Original research article

A Clinical Study of Cutaneous Small Vessel Vasculitis in a Tertiary Care Hospital

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ABSTRACT

Background: Clinical signs and symptoms of cutaneous vasculitis can vary greatly. It could be idiopathic or linked to a variety of illnesses, such as infections, medications, etc. Small vessel vasculitis (SVV) and medium vessel vasculitis both affect the skin (MVV). There are similarities between the characteristics of SVV and MVV. The histological characteristics and the clinical lesions may not always be related. The current study's objective was to assess the clinicopathological correlation and etiological variables in individuals with cutaneous vasculitis.

Methods: All patients with clinical signs and symptoms that are suggestive of small vessel vasculitis, such as palpable purpura, infiltrated erythema, hemorrhagic vesicles and bulla, ulcers, infarcts, digital gangrene, erythematous plaques and nodules, urticaria, and livedoreticularis. Clinical evaluation: Extensive systemic and general evaluations were performed. The morphology of the skin lesion, its location, symmetry, pain, and diascopy were all thoroughly examined.

Results: Based on the inclusion and exclusion criteria n=30 cases with clinical features of cutaneous small vessel vasculitis were seen during the study period. Out of the total cases seen the common types were Henoch Schonlein purpura in n=12/30 (40%), Erythema nodosum leprosum n=7/30 (23.33%), small vessel vasculitis associated with collagen vascular disease n=6/30 (20%). The less common types were urticarial vasculitis n=1/30 (3.33%) septic vasculitis n=2/30 (66.66%), essential mixed cryoglobulinemia n=1/30 (3.33%) the details of the distribution between males and females.

Conclusion: The most frequent kind of cutaneous small vessel vasculitis identified in the current investigation was Henoch Schonlein purpura, followed by Erythema nodosum leprosum. Females tended to have cutaneous small vessel vasculitis more often. The majority of the cases were people under 40 years of age. Upper respiratory tract infection was the most prevalent causative factor. The clinical diagnosis of cutaneous small vessel vasculitis is based on the presence of non-thrombocytopenic purpura.

Keywords: Cutaneous small vessel vasculitis, Henoch Schonlein purpura, hypersensitivity vasculitis, medium vessel vasculitis.

Introduction

Vasculitis is an inflammatory condition that affects the vessel walls and can impair or destroy them, causing hemorrhagic and ischemic events to occur as a result. Any organ's vessels may be involved, which can cause a wide range of symptoms. [1, 2] The participation of several organs is what makes this group special. The skin is vulnerable to regularly being impacted

by vasculitis due to its abundant vascularity. Vasculitides may have cutaneous involvement as their major symptom, as a reflection of a deadly systemic illness, or as proof of a relationship with another systemic sickness. Cutaneous vasculitic lesions provide a diagnostic window and a rapid supply of readily available tissue for histopathological analysis.

Arterioles, capillaries, and venules are only a few examples of vessels smaller than arteries that are typically affected by small vessel vasculitis. [3] The identification of a particular kind of vasculitis is made more difficult by these diverse clinical symptoms and the etiologic non-specificity of the histologic lesions. Since few kinds of vasculitis exhibit a pathognomonic laboratory or imaging result, histologic confirmation on biopsy is the gold standard for a diagnosis of vasculitis. As a clinicopathological process, vasculitis can develop both independently or as a secondary symptom of various illnesses such as collagen vascular diseases, viral diseases, metabolic problems, cancer, and Adverse Drug reactions. The skin is frequently where vasculitis first manifests, and it is the dermatologist's responsibility to identify and manage this difficult illness. The majority of instances of cutaneous vasculitis are idiopathic, despite the fact that there are several potential causes. Depending on the epidemiological variations and the prevalence of illnesses, the frequency of each cause varies. According to a pooled set of data, cutaneous vasculitis is caused by infections in 22% of cases, medications in 20%, CTD in 12%, Henoch Schonlein purpura (HSP) in 10%, and systemic inflammatory diseases such sarcoidosis and cryoglobulinemia in 5% of cases [2] Numerous histopathological findings of SVV and MVV overlap, and histopathology can be quite variable. The histological characteristics may be unrelated to the clinical lesions. Only a few research from India have been done on cutaneous vasculitis. [4, 5] Therefore, we conducted this study at a tertiary care hospital in South India to assess the aetiological variables and clinicopathological correlation with clinical lesions in patients with cutaneous vasculitis.

