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Epidemiological, Clinical, Paraclinical, and Progressive Characteristics of Idiopathic Inflammatory Myopathies in the Internal Medicine Department of the Treichville University Hospital in Abidjan

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Abstract

Introduction: Idiopathic inflammatory myopathies (IIM) are a group of rare autoimmune diseases that share the common feature of muscle inflammation. The objective was to study the epidemiological, clinical, paraclinical, and evolutionary characteristics of IIM. **Materials and Methods:** This is a Cross-sectional study conducted in the Internal Medicine Department of Treichville University Hospital from 1 January 2012 to 31 December 2022 (11 years); It included patients of all ages with a definite diagnosis of IIM based on the pathological results of muscle biopsy and/or autoantibodies specific to myositis, or a probable diagnosis according to the 2017 EULAR-ACR criteria. **Results:** The hospital prevalence of IIM was 0.33%. Women predominated in 58.3% of cases. The age group [35 to 44 years] was the most affected (42%) and the average age at diagnosis was 40 years (range 9 to 81 years). The main manifestations were cutaneous-mucosal (100%) and muscular (83.3%). Myogenic syndrome (58.3%) and suggestive signs on muscle MRI (41.7%) were found. Muscle biopsy and myositis autoantibodies mainly revealed anti-synthetase syndrome (41.7%), followed by dermatomyositis (33.3%). Treatment consisted of corticosteroid therapy (100%) combined with an immunosuppressant (75%). Mortality (25%) was mainly caused by respiratory disorders. **Conclusion:** Although rare, IIM does exist in Côte d'Ivoire. Mortality remains high and is dominated by respiratory disorders despite corticosteroid therapy often combined with immunosuppressants.

Keywords

Idiopathic Inflammatory Myopathies, Mortality, Internal Medicine, Treichville University Hospital

1. Introduction

Myopathies are a group of diseases characterized by muscle weakness (not secondary to neurological damage). They have various etiologies; the majority are hereditary. Acquired myopathies are toxic, infectious, endocrine, or autoimmune in origin. Idiopathic inflammatory myopathies (IIM) are described in this last group. IIM are very rare conditions (10/100,000 per year) belonging to the large group of connective tissue diseases [1]. With the development of histopathological and immunological techniques, new classifications have emerged that reflect the wide phenotypic and prognostic heterogeneity of IIM. IIM with specific antibodies includes: dermatomyositis (DM), inclusion body myositis (IBM), autoimmune necrotizing myopathies (AINM), anti-synthetase syndromes (ASS), and overlap myositis (OM) [2] [3]. IIM are autoimmune diseases that are rarely diagnosed in sub-Saharan Africa, particularly in Côte d'Ivoire. They remain highly debilitating conditions responsible for significant morbidity and mortality, with diagnostic tools that remain difficult to access in our working environment. The aim of our work was to study the epidemiological, clinical, paraclinical, and progressive characteristics of idiopathic inflammatory myopathies in order to optimize the care of our patients.

2. Materials and Methods

This was a descriptive longitudinal study conducted in the Internal Medicine Department of the Treichville University Hospital over a 11-year period from January 1, 2012, to December 31, 2022; It included patients of all ages with IIM whose diagnosis was confirmed on the basis of the pathological results of muscle biopsy and/or autoantibodies specific to myositis, or probable according to the 2017 EULAR-ACR criteria. For each patient included, the following data (age, sex, medical history, clinical signs, electroneuromyogram (ENMG), magnetic resonance imaging (MRI) of the muscles, muscle biopsy with histological examination, myositis-specific antibodies, antibodies associated with myositis, treatment, and progression) were collected using a standardized survey form that respects confidentiality. The data were entered into an Excel database and then analyzed using SPSS version 26 software. As this was a descriptive study, no statistical tests were performed. Frequency calculations were performed for each of the modalities of the variables studied. The anonymity and confidentiality of the information collected were preserved by assigning an anonymous number to each survey form.

Exclusion criteria: files with missing autoantibody results or patient lost to follow-up during the study period were not included in the study.

3. Results

Twelve (12) cases of IIM were identified among 3,589 patients, representing a hospital prevalence of 0.33%. We found 7 women (58.3%) and 5 men (41.7%), with a male-to-female ratio of 0.71. The 35 - 44 age group was the most represented (42%), and the average age at diagnosis was 40, with extremes of 9 and 81 years old (Table 1). The average time between positive diagnosis and the onset of clinical symptoms was 8 months.

