Original Article

Clinicopathological Study on Cutaneous Vasculitis

Siraj MS[1], Rahman MM[2], Karim ATMR[3], Wahab MA[4]

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Abstract

Introduction: Cutaneous vasculitis is an inflammatory process directed primarily at vessels which results in the destruction of the vessel walls leading to hemorrhage, ischemia, and/or infarction. Cutaneous involvement in cutaneous vasculitis may be primary or reflector of a systemic disease.

Aim: To find out the aetiology and clinicopathologic features of cutaneous vasculitis in Chattogram costal area.

Methods: A total of 50 patients diagnosed clinically and confirmed histologically as cutaneous vasculitis were selected for this descriptive cross-sectional study from March 2016 to August 2017. Detailed history, clinical examination and the baseline investigations along with special tests such as Antinuclear antibodies (ANA) profile, Antineutrophil Cytoplasmic Antibodies (ANCA) and Antistreptolysin O (ASO) titer were conducted in all patients.

Results: Out of 50 cases, 20% of the patients had (Henoch-Schonlein purpura) HSP and 10% had Urticarial vasculitis (UV), collagen vascular disease associated vasculitis were present in 2%, Cutaneous polyarteritisnodosa (C-PAN), eosinophilic vasculitis and nodular vasculitis were present in 4% of the patients each. Rests of the patients (54%) were designated as idiopathic cutaneous small vessels vasculitis (SVV). The commonest lesion was palpable purpura. Infection was the commonest cause of cutaneous vasculitis for about 22%, followed by drugs (20%), malignancy (2%) and connective tissue disease (2%). Two patients with HSP had positive ANCA, 3 without any overt manifestations and 1 with systemic sclerosis. Histologically Leukocytoclastic vasculitis (LCV) was the commonnest pattern (72%), lymphocytic in 20%, granulomatous in 4% and eosinophilic vasculitis in 4% of patients.

Conclusion: Cutaneous SVV is the commonest form of vasculitis. The heterogenecity of this group of disorders is well represented in this study. To reach an etiological diagnosis of vasculitis, clinical and pathological features need to be correlated and supplemented by laboratory investigations.

Key-words: Clinicopathological study, Cutaneous vasculitis.

Introduction

Vasculitis is a term applied to inflammation and necrosis of blood vessels. Vasculitis can be local or systemic, and can be primary or secondary to another disease process[1]. A definitive diagnosis of vasculitis requires histological confirmation, but it is correlated with clinical, physical and laboratory findings[2]. Vasculitis can range in severity from a self-limited disorder to a life threatening disease like multiple-organ failure[3]. The morbidity and mortality with these disorders can be reduced if recognized and treated early[4].

Materials and Methods

This descriptive cross-sectional study was carried out at the Department of Dermatology and Venereology, Bangladesh Naval Ship (BNS) Patenga, Chattogram during March 2016 to August 2017. Total 50 patients selected and histologically diagnosed cases of cutaneous vasculitis were included in the study. Clinical diagnosis was made on morphology, signs and symptoms. Detailed history and physical examination was done for primary selection. All information and findings were recorded in data collection sheet. Primarily selected patients were included in the study. The aims and objectives of the study were explained to patients in easily understandable local language and then informed written consent was taken from interested patients. Unwilling patients were excluded from the study. All patients were subjected to a baseline workup consisting of complete blood count (CBC), serum creatinine level, liver function test (LFT), chest X-ray, urine examination, Antistreptolysin O (ASO) titer, Antinuclear antibodies (ANA), Cytoplasmic anti-neutrophil cytoplasmic antibody (C-ANCA) Cutaneous Polyarteritis nodosa (C-PAN) and hepatitis B and C serology. Histopathological examinations of lesional skin biopsy specimen from all patients were done and histologically confirmed cutaneous vasculitis finally selected for the study. The data regarding different variables were analyzed accordingly and SPSS-17 was used for analysis.

