

Clinicoepidemiological Study Of Morphea In 57 Patients At Tertiary Care Center

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Abstract: Background: Morphea is thickening and hardening of the skin and subcutaneous tissue due to excessive collagen deposition. It may have range of manifestations varying from a very small plaque involving only the skin to disease causing functional and cosmetic deformities. We hereby report case series of 57 morphea patients with wide variety of clinical entities. Material And Methods: Retrospective study of 57 patients with clinical diagnosis of morphea attending dermatology department of P.D.U govt. medical college and hospital Rajkot between 2014 to 2022. Result: Out of 57 patients, 37(65%) were females and 20(35%) were males (figure 1) with Male to female ratio being 1:1.9. Commonest age group affected was 11 to 40 years 45(78%) patients. Most common type of morphea observed was localized morphea 23(40%) followed by generalized morphea 18(31%), linear morphea 10(18%), deep morphea 4(7%) and atrophoderma pasini and pierini 2(4%). Conclusion: Morphea is rare inflammatory immune mediated disease commonly affecting the females and young individuals. Certain predisposing factors like trauma, burn and radiation can precede morphea. [Majgul N Natl J Integr Res Med, 2023; 14(2): 22-26, Published on Dated: 15/03/2023]

Key Words: Morphea, Scleroderma, Localized Scleroderma

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Introduction: Morphea is an inflammatory, immune-mediated disease of unknown aetiology. It is characterised by excessive collagen deposition, which leads to the hardening of the dermis and subcutaneous tissues. They are largely confined to the skin and subcutaneous tissue, including fat, fascia, muscle, bones and joints¹.

It is distinguished from systemic sclerosis by absence of sclerodactyly and nail fold capillary changes. Morphea is more common in females than males although adult pansclerotic morphea appears to be more common in males². Linear morphea is the most common variant during childhood or adolescence; it begins at this age in 40% to 70% of the patients³. It mainly affects the extremities, fronto-parital region of head (en coup de sabre) and the chest.

Gliosis (proliferation of astrocytes, usually leading to scar) and sclerosis of the leptomeninges have also been reported in en coup de sabre variant of morphea⁴.

Material & Methods: We retrospectively reviewed 57 morphea cases which were diagnosed between 2014 to 2022 at P.D.U govt. medical college and hospital Rajkot. We went through each patient's medical record and

collected data about gender, age, distribution and type of morphea. Morphea is classified based on Laxer and Zulian's classification into generalized, localized, linear, deep, and mixed type. Diagnosis of these morphea cases were based on clinical presentation and histopathological examination. Diagnosis was confirmed by histopathological examination in all the cases.

Results: Out of 57 patients studied 37(65%) were females and 20(35%) were males (figure 1) making Male to female ratio 1:1.9. Commonest age group affected was 11 to 40 years 45(78%) patients with mean age of patients being 29 years (table 1.0) and 5% were children below 13 years of age. History of preceding trauma, burns and radiation was present in 4(7%), 2(4%) and 1(2%) patients respectively.

Most common type of morphea observed was localized morphea 23(40%) followed by generalized morphea 18(31%), linear morphea 10(18%), deep morphea 4(7%) and atrophoderma pasini pierini 2(4%). 30(52%) patients presented to us within 1st year of appearance of skin lesions while 13(22%) patients presented between 1 to 2 years and 14(26%) patients visited after 2 years of appearance of lesions.

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Table 1: Age Distribution

Age Distribution	No. Of Patients(N=57)	% Of Patients
11 To 20 Years	18	31%
21 To 30 Years	17	30%
31 To 40 Years	10	18%
41 To 50 Years	02	03%
51 To 60 Years	06	11%
61 To 70 Years	03	06%
> 70 Years	01	01%