Material and Methods

This cross-sectional study was conducted in the Department of Dermatology, as well as those patients to this department from other departments which included General Medicine, Rheumatology, and Pediatrics. Institutional Ethical approval was obtained for the study. Written consent was obtained from all the participants of the study after explaining the nature of the study in the vernacular language.

Inclusion criteria: All patients with clinical signs and symptoms that are suggestive of small vessel vasculitis, such as palpable purpura, infiltrated erythema, hemorrhagic vesicles and bulla, ulcers, infarcts, digital gangrene, erythematous plaques and nodules, urticaria, and livedoreticularis, must meet the inclusion criteria.

Exclusion criteria

1. Patients with abnormal bleeding parameters.
2. Patients with malignancy
3. Patients with psychological issues
4. Patients are not willing to participate in the study.

A thorough history was gathered, including details on the patient's symptoms (itching, burning, and pain), the length of skin lesions, occupational history, systemic symptoms, recent sore throat history, drug use history, and information about malignancy and collagen vascular diseases. Clinical evaluation: Extensive systemic and general evaluations were performed. The morphology of the skin lesion, its location, symmetry, pain, and diascopy

were all thoroughly examined. 1) Initial laboratory tests include a complete hemogram, serum urea, serum creatinine, liver function tests, chest X-ray, urine (routine and microscopy), Mantoux test, test for stool occult blood, ASO titer, blood culture, and skin smears for acid fast bacilli, as well as USG Abdomen and Pelvis. For high-risk individuals with a history of sexual contact or occupational exposure to blood and blood products, testing for HIV, Hepatitis B, C, and syphilis was also conducted. 3) Cryoglobulin test, serum protein electrophoresis, complement, malignancy, and collagen vascular disorders tests were performed as needed to rule out cryoglobulinemia. With a warning not to include the resolving lesion, incisional elliptical skin samples from the early painful skin lesions were performed and delivered to a pathologist for histopathology. When necessary, certain stains like AFB were used. Gram stain and blood culture are two more examinations to rule out bacterial infections. According to the Proposed working classification of vasculitis [an updated version of Gilliam's 1976 system 12], individuals in our research who had characteristics of cutaneous small vessel vasculitis were classified as having those traits.
Analysis of statistical data: In our study, a descriptive analysis of the clinical traits, laboratory results, and histological characteristics of diverse cutaneous small vessel vasculitis was carried out. Data analysis and comparison with available literature were done.

Results

Based on the inclusion and exclusion criteria n=30 cases with clinical features of cutaneous small vessel vasculitis were seen during the study period. Out of these n=9(30%) were males and n=21(70%) were females. The male-to-female ratio was 1: 2.33. The mean age of the patients in the study was 31.5 years in males and 33.5 years in females.

Out of the total cases seen the common types were Henoch schonlein purpura in n=12/30 (40%), Erythema nodosum leprosum n=7/30 (23.33%), small vessel vasculitis associated with collagen vascular disease n=6/30 (20%). The less common types were urticarial vasculitis n=1/30 (3.33%) septic vasculitis n=2/30 (66.66%), essential mixed cryoglobulinemia n=1/30 (3.33%) the details of the distribution between males and females have been depicted in table 1.

Table 1: Sex-wise distribution of the diseases diagnosed in the cases of the study.

Disease	Male		Female	
	Frequency	*Percentage*	*Frequency*	*Percentage*
Henoch Schonlein purpura (HSP)	3	28	9	72
Erythema nodosum leprosum (ENL)	4	70	3	30
Collagen vascular disease vasculitis (CVDV)	0	14	6	86
Urticarial vasculitis (UV)	1	50	1	50
Septic vasculitis (SV)	0	0	2	100
Essential mixed cryoglobulinemic vasculitis (EMCV)	1	100	0	0
Total	9	30	21	70

In this study out of n= 12 cases of Henoch Schonlein purpura most cases were belonging to the 11 – 20 years age group with n=6/12 (50%) of cases. Similarly, there were n=7 cases of Erythema nodosum leprosum most of them were in the age group 31 – 40 years. Cases of Collagen vascular disease vasculitis were n=6 and the total cases of Urticarial vasculitis and Septic vasculitis were n=2 each. There was an n=1 case of Essential mixed cryoglobulinemic

vasculitis all the distribution of these cases based on the age groups have been depicted in figure 2.