Results

Among 50 patients with cutaneous vasculitis age range was 13 to 72 years with average being 39.9 years and 30(60%) were females and 20(40%) were males (Table-I). Duration of Vasculitis ranged from 1 day to 5 years with majority had

1. **Lt Col Md Shajahan Siraj**, MBBS, FCPS, Classified Specialist in Dermatology & Venereology, Bangladesh Naval Ship (BNS) Patenga, Chattogram (*E-mail:* ssiraj_amc@yahoo.com) 2. **Maj Mohammad Maksudur Rahman**, MBBS, FCPS, MPH, Classified Specialist in Surgery, BNS Patenga, Chattogram 3. **Lt Col ATM Rezaul Karim**, MBBS, MCPS, DDV, FCPS, Associate Professor of Dermatology & Venereology, AFMC, Dhaka 4. **Lt Col Md Abdul Wahab**, MBBS, MD, Associate Professor of Biochemistry, AFMC, Dhaka.

duration less than 1 year. More than half of the patients 27(54%) were designated as idiopathic but aetiological association was found in 23(46%) patients. Infection was the commonest cause found in 11(22%) cases, followed by drugs in 10(20%) cases. Patients' presenting symptoms were itching in 26(52%), pain in 27(54%), burning in 10(20%), arthralgia and fever were present in 23(46%) each and myalgia in 11(22%) patients. Thirteen patients had overlap more than one symptoms. Elevated erythrocyte sedimentation rate (ESR) was found in 26(52%) patients, leukocytosis 11(22%), 12(24%) pyuria, 7(14%) hematuria and 2(4%) had proteinuria. Liver function tests (LFT) were abnormal in 4(8%) patients and renal function tests in 5(10%) patients. ASO titre was elevated in 7(14%) and ANA was positive in 4(8%) patients, 2(4%) patients were with ANCA positivity (Table-II).

Out of 50 patients, 29(58%) had systemic symptoms like abdominal pain 22%, diarrhea 6%, bleeding from gastro-intestinal tract 2%, hematuria 4%, paraesthesia and exertional dyspnoea 10% each, hemoptysis and oral ulcer (2% each). Various cuteneous lesions like palpable purpura was found in 86% patients, papules in 24%, plaques 12%, urticaria 10%, ulcers 10%, nodules 4%, scars in 4%, vesicles and bullae in 6% each, digital gangrene and pustules in 2% of the patients each. Twenty two (44%) patients had bilaterally symmetrical pitting pedal edema. Almost all 49(98%) patients had small vessel vasculitis and 1(2%) had large vessel vasculitis.

The different histological types in these cases included leukocyto-clastic vasculitis 35(70%), lymphocytic vasculitis 10(20%), granulomatous 3(6%) and eosinophilic vasculitis 2(4%). Occasional neutrophils were present in eosinophilic vasculitis (Figure-1). All 10(20%) patients with HSP and all 5(10%) patients with Urticarial vasculitis (UV) had Leuko-cytoclastic vasculitis (LCV) histopathologically. Connective tissue disease (CTD) associated vasculitis, C-PAN, eosinophilic vasculitis, Giant-cell arteritis (GCA) and nodular vasculitis had leukocytoclastic, lymphocytic, eosinophilic, granulomatous and lymphocytic histopathological picture in biopsy specimens respectively. Among the patients with idiopathic CSVV, 20(40%) had LCV, 6(12%) had lymphocytic and 1(2%) had granulomatous vasculitis (Table-III).

Table-I: Distribution of patient by age and sex (n=50)

Characteristics		Frequency	Percentage
Age in years	< 20	15	30
	21-40	24	48
	41 -60	5	10
	> 60	6	12
Sex	Male	20	40
	Female	30	60

Table-II: Distribution of patients by aetiology, symptoms and Laboratory findings (n=50)

Characteristics		Frequency	%
Aetiology	Drugs	10	20
	Infections	11	22
	Malignancy	1	2
	CTD associated	1	2
	Idiopathic	27	54
Symptoms	Itching	26	52
	Pain	27	54
	Burning	10	20
	Fever	23	46
	Myalgia	11	22
	Arthralgia	23	46
Laboratory findings	Anemia	9	18
	Leukocytosis	11	22
	Neutrophilia	7	14
	Eosinophilia	9	18
	Raised ESR	26	52
	Elevated S. urea	5	10
	Elevated serum creatinine	5	10
	Abnormal LFT	4	8
	Albuminuria	2	4
	Pyuria	12	24
	Hematuria	7	14
	High ASO titre	7	14
	ANA	4	8
	P-ANCA	1	2
	C- ANCA	1	2

