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General Medicine

IMMUNOLOGICAL PROFILE OF VASCULITIS PATIENTS AT TERTIARY CARE HOSPITAL

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ABSTRACT **Background:** The vasculitides are a heterogenous group of conditions characterized by blood vessel inflammation and necrosis. Vasculitides are relatively uncommon conditions whose etiology is still poorly understood. Treating vasculitis is as regarding as establishing diagnosis. In the absence of treatment, most of patients will suffer or die. With treatment most of patients improve, many will achieve remission and a few will be cured. Disease classification is the process of categorizing illnesses in a larger framework of medical conditions.**Objectives:** (i) To study the immunological profile of vasculitis patients at a tertiary care centre.**Methods:** The present hospital based observational study was conducted in the Department of Internal Medicine, SKIMS Srinagar. The study had two parts; Retrospective and prospective. Retrospective part: All patients of vasculitis who were admitted or evaluated in OPD from March 2012 to Sept. 2018, were enrolled for the analysis. Prospective part: All Patients of vasculitis admitted or evaluated in OPD from Oct. 2018 to May 2020 were enrolled for study. Patients were classified as vasculitis if they fulfill ACR / EULAR / EMA / Chapell Hill consensus classification criteria for vasculitis and biopsy.**Results:** Our study was an observational study of 77 patients. The data was collected both prospectively 50.6 and retrospectively 49.4 large vessel vasculitis was present in 22%, small vessel vasculitis was present in 50.6% and others were 27.27%. Majority of patients i.e. 41 (85.4%) had normal C3/C4 levels with decreased C3 level in 3 (6.3%), C4 in 3 (6.3%) and both C3/C4 in 1 (2.1%) ANA was positive in 16 (23.5%) patients and negative in 52 (76.5%) patients, Anti-dsDNA was positive in 5 (14.7%) patients and negative in 29 (85.3%) patients. ACL was positive in 2 (28.6%) patients and negative in 5 (71.4%) patients, APLA was negative in all the 6 (100%) patients, AntiSM was positive in 3 (33.3%) patients and negative in 6 (66.7%) patients, RF was positive in 8 (26.7%) patients and negative in 22 (73.3%) patients. Anti-CCP was positive in 2 (20%) patients and negative in 8 (80%) patients. As for immunological profile, C-ANCA was positive in 22 (35.5%) and negative in 40 (64.5%), P-ANCA was positive in 5 (8.1%) and negative in 57 (91.9%), Hepatitis B,Hepatitis C, HIV,Anti-RO,Cryoglobin was negative in all patients, Anti-LA and lupus anticoagulant were positive in 3(37.5%) and negative in 5 (62.5%) .

KEYWORDS : C3/C4,ANA,ANTI-CCP ,APLA,ANTI-DS DNA

INTRODUCTION

The vasculitides are a heterogenous group of conditions characterized by blood vessel inflammation and necrosis[1]. Vasculitides are relatively uncommon conditions whose etiology is still poorly understood. Depending on the size, distribution and severity of the affected vessel, vasculitis can result in clinical syndromes that vary in severity from minor self-limiting rash to a life-threating multisystem disorder, recognizing the fact that some vasculitides can affect a wide variety of blood vessels. They are classified as primary or secondary and have their identifiable causes such as infectious agents, drug reactions, systemic autoimmune diseases or malignancy. Since it often begins with nonspecific symptoms and signs, unfolding slowly over weeks or months, vasculitis is one the great diagnostic challenges in all of medicine. Establishing the diagonsis of vasculitis requires lab tests, biopsy of affected vessel or angiogram in some cases or serological tests.

The 1994 International Chapel Hill Consensus Conference on the Nomenclature of Systemic Vasculitides (CHCC 1994) proposed names and definitions for the most common forms of vasculitis[1]. This nomenclature was widely adopted. A second International Chapel Hill Consensus Conference was held in 2012 (CHCC 2012). The goals were to change names and definitions as appropriate, and add important categories of vasculitis not included in CHCC 1994.

