Asian Journal of Research in Dermatological Science

4(4): 23-29, 2021; Article no.AJRDES.76222

The Profile of Patients with Dermatomyositis in Dakar: A Series of 56 Cases

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Giuseppe Murdaca, University of Genova, Italy.
Reviewers:
(1) Suman Lata, Chaudhry Charan Singh University, India.
(2) Shreya Srinivasan, Sree Balaji Medical College and Hospital, India.
(3) Simin Dokht Shoaei, Shahid Beheshti University of Medical Sciences, Iran.
Complete Peer review History: http://www.sdiarticle4.com/review-history/76222

Original Research Article

Received 14 August 2021
Accepted 28 October 2021
Published 03 November 2021

ABSTRACT

Introduction: The purpose of this study was to describe the epidemiological, clinical, paraclinical, therapeutic and evolutionary aspects of patients with dermatomyositis in the dermatology department of the Aristide Le Dantec hospital.
Methods: It was a prospective and descriptive study analyzing all the files of patients with dermatomyositis followed at Le Dantec's dermatology department between January 2009 and December 2019.
Results: We collected 56 cases of dermatomyositis, i.e. a hospital frequency of 1.64%. The average age of the patients was 44 years with 2 cases of juvenile dermatomyositis (9 and 15 years). Pruritus was reported in 13 patients. Cancer was associated with dermatomyositis in 11 cases, including 7 cases of gynecological cancer and 4 cases of ENT cancer. Dermatological manifestations were found in 100% of cases with a predominance of periorbital erythroedema in 66.07%. Muscular manifestations were noted in 96.42%, articular manifestations in 32.14%, pleuropulmonary manifestations in 23.21% and cardiac involvment in 3.57%. All patients received

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treatment with oral corticosteroids, combined in 5.35% with methotrexate. The outcome was successful in 33.9% and death was noted in 11 patients.
Conclusion: Dermatomyositis (DM) tends to affect young adults more, with a high frequency of forms associated with cancer, and deaths.

Keywords: Dermatomyositis; cancer; le Dantec; Senegal.

1. INTRODUCTION

Dermatomyositis is an autoimmune pathology characterized by systemic inflammatory involvment, predominantly cutaneous and muscular, associated with vasculopathy. It can be associated with neoplasia, making the skin a paraneoplastic syndrome. Only a few studies are reported in black Africa [1,2,3]. The purpose of our work was to specify the epidemiological, clinical, paraclinical, therapeutic and evolutionary characteristics of dermatomyositis in the dermatology department of the Aristide Le Dantec hospital.

2. MATERIALS AND METHODS

This was a retrospective and descriptive study analyzing the files of patients hospitalized for dermatomyositis in the Dermatology department of the Aristide Le Dantec Hospital (HALD) in Dakar, between January 2009 and December 2019. The diagnosis was made according to the 1975 Bohan and Peter criteria [4]. Investigations were carried out, such as the electromyogram, the dosage of creatine phosphokinase (CPK) which was considered high when it was greater than 1.5 times the normal. The standard for lactate dehydrogenase (LDH) was less than 40 IU / L and aldolase between 22-59 mIU / L. Visceral involvment was looked for by clinical investigations and imaging (EKG, cardiac ultrasound, chest X-ray and pulmonary CT). Cancer was looked for by physical examination, tumor markers and medical imaging.

3. RESULTS

We collected 56 cases of dermatomyositis out of 3400 hospitalized patients; which represents a hospital frequency of 1.64% and an annual frequency of 5.6 connective tissue diseases per year. The average age of the patients was 44 years with extreme ages of 9 years and 78 years. We noted 2 cases of dermatomyositis occurring in children aged 9 and 15, respectively. The distribution of dermatomyositis cases by age group is shown in Fig. 1. There were 40 women and 16 men patients, i.e. a sex ratio of 0.4. The mean duration of evolution was 6 months with extremes of 1 and 48 months. Functionally, pruritus was reported in 13 patients (23%) including 7 patients with paraneoplastic dermatomyositis.

