Review Article

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Acquired vulvar lymphangioma: risk factors, disease associations, and management considerations: a systematic review

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ABSTRACT

Background: Acquired vulvar lymphangioma (AVL) is not well-characterized. Diagnosis is delayed and the condition is often refractory to therapy.

Objective: The objective of this study was to provide a systematic review of AVL including risk factors, disease associations, and management options.

Methods: A primary literature search was conducted using 3 databases: PubMed, CINAHL, and OVID, from all years to 2022.

Results: In total, 78 publications with 133 patients (48 ± 17 years) were included. Most studies were case reports/series. The most common disease association was prior malignancy (70 patients, 53% of cases) and inflammatory bowel disease (6 patients, 5% of cases). The most common malignancy was cervical cancer (57 patients, 43% of cases). Most patients had prior radiation or surgery, with 36% (n = 48) treated with radiation, 30% (n = 40) with lymph node dissection, and 27% (n = 36) with surgical resection. Common presenting symptoms included discharge/oozing, pain, and pruritus. Most patients underwent surgical treatment for AVL with 39% treated with excision, 12% with laser therapy (the majority used CO_2), and 11% with medical therapies. Most patients had failed prior therapies and there was a diagnostic delay.

Limitations: Retrospective nature. Most studies were limited to case reports and case series, with interstudy variability and result heterogeneity.

Conclusion: AVL is an underrecognized entity and should be considered in patients with a history of malignancy or radiation to the urogenital area. Treatment should include multidisciplinary care and address underlying lymphatic changes, manage any existing inflammatory conditions, and utilize skin-directed therapies and barrier agents while addressing symptoms of pruritus and pain. Prospective studies are needed to further characterize AVL and develop treatment guidelines.

Keywords: lymphangiectasia, lymphangioma, lymphatic anomaly, vulvar edema

Introduction

Acquired vulvar lymphangioma (AVL), also known as acquired lymphatic anomaly or lymphangiectasia, and previously known as lymphangioma circumscriptum, is an uncommon condition characterized by an abnormality in the lymphatic vessels of the vulva.[1,2] AVL is associated with obstructed or impaired pelvic lymph drainage,[1] which can occur in the setting of chronic inflammatory or neoplastic conditions, as well as in the setting of prior surgical or radiation therapy that leads to lymphatic disruption. Studies have shown that AVL is often a late complication of anogenital and pelvic malignancies, with cervical carcinoma being the most common malignancy reported, but is also reported in inflammatory conditions including Crohn's disease and infectious conditions such as tuberculosis.[1,3,4]

What is known about this subject with respect to women and their families?

- Acquired vulvar lymphangioma (AVL) is a lymphatic neoplasm that is localized to the vulva, often appearing in the setting of prior pelvic malignancy or surgery.
- Diagnosis is often delayed, and the condition is commonly refractory to treatment.

What is new from this article as messages for women and their families?

- This systematic review found that AVL is commonly associated with prior pelvic malignancy and the subsequent surgical treatment, radiation, or lymph node dissection, and is seen in the setting of inflammatory bowel disease.
- The most reported treatment modalities for AVL were excision, followed by laser therapy, but there were limited outcomes data and no information on quality-of-life following therapy.
- In most cases, patients had been treated with other therapies without improvement, and the diagnosis was delayed.

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Clinically, AVL is often described as clusters of vesicular or verrucous papules,[3] and superficial cases are sometimes described as resembling "frogspawn" with grouped clear to cloudy fluid-filled vesicles. Typically, vesicles have surrounding erythema and can appear as small confluent plaques (Fig. 1A). These features are often accompanied by surrounding edema (unilateral or bilateral) (Fig.1B) and even overlying eczematous change or lichenification. Symptoms may include pain, pruritus, discomfort, dyspareunia, and a burning sensation. In more superficial cases there may be fluid drainage and malodor.[3,5–7] Information on the clinical course of AVL is limited, primarily due to few observational studies and the rarity of this condition.

