CLINICAL PROFILE OF SEGMENTAL VITILIGO IN A GROUP OF PAKISTANI PATIENTS

Aamir Habib

Combined Military Hospital Kohat Pakistan

ABSTRACT

Objective: To determine clinical profile of segmental vitiligo in a group of patients reporting to Combined Military Hospital Kohat.

Study Design: A descriptive study.

Place and Duration of Study: Combined Military Hospital Kohat, from Sep 2016 to Aug 2018.

Material and Methods: All new patients clinically diagnosed to be having Segmental vitiligo, were included in the study. A detailed history was obtained, thorough physical examination was performed, and findings were recorded on a specially designed proforma for each patient separately. Computer programme SPSS-14 was used to manage and analyze the data.

Results: Out of 56 patients, 33 (58.9%) were male and 23 (41.1%) were female. Mean age at onset was 7.1308 \pm 3.93107 years. The most frequent age of onset of segmental vitiligo was found to be between 5 to 10 years. Average duration of the disease was 2.6271 \pm 3.4410 years. Average duration of the disease was 3.5021 \pm 4.1041 years in male patients and 1.3717 \pm 1.5185 years in female patients.

The disease most commonly involved the area of innervation of Maxillary division of Trigeminal nerve followed by thoracic, cervical, lumbar, and sacral dermatome in either sex, in descending order of frequency. Family history of vitiligo was found in 6 (10.7%) patients. Koebner phenomenon was observed in one patient (1.78%). Associated diseases were present in 2 patients (3.57%).

Conclusion: Segmental vitiligo often begins in childhood and often occurs on face.

Keywords: Clinical profile, Segmental vitiligo, Pakistan.

This is an Open Access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

INTRODUCTION

Vitiligo is an idiopathic, often heritable, acquired depigmenting disorder of skin characterized by, progressive, circumscribed hypomelanosis of the skin and hair, with total absence of melanocytes microscopically¹⁻². It affects 0.5-1% of the population worldwide without racial, regional or gender differences²⁻³. Vitiligo is classified into two main types i.e. generalized and localized. Localized vitiligo is further classified into focal and segmental vitiligo. Localized vitiligo with dermatomal distribution is called segmental vitiligo^{1-2,4}. There is marked clinical difference between segmental vitiligo and non segmental vitiligo⁵.

Clinical characteristics of vitiligo have frequently been described^{3,6-9} but only few studies

have focused on segmental vitiligo¹⁰⁻¹². Moreover important racial differences may exist in different communities. Clinical characteristics of segmental vitiligo have never been described in Pakistani population. The purpose of this study was to describe the clinical profile of segmental vitiligo in a group of Pakistani patients.

PATIENTS AND METHODS

This decriptive study was conducted at Dermatology department of Combined Military Hospital Kohat, during a period of two years from September 2016 to August 2018. All the patients with clinical diagnosis of segmental vitiligo reporting to Dermatology outpatient during this two years period were included in the study after taking informed consent. Diagnosis of segmental vitiligo was based on the clinical findings of acquired, well-demarcated white patches distributed in a segmental/dermatomal pattern with no associated inflammation. Diag-

Correspondence: Dr Aamir Habib, Dermatology Dept, Combined Military Hospital Kharian Pakistan (*Email: aamir1158@gmail.com*) *Received: 14 Jun 2018; revised received: 31 Jan 2019; accepted: 07 Feb* 2019

nosis was further confirmed by Wood's lamp examination. Clinical profile of segmental vitiligo which included the age of the patient, gender, age at onset, duration of the disease at presentation, affected dermatome, family history, presence or absence of Koebner phenomenon and the presence of associated diseases, was studied in these patients. Total number of patients with diagnosis of vitiligo regardless of type of vitiligo was also noted. The study was approved by the Ethical and Scientific Committee of the Hospital. A total of 56 patients with segmental vitiligo were enrolled. Each patient was interviewed in detail and a thorough physical examination was performed with a focus on morphology and distribution of lesions. Data regarding the age, gender, age at onset, duration of disease at presentation, affected dermatome, family history, presence or absence of Koebner phenomenon and the presence of associated diseases was collected for each patient. Data for each patient was collected on a specially designed proforma for each patient separately. Descriptive statistics (mean with standard deviation, percentages and frequency distribution) were used to evaluate the results. Frequencies and percentages were calculated for categorical variables while mean and standard deviations were calculated for numerical variables. Computer program SPSS-14 was used to manage and analyze the data.