Cancer was associated with dermatomyositis in 19.64% (n = 11) including 7 cases of gynecological cancers and 4 cases of ENT cancers with a mean age of onset of 51.45 years. Table 1 gives the distribution of the different types of cancers associated with connective tissue diseases.

Dermatological manifestations were found in 100% of cases with a predominance of periorbital erythroedema in 66.07% (n = 37) followed by erythemato-squamous lesions of the photo-exposed areas in 48.21% (Figs. 2 and 3). The different dermatological manifestations of dermatomyositis found in our study are illustrated in Table 2.

Muscle manifestations were noted in 96.42% of cases (n = 54), such as muscular syndrome in 41.66% of cases (n = 50) and isolated myalgia in 3.33% of cases (n = 4). Non-deforming inflammatory polyarthralgia was noted in 32.14% (n = 18). Pleuropulmonary manifestations were found in 23.21% (n = 13) of cases with predominance of crackles in 8.79% (n = 8). The other respiratory signs were represented by exertional dyspnea in 3.30% (n = 3) and dry cough in 2.119% (n = 2.19%). Cardiovascular involvment was present in 3.57% of cases (n = 2), the main symptom being tachycardia noted in 3.57% (n = 2). Dermatomyositis was associated with lupus in 2 cases and systemic sclerosis in 1 case.

Autoantibodies were requested in 30.35% (n = 17) with positivity of anti U1RNP antibodies in 3.35% (n = 3), anti OJ 1 antibodies in 1.78% (n = 1). Speckle type antinuclear antibodies, anti SSA/Ro antibodies and anti SSB/La antibodies were positive in 1.78% (n = 1), respectively. The main muscle enzymes requested were: AST, CPK, LDH and Aldolase. Table 3 shows the

distribution of the various muscle enzymes found during dermatomyositis.

The electromyogram performed in 6 cases was suggestive of the disease in 5 cases.
Radiological abnormalities were interstitial syndrome in 14.28% (n = 8), pulmonary fibrosis in 3.35% (n = 3) and pleural fluid effusion in 1.78% (n = 1). EKG abnormalities were dominated by tachycardia.

All the patients received treatment with oral corticosteroids combined in 5.35% with methotrexate. Among these patients, 10.71% (n = 6) were initially placed on a corticosteroid bolus. Hydroxychloroquine was prescribed in 82.14% (n = 46) of patients. In forms associated with cancer, surgery was performed in 5 cases, chemotherapy and radiotherapy in 4 and 2 cases respectively.

The outcome was good in 33.92% of patients (n = 19) on the basis of a clear improvement in clinical signs and sometimes paraclinical signs including CPK and LDH. However, complications were noted in 30.35% (n = 17) of patients, the main ones being infections (26.78%), corticosteroid-induced diabetes (3.57%), respiratory distress (1,78%). The main infectious sites found were dermatological (23.21%), such as herpetic superinfection in 5 cases (8.92%) and zoster in 2 cases (3.57%), and pleuropulmonary (12.5%) such as lung infection with common germs.

Table 1. Different types of cancers associated with dermatomyositis

Cancers		Number of cases
Gynecological cancers	Mammary adenocarcinoma	4
	Ovarian adenocarcinoma	2
	Squamous cell carcinoma of the uterus	1
ENT cancers	Squamous cell carcinoma of the nasopharynx	2
	Squamous cell carcinoma of the hypopharynx	2
Total		11

Table 2. Distribution according to dermatological manifestations

Dermatological manifestations	Population	Percentage (%)
Periorbital erythroedema	37	66,07
Erythemato-squamous lesions of the photo-exposed areas	27	48,21
Ulcero-necrotic and/or ulcero-crusted lesions	25	48
Poikiloderma	18	32,14
Peri-ungual telangiectasia « manicure sign »	11	21
Gottron's papules	8	14,28
Purpura	6	10,71