Table 1. Distribution of patients according to age group and gender.

Age group (years)	Number	Percentage (%)
Under 15 years old	01	08.3
[15 - 24 years old]	01	08.3
[35 - 44 years old]	**05**	**41.7**
[45 - 54 years old]	03	25.0
[55 - 64 years old]	01	08.3
Over 64 years old	01	08.3
Gender		
Male (M)	05	41.7
Female (F)	**07**	**58.3**

The most common reasons for admission were lilac-colored rash on the eyelids (41.7%) and muscle pain (25%). Skin and mucosal involvement was present in 100% of cases and was dominated by lilac-colored erythema of the eyelids (75%), the manicure sign (41.7%), and Gottron's papules (16.7%). Raynaud's syndrome was found in 16.7% of cases. Ten (10) patients (83.4%) had muscle involvement with proximal deficit in 58.3% of cases and diffuse myalgia in 25% of cases. Joint involvement of the polyarthralgia type was observed in 9 patients (75%). Diffuse infiltrative pneumonitis was present in 7 patients (58.7%) and four (4) patients, or 33%, had dysphagia. During follow-up, 2 patients (16.7%) developed neurological involvement (coma, seizures) and 3 patients (25%) had cardiovascular involvement (1 case of pericarditis and 2 cases of ventricular extrasystole) (Figure 1).

ENMG revealed myogenic syndrome in 7 patients (58.3%), neurogenic syndrome in 1 case (8.3%) and a normal trace in 4 patients (33.3%). Muscle MRI performed in 5 patients (41.7%) showed suggestive signs and enabled muscle biopsy to be performed in these 5 patients. A specific autoantibody for myositis was found in 11 patients (91.7%) and soluble nuclear autoantibodies in 10 patients (83.3%).

Clinical and paraclinical signs revealed (Figure 2):

- 5 cases of anti-synthetase syndrome (ASS) (41.7%), including 3 cases of Jo1 ASS (25%) and 2 cases of PL12 ASS (16.7%);
- 4 cases of dermatomyositis (DM) (33.3%), including 2 cases of MDA5 DM (16.7%), 1 case of NXP2 DM (8.3%) and 1 case of non-specific Ac DM;

- 2 cases of overlap myositis (OM), including 1 case of PL7 SAS associated with SLE and 1 case of PL12 SAS associated with Gougerot Sjogren syndrome;
- 1 case of autoimmune necrotising myopathy (8.3%).

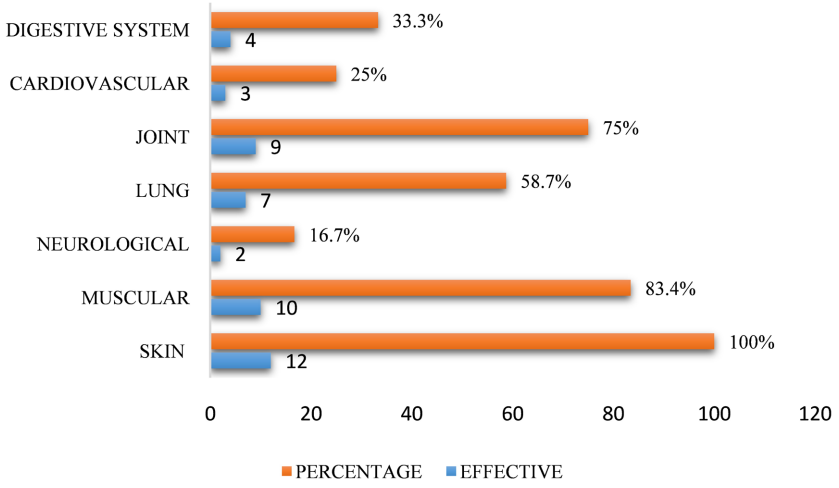

Figure 1. Distribution of patients according to organ involvement.

