ORIGINAL ARTICLE

Morphea: A single-center clinical-epidemological study in a tertiary care hospital

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ABSTRACT

Introduction: Morphea is an idiopathic inflammatory disorder characterized by intemperate collagen deposition leading to inspissation of the dermis and subcutaneous tissue. There are few research addressing the clinical-epidemiological features of morphea from India, despite the fact that the condition is common in Caucasians.

Aim: The study was aimed to elucidate the clinical-epidemiological features of Morphea in a tertiary care hospital.

Material and methods: A five-year, cross-sectional study was carried out in the dermatological outpatient department from 2018 to 2022. It included 136 clinically and histopathologically (in doubtful cases) diagnosed patients with morphea. The patients' characteristics, including age, sex, clinical subtype, duration of the disease, associations, and complications, were assessed. The obtained data was analyzed and tabulated.

Results: In our study, the average mean age of the patients was 20.8 ± 14.11 years. Statistical significance was seen when the age-wise distribution of lesions was compared with gender (P-value: 0.009), with more children and a female preponderance. Linear morphea was the most significant and common type observed when types of morphea were compared with age-wise distribution (P-value: 0.051). Extremities were the most commonly involved sites in our study.

Conclusion: Morphea is a disease of the younger age group with a female preponderance. Early age of onset and linear type predominance were the most leading findings in this study. Early diagnosis, treatment and regular monitoring will provide insight in order to prevent complications in patients (especially in children) with potentially disabling linear morphea.

KE YWORDS: Morphea, Caucasians, India

INTRODUCTION

Morphea is an idiopathic inflammatory disorder characterized by intemperate collagen deposition leading to inspissation of the dermis and subcutaneous tissue.[1] It is an uncommon disease relatively with an estimated incidence of 0.4 to 2.7 per 1,00,000 persons and is more often seen in Caucasians.[2]

According to Laxer and Zulian classification, morphea has five variants: circumscribed, linear, generalized, pansclerotic, and mixed.[3] It is associated with significant morbidity in the form of cosmetic disfigurement and/or functional impairment, as atrophy and contractures often remain after the resolution of active disease. There is a dearth of studies on clinical-epidemiological features of morphea from India, so the study was undertaken in this regard to obtain more data.

AIM

The study was aimed to elucidate the clinical-epidemiological features of morphea in a tertiary care hospital.

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MATERIAL AND METHODS

A five-year, cross-sectional study was carried out in the dermatological outpatient department from 2018 to 2022, after obtaining clearance from the institutional ethical committee.

All the morphea cases (136) were diagnosed based on the clinical features, and a biopsy was done in doubtful cases to confirm morphea after taking informed consent. CT scan, eye, and dental examinations were also done wherever needed. The patient characteristics, including age, sex, clinical subtype, duration of the disease, associations, and complications, were assessed.

STATISTICAL ANALYSIS

The Statistical Package for the Social Sciences, version 21 (SPSS Inc.), was used to analyze and tabulate the collected data. After the normality test was done, frequency tables, percentages, and means ± standard deviation were used to present the age and sex distribution results, types of morphea. A chi-square test was used for testing the age-wise distribution of lesions compared with gender and types of morphea compared with the age-wise distribution. Two-tailed tests $P < 0.05$ were considered significant statistically.

RESULTS

Our study included 136 patients, 93 (68.38%) were females and 43 (31.61%) males. The average mean age of the patients was 20.8 ± 14.11 years, with most of them in the pediatric age group, and the duration of the disease varied from two months to 22 yrs. Statistical significance was seen when the age-wise distribution of lesions was compared with gender (P-value: 0.009) with more children and female preponderance, as shown in table 1.

Table 1 Age and gender distribution of patients

Age Group	Male	Female	Total (%)	P- value
Children (≤18)	18	61	79 (58.08)	0.009
Adult (>18)	25	32	57 (41.91)	
Total	43	93	136 (100)	

Linear morphea was found more significantly in female children when compared with adults with a p-value (0.0015) in our study. It was the significant and commonest type (Fig. 1) observed when types of morphea were compared with age-wise distribution (P-value: 0.051), as shown in table 2.