RESULTS

A total of 56 patients with segmental vitiligo were enrolled. Out of these fifty six patients, 33 (58.9%) were male and 23 (41.1%) were female. The male to female ratio was 1.43:1. Age at presentation ranged from 2 years to 20 years with a mean of 9.7455 \pm 4.5600 years. Age at onset of disease ranged from 1 to 19 years with a mean of 7.1308 \pm 3.93107 years. Mean age at onset was early in male patients. Mean age at onset in male patients was 6.5757 \pm 4.4749 years while in female patients it was 7.9274 \pm 2.8948 years. Duration of the disease at presentation ranged from 2 months to 18 years with an average of 2.6271 \pm 3.4410 years. The average duration of disease at first presentation was lesser in female patients as compared to male patients. The average duration of disease at first presentation was 1.3717 ± 1.5185 years in female patients and 3.5021 ± 4.1041 years in male patients. The disease started during first five years of life in 28.6% of patients, between 5 to 10 years in 48.2% of patients and between 10 to 15 years in 21.4% of patients. The disease started in first decade in 44 (78.6%) patients. The onset was in second decade in 21.4% of patients (table-I). The disease started during first decade of life in 85% (n=28,) of male patients and in 70% (n=16) of female patients. The disease was more likely to start in first decade in male patients as compared to female patients. The most frequent age of onset of segmental vitiligo was found to be between 5 to 10 years (table-I). Family history of vitiligo was found in 6 (10.7%) patients of which 4 were male and 2 were female. The involvement of right and left sides of the body was almost equal in either sex. Left side involvement (30 patients: male, 18; female, 12) was almost the same as right side involvement (26 patients: male, 15; female, 11). The trigeminal dermatome was most commonly affected in either sex. The disease most commonly involved the area of innervation of Maxillary division of Trigeminal nerve followed by thoracic, cervical, lumbar, and sacral dermatome in either sex, in descending order of frequency (table-II). Face was the most common site of involvement, regardless of sex, followed by lower limbs and trunk (table-II). 15 of the 26 patients (57.7%) with involvement of face were female. This preponderance of female patients amongst those with involvement of face was statistically significant (p < 0.05). Koebner phenomenon was observed in one patient (1.78%). Associated diseases were present in 2 (3.57%) patients. One of these two had atopic eczema while the other had lichen planus which was found co localized to the dermatome affected by vitiligo.

DISCUSSION

Vitiligo is a multifactorial polygenic disorder characterized by patchy depigmentation which is often progressive^{2,3,13}. Different intrinsic, metabolic and functional defects appear to affect melanocytes leading to a substantial loss of functioning epidermal, and sometimes hair follicle melanocytes^{2,12}. The disease is currently classified into two major subtypes: Segmental vitiligo (type B), which usually does not cross the midline; and (2) and non-segmental vitiligo (type A), also simply called "vitiligo" without qualification^{1,3,14-16}. Recently, mixed vitiligo (MV) has been described which has an initial segmental involvement which is associated in a second step reported 6.5%, Khaitan *et al* 18 6.7%, Liu *et al* 7 9.99%, de Barros *et al*¹⁹ 11.3%, Berti *et al*²⁰ 8.0%, Hann *et al*¹² 16.1%, Koga and Tango *et al*¹⁶ 27.9%; and in this study the prevalence was 8.26%. The prevalence of segmental vitiligo is higher in childhood vitiligo^{8,10,17-21,22-24}.

