

Pattern of Autoimmune Dermatological Disorders Amongst Outpatients Attending SKIMS MCH Kashmir

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ABSTRACT

Introduction: The pattern of dermatological disorders vary from one region to another. Autoimmune entities form a significant sub group of these. However even the pattern of autoimmune dermatological disorders vary from one region to another. Objective was to study the pattern of autoimmune dermatological disorders in Kashmir amongst the outpatients attending a tertiary care teaching hospital.

Material and methods: All consecutive autoimmune dermatological disorders with a proven autoimmune etiology attending the outpatient dermatology department of a tertiary care teaching government hospital during 2015 starting from 1st Jan of the year were included in the study irrespective of duration of disease. Diagnosis was made on clinical grounds alone in a majority of cases. Only in a few cases relevant investigations like Tzanck smear and ANA titres were done. All relevant data such as history and dermatological examination were recorded.

Results: 1000 patient of autoimmune disorders were seen and they constituted 6.46% of total dermatological cases. Females (55.4%) and age group of 10-30yrs (56.9%) predominated the study. 712 patients (71.2%) had vitiligo, 145 (14.5%) had alopecia areata, 35 (3.5%) had chronic urticaria, 72 (7.2%) had collagen vascular and 36 (3.6%) had immunobullous dermatosis

Conclusion: Autoimmune dermatological patients form significant percentage of dermatological disorders and vitiligo, alopecia areata and collagen vascular disorders form the bulk of these patients.

Key words: Autoimmune, Collagen Vascular Disease

INTRODUCTION

The pattern of skin diseases in any area depends upon many factors-genetic, racial, religious, environmental, climatic and geographical. Kashmir is a high altitude area in the North of India. A number of studies regarding various autoimmune dermatological disorders can be cited from Kashmir.¹⁻⁶ But there is no study encompassing all the autoimmune disorders collectively to the best of our knowledge.

Keeping this in view an attempt was made to see and document the pattern of various autoimmune dermatological diseases in Kashmir, amongst the outpatients attending a tertiary care teaching government hospital.

MATERIAL AND METHODS

This study was a prospective hospital based study conducted in a teaching hospital associated with a Tertiary care facility in Kashmir. In addition to general OPD catering to all districts of Kashmir, in particular North Kashmir in view of its location, referrals from departments like Rheumatology,

hematology, internal medicine and endocrinology contribute to the patient flow. The OPD attendance for dermatology patients for the year of 2015 was also recorded.

All newly diagnosed ethnic Kashmiri patients with proven autoimmune dermatological disorders were included in the study irrespective of age, sex and duration of disease. Diagnosis in a majority of cases was made on clinical grounds alone. Relevant investigations like Tzanck smear, Histopathology and Direct Immunofluorescence of the skin biopsy, autologous serum skin test and ANA titres were done when required.

All relevant data such as demographic data, complete history and dermatological examination was documented and entered in the study protocol

Psoriasis, sarcoidosis and lichen planus were not included as autoimmune etiology of these disorders is still under consideration. Cutaneous Vasculitis was also excluded due to its varied etiology.

RESULTS

This study was conducted over a period of one year and during this period a total of 15, 479 patients were attended to by the consulting dermatologist and among them a total of 1000 patients fulfilled the inclusion criteria and were included in this study as autoimmune disorders. They constitute about 6.46% of the total cases seen by the dermatologist. These conditions were more common in females (55.4%) as compared to males (446/554). The most common age group to be affected by these disorders was from 10 to 30 years (56.9%) [Table-1,2].

Vitiligo was the most common condition comprising about 71.2% (712) of cases seen in our study followed by alopecia areata (14.5%) and chronic urticaria (3.5%) [Table-3].

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Disease	No. of Patients	Sex Distribution		Age Groups			
		Male	Female	<10 Years	10-30yrs	30-50 years	>50 Years
Vitiligo	712	311	401	(198)	(405)	(101)	8
Alopecia Areata	145	94	51	25	92	26	2
Chronic Urticaia	35	7	28	2	15	13	5
Bullous Disorders	36	15	21	1	22	8	5
connective tissue disorder	72	12	60	2	35	28	7
Total Patients		439	561	228	569	176	27

Table-1: Age and sex distribution of various autoimmune dermatological conditions

Diagnosis	Male	Female
DLE	6	8
SLE	-	8
SCLE	1	1
MCTD	-	5
Sjog	-	1
DMS	1	3
Morphea	2	11
Atrophoderma	1	1
Scleroderma	1	21