In early cases, AVL may have nonspecific clinical and histologic findings, often resulting in a misdiagnosis of inflammatory dermatitis. In cases where vesicles are more apparent, the differential includes herpes and autoimmune blistering disorders. When AVL presents as verrucous or vascular papules, neoplastic etiologies including condyloma acuminata, molluscum contagiosum, angiokeratomas, and even vascular lesions may be considered.[3] Because of this variability in clinical findings, diagnosis is usually confirmed by tissue biopsy.[4,8] Histologically, AVL typically presents with multiple dilated lymphatics in the superficial dermis. The epidermis may have overlying hyperkeratosis, acanthosis, and spongiosis.[9]

Studies on treatment are limited with no guidelines to date on the medical or surgical management of AVL. Various treatments have been reported including surgical excision, ablative and nonablative laser therapies, and topicals[3,10,11] and all have been associated with a high rate of recurrence. The aim of this study is to perform a systematic review on cases of AVL to summarize the epidemiologic and clinical findings and identify associated comorbidities and treatment options.

Materials and methods

Literature search

This study was performed in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (Fig. 2). A primary literature search was conducted using PubMed, CINAHL, and OVID, from all years to 2022. Two authors independently searched and cross-checked with the following search terms: (lymphangioma circumscriptum OR microcystic lymphatic malformation OR lymphangioma OR lymphangiectasia) AND (vulva OR vagina).

Article selection

Articles published in English from all years were considered for eligibility. Articles were excluded if they discussed congenital lymphangioma circumscriptum or nonvulvar presentations.

Data extraction

Included studies were summarized using a data extraction form with the following variables extracted: number of patients reported in the study, disease association, treatments for prior pelvic malignancy, patient ages, ethnicity, time between onset of AVL symptom onset and diagnosis, symptoms, clinical findings, treatment for AVL, duration/number of treatments, duration of follow-up, efficacy, reported recurrence, interval for follow-up, prior treatments, histopathology, status of quality of life measures, and inclusion of clinical or histopathological photos.

Results

Literature review

The initial literature search yielded 290 articles. Overall, 146 were nonduplicate articles. A total of 54 articles were excluded based on their title and/or abstract, including 48 that were not related to AVL and 8 that were not available in the English language. No additional articles were identified based on a search of article references. Ultimately, 78 articles,[1,3–79] comprising a total of 133 patients, met the eligibility criteria and were included in this qualitative analysis. Articles were published between 1979 and 2022. The eligible articles consisted of case series, case reports, literature reviews, and a retrospective chart review. Relevant variables extracted from each article are included in Table 1.