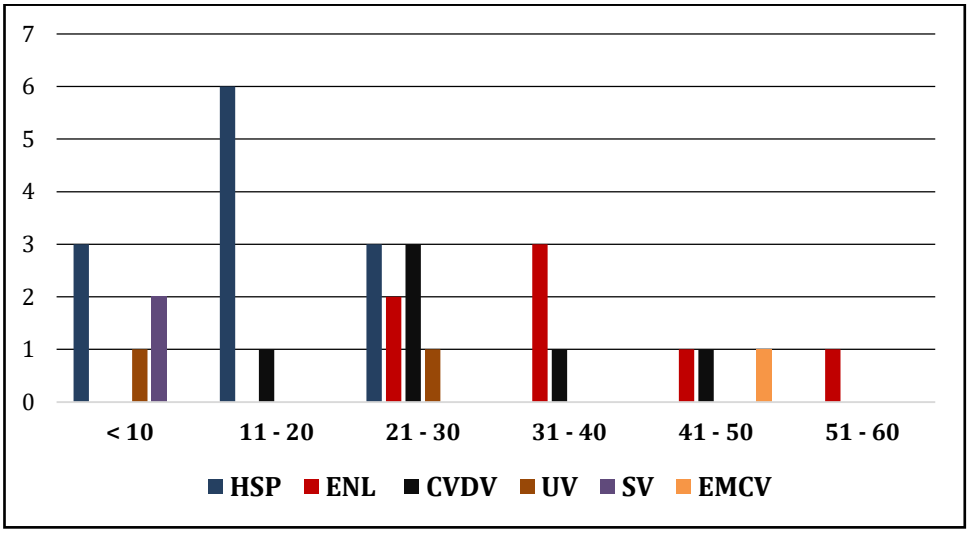

Figure 2: Age-wise distribution of the cases of small vessel vasculitis included in the study.

In this study based on the etiological factors 54% of cases were due to infections, and 20.0% of cases had positive connective tissue disease workup without any overt manifestations, N=4 cases were attributed to drugs which included NSAIDs in two cases and antibiotics in one case. The cause was not detected in 13% of cases depicted in figure 2.

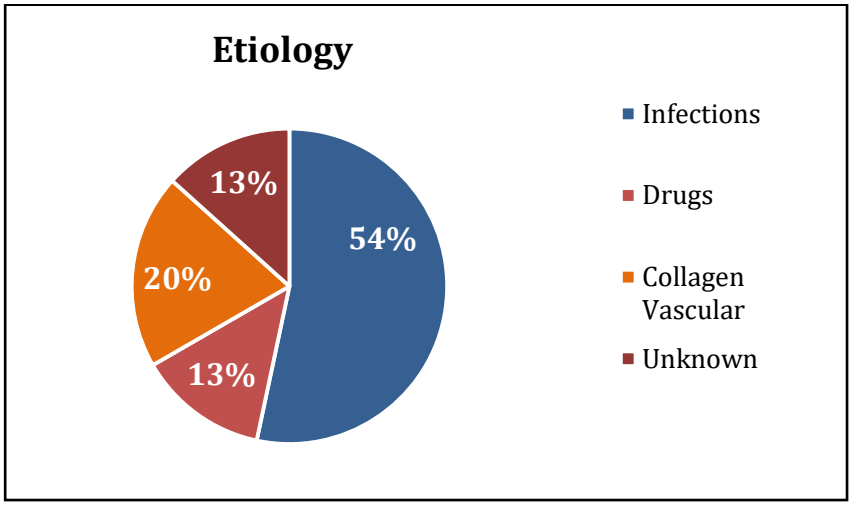

Figure 2: Showing the etiological factors in cutaneous small vessel vasculitis.

In this study, we found Itching was the common presenting symptom in n=11/30 (36.67%) of cases followed by pain and site of the lesion and fever in n=5/30 (16.67%) cases each. Burning sensations were found in n=3/30 (10%) cases and no symptoms were reported in 20% of cases depicted in figure 3.