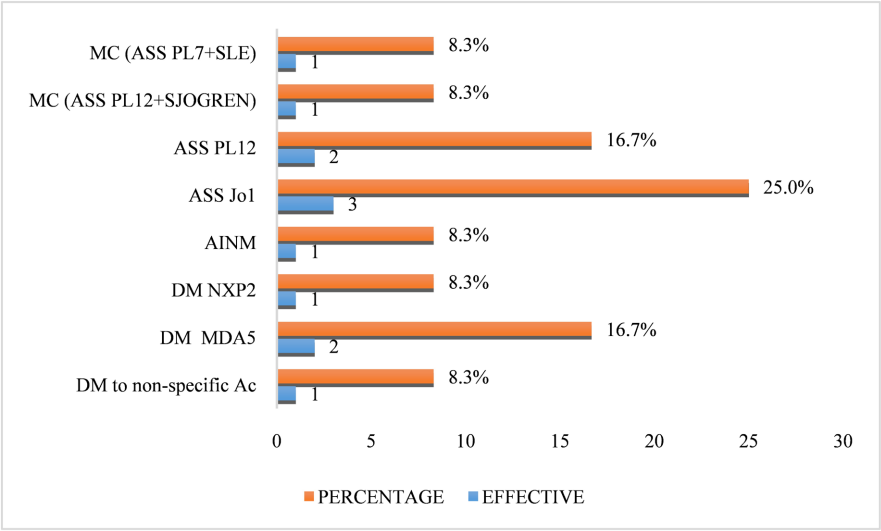

Figure 2. Distribution of patients according to specific antibodies for myositis.

Corticosteroid therapy was used in all of our patients (100%). In addition, an immunosuppressant was added in 75% of cases, including methotrexate MTX (44.4%); azathioprine AZA (22.2%); cellcept CELLC (11.1%); cyclophosphamide (11.1%) and rituximab RTX (11.1%). We used intravenous immunoglobulin in 2 patients (16.7%) (**Figure 3**).

The outcome was favourable in 75% of cases, with 2 cases of complete remission (16.7%); 4 cases of stabilisation (33.3%) and 3 cases of relapse (25%) (**Figure 4**). Mortality was 25%, caused by respiratory disorders and severe sepsis.

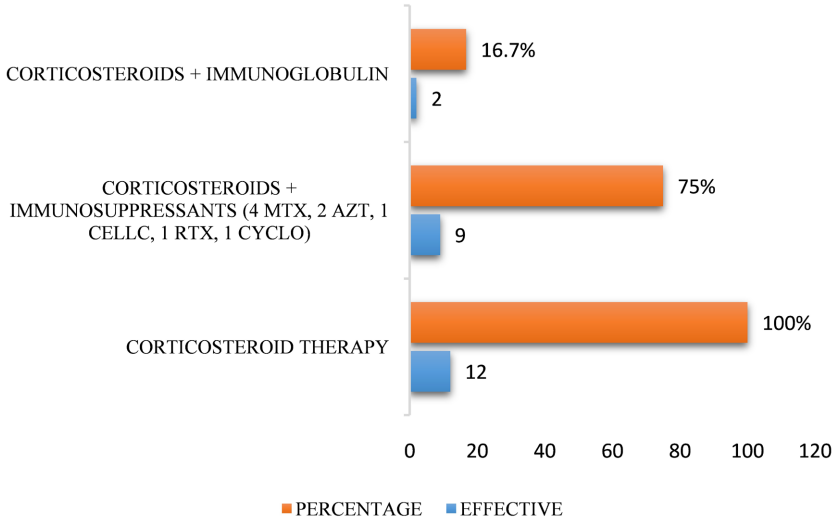

Figure 3. Distribution of patients according to treatment.

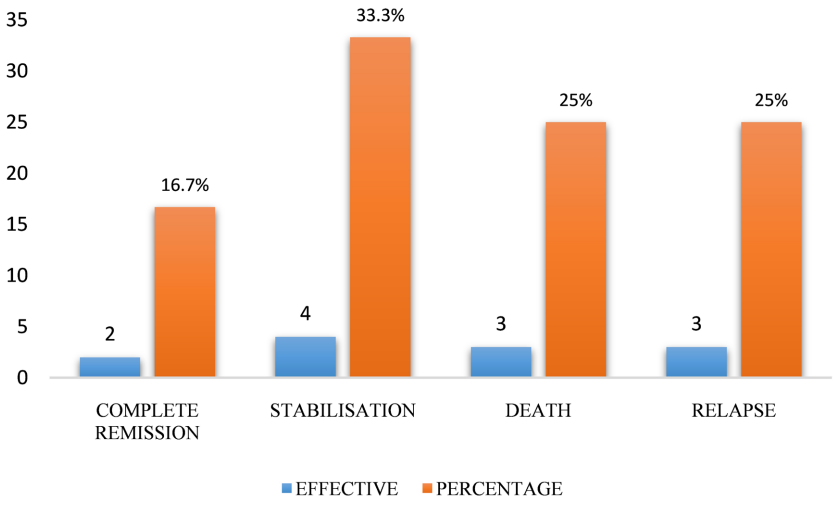

Figure 4. Distribution of patients according to progression after treatment.