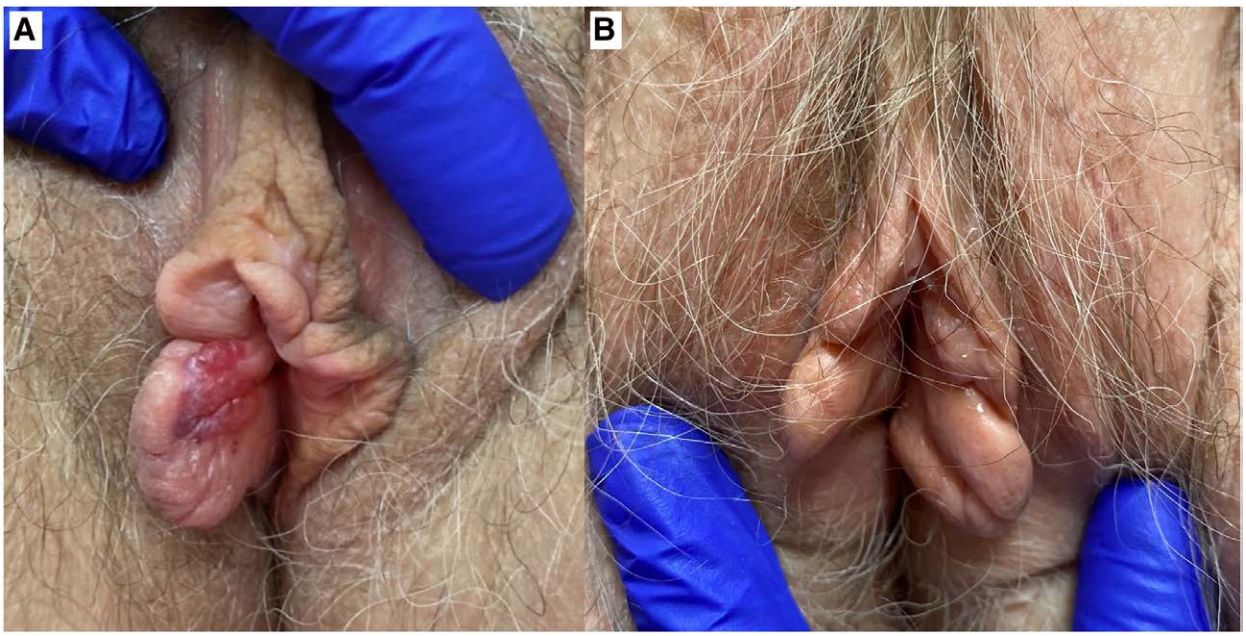

Fig. 1. Clinical images of vulvar-acquired lymphatic anomaly and edema. (A) Erythematous to purple papulovesicles on right labium minus, representing acquired vulvar lymphatic anomaly. (B) Left-sided labium minus edema, representing vulvar lymphedema.