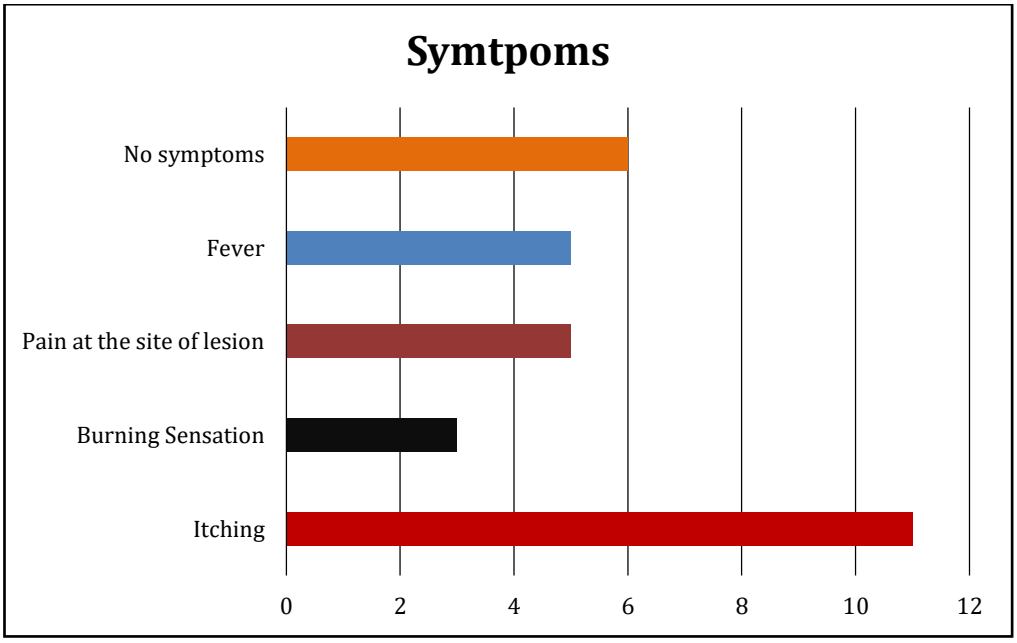

Figure 3: Distribution of symptoms reported by the cases in the study.

Systemic symptoms were present in n=25 instances (83.33%) and were shown as concomitant joint aches in n=14 cases (46.67%) of patients, with the knee joint being the most frequently affected joint (n=11). The ankle joint and smaller joints in the foot, wrists, and ankles were also affected. Five individuals had joint swelling noted. Five patients (16.67%), four (13.33%), and one (3.33%) of the patients had a history of melena, hemoptysis, or both.

The most frequent cutaneous symptom observed in n=16 (53.33%) patients was palpable purpura (12 females and 4 males). The additional skin conditions that were seen in n=14 individuals were nodules (n=6), plaques (n=2), ulcers (n=2), bullae (n=1), vesicles (n=1), toe gangrene (n=1), and urticarial lesions in (n=1). The period since lesions first appeared varied from one day to nine months.

Table 2: Showing the results of laboratory investigations in the cases of study.

Investigations	*Frequency*	*Percentage*
Anaemia	13	43.33
Leukocytosis	8	26.67
Raised ESR	23	76.67
Elevated urea	5	16.67
Elevated serum creatinine	3	10.00
Albuminuria	5	16.67
Stool for occult blood	6	20.0
Abnormal chest x-ray	1	3.33
Anti-nuclear antibody	4	13.33
Rheumatoid factor	2	6.67
ASO titer	8	26.67
Cryoglobulin Test	1	3.33
USG abdomen – abnormality	1	3.33

The results of the hematological and biochemistry workup showed in table 2. It was found that 43.33% of cases had anemia, 26.6% had leukocytosis, 76.67% had Elevated ESR, 16.67% had elevated serum urea, and 10% of cases had elevated creatinine levels. Urine microscopy revealed blood cells in seven individuals, whereas 16.67% of patients' routine urine tests revealed albuminuria. In 20% of cases were positive for occult blood in the stool. One patient had hemoptysis and the cavity was shown in chest X-rays. Serum cryoglobulins were positive in one patient, whereas anti-nuclear antibody and rheumatoid factor were positive in 13.33% and 6.67% of patients, respectively. 26.67% of cases were having increased ASO titers, and one patient had positive Mantoux results. USG showed bowel wall edema in one patient with Henoch Schonlein Purpura. According to histological results, leukocytoclastic vasculitis was diagnosed in n=25 (83.33%) of the patients, whereas perivascular lymphocytic infiltrates were seen in n=5 (16.67%) of the patients but no evidence of vasculitis was present. Endothelial edema, fibrinoid necrosis, RBC extravasation, and leukocytoclasis were all evident in the skin sample. Subepidermal bullas, hyaline thrombi, and mixed panniculitis were also identified.

