Lymphocytic thrombophilic arteritis and cutaneous polyarteritis nodosa: Clinicopathologic comparison with blinded histologic assessment

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Background: Lymphocytic thrombophilic arteritis (LTA), or macular lymphocytic arteritis, is defined by a primary lymphocytic vasculitis. However, the nosology of LTA has been controversial, with speculation that it may represent an indolent non–nodule-forming variant of cutaneous polyarteritis nodosa (cPAN).

Objective: This study compares the clinicopathologic features of patients with LTA or cPAN to assess if these conditions should be considered distinct entities.

Methods: This is a cross-sectional study of all LTA and cPAN cases at a single tertiary center using prospectively collected clinical data and blinded histologic assessment.

Results: The study included 17 patients with LTA and 13 patients with cPAN. Clinically, cases of LTA were distinguished by a more widespread pattern of livedo racemosa, which was noninfiltrated and asymptomatic. In contrast, cPAN was associated with localized starburst livedo, purpura, and episodic features including nodules, pain, and large inflammatory ulcers. When patients were separated according to the presence (>5%) or paucity (≤5%) of neutrophils on blinded histology review, they had distinct clinical features and differences in disease course.

Limitations: This was a single-center study.

Conclusion: Our data support the classification of LTA and cPAN as separate entities rather than a spectrum of the same disorder and highlight the importance of clinicopathologic correlation in distinguishing these conditions. (J Am Acad Dermatol 2020;83:501-8.)

Key words: cutaneous polyarteritis nodosa; livedo racemosa; lymphocytic thrombophilic arteritis; lymphocytic vasculitis.

INTRODUCTION

Lymphocytic thrombophilic arteritis (LTA), or macular lymphocytic arteritis, is a recently described entity defined by a primary lymphocytic vasculitis. Pathologically, the condition is characterized by dense lymphocytic inflammation affecting small- to medium-sized arteries in the reticular dermis and upper subcutis associated with fibrin deposition classically forming a distinct luminal fibrin ring.[1] Clinically, LTA presents with persistent asymptomatic widespread livedo racemosa or macular hyperpigmentation on the lower limbs and typically has a chronic indolent course,[1] though peripheral neuropathy[2] and ulceration[3-5] have been reported.

Cutaneous polyarteritis nodosa (cPAN) is a cutaneous medium vessel vasculitis characterized by neutrophilic inflammation of small to medium arteries in the reticular dermis and upper subcutis, with

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fibrinoid necrosis.[6] Clinically, cPAN presents with painful nodules, purpura, ulceration, and livedo racemosa on the lower limbs.[6] Extracutaneous features, such as fever, arthralgia, myalgia, and peripheral neuropathy, are often present.[6] Importantly, cPAN is distinct from systemic polyarteritis nodosa (PAN), which is a medium vessel vasculitis affecting multiple organ systems.[7] It is rare for cPAN to progress to systemic PAN, and only 2 such cases have been documented in the published literature.[8]

The relationship between LTA and cPAN has been controversial, with some authors speculating that LTA may represent an indolent non–nodule-forming variant of cPAN.[4,5,9,10] This has been underscored by perceived clinical similarities between these conditions and the finding that the subacute and reparative stages of cPAN may have a predominantly lymphocytic infiltrate with appearances similar to LTA.[4,5,9,10] The nosology of LTA remains a dilemma in the literature, with no gold standard criteria on how it should be distinguished from cPAN. This study aimed to compare in detail the clinicopathologic features of patients with LTA or cPAN and to assess if these conditions may be considered as distinct entities.

CAPSULE SUMMARY

- Lymphocytic thrombophilic arteritis (LTA) presents with asymptomatic widespread livedo racemosa or macules.
- In contrast, cutaneous polyarteritis nodosa (cPAN) has localized starburst livedo, purpura, and episodic features including nodules, pain, and large inflammatory ulceration.
- Clinicopathologic correlation is important in distinguishing these conditions because cPAN can sometimes have a paucity of neutrophils on histology.

METHODS

All cases of LTA or cPAN diagnosed between 2008 and 2018 at St Vincent's Hospital in Melbourne, Australia, were included in this study. Diagnoses were established based on clinical as well as histologic criteria, with examination of multiple biopsy specimens and serial sections. One patient was excluded because histologic specimens were not available for subsequent blinded assessment.