Table 1 summarizes the entities classified as vasculitis and the main subcategories according to the 2012 CHCC nomenclature

VASCULITIS CLASSIFICATION		
Primary systemic vasculitis	**Vasculitis category, subcategory or entity**	**Characteristic features**
Large-vessel vasculitis	Takayasu arteritis	Granulomatous Aorto-arteritis usually occurring before age 50 years
	Giant-cell arteritis	Granulomatous Aorto-arteritis predominantly involving the carotid and vertebral arteries occurring after age 50 years and often associated with polymyalgia rheumatica
Mediumvesselvasculitis	Poly-artertitis nodosa	Arteritis of medium/small Arteries without small vessel involvement, glomerulonephritis or antineutrophil cytoplasmic antibodies (ANCAs).
	Kawasaki disease	Childhood mucocutameus lymph node syndrome with arteritis often involving coronary arteries.
Small-vessel vasculitis ANCA associated vasculitis	Microscopic polyangitis	Vasculitis of small/medium vessel and frequent pauci. Immune glomerulonephritis and ANCAs
	Granulomatosis with polyangits (Wegener s)	Granulmatous inflammation of the respiratory tract with vasculitis of small/medium vessels and frequent. Pauci-immune glomerulonephritis and ANCAs
	Eosinophilic granulomatosis with Polyangits (Churg-Strauss)	Asthma, Eosinophilia and eosinphilic granulomatous inflammation frequently involving the respiratory tract with vasculitis of small/medium vessels and sometimes ANCAs
Immune-complex	Antiglomerular basement membrane	Pulmonary and glomerular capillaritis with depositin anti-

smallvessel vasculitis	(anti GBM) disease Cryoglobulinemic vasculitis	GBM antibodies Vasculitis with frequent skin, glomerular and peripheral nerve involvement associated with serum cryoglobulins
	Immunoglobuling (Ig) A vasculitis (Henoch-Schonlein)	Arthritis with frequent skin and gastrointestinal vasculities with IgA deposits and possible IgA neuropathy
	Hypocomplementemi c urticarial vasculitis (anti-C1q vasculitis)	Urticarial hypocomplementic small vessel vasculitis with anti C1 q antibodies and, articular, glomerular, ocular and bronchial disease
Variable vessel vasculitis	Behcet s disease	Recurrent oral and/or genital ulcers with skin, ocular, articular, gastrointestinal, and/or central venous system lesions, and possible variable vessel vasculitis.
	Cogan s syndrome	Vasculitis of small, medium or large arteries occurring in Cogan s syndrome.
Single organ vasculitis	Cutaneous leukocytoclastic angittis Cutaneous arteritis Primary central nervous system vasculitis Isolated aortitis	Vasculitis in a single organ and no features indicating a limited form of a systemic vasculitis
Vasculitis associatd with systemic disease	Lupus vasculitis Rheumatoid vasculitis Sarcoid vasculitis Other (e.g. IgG4-related aortitis)	Vasculitis secondary to a systemic disease
Vasculitis associated with probable cause	Hepatitis C virus – associated cryoglobulineic vasculitis Hepatitis B virus associated vasculitis Syphilis-associated aortitis Drug-associated immune complex vasculitis Drug associated ANCAassociated vasculitis Others	Vasculitis secondary to specific cause

ANCA = Anti-neutriphil cytoplasmic antibody

Table 2. Selected sets of classification criteria for the main vasculitis entities