Figure 1: Sex Distribution Of Patients With Morphea

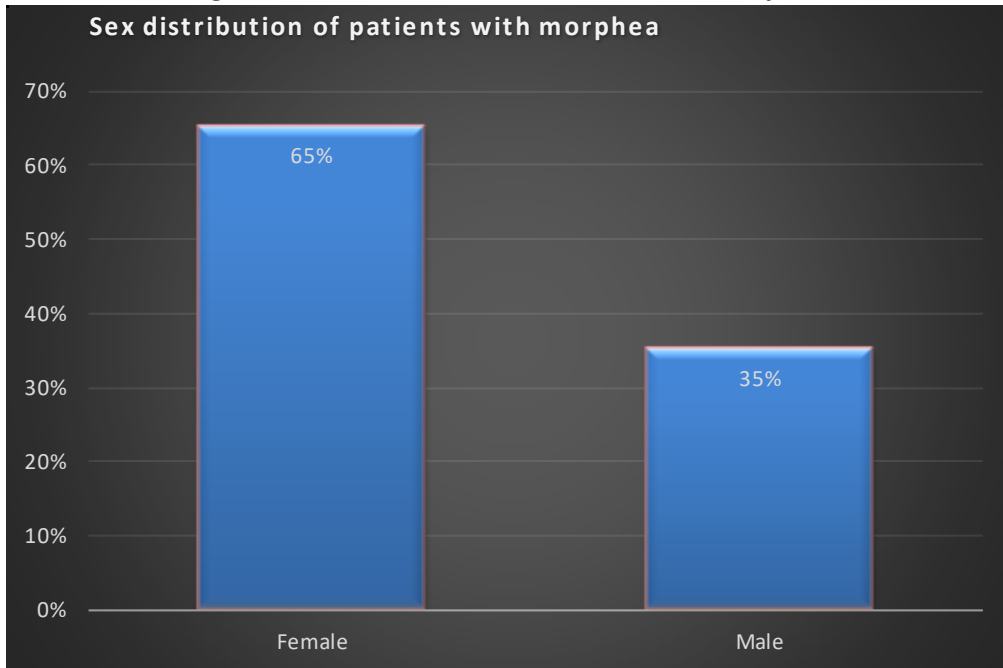
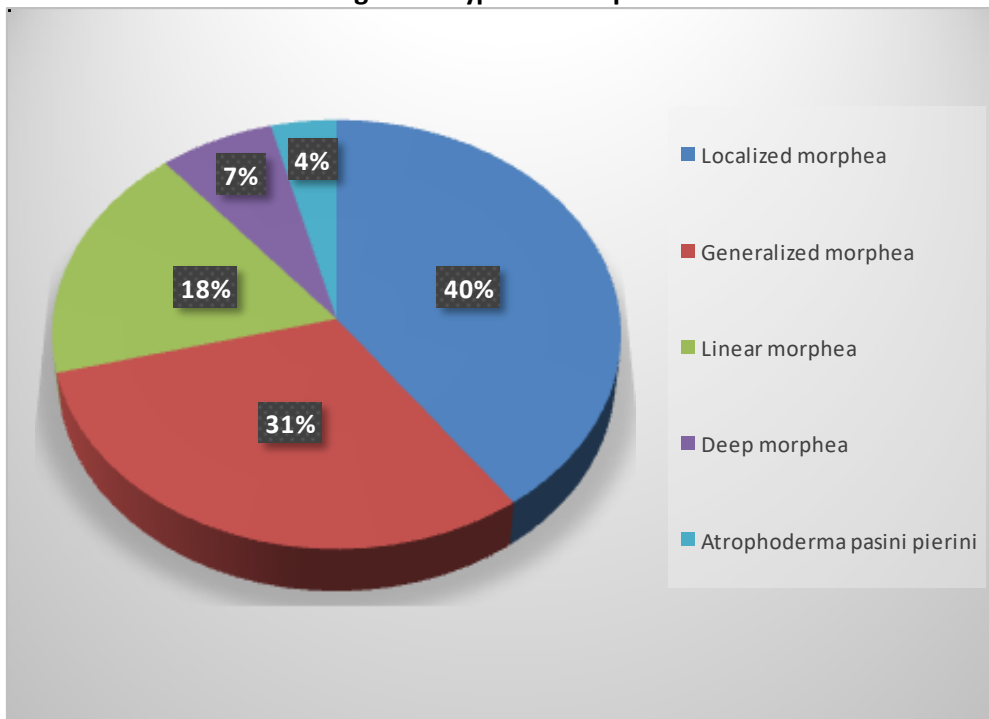


Figure 2: Types Of Morphea



Discussion: Morphea, encompasses a group of related conditions characterized by varying degrees of sclerosis, fibrosis and atrophy in the skin and subcutaneous tissues, sometimes extending deep in to muscle, bone and brain.

There is no increased mortality, but substantial morbidity may occur as a result of joint contractures, facial and limb asymmetry, extra cutaneous manifestation and psychological impact of the condition⁵.

Figure 2: Types Of Morphea: The etiology of morphea is unclear, but genetic predisposition, autoimmune dysregulation, and environmental factors play a role in the pathogenesis of morphea.

The strongest genetic associations with morphea were found with the HLA class II allele DRB1*04:04 and class I allele HLA-B*37.

The presence of auto-antibodies like antinuclear antibody (ANA), anti-single-stranded DNA (SS DNA) and anti-histone antibodies are responsible for autoimmune dysregulation⁶.

Incidence rate of morphea vary from 3.4 to 27 per 100,000, and women are more susceptible than men.

Female predominance in present study is 37(65%) in comparison to male 20 (35%) which is comparable to Ahmed Alhumidi et al study⁷ with females 34(59%) and male 24(41%) patients and tasleem arif et al⁸ study with females 37(78%) and male 10(22%) patients.

Commonest age group affected in our study was 11 to 40 years in 50 cases (76%) with mean age of patients being 29years (Table1) which is comparable to tasleem arif et al study⁷ in which the commonest age group affected was between 11 to 35years.

Multiple environmental factors like trauma, radiation, and intramuscular injection have been reported to play an important role as preceding factors for development of morphea.

