Acquired Vulvar Lymphatic Anomaly: A Systematic Review

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Background/Objective

- Acquired vulvar lymphatic anomaly (AVL) (previously known as lymphangioma circumscriptum) is a neoplastic condition of lymphatic vessels that is not well-characterized. Diagnosis is often delayed and AVL is often treatment-refractory. Studies are limited with no guidelines to date on management.
- Herein, we performed a systematic review of AVL to summarize the epidemiology and management options.

Methods

Records identified through database searching
PubMed, CINAHL, OVID

Nonduplicate case reports and case series in the English language investigating AVL were included

78 publications (133 patients) extracted for:
disease association, treatments for prior pelvic malignancy, demographics, disease prior to presentation, symptoms, clinical findings, treatment for AVL, duration/number of treatments/follow up

Results

Disease Association	N	%
Prior Pelvic Malignancy:	66	51
Cervical	57	43
Vulvar	4	3
Vaginal	1	1
Endometrial	2	2
Bladder	2	2
Inflammatory Bowel Disease	6	5
Lymphoma	3	2
Pregnancy	3	2
Rectal Malignancy	1	1
Other	12	9

AVL Treatment	N	%
Surgical	36	27
Laser	17	13
Topical*	9	7
Lymphedema therapy	1	1
Other	9	7

**Topical: clobetasol cream, other topical corticosteroids, local antiseptics*

Prior Malignancy Treatment*	N	%
Radiation	45	34
Lymph Node Dissection (LND)	39	29
Resection alone w/o LND	35	26
Chemotherapy	4	3
Other	0	0
N/A	39	29

**will not add up to total cases as some patients had both*

Conclusions

- While the mechanism of AVL development is not well-elucidated, it is thought that disruption to lymphatic drainage, through surgery, lymph node dissection, or radiation is what leads to the development of this condition.
- A thorough examination, history, biopsy, and high clinical suspicion are required to diagnose these conditions early.
- Surgical excision was found to have the lowest rate of recurrence when compared to laser and topical treatments, however, follow-up duration and reported outcomes were not standardized, making evaluation of therapy efficacy challenging.
- Information regarding duration of therapy, treatment time to resolution or recurrence , and efficacy measures were limited and variable.

Future Directions

- Prospective studies are needed to better understand true incidence, prevalence, risk factors, and pathogenesis of this condition.
- Because symptoms can be debilitating, future studies should consider incorporating quality of life outcome measures.