Vasculitis entity	Classification systems	Comments
Giant cell arteritis	ACR criteria[12]	Should be used in combination with vasculitis entry criteria
	Positive temporal artery biopsy (TAB)	No consensual histological definition for positive TAB Exclude by definition TABnegative disease
Takayasu arteritis	ACR criteria[13]	Should be used in combination with vasculitis entry criteria
	Published by Sharma et al[11] EULAR/PRINTO/PRES[14]	Expert based criteria Developed for pediatric populations. Should be used with vasculitis entry criteria
Polyarteritis nodosa PAN	CHCC definition[2,3]	Not intended as classification criteria
	ACR criteria[15]	Should be used in combination with vasculitis entry criteria. Low sensitivity and specificity
	FVSG criteria[10]	Should be used in combination with vasculitis entry criteria. Moderate sensitivity and specificity
	EMA algorithm[16]	Discriminates PAN from GPA,
	EULAR/PRINTO/PRES[14]	Developed for pediatric populations. Should be used in combination with vasculitis entry criteria
Kawasaki disease Granulomatosis with polyangitis (GPA, (Wegner"s)	American Heart Association[7]	May not work well in adult populations
	ACR criteria[17]	Should be used in combination with vasculitis entry criteria
	Modified ACR criteria[8]	Alteration of the ACR criteria17. Should be used in combination with vasculitis entry criteria
	EMA algorithm[16]	Discriminates GPA from MPA, EGPA and PAN
	EULAR/PRINTO/PRES[14]	Developed for pediatric populations. Should be used in combination with vasculitis entry criteria
Microscopic polyangitis (MPA)	EMA algorithm[16]	Discriminates MPA from GPA, EGPA and PAN
Eosinophilic granulomatosis with polyangitis (EGPA), (Churg-Strauss)	ACR criteria[6]	Should be used in combination with vasculitis entry criteria. Discriminant ability from hypereosinophilic syndrome [HES] unclear
	Published by Lanhan et al[18]	Expert-based criteria. Discriminant ability from (HES) unclear
	EMA algorithm[16]	Discriminates EGPA from GPA, MPA and PAN. Discriminant ability from HES unclear
IgA vasculitis (Henoch-Schonlein)	ACR criteria[11]	May not work well in adult populations, should be used in combination with vasculitis entry criteria

	Published by Michael et al[19]	Discriminates IgA vasculitis from hypersensitivity vasculitis
	EULAR/PRINTO/PRES[14]	Developed for pediatric population. Should be used in combination with vasculitis entry criteria
Cryoglobulinemic vasculitis Behcet"s disease	Published by de Vita et a[14]	Validation study published separately20
	ICBD[5]	More specific than the recently published ICBD criteria5
	1987 JBDRC Criteria[21]	More sensitive than former ISG criteria9 Expert based criteria. Predominantly used in the Asian content.

ACR,American College of Rheumatology; CHCC, Chapel Hill Consensus Conference; EMA,European Medicines Agency; EULAR/PRINTO/PRES, European League against Rheumatism/ Paediatric Rheumatology International Trial Organization/Paediatric Rheumatology European Society; FVSG, French Vasculitis Study Group; ICBD, International criteria for Behcet s disease; ISG, International Study Group; JBDRC, Japanese Behcet s disease research committee

AIMS AND OBJECTIVE:

To study the immunological profile of vasculitis patients at a tertiary care centre.

METHODS

The present hospital based observational study was conducted in the Department of Internal Medicine, SKIMS Srinagar. The study had two parts; Retrospective and prospective.

Retrospective part: All patients of vasculitis who were admitted or evaluated in OPD from March 2012 to Sept. 2018, were enrolled for the analysis.

Prospective part: All Patients of vasculitis admitted or evaluated in OPD from Oct. 2018 to May 2020 were enrolled for study.

Inclusion Criteria

Age > 18 to 85 years

Patients fulfilling ACR / EULAR / EMA / Chapell Hill consensus classification criteria and biopsy evidence of vasculitits.

Patient who give consent

Exclusion Criteria

Age <18 years and >85 years.

Patients who refuse to consent.

Patients were classified as vasculitis if they fulfill ACR / EULAR / EMA / Chapell Hill consensus classification criteria for vasculitis and biopsy.

No major ethical issues are involved as the study does not involve any interventional experimentation, since it is purely an observational study. However, informed consent for confidentiality and permission for publishing the data was taken.