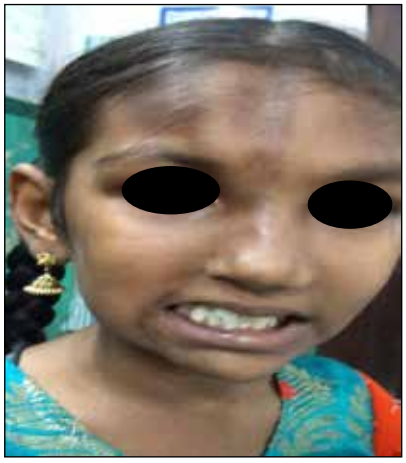

Fig. 1 Linear morphea of head & neck – En Coup De Sabre & facial hemiatrophy

Table 2 Types of morphea distribution

Type of Morphea	Children	Adult	Total (%)	P - Value
Linear	39	17	56 (41.17)	0.051
Circumscribed	19	26	45 (33.08)	
Generalized	15	10	25 (18. 38)	
Pansclerotic	0	0	0 (0)	
Mixed	7	3	10 (7. 35)	
Total	79	57	136(100)	

In our study, among 56 patients with linear morphea, 36 had a head and neck variant; among

them, 23 had En Coup De Sabre, nine had Parry Romberg syndrome, and four had both. Among four, one pediatric patient developed seizures. Her EEG was normal, but the CT brain showed hypodensity in the ipsilateral frontal area & ophthalmic examination showed pseudoretinitis and early papilloedema. Her orthopantomogram showed mandible and maxillary hypoplasia on the right side, crowding of teeth in the maxilla, retained deciduous teeth, and right condylar hypoplasia (Fig. 2). In the other three patients, the CT scan was normal, but one patient had a refractive error. Ipsilateral hemiglossal atrophy was noted in two patients.

Fig. 2 Circumscribed morphea

Circumscribed morphea was observed in 45 (33.08%) patients (Fig. 3), generalized morphea in 25 (18.38%), pansclerotic morphea 0% and mixed morphea in 10 (7.35%) (Fig. 4) patients in our study.

Among mixed morphea cases, linear with generalized type was present in 6 patients, and linear with circumscribed was present in 4 patients. Histopathology of doubtful cases (square biopsy) also showed a characteristic finding of thickening and homogenization of collagen bundles. (Fig. 5) The most common sites involved in this study were extremities (more in lower limbs), followed by the head and neck, as shown in table 3.

There were no triggering factors or associations with any other autoimmune and connective tissue disorders found in our study.

Fig. 3 Mixed morphea

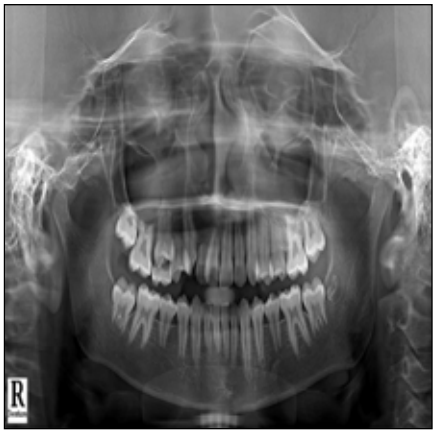

Fig. 4 Orthopantomogram

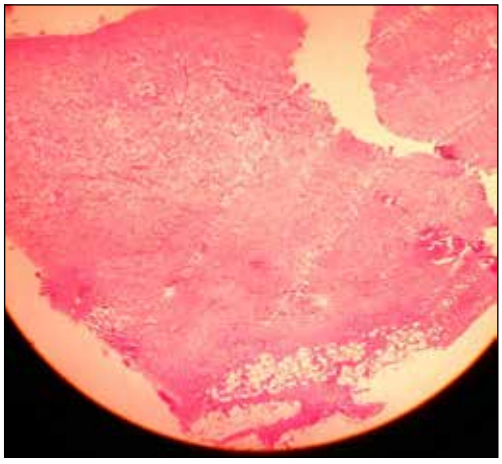

Fig. 5 Histopathology showing thickening and homogenization of collagen bundles.

Table 3 Distribution of lesions of morphea

Site of skin lesions	Linear morphea	Circumscribed morphea	Generalized morphea	Pansclerotic morphea
Head & Neck	36	8	0	0
Trunk	5	13	6	0
Extremities	15	24	10	0
> 2 sites	0	0	9	0
Total	56	45	25	0

DISCUSSION

The present study represents one of the largest data collections from south India as there is a dearth of literature. Morphea lesions generally begin with an erythematous, oedematous and inflammatory phase which may be subtle and 'bruise like' in appearance. The onset is often slow and insidious. This is followed by the development of central sclerosis associated with change in color and texture to thickened, waxy, ivory-white. This central sclerotic area may be surrounded by an erythematous to violaceous so called 'lilac ring,' widely thought to reflect ongoing active disease.[4] There may be loss of hair and absent sweating.

Lesions become atrophic and hyper or hypopigmented over months or years. In some cases, lesions may progress to the atrophic hyperpigmented stage without the sclerotic phase. Depending on depth and type of the lesion, there may be changes in the subcutis, muscle, fascia, bone, or underlying brain, which presents histologically with sclerosis of the dermis and/or subcutaneous tissue.