Mean age at presentation of Segmental vitiligo has variously been described. Habib *et al*⁹ reported 16.8 years, Khaitan *et al*¹⁹ reported 16.23

Table-I: Age at onset.					
Age at onset		Male (%age for gender)		Number of patients Female (%age for gender)	Total (% age for total patients)
Onset during first decade		28 (84.85%)		16 (69.56%)	44 (78.57%)
Onset during second decade		5 (15.15%)		7 (30.43%)	12 (21.43%)
Onset during first 5 years of age		13 (39.39%)		3 (13.04%)	16 (28.6%)
Onset between 5 to 10 years of age		15 (45.46%)		13 (56.52%)	28 (50%)
Onset between 10 to 15 years of age		4 (12.12%)		7 (30.43%)	11 (19.6%)
Onset between 15 to 20 years of age		1 (3.03%)		Nil	1 (1.8%)
Table-II: Frequency of b	ody region an	d dermatome inv	olved by	segmental vitiligo.	
				Frequency	Percentage
Body region involved	Face			26	46.4
	Neck		5		8.9
	Upper limbs		6		10.7
	Trunk		8		14.3
	Lower limbs		9		16.1
	Perineum		1		1.8
	Penis		1		1.8
Dermatomal distribution	Ophthalmic N division		8		14.3
	Maxillary division		14		25
	Mandibular division		4		7.1
	Cervical		9		16.1
	Thoracic		10		17.9
	Lumbar		8		14.3
	Sacral		3		5.4

usually after several months with bilateral NSV patches¹⁵.

Segmental vitiligo is a unique subset of vitiligo which exhibits several different features from non-segmental vitiligo^{3-5,10,12-17}. A number of studies have described the clinical profile of Segmental vitiligo^{12,14,16}. The clinical profile of segmental vitiligo in a group of Pakistani children has been described in this study.

The prevalence of segmental vitiligo within a total vitiligo population varies amongst different studies. Handa *et al*⁷ reported 5.0%, Habib *et al*⁹

years, Barona *et al*²⁵ 19.7 years, Liu *et al*⁷ 15.55, Hann *et al*¹² reported 20 years, Park *et al*²⁶ reported 26.06 years and in this study mean age at presentation was found to be 9.7455 ± 4.5600 years.

The mean onset age of Segmental vitiligo in children was reported as 6.63 years by Li *et al*¹⁰. The mean age at onset of Segmental vitiligo was found to be 14.4 years by Habib *et al*⁹, 15.6 years by Hann *et al*¹², 15.55 years by Liu *et al*⁷, 16.3 years by Barona *et al*²⁵ and 18.5 years Park *et al*²⁶. In this study mean age of onset of disease was found to

be 7.1308 ± 3.93107 years which was early as compared to the previous studies9,11-12,17,26. Mean age at onset was earlier in male patients (6.5757 \pm 4.4749 years) as compared to female patients $(7.9274 \pm 2.8948 \text{ years})$ but the difference was not statistically significant. The disease started in first decade in 44 (78.6%) patients and the most frequent age of onset of segmental vitiligo was found to be between 5 to 10 years of age (n=7, 48.2% of patients). Previously Li et al¹⁰ reported most frequent age of onset between 4 and 8 years (42.4%) and Liu et al⁷ reported most frequent age of onset between 5-14 years (48.4%). Hann et al¹² reported disease onset in first decade in 41.3% of their patients. Koga and Tango¹⁶ also reported that segmental vitiligo occurs in young patients.

Average duration of disease has been reported to be 2 years for Segmental Vitiligo by Li *et* al^{10} , 2.79 years by Ezzedine *et al*¹¹ 11, 3.67 years by Park *et al*²⁵, 4.2 years by Khaitan *et al*¹⁸, 4.8 years (male patients, 4.2 years; female, 5.3 years) by Hann *et al*¹². In this study average duration of disease was 2.6271 ± 3.4410 years which was in concordance with previous studies. The average duration of disease at first presentation was lesser in female patients (1.3717 ± 1.5185 years) as compared to male patients (3.5021 ± 4.1041 years).

