenting).

When the CVI is caused by the multilevel DVT sequelae (indolent ulcer), even a minimum correction of the obstruction/stenosis is able to assist tremendous improvement of the efficacy of the compression therapy-based conservative management following successful relief of venous hypertension.

Phlebolymphedema

Clinical Management

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- Deep vein reconstruction to relieve the CVI caused by deep vein dysplasia (e.g. aplastic/ hypoplastic iliac-femoral vein) can be beneficial when there is a clear evidence for the hemodynamic gain to improve indolent ulcers.

Primary PLE with CVI caused by the reflux of MV can be treated successfully with MV resection, while the conventional compression therapy alone for CVI by deep vein dysplasia is generally acceptable in its majority.

Secondary PLE with CVI caused by the PTS can be further improved with correction of the venous outflow obstruction with angioplasty & stent especially when the DVT sequelae is involved to the multi-levels of iliac-femoral-popliteal vein system.

Conclusion

- PLE can be managed more effectively when open and/or endovascular therapy is added to the basic compression therapy to control the CVI and CLI together.
- Primary PLE with CVI caused by the reflux of MV can be treated successfully with MV resection, while the conventional compression therapy alone for CVI by deep vein dysplasia is generally acceptable in its majority.
- Secondary PLE with CVI caused by the PTS can be further improved with correction of the venous outflow obstruction with angioplasty & stent especially when the DVT sequelae is involved to the multi-levels of iliac-femoral-popliteal vein system.

Thank you for your attention!