Statistical Methods

The recorded data was compiled and entered in a spreadsheet (Microsoft Excel) and then exported to data editor of SPSS Version 20.0 (SPSS Inc., Chicago, Illinois, USA). Continuous variables were expressed as Mean±SD and categorical variables were summarized as frequencies and percentages. Graphically the data was presented by bar and pie diagrams

RESULTS

Our study was an observational study of 77 patients. The data was collected both prospectively 39 (50.6%) and retrospectively 38 (49.4%). Mean age of study patients was 40.9±15.72 years. Our study consisted of male 33 (42.9%), female 44 (57.1%). 62 (80.5%) belonged to rural areas where as 15 (19.5%) was from urban areas. In our study, large vessel vasculitis was present in 15 (19.48%) including Takayasu arthritis in 14 (93.33%) and Giant cell arteritis in 1 (6.67%), medium vessel vasculitis was present in 3 (3.89%) including polyarteritis nodosa in 1 (33.37%) and superior mesentric arteritis in 2 (66.66%), ANCA associated small vessel vasculitis was present in 27 (35.06%) including GPA in 22 (81.48%), EGPA in 4 (14.81%) and MPA in 1 (3.70%), immune complex small vessel vasculitis (IgA vasculitis) was seen in 2 (2.59%). Variable vessel vasculitis (Behcet's disease) was present in 6 (7.79%), single organ vasculitis was seen in 6 (7.79%) patients including cutaneous vasculitis in 4 (68.66%) and CNS vasculitis in 2 (33.34%). Vasculitis associated with systemic disease was present in 4 (5.19%) patients including lupus vasculitis in 2 (50%) and rheumatoid vasculitis in 2 (50%) patients. Vasculitis associated with probable cause (NSAIDs induced vasculitis) was present in 1 (1.29%) patient. Biopsy evidence of vasculitis (not fulfilling criteria for other small vessel vasculitis) was present in 13 (16.88%).

Table 1: Distribution of study		Population
Study population	No.	%age
Prospective cases	39	50.6
Retrospective cases	38	49.4
Total	77	100

Out of total 77 patients studied, 39 (50.6%) were prospective cases and 38 (49.4%) cases were of retrospective nature.

Table 1A: Distribution of study population

Study population	LVV		SVV		Others	
	No.	%age	No.	%age	No.	%age
Prospective cases	9	60.0	22	55.0	8	36.4
Retrospective cases	6	40.0	18	45.0	14	63.6
Total	15	100	40	100	22	100

Patients were distributed in three groups viz. LVV(large vessel vasculitis), SVV(small vessel vasculitis) and others. There were 9 (60%) prospective cases and 6 (40%) retrospective cases in LVV group, 22 (55%) prospective cases and 18 (45%) retrospective cases in SVV group while as 8 (36.4%) prospective and 14 (63.6%) retrospective cases constitute others group.

Table 2: Age distribution of study patients		
Age (Years)	No.	%age
≤ 30	23	29.9
31-40	19	24.7
41-50	14	18.2
51-60	12	15.6
61-70	9	11.7
Total	77	100
Mean±SD (Range)=40.9±15.72 (18-76)		

The age of participants of the study ranged between 18-76 years with a mean age of 40.9±15.72 years.

Table 2A: Age distribution of study patients						
Age (Years)	LVV		SVV		Others	
	No.	%age	No.	%age	No.	%age
≤ 30	5	33.33	11	27.5	7	31.80
31-40	5	33.33	9	22.5	5	22.72
41-50	3	20.0	9	22.5	2	9.1
51-60	1	6.67	7	17.5	4	18.19
61-70	1	6.67	4	10.0	4	18.19
Total	15	100	40	100	22	100

When groups were distributed as per the age, it was observed that majority of patients i.e. 5 (33.33%) each in LVV group belonged to age groups of ≤30 and 31-40 years. In SVV group, majority of patients i.e. 11 (27.5%) belonged to age group of ≤30 years followed by 9 (22.5%) patients each who aged between 31-40 years and 41-50 years. There were 7 (31.80%) patients in other group who aged ≤30 years followed by 5 (22.72%) patients who belonged to the age group of 41-50 year.

Table 3: Gender distribution of study patients		
Gender	No.	%age

Male	33	42.9
Female	44	57.1
Total	77	100

There was a little female predominance in our study with 44 (57.1%) females and 33 (42.9%) males.

Table 3A: Gender distribution of study patients

Gender	LVV		SVV		Others	
	No.	%age	No.	%age	No.	%age
Male	12	80.0	8	20.0	13	59.0
Female	3	20.0	32	80.0	9	41.0
Total	15	100	40	100	22	100

When groups were distributed as per the gender, it was observed that in group LVV there were 12 (80%) males compared to 3 (20%) females. In group SVV, there were 8 (20%) males compared to 32 (80%) females while as 13 (59%) males and 9 (41%) females constituted others group.

