A STUDY OF AETIOLOGICAL FACTORS IN THE OCCURRENCE OF CUTANEOUS VASCULITIS IN A TERTIARY CARE CENTRE IN NORTH KERALA

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ABSTRACT

BACKGROUND

Cutaneous vasculitis is a condition caused by various aetiologies. They can be primary or secondary. Cutaneous lesions maybe a pointer to systemic diseases. So, it is important to identify the various aetiological factors in the occurrence of the various types of cutaneous vasculitis. The patterns and the various aetiologies of cutaneous vasculitis in Kerala is not well documented in the existing literature.

MATERIALS AND METHODS

The present study is a cross-sectional descriptive study of patients with a clinical diagnosis of cutaneous small vessel vasculitis admitted in Medicine and Dermatology Wards of Calicut, Government Medical College during January 2013 to December 2013. A detailed history and clinical examination of patients along with histopathological examination of skin biopsy was done.

RESULTS

Out of the 70 cases of cutaneous vasculitis studied, idiopathic cutaneous small vessel vasculitis was the most common type followed by Henoch-Schonlein purpura. The most common aetiology identified was drugs followed by infections. No aetiological factor was identified in 42.8% of the cases.

CONCLUSION

An aetiological association could be found in 57.8% of cases. The causes identified include drugs, infections, malignancy, connective tissue disorder associated, chronic systemic diseases and Behcet's disease in decreasing order of frequency.

KEYWORDS

Vasculitis, Cutaneous Vasculitis, Behcet's Disease, Henoch-Schonlein Purpura, Urticarial Vasculitis.

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BACKGROUND

Vasculitis is an inflammation of the blood vessel wall with wide range of clinical manifestations.[1,2] The skin is commonly involved in vasculitic disorders. Skin being one of the most common and early organs to be involved in vasculitic disorders, it offers a good window to the diagnosis and ready source of accessible tissue for diagnosis.[3]

Cutaneous vasculitis presents as a mosaic of clinical and histological findings due to varied pathogenic mechanisms.[1]It maybe idiopathic or associated with a spectrum of conditions such as infections, drugs, etc. Skin is involved in both small vessel vasculitis and medium vessel vasculitis. Identifying the cutaneous lesion is important because they may predate the involvement of other organ systems. Vasculitis as a clinical entity is also very difficult to treat without a known aetiology.The relevance of identifying the different aetiologies of cutaneous vasculitis lies in the fact that avoidance or termination of exposure to the alleged cause results in the resolution of the symptoms of vasculitis.[2,4] A definitive diagnosis of vasculitis requires histological confirmation in almost all cases. Skin biopsy is the gold standard for diagnosis of cutaneous vasculitis whose manifestations include urticaria, infiltrative erythema, petechiae, purpura, purpuric papules, haemorrhagic vesicles/bulla, nodules, deep punched out ulcers and digital gangrene.[1,5]

Proposed working classification (updated Gilliam's classification) of small vessel vasculitis include cutaneous small vessel vasculitis, Henoch-Schonlein purpura, essential mixed cryoglobulinaemia, Waldenstrom's hypergammaglobulinemic purpura, vasculitis associated with collagen vascular diseases, urticarial vasculitis, microscopic polyangiitis, erythema elevatum diutinum and oeosinophilic vasculitis.[6]

The histological diagnostic criteria for cutaneous vasculitis[1]for selection of cases included-

Dermal small vessels (venules and arterioles) (2 of 3 criteria needed).

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1. Angiocentric and/or angioinvasive inflammatory infiltrates.
2. Disruption and/or destruction of vessel wall by inflammatory infiltrates.
3. Intramural and/or intraluminal fibrin deposition ('fibrinoid necrosis').

For identifying the different causes of cutaneous vasculitis, a proper clinical history, clinical examination as well as laboratory finding along with a skin biopsy is mandatory. These patients require long-term management and follow up for early diagnosis of systemic involvement and complications. The aim of the study was to identify the various aetiological factors and triggering events in the occurrence of various types of cutaneous vasculitis.

MATERIALS AND METHODS

The present study is a cross-sectional descriptive study of 70 consecutive cases of cutaneous vasculitis presented in the Department of Medicine and the Department of Dermatology diagnosed clinically and confirmed histologically between the period of January 2013 and December 2013. A detailed history was taken and all baseline investigations done including complete blood examination, liver function and renal function tests, peripheral smear, chest x-ray, urine routine examination and other tests including ANA, ANCA, cryoglobulin levels and antibodies to hepatitis B and C virus. Exclusion criteria included those who were not willing for the study and pregnant women. Histopathological examination was done in skin biopsy and criteria for cutaneous vasculitis was applied to select the cases. A 4-mm punch biopsy was done and in suspected cases, direct immunofluorescence staining was done using IgG, IgM, IgA and C3. Ethical committee clearance was obtained from the Institutional Ethics Committee. The data obtained was analysed as ratios and proportions.