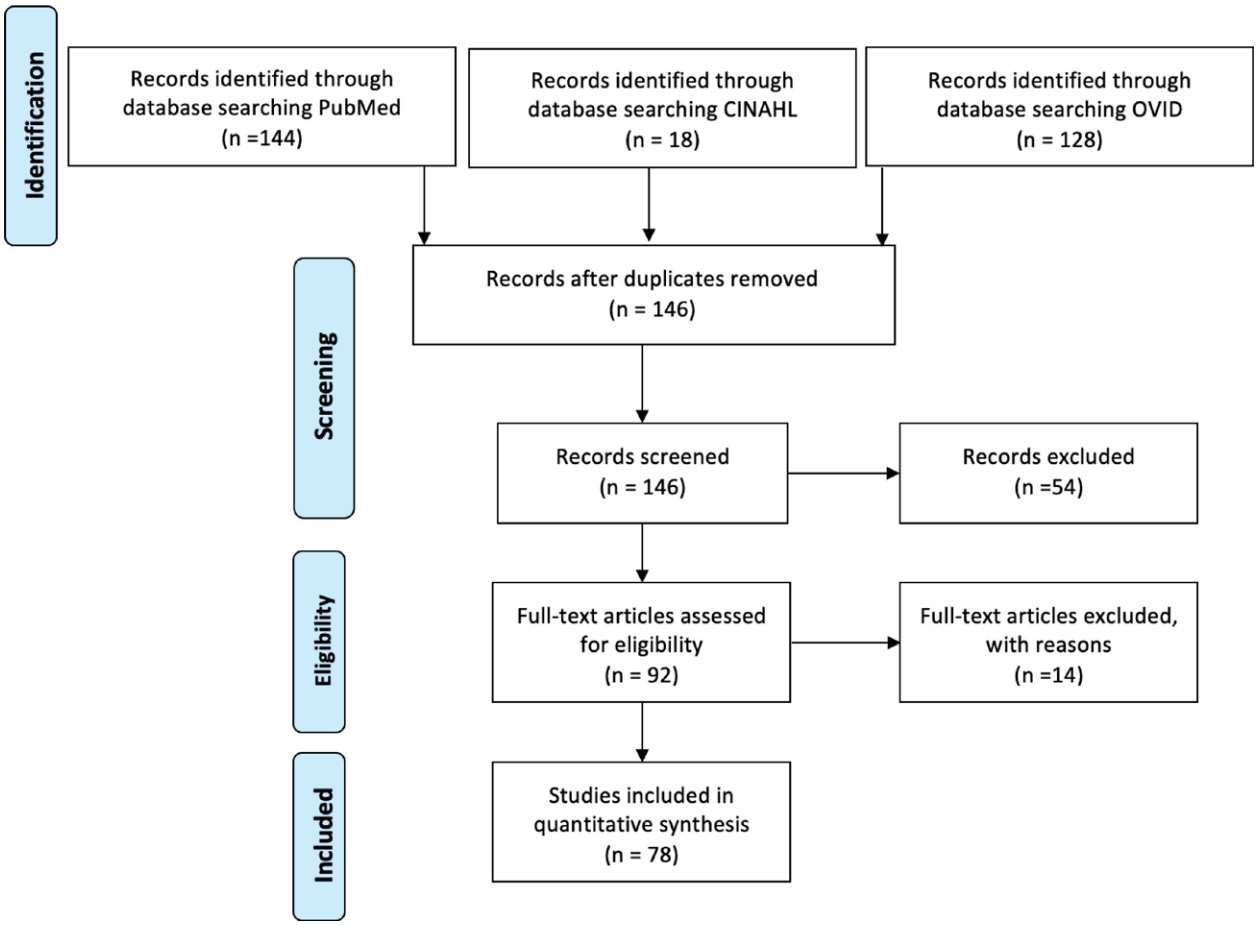

Fig. 2. Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRIMSA) flow diagram of literature search and article selection.

Clinical presentation and diagnosis

The age distribution ranged from 17 to 81 with a median of 49 years. There were 5 cases in pediatric patients that were not classified as congenital lymphangioma. Due to the lack of ethnicity data included, we could not determine any ethnic distribution for AVL. In studies that reported diagnostic delay, there was a mean delay of 5 years until the time of diagnosis. The most common symptoms reported included: clear oozing discharge, pain, and pruritus. Eleven patients were asymptomatic. The most common clinical findings included: multiple circumscribed papulovesicular lesions of varying sizes, often filled with clear fluid. Anatomical distribution was varied and involved the mons pubis, one or both labia majora, one or both labia minora, entire vulva, or entire vulva with spread to the thigh. All cases were biopsy-proven.

Treatment and outcomes

A total of 58 studies consisting of 96 patients evaluated treatment modalities. Of these, the most common treatment modality was surgery, followed by laser. Topical therapies included clobetasol cream and other topical corticosteroids, local antiseptics, isonicotinic acid hydrazide, ethambutol chloride, rifampicin, gentian violet 1% solution, emollients, silver sulfadiazine 1%, imiquimod and antihistamines. One study mentioned the use of physical therapy or lymphedema therapy. Follow-up was variable and not reported in most studies.

Of the 36 case reports that listed surgery as the treatment modality, 23 (64%) reported no evidence of recurrence for a total of 31 patients. Seven reports of 21 patients did not mention the efficacy of surgical excision, and all other reports noted some type of recurrence. There were 13 case reports which reported laser as a treatment modality. Of these, 6 reported a combination of topical or surgical treatment modalities in addition to laser therapy. Five reports of 6 patients (38%) had no evidence of recurrence. One report did not include treatment efficacy, and all other reports noted recurrence, most being minor or lesion size smaller than the primary lesions. The analysis also included 11 patients that were treated with topical therapies. Treatment outcomes were rarely reported, with some just noting symptom improvement. Most outcome measures relied on clinical improvement in signs and symptoms as evaluated by the provider. Quality-of-life outcome measures were not included in any of the studies.

Disease associations

Although 68% of cases had a disease association, the most common being a prior diagnosis of pelvic malignancy (cervical, vulvar, vaginal, endometrial, or bladder). Inflammatory bowel disease (IBD) was the second most common disease association and was seen in 5% of cases. There were 3 cases (2%) presenting with lesions associated with pregnancy. Most patients had prior radiation or surgical treatment with 36% (n = 48) treated previously with radiation, 30% (n = 40) with prior lymph node dissection, and 27% (n = 36) with prior surgical resection without lymph node dissection. While the other 32% of cases were idiopathic, many did evaluate for other disease associations or malignancies with a variety of modalities. The most used diagnostic tools were screens for sexually transmitted infections such as syphilis, human immunodeficiency virus, or hepatitis. Other