Table 3. Distribution of muscular enzymes

Muscular enzymes	Number of cases	(%)
LDH	43	(76,78)
-Very high	8	(14,28)
-Moderatly elevated	23	(41,07)
-Slightly elevated	12	(21,42)
CPK	39	(69,64)
-Very high	16	(28,57)
-Moderatly elevated	17	(30,35)
-Slightly elevated	6	(10,71)
AST	35	(62,5)
-Very high	11	(19,64)
-Moderatly elevated	12	(21,42)
-Slightly elevated	12	(21,42)
Aldolase	1	(1,78)
-Moderatly elevated	1	(1,78)
-Slightly elevated	0	(0)

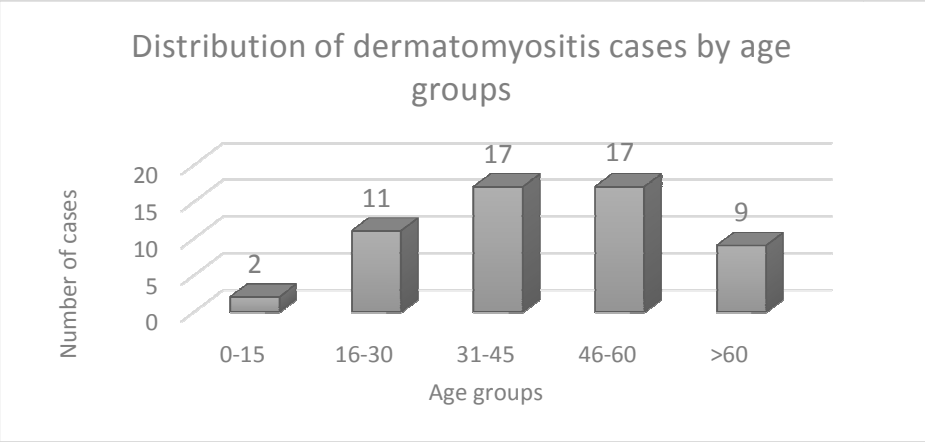

Fig. 1. Distribution of dermatomyositis cases by age groups

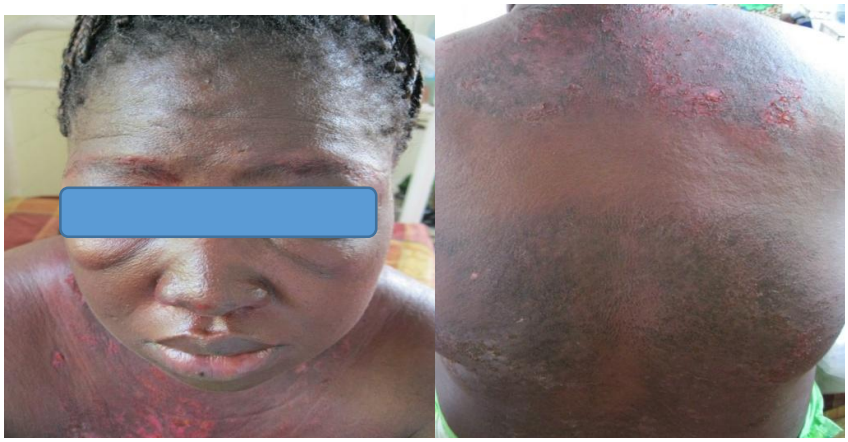

Fig. 2. Periorbital erythroedema during dermatomyositis
Collection : Dermatologie- HALD