Table 6: Immunological profile of study patients

Immunological profile	No.	%age	
C3/C4	WNL	41	85.4
	Decreased C3	3	6.3
	Decreased C4	3	6.3
	Decreased both C3 and C4	1	2.1
ANA	Positive	16	23.5
	Negative	52	76.5
Anti dsDNA	Positive	5	14.7
	Negative	29	85.3
ACL	Positive	2	28.6
	Negative	5	71.4
APLA	Positive	0	0.0
	Negative	6	100
Anti SM	Positive	3	33.3
	Negative	6	66.7
RF	Positive	8	26.7
	Negative	22	73.3
Anti CCP	Positive	2	20.0
	Negative	8	80.0

C3/c4;complement,ANA;anti-nucleic acid antibody,Anti dsDNA;Anti double DNA antibody,ACL;Anti Cardiolipin antibody,APLA;Anti phospholipid antibody,Anti SM; Anti smith antibody,RF ; Rheumatiod factor, Anti CCP; Anti cirtulite antibody

Majority of patients i.e. 41 (85.4%) had normal C3/C4 levels with decreased C3 level in 3 (6.3%), C4 in 3 (6.3%) and both C3/C4 in 1 (2.1%) ANA was positive in 16 (23.5%) patients and negative in 52 (76.5%) patients, Anti-dsDNA was positive in 5 (14.7%) patients and negative in 29 (85.3%) patients. ACL was positive in 2 (28.6%) patients and negative in 5 (71.4%) patients, APLA was negative in all the 6 (100%) patients, AntiSM was positive in 3 (33.3%) patients and negative in 6 (66.7%) patients, RF was positive in 8 (26.7%) patients and negative in 22 (73.3%) patients. Anti-CCP was positive in 2 (20%) patients and negative in 8 (80%) patients.

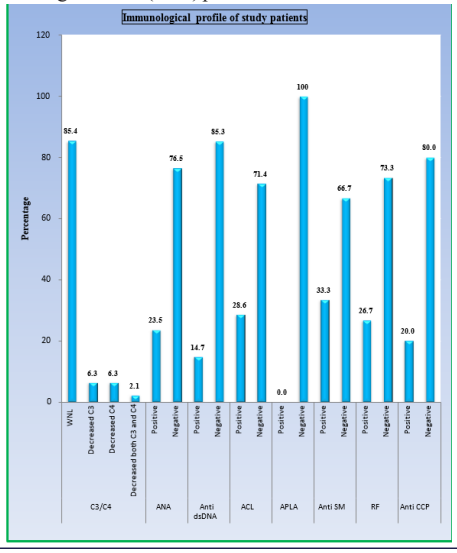
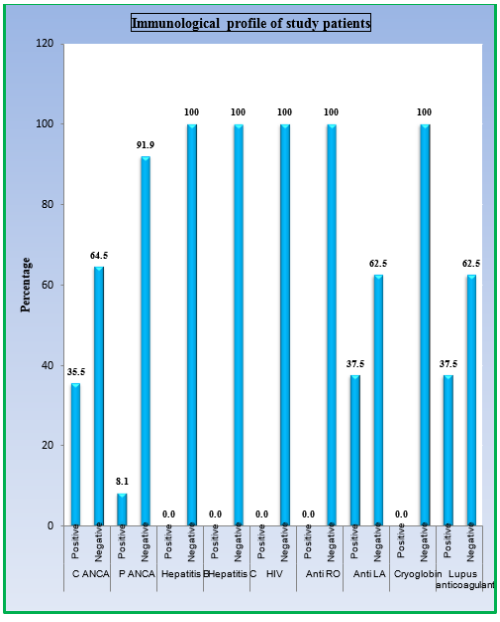