Table 1

Variables extracted from studies included in literature review

	PMID	Type of study	Number of patients	Prior pelvic malignancy	Other disease association	Prior radiation	Prior lymph node dissection	Prior surgery or chemotherapy	Patient age	AVL duration before diagnosis	Treatment type (T = topical therapy, PT = physical therapy, L = laser, S = surgical, or other)	Treatment specifics (procedure, laser settings, medication)	Interval for follow-up
1	34736616	Case report	1	None	Crohn Disease	None	None	None	56	3 years	T	Clobetasol 0.05%	None
2	34877687	Review, case Series	6	n = 6	None	n = 6	n = 5	None	Median: 61, Range: 44–72	median interval between malignancy diagnosis > acquired vulvar lymphangioma circumscriptum (AVLC) diagnosis: 10 years (0–32 years)	S	Excision (n = 5), Unknown (n = 1)	1.5 years, 3 years, 0.04 years, Unknown (n = 3)
3	14761137	Case report	1	None	Pregnancy	None	None	None	35	5 years	None	None	None
4	2644602	Case report, review	1	None	None	None	None	None	42	None	S	Excision	6 weeks
5	8408931	Case report	1	None	None	None	None	None	46	None	None	None	None
6	27329721	Case report	16	n = 9	None (n = 7)	n = 7	n = 3	Hysterectomy (n = 4), Salpingo-oophorectomy (n = 3), Wide local excision (n = 1)	Median: 55, Range: 43–81	11.3 years between cancer treatment and vulvar lymphangioma circumscriptum (VLC) development (range: 5–20 years)	Unknown	Unknown	None
7	15488131	Case report	1	None	None	None	None	None	17	4	S	Excision	2 weeks
8	22529452	Case report	1	None	Tuberculosis	None	None	None	35	1 year	S	Excision (n = 2)	None
9	8914364	Case report	1	None	Recurrent cellulitis	None	None	None	42	7 months	T	IV antibiotics and local antiseptic	5 years
10	7747544	Case report	1	None	None	None	None	None	76	2 years	T	None	None
11	26967121	Literature review and case report	5	n = 2	Crohn's Disease	n = 1	None	None	Median: 50, Range: 18–68	None	L, S	YAG Laser & Excision (n = 2), Electrodessication and curettage (n = 1), Excision (n = 1)	None
12	23595194	Case report	1	None	None	None	None	None	24	since teenager	Other	Compression therapy	None
13	25706522	Case report	1	n = 1	None	n = 1	None	Cystectomy (n = 1)	69	3 years	None	None	None
14	11270296	Case report	1	None	None	None	None	None	44	6	None	None	None
15	21464721	Case series, review	1	None	None	None	None	None	39	None	S	Excision	None
16	26374361	Case series, review	3	None	None	None	None	None	Mean: 33.3, Standard Deviation: 8.4	None	None	None	None
17	18312992	Case series, review	1	n = 1	None	n = 1	n = 1	Hysterectomy (n = 1)	63	4 years	S	Excision	28 weeks
18	22615519	Case series	2	n = 2	None	n = 2	None	Hysterectomy (n = 2)	65, 44	10 years, 16 years	S	Excision	Regular
19	30812056	Case report	1	None	None	None	None	None	61	1.5 years	S	Excision	4 weeks
20	16803506	Case series, retrospective study	1	n = 1	None	n = 1	None	None	40	6 months –2 years	S	Excision	None

(Continued)

Table1

(Continued)