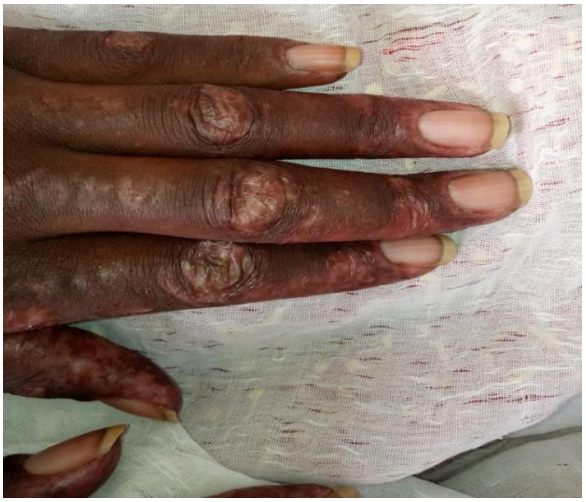

Fig. 3. Gottron's papules during dermatomyositis
Collection : Dermatologie-HALD

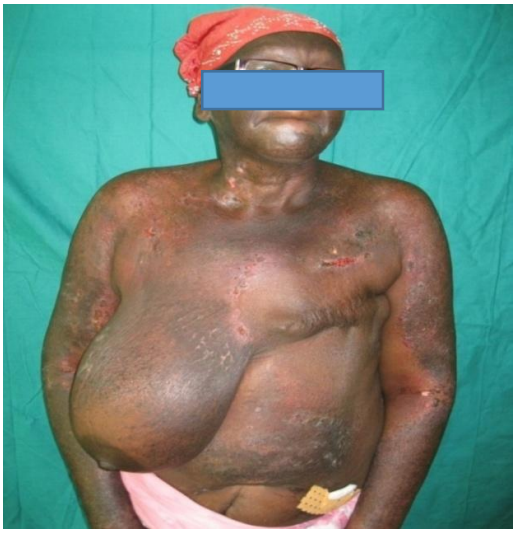

Fig. 4. Dermatomyositis + relapse of a right mammary neoplasia
Collection : Dermatologie-HALD

Death was noted in 11 patients (19.64%) secondary to severe hyponatremia (1 case), respiratory distress of undetermined etiology (1 case) and pulmonary tuberculosis. In the other cases, they were dermatomyositis associated with cancer (n = 3).

4. DISCUSSION

We conducted a retrospective study in the dermatology department of the Aristide Le Dantec Hospital over the period from January 2009 to December 2019. In total, 56 files of patients hospitalized for dermatomyositis were collected during this period, i.e. an annual frequency of 5.6 dermatomyositis / year. This result is similar to the one obtained by Badji [5] during the previous study on dermatomyositis in the same department, which found the same frequency (5.6 cases / year), but lower than Dieng's (8 cases / year) [6]. Our series is mainly made of young adults. Indeed, the average age of the patients was 44 years and the age range [31-60] years was the most representative. This age was lower in the studies of Badji in Senegal (38.28 years) and Iba in Gabon (36 years), as well as the British series by Ramirez et al. (37 years old) [6,7,8]. On the other hand, 2 cases of juvenile dermatomyositis were found. The latter is a rare entity but remains the most common chronic inflammatory myopathy in children [9]. The frequency of pruritus reported in 23% was described in previous studies and strongly predictive of associated neoplasia in subjects of African descent [5].

Neoplasia was associated in 19.64% (n = 11), compatible with the data in the literature which report the prevalence of paraneoplastic dermatomyositis between 6 and 40% [10,11,12,13]. A non-fortuitous link between dermatomyositis and cancer has already been established. Dermatomyositis can precede, accompany or follow cancer [14]. In our case, 3 patients were already known to have cancer at the time of the diagnosis of dermatomyositis while the diagnosis of other cases of cancer was made at the same time as the dermatomyositis' without the concomitant character being established with certainty. The predominance of gynecological cancers in women found in our study is usual. Likewise, ENT cancers came second with 2 cases of nasopharyngeal cancer and 2 cases of hypopharyngeal cancer with a predominance of women, even though these types of cancer are more common in men [15]. Hence the importance of expanding the search for cancers in the ENT region in women.

The dermatological manifestations were constant, dominated by the periorbital erythroedema found. This predominance of periorbital erythroedema is also reported by other series in the literature [16,17,18,19]. Likewise, Gottron's papules, described in 30% of cases in the literature [20], were found in 14.3% of cases in our study, less frequent than in Badji's study (23%) and in Iba's in Gabon [5,7]. Erythemato-squamous lesions of the photo-exposed areas during dermatomyositis have been previously described in the department [21,22] as well as in

black American subjects without a clear explanation being given.They were found in 48.21% of cases in our study.