Patients with LTA were diagnosed based on clinical and histologic criteria including: 1) persistent or slowly progressive livedo racemosa or macular pigmentation affecting the lower limbs or other sites, such as the buttocks, trunk, and upper limbs; 2) biopsy specimen evidence of inflammation of medium-sized arteries in the reticular dermis, the dermosubcutaneous junction, or the superficial subcutis with 3) a predominance of lymphocytes and absent or scant neutrophils and 4) significant intramural or intraluminal fibrin deposition in the affected vessels.

Patients with cPAN were diagnosed based on clinical and histologic criteria including: 1) livedo racemosa or nodules or ulceration and 2) biopsy specimen evidence of inflammation of medium-sized arteries in the reticular dermis, the dermosubcutaneous junction, or the superficial subcutis, with 3) prominent neutrophils in clinically acute lesions or 4) a more lymphohistiocytic infiltrate in clinically subacute lesions.

Clinical characteristics for each case were assessed prospectively (Table I). The presence, appearance, and distribution of livedo racemosa was evaluated, with a widespread pattern defined as extensive involvement of the lower limbs in addition to the buttocks, trunk, or upper limbs (Fig 1, *A*). Livedo racemosa was also assessed for features such as blanching with pressure, induration, or infiltration, as well as purpura. Starburst livedo was defined as a distinct form of livedo racemosa with well-demarcated localized patches consisting of a dense reticulate pattern (Fig 1, *B*). Ulceration was classified by morphology as either large inflammatory (≥10 mm) or small punctate (<10 mm; Fig 1, *C*). Peripheral neuropathy was confirmed through nerve conduction studies. Patients were determined to have a relapsing–remitting disease course if cutaneous features (eg, livedo, nodules, or ulcers) fluctuated or resolved over days to weeks. Serum antinuclear antibody testing was considered positive if greater than or equal to a titer of 1:160. When available, laboratory assessments for inherited and acquired thrombophilia were also reviewed.

Histologic specimens for each case were blindly reviewed by the same dermatopathologist. Specimens were assessed for a predominance of lymphocytes (>50%) as well as the presence (>5%) or paucity (≤5%) of neutrophils; this was determined as an estimated percentage and averaged across several high-power fields. Vascular inflammation relating to the overall number and density of inflammatory cells was graded from 1 to 3 (1, mild; 2,

Table I. Demographic, clinical and histologic features of patients with cutaneous polyarteritis nodosa or lymphocytic thrombophilic arteritis

Characteristic	LTA (n = 17)	cPAN (n = 13)	*P* value
Female, n (%)	13/17 (76)	9/13 (70)	.70
Median age at presentation, years (IQR)	27 (24-46)	50 (37-63)	.01*
Asian racial background, n (%)	7/17 (41)	1/13 (8)	.09
Clinical features, n (%)			
Presence of livedo racemosa	15/17 (88)	9/13 (69)	.36
Lower limb livedo racemosa	15/15 (100)	9/9 (100)	N/A
Widespread pattern of livedo racemosa†	14/15 (93)	2/9 (22)	.001*
Localized starburst livedo‡	0/15 (0)	6/9 (66)	.001*
Erythematous blanchable livedo	15/15 (100)	0/9 (0)	<.001*
Purpuric livedo	0/15 (0)	4/9 (44)	.03*
Pigmented macules	4/17 (24)	0/13 (0)	.09
Nodules or infiltration	0/17 (0)	10/13 (77)	<.001*
Presence of ulcers	3/17 (18)	5/13 (38)	.24
Large inflammatory ulceration§	0/3 (0)	5/5 (100)	.02*
Discomfort, pain, or tenderness	3/17 (18)	12/13 (92)	<.001*
Arthralgia	0/17 (0)	4/13 (31)	.03*
Fever	0/17 (0)	2/13 (15)	.18
Peripheral neuropathy	3/17 (18)	1/13 (8)	.36
Relapsing–remitting disease course	3/17 (18)	11/13 (85)	.001*
Positive ANA,‖ n (%)	7/13 (54)	7/13 (54)	1.00
Thrombophilic abnormalities,‖ n (%)	5/13 (38)	0/6 (0)	.11
Blinded histologic assessment			
Neutrophils >5% of infiltrate, n (%)	0/17 (0)	9/13 (70)	<.001*
Lymphocytes >50% of infiltrate, n (%)	14/17 (82)	6/13 (46)	.06
Mean inflammation grade (95% CI)	1.8 (1.5-2.2)	2.5 (2.0-2.9)	.02*
Mean fibrin deposition grade (95% CI)	2.5 (2.0-2.9)	1.5 (0.9-2.2)	.02*
Fibrin ring, n (%)	10/17 (59)	6/13 (46)	.71
Treatment, n (%)			
No treatment	1/17 (6)	0/13 (0)	N/A
Aspirin	15/17 (88)	1/13 (8)	<.001*
Pentoxifylline	14/17 (82)	4/13 (31)	.16
Colchicine	2/17 (12)	9/13 (69)	.002*
Prednisolone	4/17 (24)	10/13 (77)	.009*
Other immunosuppression	5/17 (29)	6/13 (46)	.45

