REVIEW

Lymphangioma Circumscriptum of the Vulva: A Review of the Literature

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OBJECTIVE: To promote proper identification and management of this rare entity by presenting a review of the literature, a case report, and illustrations of its presentation and pathology.

DATA SOURCES: Sources searched included the literature contained in the National Library of Medicine's PubMed database, using the term "lymphangioma circumscriptum" combined with "vulva." Other search terms used were "capillary lymphangioma," "lymphangiectasia," and "dermal lymphangioma." Sources also included articles predating the PubMed database that were cited by other writers.

METHODS OF STUDY SELECTION: Selections were restricted to the English-language medical literature published since 1960, and the search resulted in retrieval of 3272 published papers about lymphangioma.

TABULATION, INTEGRATION, AND RESULTS: Congenital lymphangioma circumscriptum of the vulva has been reported in 11 patients, including one case reported and illustrated in this paper, and acquired lymphangioma circumscriptum has been reported in 20. It affects females 9–76 years old (mean 42.5 years). Clinically, it is characterized by persistent clusters of thin-walled vesicles filled with clear fluid. The diagnosis is usually made by biopsy, as these lesions often mimic such infectious diseases as molluscum contagiosum. Management options have included surgical excision of the skin and subcutaneous tissue, surface abrasion by laser or sclerosing therapy, and observation.

CONCLUSION: From this comprehensive review, which includes the clinicopathologic features of both the congenital and acquired forms, illustrations from the rarest form, and a summary of treatment approaches, we conclude that lymphangioma circumscriptum poses a diagnostic challenge the risks of which are misdiagnosis and mistreatment. These risks would likely be reduced were a database of cases accessible that permitted long-term follow-up and better assessment of presenting characteristics and treatment options. (Obstet Gynecol 2003;101:946–54. © 2003 by The American College of Obstetricians and Gynecologists.)

Lymphangioma circumscriptum is a rare benign disorder of no specific etiology involving the lymphatic channels in the deep dermal and subcutaneous layers. Lymphangioma circumscriptum can occur as either a congenital abnormality or as acquired damage to previously normal lymphatic channels.[1,2] In the English-language medical literature since 1960, congenital lymphangioma circumscriptum of the vulva has been reported in 11 patients, including the one reported in this paper (Cecchi R, Bartoli L, Brunetti L, Pavesi M, Giomi A. Lymphangioma circumscriptum of the vulva of late onset [letter]. Acta Derm Venereol 1995; 75:79–80).[3–10] Acquired lymphangioma circumscriptum has been reported in 20 patients (Landthaler M, Hohenleutner U, Braun-Falco O. Acquired lymphangioma of the vulva: Palliative treatment by means of laser vaporization carbon dioxide [letter]. Arch Dermatol 1990;126:967–8).[5,6,11–26] Because lymphangioma circumscriptum can mimic infectious processes and thus lead to inappropriate invasive treatment,[12,15,19,22,27] we present the following illustrated comprehensive literature review to help clinicians meet the diagnostic challenge lymphangioma circumscriptum poses.

SOURCES

Sources searched included the literature contained in the National Library of Medicine's PubMed database. Search terms included "lymphangioma circumscriptum" combined with "vulva," "capillary lymphangioma, lymphangiectasia," and "dermal lymphangioma." Articles predating the PubMed database were drawn from works by other writers.

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Table 1. Classification of Lymphangioma Circumscriptum

Characteristic	Classification	
	Classic	Localized
Localization	Proximal limbs	Various sites
Size	≥1 cm^2	≤1 cm^2
Age	<30 y	Any age
Symptoms	Oozing of fluid from vesicles	Oozing of fluid from vesicles
	Crusting and infection	Crusting and infection
	Swelling	Swelling
	Pain	Pain
Pathology	Dilated lymphatic channels in papillary dermis and subcutaneous tissue	Dilated lymphatic channels in papillary dermis
	Acanthosis	
	Hyperkeratosis	
Treatment	Observation	Observation or surgery
	Surgical excision	
	Surface abrasion	

STUDY SELECTION

Selections were restricted to the English-language medical literature published since 1960, and the search resulted in retrieval of 3272 published papers about lymphangioma. Of these papers, at least 110 referred to lymphangioma circumscriptum directly or by other names (capillary lymphangioma, lymphangiectasia, or dermal lymphangioma).