Poikiloderma was found in previous studies in Senegal and Togo with a high frequency [5,7]. It was present in 32.14% (n = 11) in our study and reflects the chronicity of the disease due to diagnostic delay.

The frequency of joint damages in our study (32%) fits with the results of the literature where they are described between 15 and 30%. They are described as non-deforming inflammatory polyarthralgia affecting large joints.

Lung involvment in dermatomyositis is dominated by diffuse interstitial lung disease (DILD). Its frequency varies according to the series (5 to 80%). It is estimated at 39% by the study of Selva-O 'Callaghan et al. [19] against 7.14% in our series. The presence of anti-synthetase antibodies is also considered to be a predictor of the onset of DILD and can occur at any time during the course of dermatomyositis [19,23,24]. It is a factor of poor prognosis because it is often responsible for respiratory failure causing severe mortality [25,26].

Anti-nuclear autoantibodies are present in 10 to 30% of cases during dermatomyositis according to the literature [27]. They were positive in 1.78% of cases in our speckled fluorescence study.

The outcome of the patients was good in 33.92% of cases (n = 19) and however, death was noted in 19.64% (n = 11); compatible with most studies which report it between 11 and 33% [19,18,28]. The various predictive factors of mortality found by some studies were pulmonary involvment (especially DILD during DM), cardiac involvment, esophageal involvment, old age and the presence of neoplasia [19,29,30,31]. In our series, a death was associated with cancer in 3 cases.

5. CONCLUSION

Dermatomyositis is not a rare connective tissue disease in dermatology and tends to affect young adults more. An association with cancers was observed in almost a quarter of patients and dominated by gynecological neoplasia. A high frequency of deaths was noted.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Toumi S, Ghnaya H, Braham A, et al. Adult polymyositis and dermatomyositis. Tunisian multicenter study. Rev Internal Medicine. 2009;30:747-53.
2. Dieng MT, Diallo M, Dia D, et al. Dermatomyositis in Senegal. Study of 56 cases. Dakar Med. 2005;50:123 7
3. Selva-O'Callaghan A, Labrador-Horrillo M, Munoz-Gall X, et al. Polymyositis / dermatomyositis-associated lung disease: analysis of a series of 81 patients. Lupus. 2005;14:534 42.
4. Bohan A, Peter JB, Bowman RL, et al. A computer-assisted analysis of 153 patients with polymyositis and dermatomyositis. Medicine (Baltimore). LWW; 1977;56:255 86.
5. Badji CH. Dermatomyositis: retrospective study of 74 cases over a 13-year period in the Dermatology department of the Aristide Le Dantec hospital in Dakar (2004-2016). [Faculty of medicine, pharmacy and dentistry]: UCAD; 2017.
6. Dieng MT, Diallo M, Dia D, et al. Dermatomyositis in Senegal. Study of 56 cases. Dakar Med. 2005;50:123 7.
7. Iba BJ, Nzenze JR, Metoule A, et al. Dermatomyositis and polymyositis: 15 cases in Gabon. Med Afr Noire. 2013;60:223 9.
8. Ramirez G, Asherson RA, Khamashta MA, Cervera R, D'Cruz D, Hughes GR. Adult onset polymyositis / dermatomyositis: Description of 25 patients with emphasis on treatment. Sem Arthritis Rheum 1990;20:114–20.
9. Morinishi Y, Oh-Ishi T, Kabuki T, et al. Juvenile dermatomyositis: clinical characteristics and the relatively high risk of interstitial lung disease. Mod Rheumatol. Taylor & Francis; 2007;17:413 7.
10. Holden DJ, Brownell AKW, Fritzler MJ. Clinical and serologic features of patients with polymyositis or dermatomyositis. Can