ANA, Antinuclear antibody; *CI*, Confidence interval; *cPAN*, cutaneous polyarteritis nodosa; *IQR*, interquartile range; *LTA*, lymphocytic thrombophilic arteritis; *N/A*, not applicable.
*Statistically significant.
†Widespread livedo racemosa of lower limbs with additional involvement of buttocks, trunk, or upper limbs.
‡Starburst livedo defined as distinct well-demarcated localized livedo patches consisting of a dense reticulate pattern.
§Ulceration defined as either large inflammatory (≥10 mm) or small punctate (<10 mm).
‖ANA and thrombophilia assessments were not performed in all patients.

moderate; and 3, severe). The amount of vessel mural and luminal fibrin deposition was graded from 0 to 3 (0, absent; 1, mild; 2, moderate; and 3, severe), and each case was assessed for the presence of a hyalinized fibrin ring.

The Fisher's exact test was used to compare qualitative data. The 2-sided *t* test was used to compare quantitative data. Statistical analyses were performed with Stata software (version 14; StataCorp, College Station, TX). Institutional review board approval was obtained from St Vincent's Hospital Melbourne.

RESULTS

In total, 17 patients with LTA and 13 patients with cPAN were included in the study. The LTA group included 5 patients that we have previously reported.[2,3,11] The clinical, demographic, and histologic features of these patients are summarized in Table I.

Clinical characteristics

Most patients with LTA or cPAN were female. Patients with LTA presented at a younger age (median 27 years; range 19-63 years) than those with