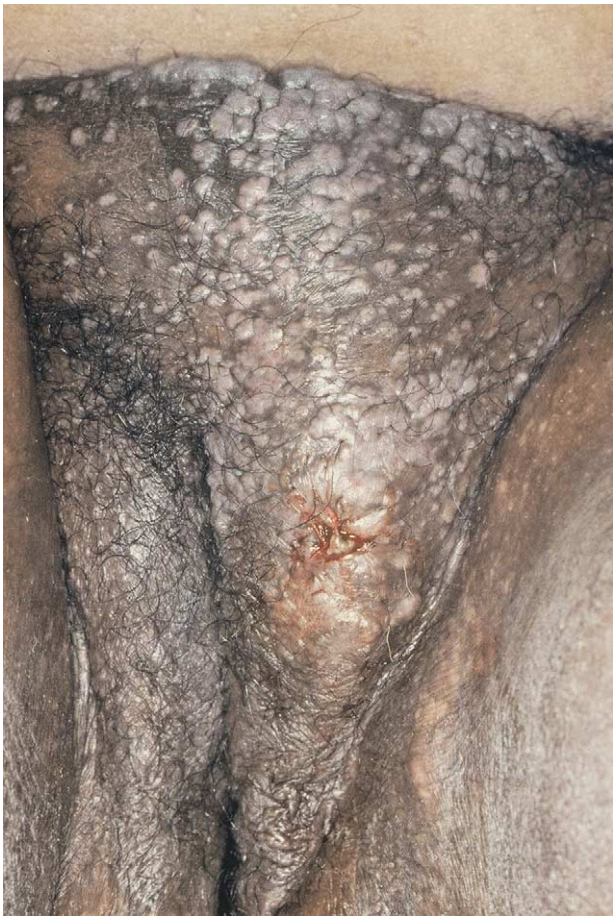

Figure 1. Lymphangioma circumscriptum of the vulva.
Vlastos. Vulvar Lymphangioma Circumscriptum. Obstet Gynecol 2003.

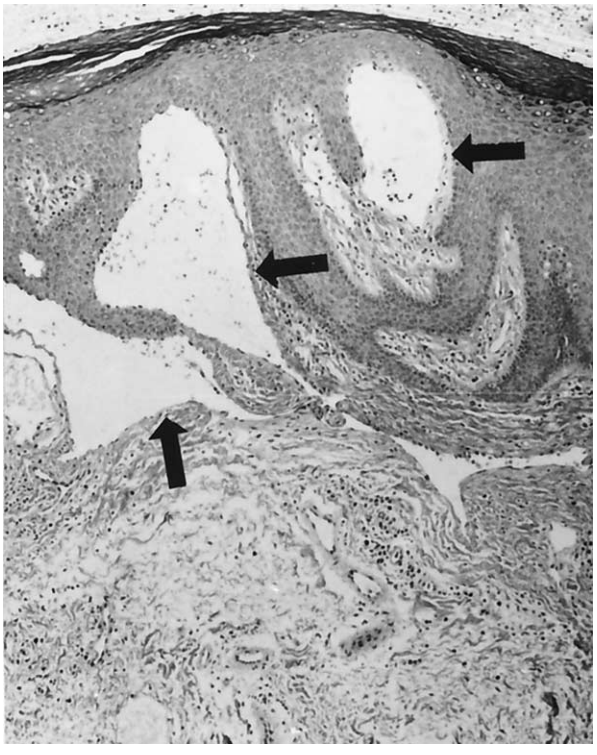

Figure 2. Microphotograph of the vulvar lesion demonstrating dilated lymphatic channels *(arrows)* in papillary dermis and an acanthotic epidermis (× 100, original magnification; hematoxylin–eosin). Hyperkeratosis was also present in this case of congenital lymphangioma circumscriptum.
Vlastos. Vulvar Lymphangioma Circumscriptum. Obstet Gynecol 2003.

Table 2. Literature Review of Congenital Lymphangioma Circumscriptum of the Vulva

Reference	Patient age (y)	Previous treatment	Treatment
1981 Bauer et al[4]	14	Excision (3 times), dates unreported	Radical vulvectomy and abdominoplasty
1989 Abu-Hamad et al[5]	42	Excision, 20 y previously	Partial vulvectomy and Z plasty
1991 Johnson et al[6]	32	No	Excision and CO_2 laser therapy
	35	No	Vulvectomy
1992 Murugan et al[7]	20	No	Vulvectomy
1995 Cecchi et al (letter)	76	No	Observation (patient refused treatment)
1995 Short and Peacock[9]	20	Excision (2 times), 6 and 3 y before	Excision of squamous cell carcinoma
1998 Nishi[8]	28	No	Sclerosing therapy planned (patient discontinued treatment)
1999 Gordon and Kaufman[3]	24	Excision and laser therapy, dates unreported	Laser therapy or observation proposed
2001 Sah et al[10]	48	No	Simple vulvectomy with wide excision
2002 Vlastos et al (current study)	30	No	Radical wide local excision

NA = not available.