RESULTS

A total of 70 patients were studied. Out of this, 44 patients were females and 26 were males. The female-to-male ratio was 1.69:1. The age distribution of the cases were as follows (Table 1). The maximum number of patients were in the age group of 21 to 40 years.

Age (Years)	Number of Patients	Percentage
<20	14	20
21-40	31	44.28
41-60	19	27.14
>60	6	8.5
Total	**70**	**100**

Table 1. The Age Distribution of Patients Presenting with Cutaneous Vasculitis

Out of the total cases of cutaneous vasculitis studied, the distribution of the different type of vasculitis by clinical and histopathology are given in Table 2.The majority of patients included idiopathic cutaneous small cell vasculitis (30 patients) followed by Henoch-Schonlein purpura (22 patients) and urticarial vasculitis (6 patients) (Table 2).

Type of Vasculitis	Number of Patients	Percentage
Idiopathic cutaneous small cell vasculitis	30	42.85
Henoch-Schonlein purpura	22	31.42
Urticarial vasculitis	6	8.57
Churg-Strauss vasculitis	2	2.85
Vasculitic ulcer	2	2.85
Nodular vasculitis	2	2.85
CTD associated vasculitis	2	2.85
Behcet's disease	2	2.85
Microscopic polyangiitis	1	1.43
Erythema nodosum necroticans	1	1.43

Table 2. The Number of Patients Presenting with the Different Types of Cutaneous Vasculitis

The aetiology of the different types of cutaneous vasculitis are summarised in the table given below (Table 3).

Aetiology	Number of Patients	Percentage
Drugs	16	22.85
Infection	13	18.57
Systemic disease associated	4	5.71
Connective tissue disorders	3	4.28
Malignancy	2	2.85
Behcet's disease	2	2.85
Idiopathic	30	42.85

Table 3. The Aetiology of the Different Types of Small Vessel Vasculitis

In the present study, the most common aetiology observed was exposure to drugs (16 patients, 22.85%) followed by infection (13 patients, 18.57%). Drug intake of a minimum duration of one month prior to the diagnosis of the vasculitis was taken into consideration. All the cases of drug-induced vasculitis was due to nonsteroidal anti-inflammatory drug intake. There was no difference in the clinical course of the disease in those caused by drugs and otherwise. In 30(42.85%) patients, the aetiology could not be identified and hence designated as idiopathic.

DISCUSSION

In the current study of 70 patients, a significant majority of the patient were females (44 out of 70) with 31 patients in the younger age group of 21-40 years old. 16 patients (22.85%) had a history of drug intake prior to the onset of the illness. 13 patients (18.57%) had a history of infection prior to the onset of the disease, which included upper respiratory tract infection, urinary tract infection and infectious mononucleosis in that order of frequency. 30 patients (42.85%) had apparently no causes after extensive investigations were classified as idiopathic. So, an aetiological association was found in 57.15% of the

cases. Two cases of vasculitis was caused by non-Hodgkin's lymphoma (diffuse large B cell variant). Three cases of cutaneous vasculitis occurred associated with connective tissue disorder (two cases being Systemic Lupus Erythematosus (SLE) and the other Mixed Connective Tissue Disorder (MCTD)). Of the four cases of cutaneous vasculitis associated with systemic diseases include one case of hypothyroidism, one case of chronic liver disease and 2 cases of chronic kidney disease due to IgA nephropathy and membrano proliferative glomerulonephritis.

Although, many previous studies had shown female predominance in vasculitis occurrence, some recent studies had shown equal gender predilection of cutaneous vasculitis.[7]Many previous studies had put the average age of patients around 40 to 45 years with female preponderance.[8]

Previous studies had found that leukocytoclastic vasculitis formed the majority of cases of cutaneous vasculitis.[9,10] A causal agent or an underlying condition has been reported in 20-85 percent of the cases.[11,12,10,13] Similar to our findings, a previous study from India had found that drugs were the most common aetiological factor in vasculitis.[14] In contrast to our study, some previous studies had shown that antibiotics rather than NSAIDs were responsible for drug-induced vasculitis.[12,13,15] Unfortunately, no tests are available to definitely incriminate drugs as a causation except for the temporal relationship of the drug and the subsidence of lesion after the withdrawal of the drug. The overall frequency of infection was 18.57% in our study, which is slightly higher than that observed in reports from Belgium (9.5%) and Mexico (6.8%); while higher frequency has also been reported from Australia (26%), Spain (19.8%) and Kuwait (14%).[12,13,15,4,16]

SUMMARY AND CONCLUSION

In the present study of 70 consecutive cases of cutaneous vasculitis who presented between January 2013 and December 2013, females outnumbered males by a ratio of 1.69:1 and 44.28% of patients were in the age group of 21 to 40 years. Of the various causes of cutaneous vasculitis, 21.4% had a history of prior drug therapy (all due to NSAIDs), 17.14% had a history of infection of which upper respiratory infection was prominent. 37.14% of patients had no apparent cause evident and were classified as idiopathic. Other aetiologies included malignancy related, systemic disease related, trauma related chronic ulcers, connective tissue related and Behcet's disease.

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