Table 6A: Immunological profile of study patients

Immunological profile		LVV		SVV		Others	
		No.	%age	No.	%age	No.	%age
C3/C4	WNL	7	100	24	80.0	10	90.9
	Decreased C3	0	0.0	3	10.0	0	0.0
	Decreased C4	0	0.0	3	10.0	0	0.0
	Decreased both C3 and C4	0	0.0	0	0.0	1	9.1
ANA	Positive	3	27.3	8	21.1	5	26.3
	Negative	8	72.7	30	78.9	14	73.7
Anti dsDNA	Positive	0	0.0	3	15.8	2	25.0
	Negative	7	100	16	84.2	6	75.0
ACL	Positive	0	0.0	0	0.0	2	66.7
	Negative	3	100	1	100	1	33.3
APLA	Positive	0	0.0	0	0.0	0	0.0
	Negative	1	100	2	100	3	100
Anti SM	Positive	1	100	1	33.3	1	20
	Negative	0	0.0	2	66.7	4	80
RF	Positive	0	0.0	6	35.3	2	20
	Negative	3	100	11	64.7	8	80
Anti CCP	Positive	0	0.0	1	16.7	1	25
	Negative	0	0.0	5	83.3	3	75

There were 7 (100%) had normal C3/C4 levels in LVV group, 24 (80%) patients in SVV group with 3 having decreased C3 levels and 10 (90.9%) patients in others group have normal C3/C4 levels. ANA was positive in 3 (27.3%) patients, 8 (72.7%) negative patients in LVV group, 8 (21.1%) positives and 30 (78.9%) negative in SVV patients, with 5 (26.3%) positive ANA patients and 14 (73.7%) negative patients in others group. Anti-dsDNA was positive in 3 (15.8%) in SVV group and 2 (25%) in others group with 7 (100%) negative in LVV group. 16 (84.2%) in SVV group and 6 (75%) in Other group. ACL is negative in all patients with SVV and LVV and in 1 (33.3%) in Other group and positive in 2 patients (66.7).APLA was negative in all groups.Anti SM was positive in 1 (100%) in LVV group with 1(33.3%) having positive and 2 (66.7%) negative in SVV group.In Other group Anti SM is positive in 1 (20%) and negative in 4 (80%).RF was positive in 6 (35.3%) and negative in 11(64.7%) in SVV .while 2 (20%) are positive and 8 (80%) are negative in Other group.Anti CCP was positive in 1 (16.7%) and negative in 5 (83.3%) in SVV group while other group was having negative in 3 (75%) and positive in 1 (25%).



As for immunological profile, C-ANCA was positive in 22 (35.5%) and negative in 40 (64.5%), P-ANCA was positive in 5 (8.1%) and negative in 57 (91.9%), Hepatitis B,Hepatitis C, HIV,Anti-RO,Cryoglobin was negative in all patients, Anti-LA and lupus anticoagulant were positive in 3(37.5%) and negative in 5 (62.5%) .

C- ANCA was positive in 22 (56.4%) patients in SVV group, P-ANCA was positive in 5 (12.8%) patients in SVV group, Hepatitis B was negative in all three groups, Hepatitis C was negative in all patients. HIV was negative in all the patients. Anti – RO was negative in all three

study groups. Anti-LA was positive in 3 (75%) patients in SVV group. Lupus anticoagulant was positive in 2 (33.3%) in SVV group and 1 (100%) patients in Others group

Table 7: Immunological profile of study patients			
Immunological profile		**No.**	**%age**
C ANCA	Positive	22	35.5
	Negative	40	64.5
P ANCA	Positive	5	8.1
	Negative	57	91.9
Hepatitis B	Positive	0	0.0
	Negative	77	100
Hepatitis C	Positive	0	0.0
	Negative	77	100
HIV	Positive	0	0.0
	Negative	77	100
Anti-RO	Positive	0	0.0
	Negative	7	100
Anti-LA	Positive	3	37.5
	Negative	5	62.5
Cryoglobin	Positive	0	0.0
	Negative	6	100
Lupus anticoagulant	Positive	3	37.5
	Negative	5	62.5

