

Dermatomyositis: A Retrospective Study of Sixteen Cases

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Abstract

Introduction: Dermatomyositis is an inflammatory multisystemic disease characterized by muscle weakness with a characteristic and pathognomonic cutaneous eruption. Our purpose was to describe the common cutaneous, extra-cutaneous features and course of dermatomyositis in our institution.

Method: In this retrospective study we reviewed the medical data of dermatomyositis registered cases in dermatology department at Treichville teaching hospital in Côte d'Ivoire from 2004 to 2013. Diagnosis was assessed for each case using the Bohan and Peter criteria for dermatomyositis.

Result: Sixteen cases which fulfilled at least three out of four of the Bohan and Peter criteria for dermatomyositis were analysed. Prevalence was 15.3 for 10000 consulting patients. The delay to establish the diagnosis after the onset of the disease was 4 months. The average age of patients was 39 years. The sex ratio was 1.2. Features were: hyperpigmentation of sun exposed sites in 100% of cases, symmetric erythema in 93.7% of the cases, facial oedema in 56.2% of the cases, poikilodermatous lesions in 25% of the cases. Telangiectasia was noted in 6.2% of the cases. The average delay for occurrence of muscular symptoms after the onset of cutaneous symptoms was 1 month. The longest delay was 4 months. All cases had at least proximal muscles weakness. Three cases representing 18.7% of cases displayed oesophageal muscles involvement. Muscular biopsy analysis performed in 10 cases that represent 62.5% of cases was specific in 4 cases (40%) and not in 6 cases (60%). Electromyogram was performed in 75% of cases and displayed myogenic deficit in all of them. Serum LDH level has been abnormal in all cases. Whereas serum CPK level has been abnormal in 87.5% of the cases. Two cases representing 12.5% of the cases had interstitial pneumonia and pericarditis. In an average period follow-up of 12 months death occurred in 6.2% of the cases and invalidity was constant. We used oral prednisone in 93.7% of cases. The maximal dose useful to induce remission was 60 mg of prednisone a day. The minimal dose useful for remission control was 10 mg a day. The average delay for partial remission was 10 days. Relapse occurred in all cases when decreasing doses of prednisone. Conclusion: We confirm that dermatomyositis is an uncommon disease. Our cases of dermatomyositis are relatively younger than those classically described. There was no sex predominance.

Keywords: Dermatomyositis; Africa; Skin; Muscle

Introduction

Dermatomyositis is an inflammatory multisystemic disease characterized by muscle weakness with a characteristic and pathognomonic cutaneous eruption. Pathogeny relies on muscle fiber necrosis induced by interstitial inflammation. Diagnosis is made on the basis of precise criteria. Cutaneous manifestations are pathognomonic. The skin lesions may precede the characteristic progressive muscle weakness. In a retrospective study we reviewed the medical data of cases registered as dermatomyositis at Treichville teaching hospital from 1993 to 2003. Our purpose was to describe the common cutaneous, extra-cutaneous features and course of dermatomyositis in our institution. This in order to contribute to the evaluation and improvement of diagnosis tools.

Method

We realized a descriptive and retrospective study of all registered cases of dermatomyositis from 2004 to 2013 in the dermatology department of Treichville Teaching hospital in Côte d'Ivoire. Diagnosis

was assessed for each case using the Bohan and Peter criteria for dermatomyositis. The data concerning the sex the age, professional activity were noted. We systematically considered the notion of existence of the following data:

Suggestive features on the skin

Pruritus, Edema, Symmetric purplish erythema of knees, Elbows or Face, Gottron's sign, Orbital erythema, Poikilodermatous lesions, Telangiectasia, and Erythema of the sun exposed areas. The signs of muscular disorder:

Weakness, myalgia, atrophy. All associated symptoms were noted. Were analysed lung radiography, the electromyogram, skin histopathology, Creatine-phosphokinases, lactic-dehydrogenases and aldolases serum level had been systematically reviewed.

The criteria of the diagnosis were:

1. First of all Weakness of proximal muscles.
2. Second of all elevated serum muscle enzymes (CPK and LDH).

3. Abnormal electromyogram.

4. Muscle histopathology with inflammation involving muscle fibers.

5. Any pathognomonic skin lesions.

The diagnosis was considered as confident when a minimum of 3 non cutaneous criteria were associated to any pathognomonic skin lesion.

All therapeutic option was listed and their total duration noted. The maximal duration of follow-up was noted as well as the evolution in the various controls (worsening, still state, improvement or any specific sign).

Results

On 20 files registered as dermatomyositis in ten years of activity, 16 having satisfied the criteria of the diagnosis were analysed. The average delay for establishment of the diagnosis diagnostic after the beginning of the first clinical sign was 4 months. During the period of study 104131 patients were received in consultation. Prevalence was thus 15.3 per 10000 patients. The center of study being the only center of national reference and the national population being 15000000 inhabitants, global prevalence was estimated at 1 for 1000000 inhabitants.