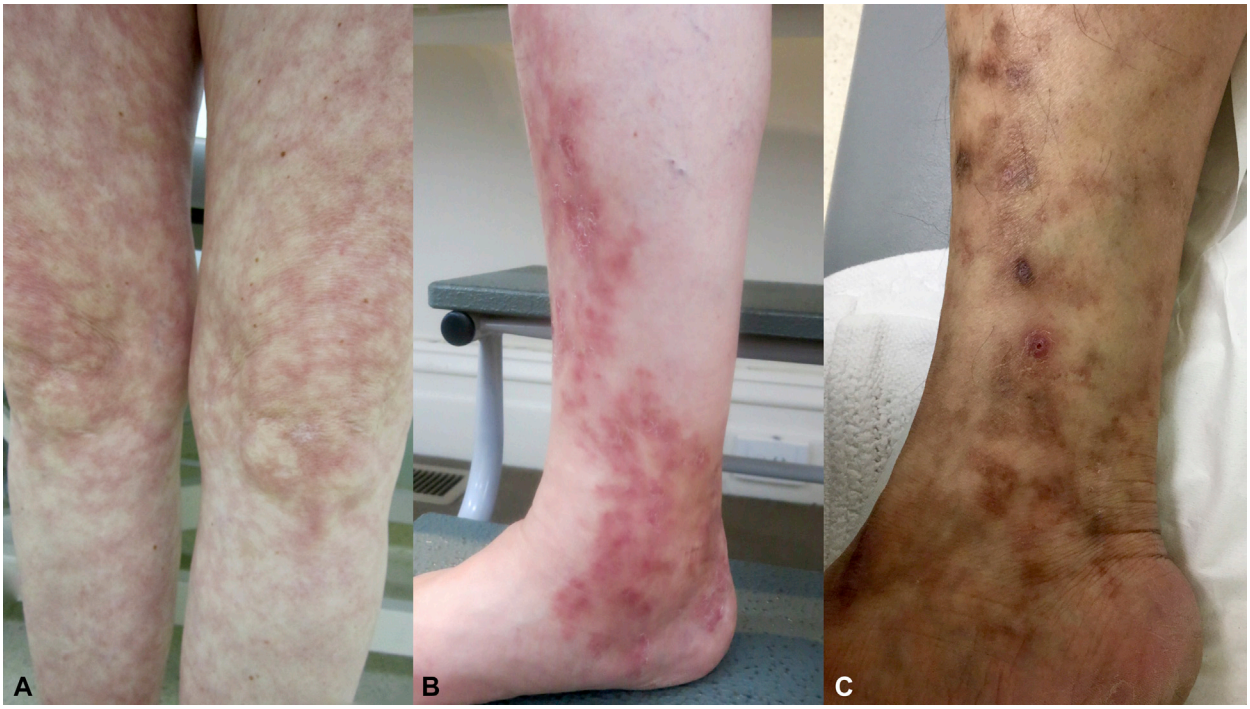

Fig 1. Clinical features of patients with lymphocytic thrombophilic arteritis (LTA) and cutaneous polyarteritis nodosa (cPAN). **A**, Widespread noninfiltrated blanchable livedo racemosa on the thighs and lower leg in a patient with LTA. **B**, Localized starburst livedo racemosa with nodules and infiltration on the right shin and ankle in a patient with cPAN. **C**, Small punctate ulcers and background livedo racemosa on the right ankle in a patient with LTA.

cPAN (median 50 years; range 30-74 years; $P = .008$). The median follow-up period for all patients was 4 years (range 0.5-10 years). A higher proportion of patients with LTA (7/17; 41%) than cPAN (1/13; 8%) were Asian, but this did not reach statistical significance ($P = .09$).

Most patients who had either condition presented with livedo racemosa involving the lower limbs. However, compared to those with cPAN (2/9; 22%), LTA (14/15; 93%) was associated with widespread livedo racemosa (Fig 1, *A*) also affecting the upper limb, trunk, or buttocks ($P = .001$). The localized pattern of starburst livedo (Fig 1, *B*) was exclusively found in patients with cPAN (6/9; 67%) and not in those with LTA ($P = .001$). Purpuric livedo was also only present in patients with cPAN (4/9; 44%) and not in those with LTA ($P = .03$). The livedo racemosa in all patients with LTA was erythematous and blanched with pressure, a feature that was not present in those with cPAN ($P < .001$). Four patients with LTA had pigmented macules, including 3 with a macular annular eruption we have previously described.[3]

Nodules or infiltration and induration of livedo was noted in the majority of patients with cPAN (10/13; 77%), but not in those with LTA ($P < .001$). Most patients with cPAN (12/13; 92%) had cutaneous discomfort, pain, or tenderness, while in LTA this was only a feature in 3 patients when ulceration was present ($P < .001$). Livedo racemosa or macular pigmentation in all patients with LTA was noninfiltrated and asymptomatic.

Ulceration was present in 3 patients with LTA and 5 patients with cPAN. The ulcers in the 5 patients with cPAN were large and inflammatory in contrast to the small punctate ulcers (Fig 1, *C*) seen in the 3 patients with LTA. Systemic symptoms, such as fever and arthralgias, were only noted in patients with cPAN ($P = .18$ and $P = .03$, respectively). A symmetric bilateral lower limb peripheral neuropathy was detected in 3 patients with LTA and 1 patient with cPAN.

A similar proportion of patients with LTA or cPAN had serum antinuclear antibody positivity. However, a proportion of LTA patients had underlying thrombophilic abnormalities, including factor V Leiden heterozygosity (3 patients), low antithrombin III (1 patient), and anticardiolipin immunoglobulin M positivity (1 patient) in the absence of thromboembolic events. No thrombophilic abnormalities were noted in the 6 patients with cPAN who were tested.