Table 7A: Immunological profile of study patients							
Immunological profile		**LVV**		**SVV**		**Others**	
		No.	**%age**	**No.**	**%age**	**No.**	**%age**
C ANCA	Positive	0	0.0	22	56.4	0	0.0
	Negative	11	100	17	43.6	12	100
P ANCA	Positive	0	0.0	5	12.8	0	0.0
	Negative	11	100	34	87.2	12	100
Hepatitis B	Positive	0	0.0	0	0.0	0	0.0
	Negative	15	100	40	100	22	100
Hepatitis C	Positive	0	0.0	0	0.0	0	0.0
	Negative	15	100	40	100	22	100
HIV	Positive	0	0.0	0	0.0	0	0.0
	Negative	15	100	40	100	22	100
Anti RO	Positive	0	0.0	0	0.0	0	0.0
	Negative	1	100	3	100	3	100
Anti LA	Positive	0	0.0	3	75.0	0	0.0
	Negative	1	100	1	25.0	3	100
Cryoglobin	Positive	0	0.0	0	0.0	0	0.0
	Negative	1	100	3	100	2	100
Lupus anticoagulant	Positive	0	0.0	2	33.3	1	100
	Negative	1	100	4	66.7	0	0

Table 8: Final diagnosis of study patients		
Diagnosis	**No.**	**%age**
TA	14	18.2
GPA	22	28.6
EGPA	4	5.2
MPA	1	1.3
Behcets disease	6	7.8
Ig A Vasculitis	2	2.6
NSAID induced vasculitis	1	1.3
Lupus vasculitis	2	2.6
SLE with SVV	1	1.3
Cutaneous vasculitis	3	3.9
SLE with cutaneous vasculitis	1	1.3
CNS vasculitis	2	2.6
Rheumatoid vasculitis	1	1.3
RV with intersitial pheumonia	1	1.3
GCA	1	1.3
Isolated SMA vasculitis	1	1.3
SVV	10	13.0
PAN with ILD	1	1.3
SVV with IgA nephropathy	1	1.3
SLE with SMA vasculitis	1	1.3
UCTD with SVV	1	1.3
Total	77	100

Final diagnosis was GPA in 22 (28.6%) patients, TA in 14 (18.2%) patients, biopsy evidence of SVV in 13 (16.88%) patients, Behcets disease in 6 (7.8%) patients, cutaneous vasculitis in 4 (5.1%) patients, EGPA in 4 (5.1%) patients. Two patients each were diagnosed as IgA Vasculitis, lupus vasculitis and CNS vasculitis. PAN, GCA and NSAID induced vasculititis were the diagnosis of 1 (1.3%) patient each.

DISCUSSION

In our study, majority of patients i.e. 41 (85.4%) had normal C3/C4 levels, ANA was positive in 16 (23.5%) patients and negative in 52 (76.5%) patients, Anti-dsDNA was positive in 5 (14.7%) patients and negative in 29 (85.3%) patients. ACL was positive in 2 (28.6%) patients and negative in 5 (71.4%) patients, APLA was negative in all the 6 (100%) patients, Anti-SM was positive in 3 (33.3%) patients and negative in 6 (66.7%) patients, RF was positive in 8 (26.7%) patients and negative in 22 (73.3%) patients. Anti-CCP was positive in 2 (20%) patients and negative in 8 (80%) patients. There were 7 (100%) had normal C3/C4 levels in LVV group, 24 (80%) patients in SVV group and 10 (90.9%) patients in others group. ANA was positive in 3 (27.3%) patients, 8 (72.7%) negative patients in LVV group, 8 (21.1%) positives and 30 (78.9%) negative in SVV patients, with 5 (26.3%) positive ANA patients and 14 (73.7%) negative patients in others group. Anti-dsDNA was positive in (15.8%) in SVV group and 2 (25%) in others group with 7 (100%) negative in LVV group. Ahn SS et al (2020)[25] conducted a study in which mean C3 (mg/dL) was 112.0±25.0 and mean C4 (mg/dL) was 24.5±8.9. Nooshin D et al (2013)[22] conducted a study in whichall the patients had negative results for RF and ANA. Fauci AS et al (1983)[23] conducted a study in which rheumatoid factor (latex fixation) was measured in 44 patients before institution of therapy or during periods of disease activity and was positive in 27 patients. The mean titer was 1:128 and was never greater than 1:4096. These studies are consistent with the finding of the present study.