	PMID	Type of study	Number of patients	Prior pelvic malignancy	Other disease association	Prior radiation	Prior lymph node dissection	Prior surgery or chemotherapy	Patient age	AVL duration before diagnosis	Treatment type (T = topical therapy, PT = physical therapy, L = laser, S = surgical, or other)	Treatment specifics (procedure, laser settings, medication)	Interval for follow-up
21	22199085	Case report (letter)	1	None	None	None	None	None	55	2 years	S, L	Hysterectomy, CO_2 laser vaporization (defocused mode with a 2 mm spot size)	15 weeks
22	23130218	Case report (letter)	1	n = 1	None	None	n = 1	None	50	3 months	None	None	None
23	2684843	Case series	3	n = 1	Crohn's Disease (n = 2)	n = 1	None	Hysterectomy (n = 1)	Median: 38, Range: 37–38	2 years (n = 3)	L, S, Other	Excision, cryotherapy	2 months (n = 1)
24	8286236	Case report	2	n = 2	None	n = 2	None	Hysterectomy (n = 2)	49, 50	None	S	Excision	18 months
25	761961	Case report	1	None	Tuberculosis (n = 1)	None	None	Lymph node incision (n = 1)	62	30 years	T, L	Isonicotinic acid hydrazide, Ethambutolcholride, Rifampicin, Electocoagulation	6 months
26	30289772	Case report (letter)	1	n = 1	None	n = 1	None	Hysterectomy (n = 1)	78	10 years	None	None	None
27	16219410	Case series, review	3	n = 2	Tuberculosis (n = 1)	n = 1	n = 1	Complete surgical resection (n = 2), Hysterectomy (n = 2)	Median: 67, Range: 53–71	None	L	Laser therapy (n = 1); CO2 laser vaporization (n = 2)	4 months, 22 months, 90 months
28	21547888	Case report	2	n = 2	None	n = 2	None	Hysterectomy (n = 2)	56, 68	None	S	Excision	7 months, 20 months
29	24133609	Case report	1	None	Crohn's Disease	None	None	None	35	Several years	None	None	None
30	12218845	Case Report	2	n = 2	None	n = 2	n = 2	Hysterectomy (n = 2)	75, 46	10 years	L	CO_2 laser vaporization (n = 2)	3 months 8 months
31	28461089	Case series	1	None	None	None	None	None	68	None	None	None	None
32	25099515	Case report (letter)	1	None	Pregnancy (n = 1)	None	None	None	28	1 month	None	None	2 months after delivery
33	22802470	Case report	1	None	Breast Cancer (n = 1)	None	None	Masectomy (n = 1), Chemotherapy (n = 1)	46	8 years	S	Excision (n = 2)	4 weeks post-op, 3 month return
34	1694423	Case report (letter)	1	n = 1	None	n = 1	None	Complete surgical resection (n = 1)	75	2 years	L	CO_2 laser vaporization-multiple sessions.	18 months
35	4065708	Case report	1	n = 1	None	None	n = 1	Hysterectomy (n = 1)	51	None	None	None	None
36	29451158	Case report (letter)	1	None	Klippel-Trenaunay syndrome (n = 1)	None	None	None	31	4 years	None	None	None

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Table1
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	PMID	Type of study	Number of patients	Prior pelvic malignancy	Other disease association	Prior radiation	Prior lymph node dissection	Prior surgery or chemotherapy	Patient age	AVL duration before diagnosis	Treatment type (T = topical therapy, PT = physical therapy, L = laser, S = surgical, or other)	Treatment specifics (procedure, laser settings, medication)	Interval for follow-up
37	10430005	Case report	1	n = 1	None	n = 1	None	Complete surgical resection (n = 1)	70	4 years	L	CO2 laser vaporization	4 weeks after wound healing: 5 more laser sessions
38	34621964	Case series	2	None	Hiradenitis suppurativa (n = 2)	None	None	None	44, 46	0	S	Excision	3 years (n = 2)
39	15752314	Case report	1	None	Tuberculosis (n = 1)	None	None	None	22	5 years	Other	Oral isoniazid, rifampicin, pyearazinamide and ethambutol	None
40	34837392	Case report	1	n = 1	None	n = 1	None	Hysterectomy (n = 1)	61	None	L	CO2 laser vaporization	None
41	17684378	Case report	1	n = 1	None	None	n = 1	Hysterectomy (n = 1)	73	2 years	S	Lymphaticovenular Anastomosis (LVA) surgery	6 months
42	10235381	Case report	1	None	Crohn's Disease	None	None	None	44	7 months	Other	Podophyllin	None
43	1427807	Case report	1	None	None	None	None	None	20	2 years	S	Vulvectomy	9 months
44	28283172	Case report	1	None	None	None	None	None	47	None	S	None	None
45	11270298	Case report	1	n = 1		n = 1	None	None	79	1 year	T	Topical mid-potency corticosteroid	None
46	15250899	Case report	1	None	None	None	None	None	20	10 years	None	None	None
47	22901902	Case report	1	None	None	None	None	None	44	3 years	T, Other	Gentian Violet 1% solution, Cetrizine hydrochloride	None
48	17656926	Case Report	1	None	None	None	None	Surgical resection (n = 1)	48	5 years	S	Excision	1 year
49	11776513	Case report, review	1	None	None	None	None	None	48	3 years	S	Excision	16 months
50	22361479	Case report	1	None	None	None	None	None	44	None	T	Antihistamines, emollients	None
51	34149227	Case report	1	None	None	None	None	None	25	1 month	S	Excision	None
52	26167062	Case report	1	None	None	None	None	None	60	4–5 years	S	Excision	1 year
53	10609498	Case report, review	1	n = 1	None	n = 1	n = 1	Hysterectomy (n = 1), Salpingo-oophorectomy (n = 1)	65	10 years	L, T, Other	CO_2 laser vaporization (multiple procedures), Silver sulfadiazine	6 weeks
54	19396719	Case series	8	n = 8	None	n = 7	n = 7	Surgical excision (n = 7)	Median: 48, Range: 39-63	None	S	Excision (n = 5)	None
55	34263326	Case report	1	n = 1	None	n = 1	n = 1	Vulvectomy (n = 1), chemotherapy (n = 1)	70	3 years	Other	Sclerotherapy	4 weeks