RESULTS

Lymphangioma circumscriptum can occur anywhere in the body: intraperitoneally,[28–33] retroperitoneally,[34,35] on the skin surface (Kavanagh GM, Marshman G, Wesley J. Acquired scrotal lymphangiomata [letter]. Clin Exp Dermatol 1995;20:359; Drago F, Rampini P, Muzio G, Rebora A. Lymphangioma circumscriptum of the penis [letter]. Acta Derm Venereol 1997;77:252; Bardazzi F, Orlandi C, D'Antuono A, Patrizi A. Lymphangioma circumscriptum of the penis [letter]. Sex Transm Infect 1998;74:303–4),[36–42] or in bone.[43] However, the sites most often cited are the chest,[44] thigh,[45] and buttock.[46] The vulvar presentation is uncommon (only 31 cases, including one described here, have been reported) and can be either acquired or congenital (Cecchi et al. Acta Derm Venereol 1995;75:79–80; Landthaler et al. Arch Dermatol 1990;126:967–8).[3–19,21–27,47]

Several classifications of lymphangioma have been proposed, but they are often confusing. The most accepted one, that of Peachey et al,[1] distinguishes two main forms of lymphangioma circumscriptum: localized and classic. Both forms have a similar clinical appearance marked by clustered or diffuse thin-walled, translucent vesicles 1 to 5 mm in diameter and filled with clear lymphatic fluid. The presence of varying amounts of blood may give the vesicles a reddish color. The vesicles may develop on normal skin or on top of preexisting papules; hyperkeratosis may sometimes give them a verrucous appearance. Though similar in terms of clinical features, the classic and localized forms of lymphangioma circumscriptum differ in terms of patient's age at first appearance, size of the lesion, distribution over the body, histology, and symptoms (reviewed in Table 1).

Most of the vulvar lymphangioma circumscriptum cases reported in the literature have been classified as the classic type; however, localization and age at presentation seen in those cases are unusual for the classic type, which raises the possibility of misdiagnosis. Moreover, hyperkeratosis of the subcutaneous tissue can mimic external genital warts, adding to the difficulty of the diagnosis.[10,12,15,19,22,27]

Frequent complications of lymphangioma circumscriptum include swelling of the vulva, pain, recurrent cellulitis caused by excoriation or spontaneous oozing of the vesicles, and subsequent infection of the vulvar area (Landthaler et al. Arch Dermatol 1990;126:967–8).[6,15,17–19,21] The added complication of psychosexual dysfunction may lead to cessation of sexual activity.[6,8,21] A rare major complication is lymphangiosarcoma arising at the site of a preexisting lymphangioma circumscriptum, sometimes after radiation therapy[48,49] and sometimes not.[50] However, neither complication has been reported in a vulvar lymphangioma circumscriptum. One case of perianal squamous cell carcinoma was reported after surgery to remove lymphangioma circumscriptum that involved the vulvar and perianal areas.[9]

The etiologic factors for lymphangioma circumscriptum are not clear and may differ between the congenital and acquired forms. Congenital lesions seem to mature as patients mature and their external genital organs develop, until the lesions finally become apparent in adulthood.

Persistence or recurrence	New treatment	Total follow-up
12 mo	Treatment unreported	12 mo
NA	NA	NA
7 mo	Reexcision	21 mo
No		18 mo
9 mo	Reexcision	9 mo
		NA
No		81 mo
NA	NA	NA
NA	NA	NA
No	No	16 mo
No	No	18 mo

Considered as a circumscribed developmental defect of the lymphatics, congenital lymphangioma circumscriptum consists essentially of multiple lymphatic cisterns lying deep in the subcutaneous tissue[4,8] without direct communication to the general lymphatic system.[2] As cisterns communicate through abnormal muscle-coated channels that contact each other tonically, the pressure in the sequestered system increases, leading to the formation of saccular dilatations of the thin superficial lymphatics. Apparent vesicles represent the projection into the dermal papillae of these dilated superficial lymphatic channels.[2]