As for immunological profile, C-ANCA, P-ANCA, Hepatitis B, Hepatitis C, HIV, Anti-RO, Anti-LA,

Cryoglobin and lupus anticoagulant were all negative in majority of patients. C ANCA was positive in 22 (56.4%) patients in SVV group, P ANCA was positive in 5 (12.8%) patients in SVV group, hepatitis B was negative in all three groups, hepatitis C was negative in all patients. HIV was negative in all the patients. Anti – RO was negative in all three study groups. Anti-LA was positive in 3 (75%) patients in SVV group. Lupus anticoagulant was positive in 2 (33.3%) in SVV group and 1 (100%) patients in others group.

Lane SE et al (2005)[24] conducted a study in which ANCA were positive for 73/94 PSV cases (77.7%), results being unavailable for five patients (3 WG, 1 MPA, 1 CSS). In WG, 88.9% of patients were ANCApositive: 31 (57.4%) cANCA/PR3-ANCA, 9 (16.7%) pANCA/MPOANCA, 8 (14.8%) undetermined ANCA. Nooshin D et al (2013)[22] conducted a study in which all the patients had negative results for ANCA.

Choi H et al (2020)[26] conducted a study in which they investigated the frequency of ANCA positivity and its clinical implications in patients with TAK. In their P-ANCA was seen in 8 (6.6%) patients and C-ANCA in 2 (1.7%) patients. In a previous study, Cohen Tervaert and colleagues (14) suggested that in stable patients with Wegener granulomatosis, a rise in C-ANCA titer usually portends a clinical exacerbation. In line with other studies[27,2,27] also, results were consistent with our study.

CONCLUSION

- Out of total 77 patients studied, 39 (50.6%) were prospective cases and 38 (49.4%) cases were of retrospective nature. There were 9 (60%) prospective cases and 6 (40%) retrospective cases in LVV group, 22 (55%) prospective cases and 18 (45%) retrospective cases in SVV group while as 8 (36.4%) prospective and 14 (63.6%) retrospective cases constitute others group.
- Majority of patients i.e. 5 (33.33%) each in LVV group belonged to age groups of ≤30 and 31-40 years. In SVV group, majority of patients i.e. 11 (27.5%) belonged to age group of ≤30 years followed by 9 (22.5%) patients each who aged between 31-40 years and 41-50 years.
- In group LVV there were 12 (80%) males compared to 3 (20%) females. In group SVV, there were 8 (20%) males compared to 32 (80%) females while as 13 (59%) males and 9 (41%) females constituted others group.
- Majority of patients i.e. 41 (85.4%) had normal C3/C4 levels, ANA was positive in 16 (23.5%) patients and negative in 52 (76.5%) patients, Anti-dsDNA was positive in 5 (14.7%) patients

and negative in 29 (85.3%) patients. ACL was positive in 2 (28.6%) patients and negative in 5 (71.4%) patients, APLA was negative in all the 6 (100%) patients, Anti-SM was positive in 3 (33.3%) patients and negative in 6 (66.7%) patients, RF was positive in 8 (26.7%) patients and negative in 22 (73.3%) patients. Anti-CCP was positive in 2 (20%) patients and negative in 8 (80%) patients.

- There were 7 (100%) had normal C3/C4 levels in LVV group, 24 (80%) patients in SVV group and 10 (90.9%) patients in others group. ANA was positive in 3 (27.3%) patients, 8 (72.7%) negative patients in LVV group, 8 (21.1%) positives and 30 (78.9%) negative in SVV patients, with 5 (26.3%) positive ANA patients and 14 (73.7%) negative patients in others group. AntidsDNA was positive in 3 (15.8%) in SVV group and 2 (25%) in others group with 7 (100%) negative in LVV group.
- As for immunological profile, C-ANCA, P-ANCA, Hepatitis B, Hepatitis C, HIV, Anti-RO, Anti-LA, Cryoglobin and lupus anticoagulant were all negative in majority of patients. C ANCA was positive in 22 (56.4%) patients in SVV group, P ANCA was positive in 5 (12.8%) patients in SVV group, hepatitis B was negative in all three groups, hepatitis C was negative in all patients. HIV was negative in all the patients.

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