Age

The average age at onset of the disease was 39 years. Two main groups existed. The first group with age varying from 25 years to 35 years representing 25% of patients. The second group with age varying from 45 years to 55 years representing 31.5% of patients. (Figure 1). The sex-ratio was 1:2.

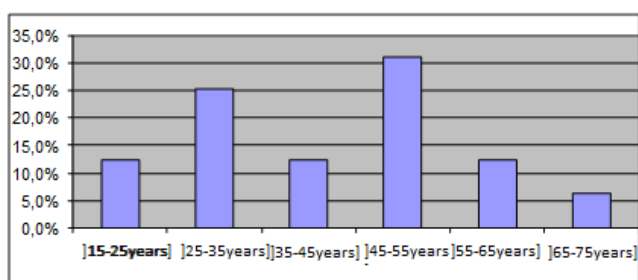


Figure 1: Showing age cluster.

The cutaneous manifestations in decreasing frequency order were: Hyperpigmentation of sun exposed areas in all cases; symmetric erythema of sun exposed existing in 93.7% of the cases, edema of the face or the extremities present in 56.7% of cases. Poikilodermatous lesions were observed in 25% of cases. The less common cutaneous manifestation was telangiectasia, observed in one case representing 6.2% of the cases (Table 2). The precession of cutaneous manifestations on muscular manifestation was observed in 100% of patients (Figure 2-3).



Figure 2: Showing hyperpigmentation of sun exposed area.



Figure 3: Showing oedema of the face.

Estimated by disease history and the follow-up, the average delay for occurrence of muscular symptoms after the onset of cutaneous symptoms was 1 month. The longest delay was 4 months. All cases had at least proximal muscles weakness. In all cases the muscular manifestations were observed in pelvic or scapular muscles. Muscle weakness was noted in all cases. Muscular fasciculation was noted in 6 cases representing 37.5% of cases. Because of this weakness, none of the cases was able to resume a normal activity during the period of follow-up. Three cases representing 18.7% of cases had dysphagia sign of

esophageal muscles involvement. Symptoms were difficulty to swallow solid food for two of them and inability to swallow any solid or liquid food for one of them. This case required the pose of a stomach tube for feeding. This case was associated with the highest serum level of muscular enzymes: CPK 1760 UI/L; LDH 680 UI/L. Death occurred in a short delay. Statistics showed that the delay for muscles manifestation occurrence after skin manifestation was not associated with the existence of systemic disorders. Chi² was equal to 0.04. P=0.84. The Fisher exact test was equal to 0.70 (Table 1). Three cases representing 18.7% of cases displayed oesophageal muscles involvement. Two cases representing 12.5% of the cases had interstitial pneumonia and pericarditis. Fourteen cases (87.5% of the cases) did not develop visceral manifestations.

Occurrence delay after cutaneous manifestations onset	Existence of systemic symptoms	No systemic symptoms
<1 mois	1	8
>1 mois	1	6
Total	2	14
Chi ² = 0.04. P=0.84	Fisher exact test=0.70	

Table 1: Systemic symptoms are not associated to delay of muscles symptoms occurrence after cutaneous manifestations onset.

A case was associated with a vitiligo secondarily developed. Muscular biopsy analysis performed in 10 cases representing 62.5% of cases was specific in 4 cases (40%) and not in 6 cases (60%). Electromyogram was performed in 75% of cases and displayed myogenic deficit in all of them. Serum LDH level has been abnormal in all cases. Whereas serum CPK level has been abnormal in 87.5% of the cases. In an average follow-up period of 12 months death occurred in 6.2% of the cases and invalidity was constant. We used oral prednisone in 93.7% of the cases. The maximal dose required to induce remission was 60 mg of prednisone a day. The minimal dose required for remission control was 10 mg a day. The average delay for partial remission was 10 days. Relapse occurred in all cases when decreasing doses of prednisone.

Cutaneous manifestations	number	percentage
Hyperpigmentation on sun exposed areas	16	100
Symétrique erythema	16	100
Edema	9	56.2
Pruritus	7	43.7
Gottron's sign	5	31.2
Poikilodermatous lesions	4	25
Vitiligo	1	6.2
Telangiectasia	1	6.2

Table 2: Showing cutaneous signs.

Discussion

We observe that dermatomyositis is an uncommon disorder. Porkodi [1] noted 26 cases in 10 years, Drouet 19 cases in 23 years [2], and Sakata 28 cases in 30 years [3]. Prevalence of the disease varies between 1 and 10 for 1000000 inhabitants [4]. This is coherent with our data. Considering the age, it is necessary to distinguish two clinical entities of dermatomyositis: the dermatomyositis of the adult surveying between 45 and 65 years, associated in 20% to 30% of cases with a cancer, and dermatomyositis in youth surveying in child. This form is characterized by a frequent association of widespread calcinosis we observed only cases dermatomyositis of the adult.