Table-2: Disease distribution of various autoimmune connective tissue disorders

Type of disease	Number of patients	% age of total
Vitiligo	712	71.2
Alopecia areata	145	14.5
Chronic urticaria	35	3.5
Bullous diseases	36	3.6
Connective tissue disorders	72	7.2
Total		

Table-3: Disease wise distribution of patients

Type of vitiligo	No of patients seen
Focal	20(2.8%)
Vulgaris	536(75.28%)
Acrofacial	81(11.37%)
Segmental	71(9.97%)
Universal	4(0.5%)

Table-4: Clinical variants of vitiligo

Type of alopecia areata	No of patients
Areata	130(89.65%)
Universalis	3(2.06%)
Totalis	5(3.44%)
Ophiasis	7(4.82%)

Table-5: Clinical variants of alopecia areata

Among vitiligo patients, vitiligo vulgaris (75.28%) was the commonest [Table -4] and areata type was the most common presentation of patients with alopecia (89.65%) [Table-5]. Pemphigus vulgaris (44.44%) was the commonest bullous disorder. (Table-6). 35 cases of chronic urticaria with positive autologous serum skin test (ASST) were seen during the study period. 12 out of 35 (34.28%) patients had an identifiable underlying cause, with 50% of these being positive for anti nuclear antibody (ANA) (Table-7). The

Type of Bullous Disorders	No:of patients seen
Pemphigus Vulgaris	16 (44.44%)
Pemphigus Vegetans	1(2.77%)
IgA Pemphigus	2(5.55%)
Paraneoplastic Pemphigus	1(2.77%)
Sub Corneal Pustular Dermatoses	1(2.77%)
Bullous Pemphigoid	4(11.11%)
Dermatitis Herpetiformis	8(22.22%)
Linear IgA disease	2(5.55%)
EBA Acquisita	1(2.77%)

Table-6: Clinical variants of bullous disorders

Case 1	Age/sex	ANA	Associated diagnosis
Case 1	40/f	+	Anti jo antibody + Leucopenia
Case 2	35/f	+	MCTD
Case 3	20/f	-	Anti TPO+
Case 4	7/f	+	JRA
Case 5	40/f	-	M band in serum neutrophilia CBC
Case 6	35/m	-	Adult onset Stills
Case 7	20/f	+	C4 C 1 esterase low
Case 8	20/f	-	Anti TPO +
Case 9	20/f	+	DMS
Case 10	20/f	-	C4 c1 esteraseslow
Case 11	40/m		Anti TPO +

ASST +ve chronic urticaria with identifiable causes

Table-7: Identifiable cause of chronic urticaria

Diagnosis	No:	Variants
Discoid lupus erythematosus	14	Localized 5
		Disseminated 2
		Chilblain 6
		Profundus 1
Systemic lupus erythematosus	8	
Subacute cutaneous lupus	2 (annular)	
MCTD	5	
Sjogrens	1	
DMS	4	
Morphea	13	Linear 6
		Encoup 2
		Generalized 1
		Plaque 4
Atrophoderma	2	
Scleroderma gp	22	Lcsc 14
		Dessc 6

Table-8: Clinical variants of connective tissue disorders

chilblain variant was the commonest variant (42.85%) of DLE observed as was the linear variant of morphea (46.15%) (Table -8).

DISCUSSION

This study was conducted over a period of one year till consecutive total of 1000 patients were seen in the dermatology OPD fulfilling the inclusion criteria for this study. These autoimmune dermatological conditions were more common in females (55.4%) as compared to males. The most common age group to be affected by these disorders was from 10 to 30 years (56.9%). This is in consonance with other studies like the ones conducted by Rajpal S et al⁷ and Arycan O et al⁸.

Vitiligo was the most common condition seen in our cases. 712 patients (71.2%) had vitiligo. Vitiligo is a very common condition in Indian subcontinent and in this region the highest incidence is seen in India⁹. This explains the high incidence of vitiligo in our patients. Vitiligo vulgaris was the most common type of vitiligo in our study, seen in 75.28% cases of vitiligo, followed by acrofacial. Vitiligo vulgaris has been mentioned as the most common variant by various other studies like Suman S et al⁹ and Shah et al¹⁰. The disease distribution for vitiligo was almost same in both genders (male: female 43.67%: 56.3%). Various studies have shown the disease distribution to be same in both sexes^{11,12,13}, though a few studies have documented a higher incidence in females as was seen by Shah et al¹⁰ and who attributed it to early seeking of medical attention by females in view of cosmetic reasons. We saw that vitiligo was most common in the young age group of 10 to 30 years. This indicates the early onset of disease as was also seen by various studies like Shah et al¹⁰.