Note: ANA: Antinuclear antibodies; ASO: Anti-streptolysin O; C-ANCA: Cytoplasmic anti-neutrophil cytoplasmic antibody C-PAN: Cutaneous Polyarteritisnodosa;

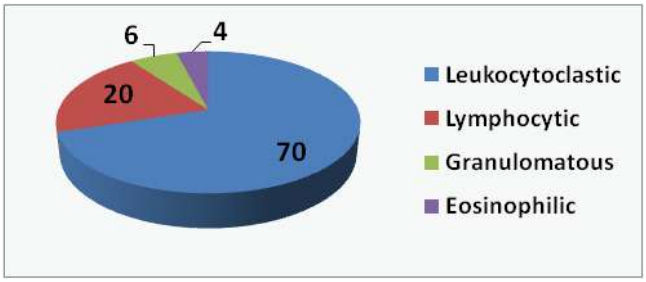

Figure-1: Histopathological types of vasculitis

Table-III: Clinicohistopathological correlation

Clinical diagnosis	n(%)	Histopathological diagnosis	n(%)
HSP	10(200	Leukocytoclastic	10(20)
UV	5(10)	Leukocytoclastic	5(10)
GCA	1(2)	Granulomatous	1(2)
CTD associated vasculitis	1(2)	Leukocytoclastic	1(2)
C-PAN	2(4)	Lymphocytic	2(4)
Eosinophilic Vasculitis	2(4)	Eosinophilic	2(4)
Nodular Vasculitis	2(4)	Lymphocytic	2(4)
Idiopathic CSVV	27(4)	Leukocytoclastic	20(40)
		Granulomatous	1(2)
		Lymphocytic	6(12)

Note: HSP:Henoch-Schonlein purpura; GCA: Giant-cell (temporal) arteritis; CTD: Connective tissue disease; C-PAN: Cutaneous polyarteritisnodosa; CSVV: Cutaneous small vessel vasculitis; UV: Urticarial vasculitis

Discussion

Total 50 patients with cutaneous vasculitis were analyzed; their age was ranged from 13 to 72 years and average being 39.9 years. Duration of vasculitis ranged from 1 day to 5 years with majority having duration less than 1 year which is similar to earlier studies[5,6]. Aetiological association was seen in 23(46%) patients. This is similar to previous studies[6,7]. In this study, infection was found as the commonest factor (22%), followed by drugs in 20% cases. But drugs (19.7%) followed by infections (11.4%) were the common causes in a previous study[6,8]. Commonest symptom was pain (54%), followed by itching (52%) and burning (20%). In a study conducted by Sais and colleagues[9], 41.4% complained of itching and 30% complained of painful lesions. In this study, constitutional features were present in 34(68%) patients, arthralgia and fever were present in 23(46%) each and myalgia in 11(22%) patients which is consistent with a study conducted by Khetan et al[6]. Systemic features were 50% of patients in a study conducted by Gupta et al[2] and 51% patients in a study conducted by Ekenstam et al[7]. In this study, 29(58%) had associated systemic symptoms. The commonest lesion was palpable purpura seen in 86% of the patients, which is similar to previous studies[2,6,9].