As for acquired lymphangioma circumscriptum, one proposed etiology is the architectural disruption of previously normal channels leading to sequestration and further dilatation of the lymphatics.[2] This theory is supported by the existence of diverse predisposing factors such as lymphangioma after dissection and radiation therapy at axillar sites in breast cancer,[36,38,41] at vulvar sites in cervical cancer (Landthaler et al. Arch Dermatol 1990;126:967–8),[11,13,16,18–20,26] or at different sites in tuberculosis at different localizations.[12]

Typical of congenital lymphangioma circumscriptum is a case that appeared in a 30-year-old black woman who presented at our clinic with a 3-year history of multiple painless vulvar papules. The patient reported no oozing, itching, dyspareunia, changes in her sexual life, or referred psychosexual dysfunction because of these lesions. She had undergone cesarean delivery 6 years before. The patient had no history of cancer surgery, radiation therapy, previous infections, or sexually transmitted diseases.

Physical examination showed a diffuse swelling of the vulva including both labia majorum and minorum from the mons pubis to the perineum. A thick subcutaneous tissue containing gray, coalescent, hyperkeratotic 1- to 5-mm vesicular papules was seen (Figure 1). No excoriation, crushing, or oozing was observed. The remaining findings on genital and physical examination were normal. Biopsy of the lesion revealed the features of lymphangioma circumscriptum, namely, dilatation of lymphatic channels in the papillary dermis, acanthosis, and hyperkeratosis (Figure 2).

The patient was offered several treatment options, including surgical excision or observation, and she chose observation initially. She was not sexually active at this time. Six months later, she became engaged and decided to marry. Once becoming sexually active, she experienced abrasion of the vesicles. At times, the abrasion was accompanied by cellulitis. After repeated episodes, she requested surgical therapy. She was seen in the gynecologic oncology clinic and the plastic surgery clinic because a large area of the vulva, mons pubis, and the skin overlying the pelvic brim were involved. A radical wide local excision of the involved areas was performed. Sufficient redundant skin was available to close the defect, and she has been followed for 18 months without evidence of recurrence.

The patient's age (30 years) places her near the mean age for women presenting with congenital lymphangioma circumscriptum (mean 33.5 years, range 14–76 years) (Cecchi et al. Acta Derm Venereol 1995;75:79–80)[3–10] (Table 2). In contrast, patients with acquired lymphangioma circumscriptum tend to be older (mean 47.5 years, range 9–75 years) (Landthaler et al. Arch Dermatol 1990;126:967–8)[5,6,11–26] (Table 3). Overall, lymphangioma circumscriptum affects women from childhood to old age (mean 42.5 years, range 9–76 years) (Cecchi et al. Acta Derm Venereol 1995;75:79–80; Landthaler et al. Arch Dermatol 1990;126:967–8).[3–26] Among the 20 published cases of acquired lymphangioma circumscriptum, the most frequent predisposing conditions are radical surgery and/or radiation for cervical neoplasia (from carcinoma in situ to stage III) (11 cases, 55.0%) (Landthaler et al. Arch Dermatol 1990; 126:967–8),[11,13,16,18,19,25,27] genital tuberculosis (three cases, 15.0%),[12,17,22,24] Crohn disease with vulvar or peritoneal fistulae (three cases, 15.0%),[15,18] acute cellulitis with group G β hemolytic streptococcus (one case, 5.0%),[20] varicose veins in both legs (one case, 5.0%),[23] and rhabdomyosarcoma (one case, 5.0%).[14]

In the 28 cases in which vesicular localizations were reported, the disease was limited to the vulva in 22 (78.6%) (Cecchi et al. Acta Derm Venereol 1995;75:79–80; Landthaler et al. Arch Dermatol 1990;126:967–