Med Assoc J. Canadian Medical Association. 1985;132:649.
11. Lynn SJ, Sawyers SM, Moller PW, et al. Adult-onset inflammatory myopathy: North Canterbury experience 1989–2001. Intern Med J. Wiley Online Library; 2005;35:170 3.
12. Wakata N, Kurihara T, Saito E, Kinoshita M. Polymyositis and dermatomyositis associated with malignancy: A 30-year retrospective study. Int J Dermatol. 2002;41:729–34.
13. Sparsa A, Liozon E, Hermann F, Ly K, Lebrun V, Soria P, et al. Routine vs extensive malingnancy search for adult dermatomyositis and polymyositis: A study of 40 patients. Arch Dermatol. 2002;138:885–90.
14. Hill CL, Zhang Y, Sigurgeirsson B, Pukkala E, Mellemkjaer L, Airio A, et al. Frequency of specific cancer types in dermatomyositis and polymyositis: A population-based study. Lancet 2001;357:96–100.
15. Doumbia MY. Epidemiological, clinical, paraclinical aspect of hypopharyngeal cancer in the CCF ENT department of the CHU Gabriel TOURE ; 2019.
16. Toumi S, Ghnaya H, Braham A, et al. Adult polymyositis and dermatomyositis. Tunisian multicenter study. Rev Internal Medicine. 2009;30:747 53.
17. Roebker JJ, Case JD, Lear SC, et al. Immunoperoxidase technique on paraffin sections fails to demonstrate immunoglobulin deposition in cutaneous lesions from patients with dermatomyositis. Am J Dermatopathol. 1988;10:297 9.
18. Holden DJ, Brownell AKW, Fritzler MJ. Clinical and serologic features of patients with polymyositis or dermatomyositis. Can Med Assoc J. Canadian Medical Association. 1985;132:649.
19. Selva-O'Callaghan A, Labrador-Horrillo M, Munoz-Gall X, et al. Polymyositis / dermatomyositis-associated lung disease: analysis of a series of 81 patients. Lupus. 2005;14:534 42.
20. Dalakas MC. Polymyositis, dermatomyositis, and inclusion-body myositis. N Engl J Med. Mass Medical Soc 1991;325:1487 98.
21. Dieng MT, Diallo M, Dia D, et al. Dermatomyositis in Senegal. Study of 56 cases. Dakar Med. 2005;50:123 7.
22. Faye I, Privat Y, Bellossi A, et al. Dermatomyositis. Bull Soc Med Afr Noire Lang Fr. 1969;14:346-8.
23. Clawson K, Oddis CV. Adult respiratory distress syndrome in polymyositis patients with the anti-Jo-1 antibody. Arthritis Rheum. 1995;38:1519–23.
24. Dejardin Botelho A, Perez T, Pouwels S, Wallaert B, Hachulla E, Tillie-Leblond I. Myopathy of inspiratory muscles in anti-synthetase syndrome. Rev Med Internal 2008;29:325–7.
25. Shinohara T, Kidaka T, Matsuki Y, Ishizuka T, Takamizawa M, Kawakami M, et al. Rapidly progressive interstitial lung disease associated with dermatomyositis responding to intravenous cyclophosphamide pulse therapy. Intern Med. 1997;36:519–2.
26. Fathi M, Lundberg IE, Tornling G. Pulmonary complications of polymyositis and dermatomyositis. Semin Respir Crit Care Med. 2007;28:451–8.
27. Goulvestre C. Antinuclear antibodies. Medical Press. 2006;35:287 95.
28. Koh ET, Seow A, Ong B, Ratnagopal P, Tjia H, Chng HH. Adult onset polymyositis / dermatomyositis: Clinical and laboratory features and treatment response in 75 patients. Ann Rheum Dis 1993;52:857–61.
29. Danko K, Ponyi A, Constantin T, Borgulya G, Szegedi G. Long-term survival of patients with idiopathic inflammatory myopathies according to clinical features. A longitudinal study of 162 cases. Medicine. 2004;83:35–42.
30. Airio A, Kautiainen H, Hakala M. Prognosis and mortality of polymyositis.

Peer-review history:
The peer review history for this paper can be accessed here:
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