In this study, 22(44%) patients had bilaterally symmetrical pitting type of pedal edema; this was in contrast to Sais et al[9]. Commonest laboratory abnormality was elevated ESR found in 26(52%) patients which is similar to the previous studies[6,9,10]. Gupta et al[2] reported anemia in 48% and leukocytosis in 12% patients, wheras Sais et al[9] observed anemia in 37%, eosinophilia in 2.5%, leukocytosis in 18%. In this study, anemia 18%, leukocytosis 22%, eosinophilia was present in 30% patients respectively. Gupta et al[2] observed these parameters to be 2%, 4% and 10% respectively. LFT was abnormal in 4 patients (8%), out of which 1 had history of chronic liver disease, and other 3 had elevated transaminase levels with no signs of chronic liver disease. LFT were within normal limits in all patients in the study conducted by Gupta et al[2] while Sais et al[9] observed elevated transaminase levels in 18% of patients. In my study, renal function was abnormal in 5(10%) patients which was in the form of elevated serum urea and serum creatinine. The renal functions were altered in 6% patients and 26 % in the previous studies[2,9]. ASO titre was elevated in 7(14%), though 22 had history of recurrent sore throat as compared to a previous study where only 2% had elevated ASO titre[5]. ANA was positive in 8% patients, 1 had scleroderma. Gupta et al[2] reported ANA positivity in 6% and Sais et al[9] in 28.5%. In this study, 2 patients with ANCA positivity, 1 was p- ANCA and another was c-ANCA positive but diagnosed as HSP. This indicates that ANCA positivity is not specific for ANCA associated vasculitis syndromes. Study conducted by Sais et al out of 160 patients, 21% were p- ANCA positive and none had c-ANCA[9]. Histopathology revealed different features in vasculitis included leukocytoclastic vasculitis, lymphocytic vasculitis, granulomatous and eosinophilic vasculitis. Out of 50 patients, 49 had SVV and 1 had large vessel vasculitis. This was in contrast to Sais et al[9] who reported SVV and MVV in 60 and 40 per cent respectively and Khetan et al[6] who reported SVV and MVV in 96% and 4% respectively. In the study conducted by Alexander et al[11] LCV was reported in 68%, Lymphocytic vasculitis in 6%, granulomatous and subepidermal bulla in 2% each and the rest that is 22% had non specific features. Table-V shows the clinicohistopathological correlation in this study. All the 10 patients with HSP had LCV on histo- pathological examination. This is similar to a previous study[6]. Similarly, all the 5 patients with UV had LCV on histo- pathological examination, CTD associated vasculitis, cutaneous PAN, eosinophilic vasculitis, GCA and nodular vasculitis had leukocytoclastic, lymphocytic, eosinophilic, granulomatous and lymphocytic histopathological picture in their biopsy specimens respectively. Gupta et al[2] conducted a study on total of 50 patients diagnosed clinically as cutaneous vasculitis, 41 were classified as LCV, 2 as HSP, 2 as UV and one each as nodular vasculitis, C-PAN and PLEVA. Based on histopathological findings, 72% patients were given a diagnosis of cutaneous SVV, 12% were diagnosed as lymphocytic vasculitis, 2% was diagnosed as PLEVA, while 16% patients did not show any evidence of vasculitis[2].

In this study, 20% patients had HSP and 10% had UV. CTD associated vasculitis that is scleroderma and GCA were present in 2% patient each. C-PAN, eosinophilic vasculitis, and nodular vasculitis were present in 2(4%) patient each. Rest of the patients that is 54% patients were categorised as idiopathic cutaneous small vessel vasculitis. Khetan et al[6] reported hypersensitivity vasculitis (HSV) in 37.7%, HSP in 26.2%, CTD and UV in 6.5% each, 1.6% each had microscopic polyangiitis, Wegener's granulomatosis, PAN and Takayasu's arteritis and the rest of the patients were designated as unclassified vasculitis. Hence, the study showed that an etiological, clinical, histopathological and laboratory features of cutaneous vasculitis are similar in coastal area of Chattogram, Bangladesh like any other area[12].

Conclusion

Cutaneous vasculitis may range from self-limiting to progressive systemic disease. Most patients with cutaneous vasculitis presented with polymorphic lesions though majority of them presented with palpable purpura and histologically LCV. The morbidity and mortality associated with vasculitis can be reduced if recognized and treated early. In this study, etiology could not be identified in majority of the patients. The investigations conducted in this study aided in confirming certain etiologies like infections. Patients with drug induced vasculitis had eosinophilia. Treating the underlying cause of the vasculitis is needed for its management. Hence thorough assessment of the history and correlation with the clinical pattern of involvement is crucial for the diagnosis.

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