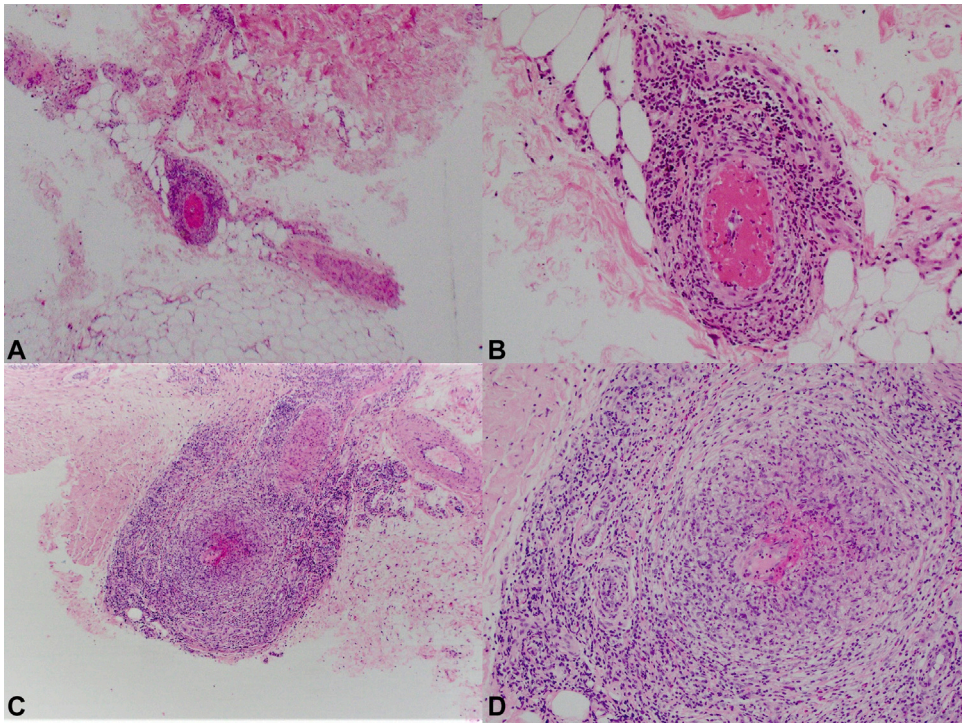

Fig 2. Histologic features of patients with lymphocytic thrombophilic arteritis (LTA) and cutaneous polyarteritis nodosa (cPAN). Histology from a patient with LTA under (**A**) low (×4) and (**B**) high (×20) magnification demonstrating medium-size vessel lymphocytic inflammation with prominent fibrin deposition. Histology from a patient with cPAN under (**C**) low (×4) and (**D**) high (×20) magnification showing intense neutrophilic inflammation of a medium size vessel with leukocytoclasis.

Disease course and treatment

Compared with patients with LTA, most patients with cPAN (11/13; 85%) had an acute relapsing–remitting disease course (P = .001) with episodic features, such as painful or tender nodules, ulceration, arthralgias, and fevers occurring over a period of days to weeks. Most patients with cPAN (9/13; 70%) were managed with courses of prednisolone and later transitioned to maintenance colchicine. Six patients were also treated with steroid-sparing agents, such as cyclosporine and mycophenolate mofetil. The episodic features in all patients with cPAN demonstrated a response to immunosuppressive or antiinflammatory agents, with early resolution or a reduction in frequency of exacerbations. Two patients with cPAN (2/13; 15%) did not have episodic features but had persistent tender livedo racemosa, which subsequently improved with a reduction in discomfort and infiltration after colchicine therapy. Three patients with cPAN showed a recurrence of nodules and pain when colchicine or prednisolone was ceased. Aspirin or pentoxifylline was trialed in 5 patients with cPAN, but no improvement in symptoms was noted.

Most patients with LTA (14/17; 82%) had a chronic indolent disease course with persistent asymptomatic noninfiltrated livedo racemosa or pigmented macules. Two patients with LTA had annular pigmented macules that responded well to treatment with aspirin and pentoxifylline.[3] Three patients with LTA (3/17; 18%) had additional episodic punctate lower limb ulcers that healed after treatment with aspirin and pentoxifylline.[3] Colchicine and prednisolone were trialed to treat ulcers in 1 patient with LTA but this was found to be ineffective. As previously reported,[11] 1 patient with LTA subsequently developed mononeuritis in addition to testicular infarcts, though investigations did not reveal any evidence of other organ involvement. In addition to pentoxifylline and aspirin, this patient was anticoagulated using rivaroxaban and underwent immunosuppressive therapy with systemic steroids and rituximab, with a subsequent improvement in the extracutaneous symptoms.