History of preceding trauma, burns and radiation was present in 4(7%), 2(4%) and 1(2%) patients respectively which is comparable to tasleem arif et al study having history of trauma in 3 patients while they also reported development of morphea following intramuscular injection and herpes zoster in one patient each.

Radiation can cause localized secretion of interleukin 4 and 5, which induces TGF-B mediated fibrogenesis⁶.

Most common type of morphea in this study was localized morphea 23(40%) followed by generalized morphea 18(31%), linear morphea 10(18%), deep morphea 4(7%) and atrophoderma pasini pierini 2(4%) which is comparable to Ahmed Alhumidi et al study and tasleem arif et al study in which localized morphea was commonest type observed while second most common type in tasleem arif et al study was linear morphea.

30(52%) patients presented to us within 1st year of appearance of skin lesions in comparison to 22(46%) in tasleem arif et al study.

13(22%) and 14(26%) patients visited between 1 to 2 years and 2 years of appearance of lesions respectively in our study.

Diseases mimicking the presentation of morphea such as extra genital lichen sclerosis et atrophicus, inflammatory granuloma annulare and scleromyxedema needs to be differentiated by thorough history, clinical and histopathological examination.

Table 2: Histopathological Findings In Our Study

Sr. No	Histopathological Findings	No Of Patients (N=57)	% Of Patients
1	Thickened Collagen Bundles	43	78%
2	Dense Perivascular And Periadnexal Lymphocytic Infiltration	39	71%
3	Loss Of Rete Ridges	22	40%
4	Atrophy Of Adnexal Structure	14	25%
5	Basal Layer Vacuolar Degeneration	09	16%

Histopathological findings are helpful to confirm the diagnosis and to correlate with the clinical state of morphea. Histology of morphea depends on two factors: the stage of the disease and the depth to which it extends. Most common features seen in histology of morphea is dermal sclerosis, collagen homogenization, dermal thickening, and periadnexal fat loss/ decreased skin appendages⁹. At later stages, the inflammatory infiltrate decreases or disappears, and loss of appendages, flattened dermo-epidermal junction and collagen bundles in the reticular dermis are observed, with a packaged

appearance and alignment parallel to the dermo-epidermal junction(Figure 6)¹⁰. Deep morphea primarily affects the subcutaneous cellular tissue, and produces extensive sclerosis and hyalinization, which extends to the underlying fascia. In our study, most common histopathological finding was thickened collagen bundles in 78% followed by dense perivascular and peri-adnexal lymphocytic infiltration in 71% and 65% histology revealed both findings concurrently. There is tendency for lesions of morphea to spontaneously regress over 3-5 years with residual atrophic and pigmentary changes¹¹.

Table 3: Comparison Of Demographic Details With Other Studies

	Present Study (N=57)	Ahmed Alhumidi et al ⁸ (N=58)	Tasleem Arif et al ⁷ (N=47)
Gender Distribution	F 37 (65%) M 20 (35%)	F 34 (59%) M 24 (41%)	F 37 (78%) M 10 (22%)
M:F Ratio	1:1.9	1:1.5	1:3.7
Most Common Age Group	11 To 40 Years	20 To 50 Years	11 To 35 Years
Most Common Type Of Morphea	Localized Morphea 23(40%) ↓ Generalized Morphea 18(31%) ↓ Linear Morphea 10(18%) Deep Morphea 4(07%)	Localized Morphea 48 (83%)	Localized Morphea 31 (65%) ↓ Linear Morphea 9 (19%)

Figure 3: En Coup De Sabre Of Scalp



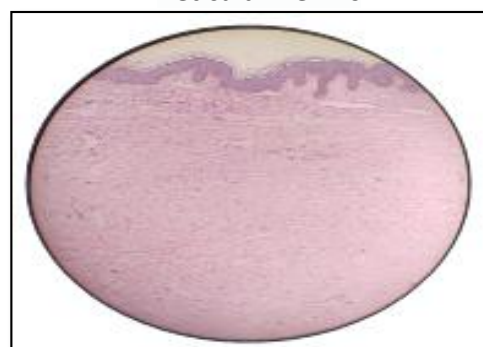
Figure 4: Plaque Morphea Involving Abdomen



Figure 5: Deep Morphea Involving Upper Back



Figure 6: Histopathology Of Morphea Showing Loss Of Epidermis, Flattened Dermo-Epidermal Junction And Dense Collagen Bundles In Reticular Dermis



Conclusion: Morphea is an inflammatory disease of common onset in childhood and predominance in females, where localized sclerosis of the skin and underlying tissue occurs.

Certain predisposing factors like trauma, burn and radiation can precede development of morphea. Early detection and treatment are cardinal to limit its progression and morbidity in morphea patients.

Histopathological examination of Morphea is helpful to diagnose the stage of the disease and helps in initiating the appropriate treatment protocol, but lesion location and the degree of potential disability that the disease can generate in the patient should always be considered in order to choose the treatment, which should be individualized and promptly started.

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