In our study, we saw 145 patients of alopecia areata. This formed 14.5% of total patients with autoimmune dermatological conditions seen in this study. Most of the patients were males (64.82%). Similar results have been reported by various studies like Maitreyee Panda et al¹⁴ who report that sex ratio is almost equal in first two decades but male predominance in subsequent decades. Most of our patients were in the age group of 11 to 30 years (63.44%). Similar results have been reported by other studies like Manzoor S et al⁶, who reported that the maximum number of cases presented in the third decade of life. But the disease may manifest at any age. The youngest patient we saw was 2 year old and oldest patient we saw was 60 year old. Seetharaman et al¹⁵ in their study saw the age range of these patients from 4 months to 70 years. In our study, alopecia areata was the most common type of alopecia seen (89.65%). Areata is the most common form recognized in other studies as well.^{14,16} The prevalence of other variants varies in different studies, but these variants form only a miniscule of patients.

Bullous disorders are a common group of dermatological disorders. These disorders are diagnosed by clinical features and histopathologic and immunologic investigations. In our study, we saw a total of 36 patients with bullous disorders. These formed 3.6% of the study group. Amongst these disorders, Pemphigus vulgaris was the most common

disorder (44.44%), followed by Dermatitis Herpetiformis (22.22%) and Bullous Pemphigoid (11.11%). Other bullous disorders seen included Pemphigus Vegetans, IgA Pemphigus, Paraneoplastic Pemphigus, EBA Acquisita, Linear IgA disease and Sub Corneal Pustular Dermatoses. Pemphigus vulgaris is the most common variant of bullous disorder as seen in the literature¹⁷. Pemphigus vulgaris (PV) is a potentially life-threatening autoimmune disease affecting the skin and/or mucosa.^{18,19} Dermatitis Herpetiformis is a rare disease epidemiologically.¹⁹ This was the second most common bullous disorder in our study. The reason for the same could be the variation in prevalence of this disease from country to country, as has been reported in literature.^{20,21,22} Bullous pemphigoid is the most common pemphigoid disorder. In Indian population, this disorder is less common than Pemphigus vulgaris²³ and we saw similar results in our study. Just like the other world literature, other bullous disorders were seen in a lesser frequency.

DLE was seen almost exclusively in patients belonging to Gujjar tribe. The chilblain variant was seen here and this could be attributable to cold weather conditions. A good number of morphea patients were seen overwhelmingly young females the linear one being the most common. Interestingly one patient had a female sibling with pemphigus vulgaris.

35(3.5%) of our cases had chronic urticaria with a positive ASST. Chronic urticaria is a common dermatologic disorder. As has been reported by various studies^{24,25}, the patients had a range of presentations. Most of the patients presented between second and third decade of life, as is the trend seen in literature²⁵. Another trend seen in these patients was that they had sought consultations from many doctors prior to visiting us. This is also a known case scenario in case of chronic urticaria²⁶. Extensive laboratory tests are not required in the vast majority of patients.²⁶ Although the functional in-vitro donor basophil histamine assay is the gold standard for detecting autoimmune urticaria,²⁷ this bioassay is difficult to standardize because it requires fresh basophils from healthy donors and is time consuming. Autologous serum skin test (ASST) is a simple in-vivo clinical test for the detection of this basophil histamine releasing activity.²⁸ We in our study included the patients with a positive ASST as autoimmune urticaria.

Our study was a first of its kind from our part of the world, to the best of our knowledge. But it had its limitations also. Coexistence of other autoimmune disorders of thyroid was not routinely included in the study proforma. Familial association of autoimmune disorders was not sought. An underrepresentation of SLE may be seen as most SLE patient report to a rheumatologist directly. An exclusion of vasculitis was done as cutaneous vasculitis is not always autoimmune and it is only after detailed workup that a plausible cause can be diagnosed. Also HSP cases are catered by paediatricians usually.

CONCLUSION

Autoimmune dermatologic disorders are quite common in dermatologic practice. Vitiligo was the most common

autoimmune dermatologic disorder seen in our patients, followed by alopecia areata, chronic urticaria and bullous disorders. Most of the cases are diagnosed clinically and do not require extensive investigations.

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