Family History of vitiligo was found to be present in 26.7% of cases of Segmental vitiligo by Habib *et al*⁹. Barona *et al*²⁵ reported family history of vitiligo in 9.5% of their patients. Hann *et al*¹² found family history of vitiligo in 11.5% of their patients. Khaitan *et al*¹⁸ reported family history of vitiligo in 13.2% of their patients. In this study family history of vitiligo was observed in 10.7% of patients which was in concordance with previous reports.

The trigeminal dermatome was most commonly affected in either sex. The disease most commonly involved the area of innervation of Maxillary division of Trigeminal nerve followed by thoracic, cervical, lumbar, and sacral dermatome in either sex, in descending order of frequency. This was in concordance with previous studies¹⁰⁻¹². Face was the most common site of involvement (n=26, 46.4%) regardless of sex, followed by lower limbs and trunk. The pre-dominant occurrence of disease on head and neck areas has been reported previously^{12,17,24,26} but an explanation for this predilection for head and neck areas has never been given. One possibility could be greater cosmetic awareness of the patients about more visible lesions or for that matter parents in case of children. This is further supported by preponderance of female patients amongst those with involvement of face.

Associated diseases were found in 2 (3.57%) patients in this study. One of these two had atopic eczema while the other had lichen planus. Occurrence of autoimmune disease in patients with segmental vitiligo has been reported variously by previous studies. Previously Li *et al*¹⁰ reported associated diseases in 4.6% of their patients. Hann *et al*¹² reported associated diseases in 6.7% of their patients. Khaitan *et al*¹⁸ reported associated diseases in 1.06%, Park *et al*²⁵ showed that about 9.5% of Segmental vitiligo cases were associated with other diseases while autoimmune diseases, in 11% of their patients.

The frequency of Halo nevi in patients with Segmental vitiligo was reported to be 10% by van Geel *et al*²⁷, 10.2% by Ezzedine *et al*¹¹ and 6.4% by Barona *et al*²⁴ and has been considered to be a marker for progression of segmental vitiligo to non-segmental vitiligo. Halo nevi were not found in any of the patients in this study. Previously Hu *et al*⁸ also did not find Halo nevi in any of their patients with Segmental vitiligo.

Koebner phenomenon was reported in 4.4% of their patients by Barona *et al*²⁵, in 5.3% of patients by Khaitan *et al*¹⁸ and Hann *et al*¹². In this study Koebner phenomenon was observed in one patient (1.8%).

CONCLUSION

Segmental vitiligo often begins in childhood and often occurs on face. The disease is characterized by significantly lower rate of association with other autoimmune diseases.

LIMITATION OF STUDY

Sample size was relatively smaller for such kind of epidemiological studies.

CONFLICT OF INTEREST

This study has no conflict of interest to be declared by any author.