Blinded histology review and clinical correlation

Cutaneous medium vessel inflammation with the presence of neutrophils (>5% of infiltrate) was noted

Table II. Patient demographic and clinical features according to the presence (≥5% of infiltrate) or paucity (<5% of infiltrate) of neutrophils seen on blinded histologic assessment

Characteristic	Neutrophils <5% (n = 21)	Neutrophils ≥5% (n = 9)	*P* value
Female (%)	16/21 (76)	6/9 (67)	.67
Median age at presentation, years (IQR)	32 (25-48)	56 (37-70)	.008*
Asian racial background, n (%)	8/21 (38)	0/9 (0)	.07
Clinical features, n (%)			
Presence of livedo racemosa	19/21 (90)	5/9 (56)	.05
Lower limb livedo racemosa	19/19 (100)	5/5 (100)	NA
Widespread pattern of livedo racemosa†	15/19 (79)	1/5 (20)	.03*
Localized starburst livedo‡	3/19 (16)	3/5 (60)	.08
Erythematous blanchable livedo	15/19 (79)	0/5 (0)	.003*
Purpuric livedo	2/21 (10)	2/9 (22)	1.00
Pigmented macules	4/21 (19)	0/9 (0)	.22
Nodules or infiltration	4/21 (19)	6/9 (67)	.03*
Presence of ulcers	4/21 (19)	4/9 (44)	.20
Large inflammatory ulceration§	1/4 (25)	4/4 (100)	.14
Discomfort, pain, or tenderness	7/21 (33)	8/9 (89)	.01*
Arthralgia	1/21 (5)	3/9 (33)	.07
Fever	1/21 (5)	1/9 (11)	.52
Peripheral neuropathy	4/21 (19)	1/9 (11)	1.00
Relapsing–remitting disease course	6/21 (29)	8/9 (89)	.004*
Positive ANA,‖ n (%)	10/17 (59)	4/9 (44)	.68
Thrombophilic abnormalities,‖ n (%)	5/14 (36)	0/6 (0)	.26

ANA, Antinuclear antibody; *IQR*, interquartile range.
*Statistically significant.
†Widespread livedo racemosa of lower limbs with additional involvement of buttocks, trunk, or upper limbs.
‡Starburst livedo defined as distinct well-demarcated localized livedo patches consisting of a dense reticulate pattern.
§Ulceration defined as either large inflammatory (≥10 mm) or small punctate (<10 mm).
‖ANA and thrombophilia assessments were not performed in all patients.

on blinded histology review in most patients with cPAN (9/13; 70%) and not in those with LTA ($P < .001$). Four patients with cPAN had a paucity of neutrophils (≤5% of infiltrate) on histology, though clinical features of cPAN (eg, nodules, pain or tenderness) were present.

Patients with LTA had less intense medium vessel inflammation ($P = .02$) and greater relative fibrin deposition ($P = .02$) compared with patients with cPAN (Table I; Fig 2). Interestingly, the presence of an intramural or luminal fibrin ring did not distinguish either condition. Likewise, a predominance of lymphocytes (>50%) in the perivascular inflammatory infiltrate was not specific for LTA.

When patients were separated according to the presence (>5% of infiltrate; n = 9) or paucity (≤5% of infiltrate; n = 21) of neutrophils on blinded histology review, there were clear differences in clinical features between the groups (Table II). The presence of neutrophils (>5% of infiltrate) was associated with an older age of disease onset ($P = .01$), nodules ($P = .03$), pain or tenderness ($P = .01$), and an episodic relapsing remitting course of disease ($P = .004$). A paucity of neutrophils (≤5% of infiltrate) was associated with widespread livedo racemosa ($P = .03$), which was erythematous and blanchable ($P = .003$), with a persistent disease course lacking episodic features.

DISCUSSION

To date, 50 previous cases of LTA have been reported in the literature. Most have involved young female patients, similar to our cohort.[12] The patients with LTA in this study were on average younger than those with cPAN. Like other reports,[12] a number of our patients with LTA were of Asian descent. Similarly, some patients with LTA also had thrombophilic abnormalities.