(Continued)

Table1
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	PMID	Type of study	Number of patients	Prior pelvic malignancy	Other disease association	Prior radiation	Prior lymph node dissection	Prior surgery or chemotherapy	Patient age	AVL duration before diagnosis	Treatment type (T = topical therapy, PT = physical therapy, L = laser, S = surgical, or other)	Treatment specifics (procedure, laser settings, medication)	Interval for follow-up
56	22026919	Case report	1	n = 1	None	n = 1	n = 1	Hysterectomy (n = 1)	42	4 years	S, L	Excision & CO_2 laser vaporization (1st treatment), excision (2nd treatment)	None
57	25468056	Case report	1	None	None	None	None	None	67	3 years	None	None	None
58	15228436	Case report	2	n = 2	None	n = 2	None	None	30, 45	1 week, unknown (max 2 years)	Other- penicillin	Penicillin	6 months
59	28242996	Case report	1	None	Tuberculosis (n = 1)	None	None	None	25	None	None	None	None
60	24396614	Case report	2	None	None	None	None	None	55, 60	25 years, unknown	T	Topical antihistamine	None
61	27331134	Case report	1	n = 1	None	n = 1	None	Chemotherapy (n = 1)	55	3–4 years	S	Excision	5 months
62	12495108	Case report	1	None	None	None	None	None	48	None	S	Oral antibiotics followed by excision	2years
63	18397567	Case report	1	None	Pregnancy	None	None	None	30	4 months	None	None	None
64	20004630	Case series	4	Cervical cancer (n = 1)	Hodgkin lymphoma (n = 3)	None	None	None	Median: 44.5, Range: 28–57	32.3 years, 10.6 years, 2.9 years, 0.2 years	S	Prophylactic antibiotic therapy (amoxicillin clavulanic acid) and excision	Median: 53 months
65	12738156	Case report, review	1	None	None	None	None	None	30	3 years	S	Excision	None
66	367022	Case report	1	None	None	None	None	None	38	7 years	S	Excision	None
67	25190008	Case report	1	n = 1	None	n = 1	None	Hysterectomy (n = 1)	76	6 years	Other	Cryotherapy	8 months
68	26156111	Retrospective chart review	8	n = 8	None	n = 4	n = 8	None	Median: 61.5, Range: 36-77	None	S	Excision (n = 8)	None
69	1765960	Case report	2	None	None	None	None	None	32, 35	1 year, 7 years	S (n = 2), L (n = 1)	Excision with laser vaporization, Excision	4 months
70	27502262	Case report	1	None	None	None	None	None	20	10 years	None	None	None
71	1669286	Case report	1	None	Tuberculosis (n = 1)	None	None	None	40	3 months	S	Excision	None
72	33423429	Case report	1	n = 1	None	n = 1	n = 1		71	None	None	None	None
73	31757874	Case report	1	None	HSV (n = 1)	None	None	None	43	6 years	Other	Lymphatic drainage, shave excision	None
74	28791276	Case report	1	Rectal cancer (n = 1)	None	n = 1	None	None	68	1year	L, S	Electrocautery and CO2 laser vaporization	6 months
75	20580481	Case report, letter	1	None	None	None	None	None	36	8 years	S	Excision	1 year
76	20300370	Case report	1	None	None	None	None	None	18	5 years	L, Other	Radiofrequency ablation, Sclerotherapy	Monthly x3, 6 months, 2 years
77	18319007	Case report	1	None	None	None	None	None	45	8 years	T, other	Topical antihistamines and emollients, Cryotherapy	None
78	6931304	Case report	1	n = 1	None	n = 1	n = 1	None	54	22 years	None	None	None