Table 3. Literature Review of Acquired Lymphangioma Circumscriptum of the Vulva

Reference	Patient age (y)	Predisposing conditions	Previous treatment
1970 Fisher and Orkin[27]	73	Squamous carcinoma of the cervix stage III Treatment by radiation 14 y before	Treatment for genital wart 4 y before*
1977 Rabinowitz et al[14]	9	Bladder rhabdomyosarcoma Treatment by pelvic exenteration and radiation	No
1979 Heuvel et al[17]	62	Tuberculosis	No
1980 Young et al[11]	54	Squamous carcinoma of the cervix stage Ib Treatment by lymphadenectomy and radiation 14 y before	NA
1985 LaPolla et al[16]	51	Squamous carcinoma of the cervix stage Ia Treatment by radical hysterectomy and radiation	No
1989 Handfield-Jones et al[18]	38	Squamous carcinoma of the cervix Treatment by radical hysterectomy and radiation 9 y before	No
	38	Crohn disease Peritoneal fistulae	No
	37	Crohn disease Perianal fistulae	Treatment for genital wart by cryotherapy*
1990 Kennedy[25]	41	Cervical carcinoma in situ treated with cone excision Recurrence treated by hysterectomy and radiotherapy Leg edema	Unreported
1990 Landthaler et al (letter)	75	Squamous carcinoma of the cervix Treatment by radical hysterectomy and radiation	No
1991 Sood et al[12]	40	Tuberculosis	No
1993 Akimoto et al[23]	46	Varicose veins of legs, hemorrhoidal status	No
1993 Harwood and Mortimer[19]	49	Squamous carcinoma of the cervix Treatment by radical hysterectomy, radiation, and colonic bladder	Treatment for genital wart by cryotherapy*
	50	Squamous carcinoma of the cervix Treatment by radical hysterectomy and radiation	No
1995 Begum et al[22]	22	Tuberculosis	Treatment for genital wart 3 y before*
1996 Buckley and Barnes[21]	42	β-hemolytic group G streptococcus with recurrent cellulitis	
1999 Celis et al[24]	69	Squamous carcinoma of the cervix Treatment by radical hysterectomy and radiation	No
1999 Mu et al[15]	44	Crohn disease Fistula	Treatment for genital wart with podophillin*
1999 Smith et al[13]	65	Squamous carcinoma of the cervix stage Ib Treatment by radical hysterectomy and radiation	No
2001 Gómez et al[26]	44	Lymphedema in lower right limb appeared 3 y before lesions on vulva	–

NA = not available.

* Possibly misdiagnosed as genital warts and treated as such before treatment for lymphangioma circumscriptum.

Treatment	Persistence or recurrence	New treatment	Total follow-up
Observation	–	–	36 mo
Vulvectomy	No	–	6 mo
Electrocoagulation	6 mo	NA	6 mo
NA	NA	NA	NA
Excision	NA	NA	NA
Skinning laser vulvectomy	2 mo	NA	2 mo
Local resection	No	–	36 mo
Liquid nitrogen	Yes	Surgery proposed	NA
Unreported	Unreported	Unreported	Unreported
CO_2 laser vaporization	Yes	Retreated, date unreported	18 mo
Simple vulvectomy	No	–	36 mo
None (patient refused) Observation	–	–	NA
Surgical excision planned	NA	NA	NA
Surgical excision	No	–	18 mo
Partial vulvectomy and skin grafting	No	–	36 mo
Observation	–	–	NA
CO_2 laser therapy (patient discontinued treatment)	NA	NA	NA
5-fluorouracil chemotherapy proposed (patient discontinued treatment)	NA	NA	NA
CO_2 laser therapy	3 and 12 mo	CO_2 laser therapy	36 mo
–	NA	NA	NA

Table 4. Treatment Summary

Treatment	Patients	Patients with recurrent or persistent disease	Patients with follow-up information	Patients with persistent or recurrent disease/patients with follow-up	
				n	%
Surgery	15	3	12	3/12	25
Abrasion	9	5	5	5/5	100
Observation	4	2	2	2/2	100
Not available	3	0	0	0	
Total	31	10	19	10/19	53

8),[3,5–12,15–19,23–26] extended to the mons pubis in four (including the present case) (14.3%),[4,13,22] extended to the perineum in one (3.6%),[21] and extended to the groin in one (3.6%).[4] Symptoms included fluid oozing in 16 of 28 cases (57.1%) (Cecchi et al. Acta Derm Venereol 1995;75:79–80; Landthaler et al. Arch Dermatol 1990; 126:967–8)* and swelling in 17 (60.7%) (Cecchi et al. Acta Derm Venereol 1995;75:79–80; Landthaler et al. Arch Dermatol 1990;126:967–8).[3,4,7–13,15,16,18–20]