Discussion

Cutaneous small vessel vasculitis is a poorly understood condition due to its complex clinical appearance and overlaps with many infections, vascular abnormalities of the collagen, and cancers. In our study, we looked at cutaneous small vessel vasculitis patients that had been identified by a combination of laboratory testing, clinical characteristics, and history. A skin biopsy was used to support the clinical diagnosis. Our research sheds light on several novel characteristics of cutaneous small vessel vasculitis while confirming a number of previously known facts. Gupta et al., [6] in Chandigarh, India, in a similar study on n=50 patients found there were n=20 male and n=30 female patients. The research group's average age was 35.9 years for women and 41.1 years for men. In the present study, we had n=25 patients out of which n=9(30%) were males and n=21(70%) were females. The mean age of the patients in the study was 31.5 years in males and 33.5 years in females. In our study, females were more frequently affected than men, which is in line with the findings of Gupta et al., [6] findings. In this study, 76% of our patients had suspicions about a potential etiological link, which is identical to the 67.2% seen by Sais et al., [7] Upper respiratory tract infections made up 56% of the most frequent causes in our analysis, which is consistent with research by Martinez-Taboada et al., [8] In our investigation, other factors that contributed to cutaneous small vessel vasculitis were medications (13%) and collagen vascular diseases (20%). The most frequent collagen vascular disease identified in our investigation was lupus erythematosus. The most popular medications were NSAIDS, then antibiotics. The findings of the research by Sams et al., [9] Callen JP et al., [10] and Hautmann et al., [11] were comparable with our findings. This was analogous to prior research by Sais et al., [7] where 30.0% of patients experienced painful lesions and 41.3% of patients reported itching. Of our patients, 15% had a fever, compared to 31.6% in Sais et al., [7] study. In this study, the common presenting symptom was Itching in (36.67%) of cases followed by pain and site of the lesion and fever (16.67%) cases each. Burning sensations were found in (10%) of cases. The majority of patients in our research had crops of non-thrombogenic palpable purpura, which mostly affected dependent regions such as the legs, ankles, feet, and buttocks. This was comparable to prior investigations by Sais et al., [7] Ekenstam et al., [13] and Gupta et al., [6]. Skinny nodules, which made up 17.5% of patients in our sample as opposed to 2% noted by Sais et al., [7] were the second most frequent kind of lesion. This might be attributed to the high incidence of multibacillary leprosy patients in India as well as the high percentage of reactive leprosy cases in our

research. Systemic symptoms were present in 25 instances (83.33%) and were most frequently shown as concomitant joint aches in 14 cases (46.67%) of patients. The musculoskeletal system was most often affected, and this was also consistent with the systemic involvement noted by Ekenstam et al., [12] in 51% of patients. However, Sais et al., [7] found that joint participation was present in 36.7% of all instances whereas systemic involvement was only present in 20% of the cases. N=6 (20.0%) of the patients had gastrointestinal involvement, which mostly manifested as melena, occult blood in stools, and colon edema on abdominal ultrasound. This was comparable with the 9.5% of instances described by Sais et al., [7] In 26.67% of the patients in our study had renal involvement, compared to Winkelmann RK et al., [13] 61% of patients. In our investigation, proteinuria, microscopic hematuria, increased blood urea and creatinine, and collagen vascular abnormalities were all signs of renal involvement. In this study, the lab parameters indicating inflammatory response were elevated 76.67% had Elevated ESR 43.33% of cases had anemia, and 26.6% had leukocytosis. Sais et al., [7] in a similar study found elevated ESR (52.4%), anemia (37%), and leukocytosis (18%) were observed in their cases. The collagen vascular disease workup revealed positive Anti-nuclear antibodies in 13.33% and rheumatoid factor in 6.67% of patients, which was consistent with study results reported by Gupta et al., [6] Histopathology showed features of leukocytoclastic vasculitis in n=25 (83.33%) of the patients. In the remaining n=5, perivascular lymphocytic infiltrate with no evidence of vasculitis was observed which was consistent with a study by Gupta et al., [6] study found n=42 patients, n=5 patients did not show evidence of vasculitis. Hence the diagnosis of cutaneous small vessel vasculitis was considered based on a high index of clinical suspicion. Moreover, histopathology was non-contributory in these cases, probably due to the biopsy of the lesion at a late stage in the disease evolution as mentioned in the literature. [14-16]

Conclusion

The most frequent kind of cutaneous small vessel vasculitis identified in the current investigation was Henoch Schonlein purpura, followed by Erythema nodosum leprosum. Females tended to have cutaneous small vessel vasculitis more often. The majority of the cases were people under 40 years of age. Upper respiratory tract infection was the most prevalent causative factor. The clinical diagnosis of cutaneous small vessel vasculitis is based on the presence of non-thrombocytopenic purpura. Widespread cutaneous symptoms might be a sign of significant systemic involvement on the skin. The diagnosis of cutaneous small vascular vasculitis requires an early biopsy and clinicopathological correlation**.**

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