tests included ultrasound, abdominal and pelvic computed tomography or magnetic resonance imaging, and pap smears.

Discussion

In this review, we summarize the clinical findings, treatment modalities, and disease associations of AVL. This is an underrecognized condition and diagnosis is often delayed with 84% of cases in our study reporting diagnostic delay. Prior studies have evaluated the association between AVL and malignancy and reported that AVL is a late complication of prior surgery or chemotherapy to treat anogenital and pelvic malignancies, with the most common preceding malignancy being cervical carcinoma.[3] AVL is likely more common in pelvic and anogenital cancer survivors than reported as it is often misdiagnosed. Because it can present initially with nonspecific inflammation and symptoms of pruritus/burning, it may be misdiagnosed as other vulvar conditions including the genitourinary syndrome of menopause, lichen sclerosus, and contact dermatitis.

AVL is thought to develop due to disruption in lymphatic drainage, through surgical resection, lymph node dissection, radiation, or even mass effect of anogenital/pelvic malignancies.[3] Because of the changes in drainage of lymphatic fluid, many cases of AVL have unilateral or bilateral vulvar edema. Vulvar edema alone may be an early presenting sign. Other conditions that lead to systemic inflammation, including IBD, specifically Crohn's disease, have been associated with AVL. The mechanism is likely through long-standing edema, fibrosis, and inflammatory changes. Both vulvar edema and AVL are nonspecific findings that can be a presenting sign in Crohn's disease or occur even in the setting of Crohn's disease where the gastrointestinal disease is well-controlled or not symptomatic.[1,10,29,34]

Overall, a thorough examination, history, and high clinical suspicion is required to diagnose these conditions early. We recommend that in patients presenting with AVL with no prior diagnosis of IBD or malignancy, a thorough review of systems and age-appropriate screening should be performed. If there is a concern for pelvic malignancy, imaging, and appropriate lab work should be considered in the appropriate patient. If there is suspicion of IBD, appropriate workup including a fecal calprotectin level, may be considered.

Evidence-based guidelines for the management of AVL are lacking, leaving no clear standards for treatment. 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 the evaluation of therapy efficacy challenging. Additionally, it is important to note that wide local excisions/labiectomy/vulvectomy may carry increased morbidity and functional impairment compared to nonsurgical methods. Overall, information regarding the duration of therapy, treatment time to resolution or recurrence, and efficacy measures were limited and variable.

Treatment requires multidisciplinary care and should focus on addressing any underlying lymphatic changes, managing any underlying inflammatory condition, and utilization of skin-directed therapies and barrier agents, as well as addressing symptoms of pruritus and pain. While only mentioned in one of the articles click or tap here to enter text., these authors suggest in addition to the above, referral to a physical therapist with specialized training in the management of genital edema.[19]

The main limitation of this review is the study design as data was derived predominantly from case reports and case studies and thus, our study is limited by interstudy variability and heterogeneity of results. Many studies were missing variables that are useful for providing diagnostic or therapeutic recommendations.

Summarizing the data from existing studies provides further information on the characteristics and treatment modalities of AVL. However, prospective studies are needed to further characterize this condition to better understand true incidence and prevalence, risk factors, and pathogenesis, with the goal of earlier diagnosis and development of treatment guidelines. Additionally, because symptoms can be debilitating, future studies should consider incorporating quality-of-life outcome measures.

Conflicts of interest

None.

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Study approval

N/A

Author contributions

AD, AB, and CK participated in screening articles, writing the manuscript, and editing the draft.

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