Clinically, cases of LTA in our study were distinguished from cPAN by the presence of asymptomatic, widely distributed, noninfiltrated blanching livedo racemosa or pigmented macules; most had a persistent indolent course lacking episodic features. In contrast, patients with cPAN were characterized by purpuric livedo racemosa localized to the lower limb in a starburst pattern; episodic features, such as painful nodules or infiltration, large inflammatory ulcers, and arthralgia were frequently present. A

small number of patients with LTA had small punctate ulceration with morphology distinct from the large inflammatory ulcers of cPAN. One previously reported patient with LTA developed testicular infarcts and mononeuritis in the absence of other organ involvement, highlighting the importance of monitoring in this condition.[11] Importantly, no patients in the study progressed from one pattern of disease to the other; none of those diagnosed with LTA subsequently developed cutaneous features more consistent with cPAN or vice versa.

Histologically, the defining difference between cPAN and LTA is the primary inflammatory cell underlying medium vessel inflammation.[12] LTA is described as a primary lymphocytic vasculitis with a relative paucity of neutrophils,[12] while cPAN is characterized by neutrophilic inflammation.[13] Consistent with this, when patients in our study were separated according to the presence (>5% of infiltrate) or paucity (≤5% of infiltrate) of neutrophils on blinded histology review, the groups had distinct clinical features and differences in disease course.

There were other histologic differences apparent on blinded assessment. Patients with LTA had more fibrin deposition and less intense inflammation than patients with cPAN. However, the previously described vascular fibrin ring of LTA was not found to be a distinguishing feature, as suggested by some authors.[14] While not assessed in our cohort, one recent pathologic study demonstrated that most but not all cases of LTA have a preserved internal elastic lamina, suggestive of an endovasculitic process distinct from the transmural arteritis of cPAN.[15]

The importance of using clinicopathologic correlation when distinguishing LTA and cPAN was highlighted by this study. Biopsy specimens in the study were preferentially obtained from the most recent or inflamed skin lesions. However, while neutrophils (>5% of infiltrate) were readily identified in most of our patients with clinical features of cPAN, a small number showed a paucity (≤5% of infiltrate) of neutrophils on histology. In addition, some patients with cPAN had a lymphocyte predominant infiltrate (>50%). This is reflective of the histologic changes found in different stages of cPAN, with subacute and reparative phases potentially having fewer neutrophils and being more lymphohistiocytic.[13] In such cases, consideration of clinical features is crucial in reaching the appropriate diagnosis.

Notably, some authors have speculated that LTA is a variant of cPAN on the basis of histologic similarities between LTA and subacute or reparative cPAN.[5,9,10] However, many of these studies lacked clinicopathologic correlation and their conclusions did not account for the presence of distinguishing clinical features of cPAN.[5,9,10] Specific features of cPAN identified by our study, such as cutaneous tenderness or pain, as well as a relapsing–remitting disease course, were not assessed or highlighted.[9,10,16,17] In addition, these were studies in cohorts of patients with a diagnosis of cPAN and comparisons were limited by the inclusion of few, if any, patients with LTA.[9,10]

The importance of correctly distinguishing LTA from cPAN is underscored by differences in management, with misdiagnosis potentially leading to unnecessary and harmful immunosuppressive therapy. Similar to other cohorts, episodic features such as painful or tender nodules and ulceration in our patients with cPAN responded well to systemic corticosteroids, colchicine, or immunosuppressive therapy.[8] In contrast, most of our patients with LTA had an indolent disease course with persistent asymptomatic livedo racemosa or pigmented macules despite therapies trialed. However, as previously described,[3] some of our patients with LTA responded to aspirin and pentoxifylline with resolution of pigmented macules or punctate ulcers, suggesting that hypercoagulability may be a contributing factor in the pathophysiology of this condition.

In summary, our findings show that LTA and cPAN have distinct clinical and histologic characteristics, supporting their classification as separate entities rather than a spectrum of the same disorder. Clinicopathologic correlation is needed when distinguishing LTA from cPAN. These conditions respond to different treatments, and an accurate diagnosis is important to avoid unnecessary immunosuppressive therapy. The main limitation of this study was its single-center design; larger multicenter studies with long-term follow-up will help clarify the differences between these two conditions.

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