REFERENCES

- 1. Faria AR, Tarle RG, Dellatorre G, Mira MT, Castro CC. Vitiligo-Part 2-Classification, histopathology and treatment. An Bras Dermatol 2014; 89(5): 784-90.
- 2. Ezzedine K, Sheth V, Rodrigues M, Eleftheriadou V, Harris JE, Hamzavi IH, et al. Vitiligo working group. Vitiligo is not a cosmetic disease. J Am Acad Dermatol 2015; 73(5): 883-5.
- Ezzedine K, Eleftheriadou V, Whitton M, van Geel N. Vitiligo. Lancet 2015; 386(9988): 74-84.
- 4. Rodrigues M, Ezzedine K, Hamzavi I, Pandya AG. Vitiligo Working Group. New discoveries in the pathogenesis and classification of vitiligo. J Am Acad Dermatol 2017; 77(1): 1-13.
- 5. van Geel N, Speeckaert R. Segmental Vitiligo. Dermatol Clin 2017; 35(2): 145-50.
- Handa S, Kaur I. Vitiligo: Clinical findings in 1436 patients. J Dermatol 1999; 26(10): 653-7.
- Liu JB, Li M, Yang S, Gui JP, Wang HY, Du WH, et al. Clinical profiles of vitiligo in China: An analysis of 3742 patients. Clin Exp Dermatol 2005; 30(4): 327-31.
- 8. Hu Z, Liu JB, Ma SS, Yang S, Zhang XJ. Profile of childhood vitiligo in China: An analysis of 541 patients. Pediatr Dermatol 2006; 23: 114-16.
- 9. Habib A, Raza N. Clinical pattern of vitiligo. J Coll Physicians Surg Pak 2012; 22(1): 61-2.
- Li Y, Xu A. Segmental vitiligo in children: A clinical epidemiologic study in China. J Eur Acad Dermatol Venereol 2013; 27(8): 1056-7.
- Ezzedine K, Diallo A, Léauté-Labrèze C, Mossalayi D, Gauthier Y, Bouchtnei S, et al. Multivariate analysis of favtors associated with early onset segmental and nonsegmental vitiligo: a prospective observational study of 213 patients. Br J Dermatol 2011;

165(1): 44-9.

- 12. Hann SK, Lee HJ. Segmental vitiligo: clinical findings in 208 patients. J Am Acad Dermatol 1996; 35: 671-4.
- Gianfaldoni S, Tchernev G, Wollina U, Lotti J, Satolli F, Franca K, et al. Vitiligo in Children : A better understanding of the disease. Open Access Maced J Med Sci 2018; 6(1): 181-84.
- Koga M. Vitiligo: A new classification and therapy. Br J Dermatol 1977; 97(3): 255-61.
- 15. Ezzedine K, Gauthier Y, Léauté-Labrèze C, Marquez S, Bouchtnei S, Jouary T, et al. Segmental vitiligo associated with generalized vitiligo (mixed vitiligo): a retrospective case series of 19 patients. J Am Acad Dermatol 2011; 65: 965-971.
- 16. Koga M, Tango T. Clinical features and course of type A and type B vitiligo. Br J Dermatol 1988 Feb; 118(2): 223-8.
- Hann SK, Gauthier Y, Benzekri L. Segmental Vitiligo In: Vitiligo, eds: Picardo M, Taïeb A., Springer Heidelberg Dordrecht London New York, 2010: 41-50.
- Khaitan BK, Kathuria S, Ramam M. A descriptive study to characterize segmental vitiligo. Indian J Dermatol Venereol Leprol 2012; 78(6): 715-21.
- De Barros JC, Machado Filho CD, Abreu LC, de Barros JA, paschoal FM, Nomura MT, et al. A study of clinical profile of vitiligo in different ages: an analysis of 669 outpatients. Int J Dermatol 2014; 53(7): 842-8.
- Berti S, bellandi S, bertelli A, Colucci R, Lotti T, Morreti S. Vitiligo in an Italian outpatient center : a clinical and serological study of 204 patientsin Tuscany. Am J Clin Dermatol 2011; 12(1): 43-9.
- 21. Taieb A, Seneschal J, Mazereew-Hautier J. Special considerations in Children with Vitiligo. Dermatol Clin 2017; 35(2): 229-33.
- 22. Habib A. Vitiligo in Children: A Distinct Subset. J Coll Physicians Surg Pak 2016; 26(3): 173-6.
- Halder RM, Grimes PE, Cowan CA, Enterline JA, Chakrabarti SG, Kenney JA. Childhood vitiligo. J Am Acad Dermatol 1987; 16: 948-54.
- 24. Lin X, Tang LY, Fu WW, Kang KF. Childhood vitiligo in China: clinical profile and immunological findings in 620 cases. Am J Clin Dermatol 2011; 12(4): 277-81.
- 25. Barona MI, Arrunatequi A, Falabella R, Alzate A. An epidemiologic case-control study in population with vitiligo. J Am Acad Dermatol 1995; 33: 621-5.

.....