Morphea can occur at any age; however, the peak age of onset varies for different clinical subtypes, with linear morphea more often seen in children and plaque morphea in adults according to Johnson W et al. study[5] and our study was also in accordance with it. In general, 75% of plaque-type occurs between 40 and 50 years, whereas 75% of linear type occurs between 2 and 14 years.[6] In our study, the mean age of presentation was 20.8 ± 14.11 years, with 58.08% of patients in the children age group.

Most studies suggest that morphea is common in women, with females to males ratio between 7: 1 and 2.6: 1.[7, 8] Our study had females to male sex ratio of 2.1:1 and was in accordance with Tasleem Arif et al.[9] Though morphea can affect individuals of any race, a predisposition for Caucasians is noted, and the disease seems to be less common in blacks and Asians.[10, 11] All patients in our study were Indians.

Linear morphea appears to follow Blaschko lines in most cases, suggesting that genetic mosaicism is important in pathogenesis.[12, 13, 14] It accounted for 20% of morphea cases in adults and 40–60% of cases in children,[9] whereas in our study 69.64% were children. Linear morphea may be subdivided into the limb, trunk, and head variants. In our study, we found the head and neck variant to be present in 36 cases of linear morphea, though other studies say that the limb and trunk variants are more common.[7, 15]

The head variant includes morphea en coup de sabre (ECDS) and progressive hemifacial atrophy (PHA), also known as Parry–Romberg's syndrome. In children, ECDS lesions are commoner than PHA, representing 87% of head variant lesions.[7] En coup de sabre morphea involves the median and paramedian frontoparietal scalp along Blaschko's lines. It may extend to the underlying fascia up to the bones and cause scarring alopecia and might be associated with neurological, auditory, and ocular complications.[16]

In Parry Romberg syndrome, there is hemifacial atrophy with no sclerosis of the skin.

In our study, four patients had both En Coup De Sabre and facial hemiatrophy, one amongst them developed seizures and showed CT, ocular and dental changes. Thus, in linear morphea, neuroimaging is suggested usually when clinical evidence of CNS or orbital involvement is suspected but in few cases, even in the absence of neurological symptoms there might be certain structural changes in CNS or orbit. Hence regular follow-up of such patients is required.

Circumscribed morphea is defined as the presence of single or multiple, oval or round lesions, not diffuse enough to be classified as a diffuse disease. We had 45 (33.08%) patients with circumscribed morphea, which was most commonly observed in adults. However, studies focused on children point out that circumscribed is the second most common after linear type in children, [7, 17] and our study was in accordance with it. (39 linear and 19 circumscribed morphea cases) Laxer and Zulian[3] defined generalized morphea as induration of the skin starting as four or more individual plaques, larger than 3 cm in size, that become confluent and involve at least two out of seven anatomical sites (head–neck, right upper extremity, left upper extremity, anterior trunk, posterior trunk, right lower extremity and left lower extremity). Peterson et al.[2] and Kreuter et al.[18] define it more simply as plaques involving three or more of these seven anatomical sites. Based on these definitions, the literature suggests that generalized morphea accounts for less than 10% of children and 10-50% of adult patients.[7,19] We found generalized morphea in 18.38% of patients, and it was not associated with any extracutaneous manifestations (myalgia, arthralgia, dyspnoea and dysphagia).

Pansclerotic morphea is rare and is characterized by extensive, often circumferential involvement of most body surface areas with sparing of the fingers and toes.[3] No cases of it were observed in our study.

Mixed morphea describes 4% of adults and 15–23% of children with the coexistence of more than one subtype of morphea.[4] The commonest combination is linear limb/trunk and plaque morphea, but any combination can occur. In our study, 7.35% had mixed morphea, but common was linear with generalized type and it was found mainly in children.

The etiology of morphea is unclear. Idiopathic vascular dysfunction, autoimmunity and genetic factors are considered to play role in etiopathogenesis with various triggering factors (radiation, infection, vaccination,[20] drugs, trauma[21] and chemicals) but in our study, no such history of triggering factors was found.

Our study attempts to describe the clinical-epidemiological trends of morphea from tertiary care centre in India. However, it is a single-center study with few number of patients in each subgroup, limiting the subgroups' detailed clinical description. Moreover, in morphea no antibodies are specific to the disease. As their clinical & prognostic significance remains unclear, they were not done in our study, and the response to treatment was also not assessed.

CONCLUSION

Morphea is a disease of younger age group with female preponderance. Early age of onset and preponderance of linear subtype were the most common findings in this study with no much extracutaneous manifestations. Early diagnosis,

treatment and regular monitoring will provide insight in order to prevent complications in patients (especially in children) with potentially disabling linear morphea.

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