Overall, there were cellulitis in nine of 31 cases (29.0%) (Landthaler et al. Arch Dermatol 1990;126:967–8),[6,15,17–19,21] pain in nine (29.0%),[3,6,12,15,17,22,27] pruritus in six (19.4%),[9,10,12,13,17,26] and lower-member lymphedema in seven (22.6%).[9,12,17,25,26] At time of consultation, patients reported that symptoms had been present for as few as 6 months to as long as 30 years. Psychosexual dysfunction was reported in four cases (12.9%),[4,6,8,21] sexual inactivity in five cases (16.1%),[3,7,13,15,22] and no perturbation of sexual activity in one case (ie, our case) (3.2%). Sexual activity was not mentioned in 20 cases (64.5%). In all 31 cases, the diagnosis was made by biopsy and histologic analysis. Misdiagnosis of and subsequent treatment for genital lesions occurred in five of 31 cases (16.1%).[15,18,19,22,27]

The literature review revealed that treatment modalities are the same for both congenital and acquired vulvar lymphangioma circumscriptum. These modalities include sclerotherapy,[8] electrocoagulation,[17] liquid nitrogen therapy,[18] carbon dioxide laser therapy (Landthaler et al. Arch Dermatol 1990;126:967–8; Blum EJ. Lymphangioma circumscriptum [letter]. J Dermatol Surg Oncol 1988;14:1033),[6,13,17,18,24,51,52] and surgical excision.[2,10,51,53] Because recurrence is the rule when using any of these methods, observation of an asymptomatic patient can be proposed.[3]

Treatments are summarized in Table 4. In 15 of 31 cases (48.4%), including ours, surgical management (wide local excision, simple vulvectomy, or radical vulvectomy) was chosen.† Follow-up ranged from 6 to 81 months (median 23.1 months), and the recurrence rate was 25%.[4,7,9] Abrasive modalities, including carbon dioxide laser (Landthaler et al. Arch Dermatol 1990;126:967–8),[3,6,13,18,29] liquid nitrogen,[18] electrocoagulation,[17] 5-fluorouracil,[15] or sclerosing therapy[8] were used in nine patients (29.0%). One of those patients (11.1%) discontinued treatment,[8] and in another four for whom follow-up was reported, disease recurred within 7 months (mean 4.5 months). Observation was proposed in two of 31 cases (6.5%).[20,21] Two others (6.5%) refused to be treated (Cecchi et al. Acta Derm Venereol 1995;75:79–80),[23] and they were observed. Data were not available for treatment in three cases (9.7%)[11,25,26] and for follow-up in 13 (Cecchi et al. Acta Derm Venereol 1995; 75:79–80).‡

In summary, six of 31 patients (19.4%)[12,14,18,19,22] were cured by a first treatment, six (19.4%) were cured by a second (Landthaler et al. Arch Dermatol 1990;126: 967–8),[3–7] and one (3.2%) was cured after a third.[9] Disease recurred in five of nine patients (55.6%) undergoing abrasion therapy and three of 13 patients (23.1%) undergoing surgical therapy. The numbers were too small to identify conclusively the best treatment; however, inasmuch as three patients (three of 31, 9.7%)[8,15,24] discontinued treatment, observation seems warranted in the nonmotivated asymptomatic patient. Although no therapies have been subjected to a randomized clinical trial, surgical therapy was far more successful than abrasive therapy. In some cases, the surgical therapy was described as a simple vulvectomy, whereas in other cases, it was described as a radical vulvectomy. Clearly, the surgeon must tailor the procedure to fit the extent and distribution of the disease.

CONCLUSION

Lymphangioma circumscriptum rarely occurs in the vulva. Only 31 cases have been described (11 congenital and 20 acquired). This uncommon pathology poses a diagnostic challenge because its clinical appearance at

*References 4–6, 8–10, 12, 13, 18, 21, 22, 24–26.

†References 4–7, 10, 12, 14, 16, 18, 19, 22.

‡References 3, 5, 8, 11, 15, 16, 19, 21, 23, 24–26.

presentation can mimic such infectious processes as molluscum contagiosum or genital warts and lead to improper therapy. Observation of this benign entity is an acceptable strategy. Surgical therapy can be curative and in the literature appears the best option if the patient desires therapy. A database of these cases should be created so that long-term follow-up of several treatment options can be evaluated. The database should include such variables as symptoms, characteristics of lesions, etiologic factors, predisposing medical conditions, treatment, complications, follow-up record, recurrence, retreatment, and results.

From this comprehensive review, which includes the clinicopathologic features of both the congenital and acquired forms, illustrations from the rarest form, and a summary of treatment approaches, we conclude that lymphangioma circumscriptum poses a diagnostic challenge the risks of which are misdiagnosis and mistreatment. These risks would likely be reduced were a database of cases accessible that permitted long-term follow-up and better assessment of presenting characteristics and treatment options.

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