Journal of Pakistan Association of Dermatologists. 2017; **27(4)**: 381-383.

Original article

Cutaneous vasculitis: a series of 56 patients

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Abstract

Objective To document the clinical features, type of vasculitis and underlying causative factors in cutaneous vasculitis.

Methods In this retrospective study we reviewed the medical records of 56 patients with biopsy-proven cutaneous vasculitis. Age and sex, age of onset, past medical history, recent drug history, laboratory data, site of involvement, symptoms of lesions and pathological findings.

Results Female patients outnumbered males (35 vs. 21). The mean age of onset was 37.7±18.6 year for women and 41.7±16.4 year for men. Lower extremities were the most frequent affected site (100%). Purpura (58.6%) and papules/plaques (55.4%) were the predominant lesions. The most common type of biopsy proven cutaneous vasculitis was cutaneous leukocytoclastic angiitis followed by vasculitis associated with probable etiology.

Conclusion
Females were frequently affected. The disease predominantly affected lower legs as purpuric or papular eruption. Cutaneous leukocytoclastic angiitis and vasculitis associated with probable etiology were the frequent types.

Key words
Vasculitis, cutaneous leukocytoclastic vasculitis.

Introduction

Cutaneous vasculitis (CV) is defined as the inflammation of the blood vessels of the dermis and is a manifestation that can be seen in a variety of settings.[1] Clinically and etiologically, CV is a heterogeneous group. CV may affect skin only or can involve the internal organs, as well. About 50% of cases are idiopathic in origin whereas in the rest, the underlying causes may be infections, drugs or chemicals, autoimmune diseases and neoplastic diseases. Although, palpable purpura is said to be the key clinical feature of CV, the disease may present as erythematous macules, hemorrhagic vesicles and bullae, pustules, papulonodular lesions and ulcers. Similarly, legs are the predominant affected site but other anatomical regions of the body can be affected.

This study was undertaken to document clinical manifestations, types, sites of involvement and underlying causes in LV.

Methods

In this retrospective study, we reviewed the clinical manifestations, laboratory findings, pathological changes and etiologic associations of CV. The study protocol was approved by the

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Institutional Review Board (IRB) of Tehran University of Medical Sciences.

We studied 56 patients with biopsy proven CV visiting the dermatology department of Razi Hospital, Tehran, Iran.

The following data were extracted from the patients' medical records: age and sex, age of onset, past medical history, recent drug history, laboratory data, site of involvement, symptoms of lesions and pathological findings.

Independent samples t-test was used to compare the means. Statistical analysis was performed using SPSS 15.0 (SPSS Inc, Chicago, IL). A *p* value <0.05 was considered statistically significant.

Results

Of 56 patients with CV, 35 (62.5%) were female patients, and 21 (37.5%) were males. The age of onset ranged from 5 to 79 year with a mean age of 39.2±17.8 year. The mean age of onset was 37.7±18.6 year for women and 41.7±16.4 year for men ($p = 0.41$).

The most common site of involvement was lower extremities, presenting in all patients. The different sites of involvement and their prevalence are shown in **Table 1**.

The patients' symptoms were also recorded. Thirty-nine (69.6%) patients had painful lesions, 21 (37.5%) had a burning sensation and 15 (26.8%) had itching. Two of the patients (3.6%) had no symptoms.

The most common types of lesions were petechiae/purpura/ecchymosis followed by papule/plaque and ulcer. More details are shown in **Table 2**.

Table 1 Site of lesions (n=56).

Site of lesions	*N (%)*
Lower extremities	56 (100)
Upper extremities	41 (73.2)
Anterior surface of the trunk	31 (55.4)
Posterior surface of the trunk	23 (41.1)
Head and neck	18 (32.1)
Mucous membranes	9 (16.1)
Palms and soles	8 (14.3)
Genitalia	2 (3.6)

Table 2 Types of lesions (n=56).

Type of lesions	*N (%)*
Petechiae/purpura/ecchymosis	33 (58.9)
Papule/plaque	31 (55.4)
Ulcer	29 (51.8)
Macule/patch	22 (39.3)
Vesicle/bulla	19 (33.9)
Target lesion	3 (5.4)

Table 3 Site of lesions (n=56).

Subtype of disease	*N (%)*
Cutaneous leukocytoclastic angiitis	21 (37.5)
Vasculitis associated with probable etiology	20 (35.7)
IgA vasculitis	6 (10.7)
Hypocomplementemic urticarial vasculitis	5 (8.9)
Polyarteritis nodosa	3 (5.4)
Vasculitis associated with systemic disease	1 (1.8)

Table 4 Types of lesions (n=56).

Probable etiology of vasculitis	*N (%)*
Drugs	10 (50)
Infection	7 (35)
Malignancy	2 (10)
Pregnancy	1 (5)

Regarding symptomatology, 29 (69.6%) patients had painful lesions, 21 (37.5%) had a burning sensation and 15 (26.8%) had itching. 2 (3.6%) patients had no symptoms. Edema in the lower extremities and arthralgia were the most common accompanying symptoms seen in 25 (44.6%) and 20 (35.7%) patients, respectively.

The prevalence of different subtypes of disease among patients is shown in **Table 3**. The diagnosis was made based on a combination of clinical and histopathological findings.

Pathological features were also reviewed. Lymphocyte was the most common cell mentioned in pathology reports. We also assessed probable etiologic associations of the disease **(Table 4)**. All vasculitides with probable etiology were named as 'vasculitis associated with probable etiology' according to 2012 revised international Chapel Hill Consensus Conference on the nomenclature of vasculitides (CHCC 2012).[2] Moreover, one patient had vasculitis associated with systemic disease which was a separate category in CHCC 2012 classification.

The most common laboratory findings were high erythrocyte sedimentation rate and low hemoglobin levels seen in 27 (48.2%) and 15 (26.8%) patients, respectively. Fourteen (25%) patients had abnormal urinalysis results including hematuria or proteinuria.

Discussion

Our results showed that mucous membranes were involved in 16% of the patients with CV, while previous studies reported rare mucous membrane involvement in CV.[3] This could reflect a higher rate of mucous membrane involvement among Iranian CV patients. Furthermore, nephropathy was present in 25% of our patients, while Martinez *et al*.[4] reported nephropathy only in 7.4% of their cases.

Our results demonstrated that cutaneous leukocytoclastic angiitis was the most common type of biopsy proven CV which was in accordance with the study conducted by Blanco *et al*.[5] Moreover, Blanco *et al*.[5] suggested that 10% of the patients with biopsy-proven CV had vasculitis associated with systemic disease, while this type of vasculitis was less prevalent in our study.

Since Razi hospital is a specialized dermatology hospital, vasculitis associated with systemic disease and variable vessel vasculitis are probably underestimated in our study. According to CHCC 2012 classification, it seems that more patients with cutaneous leukocytoclastic angiitis will be named as vasculitis associated with probable etiology in the future.

Cutaneous vasculitis is a relatively rare disease. In this study, we demonstrated that in a patient with lower extremity purpura, even with no history of recent infections or medications, vasculitis should be considered. Furthermore the involvement of head and neck or mucosal membranes does not rule out vasculitis. Moreover, ulcer-type lesions can be the prominent feature of the disease.

Our study has its limitations, including its retrospective nature and missing data.

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