Our population presents a bimodal distribution considering the age of onset. But this do not corresponds to any to no clinical peculiarity. On the other hand this population is younger than those classically described. This could explain the absence of associated cancers. Indeed it is demonstrated that the risk of cancer survey in the dermatomyositis is associated with high age [5,6].

There are two clinical criteria of the diagnosis: cutaneous pathognomonic manifestations and the muscular weakness. Cutaneous lesions can constitute the only manifestation of the disease during a variable duration. This, reaching in our cases the maximum of four months before occurrence of muscles manifestations. Longer delay up to 9 years has been reported [7]. The one month delay of occurrence of muscles disorders after initial cutaneous signs observed in our study does not constitute an indicator of the disease severity. Indeed there is no correlation between an appearance of the muscular signs in the first month of the disease and the arisen of systematic manifestations. This distribution on sun exposed area is particularly suggestive for diagnosis. Skin disorders correspond to release of autoantigens in the dermis and apoptosis of keratinocytes with the influence of sun rays [8]. On black skin the generally little perceptible erythema is quickly replaced by a characteristic residual hyperpigmentation. This manifestation should lead to evoke the diagnosis of dermatomyositis. On the other hand we observe that telangiectasia usually observed on white skin is exceptional on black skin.

The muscular manifestations are constantly and initially indicated as a muscular weakness induced by activity: This weakness in our experience is definitively invalidating. The muscular fasciculation although uncommon are very suggestive of muscular trouble. The dysphagia caused by paralysis of the esophageal muscles was less common than the 33% esophageal involvement reported by Porkodi [1]. Amyopathic dermatomyositis characterized by the absence of muscular weakness is difficult to be recognized using the criteria of Bohan and Peter. It requires the satisfaction of all other criteria. We observed any in our cases.

In the initial stage and on certain sites of the muscle histopathology may not be contributive to diagnosis. Repeating biopsies can be useful for diagnosis.

Every relapse of the dermatomyositis was characterized by the recurrence of cutaneous signs and a greater muscular weakness they result in muscle degeneration increasing weakness. Long time course and many relapse can lead to invalidity. The systematic manifestations were uncommon in our experience. Life threatening manifestation was uncommon, but invalidity was constant. Life-threatening manifestation is usually interstitial pneumonia [3] present at 18.7% of our cases. There is a tendency to individualize as clinical entity the dermatomyositis with interstitial pneumonia. It ends in an interstitial fibrosis. Evolution can be quickly progressive and lethal [9]. Antibody

Anti Jo-1 establishes constitutes a biological marker of the dermatomyositis with lung involvement [10]. Their management requires aggressive treatments by immunosuppresses or very high dose of corticoids [9-11]. The interstitial pneumonia is not including in Bohan and Peter criteria for diagnosis. However the vital risk which it represents should make it consider as a factor of gravity. In increasing order of gravity we can consider the little invalidating essentially cutaneous dermatomyositis, the invalidating dermatomyositis interesting the skin and the muscles finally, and life threatening dermatomyositis involving the lung. The whole spectra of gravity are found in our cases. It is likely that if untreated any dermatomyositis identified according to the criteria of Bohan and Peter will evolve gradually towards these successive stages of gravity.

Per os corticosteroid therapy usually allows a remission. This is confirmed in our cases. Relapses can always occur when reducing corticoids doses. A refractory evolution in treatments can exceptionally be observed. This needs the switch to immunosuppressors. The non cortico- responsive dermatomyositis we observed died in a short delay. Somehow mortality in the first year of the disease is lower in our cases. Usually corticosteroids are efficient in dermatomyositis management. The mortality generally varies with the duration of the disease. A longer follow-up of the cases allowed Maugars to show that the mortality linked dermatomyositis is more important in the first five years then declines before nullifying after nine years of evolution [12]. Generally patients survive beyond several years. However they present a muscular deficit which increases with relapse. These must be consequently avoided. What imposes to find the minimal efficient dose and avoid side effects of long term of corticotherapy. The association of corticoids and immunosuppressors particularly methotrexate allows reduction of corticoids doses. By this way the risk of corticotherapy can be minimized. Analyzing our observations in monotherapy by per corticoid is usually efficient in the disease control. But in our experience stopping the treatment was always followed by relapse in a duration that did not exceed one month. Decreasing doses of corticoids should be very progressive and associated with a strict supervision of the serum muscles enzymes level. In fact the severity of the disease due to its own evolution or to its treatment increases in time.

Conclusion

We confirm that dermatomyositis is an uncommon disease. Our cases of dermatomyositis are relatively younger than those classically described. This may explain that there are not associated to any cancer.

Sun exposed sites hyperpigmentation; myasthenia, abnormal LDH serum level, and improvement by oral prednisone were the commonest aspects. Histological proved muscle involvement was the less fulfilled criteria. Mortality was relatively low during the first year of the disease. However the disease was invalidating. Remission control will require continuous corticotherapy.

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