4. Discussion

Our study has some shortcomings mainly related to the small sample size, the single-center study, which limits generalization, the difficulty in accessing sophisticated immunological and radiological explorations, and the sometimes incomplete follow-up of patients due to the socio-economic context. Despite these limitations, the analysis of this study has revealed some relevant results.

The prevalence of IIM in our study was 0.33%. These are rare diseases with an annual incidence of 5 to 10 cases per million inhabitants. The prevalence is 6 to 7 cases per 100,000 people. A female predominance was observed, as in all studies found in the literature [4]-[6]. This could be due to the influence of female hormones on the immune system. The average age of our patients at the time of di-

agnosis is comparable to that in sub-Saharan African countries [5] and younger than that of patients in the Maghreb [6]. This age difference can be explained by the large young population in Côte d'Ivoire and sub-Saharan African countries and also by the higher life expectancy in North African countries.

None of our patients had a history of Autoimmune diseases (AID). The main reasons for admission were extra-muscular signs such as lilac-coloured erythema of the eyelids (41.7%) and myalgia (25%), as in the study by Y Allenbach [7]. This is because muscle weakness was probably mistaken for tiredness, given the inherent subjectivity bias of the examiner. The dermatological signs found in 100% of cases were dominated by lilac-coloured erythema of the eyelids (75%) and the manicure sign (41.7%), followed by muscular signs found in 83.3% of cases with proximal deficit (58.3%) and diffuse myalgia (25%); Inflammatory polyarthralgia-type joint involvement was predominant in 75% of cases; diffuse infiltrative pneumopathy was present in 58.3% of cases; digestive involvement was found in 33.3% of our patients; cardiovascular involvement was found in 25% of cases; dry eye syndrome, Raynaud's phenomenon and paraesthesia were found in 16.7% of cases. Our results are consistent with those reported in the literature [6] but differ in terms of their prevalence. The greater severity of the symptoms in our patients can be explained by delayed diagnosis. Creatine phosphokinase (CPK) levels were elevated in the majority of our patients, as reported in the literature. Signs suggestive of inflammatory muscle diseases were found in ENMG (58.3%), MRI (41.7%) and muscle biopsy (16.7%) in small proportions, unlike in North African countries [5] [7]. This low rate of testing can be explained by the unavailability of these investigations in most of our public and private centres, as well as their high cost. Immunological testing carried out on all our patients (100%) revealed myositis-specific autoantibodies (MSA) positive in 91.7% of cases, with a predominance of anti-Jo1 antibodies (25%); anti-PL12 antibodies (25%) and anti-MDA5 antibodies (16.7%). Our results corroborate the data in the literature [8] [9]. The high percentage of positive myositis-specific autoantibodies in our study could be explained by the fact that our patients come to us at an advanced stage of the disease.

In our study, we found a predominance of antisynthetase syndrome (41.7%) followed by dermatomyositis (33.3%). Our results are similar to those from sub-Saharan African countries [10] but differ from those from Maghreb countries [5]. This difference could be explained by the region and race of the study populations. A single case of underlying neoplasia (colon adenocarcinoma) was observed in dermatomyositis. Our result is similar to the data in the literature [5] [11]. Indeed, the presence of anti-SAE1 antibodies, anti-TIF1-γ antibodies and anti-NXP2 antibodies are all associated with an increased risk of cancer in patients with IIM [12]. Systemic lupus erythematosus (08.3%) and Gougerot Sjögren syndrome (08.3%) were the connective tissue diseases associated with myositis in our study.

High-dose corticosteroid therapy (1 mg/kg/day) or bolus therapy was used as first-line treatment in all our patients, as reported in the literature [5] [13] [14]. It is the cornerstone of treatment. This was combined with an immunosuppressant

in 75% of cases, including methotrexate (44.4%) followed by azathioprine (22.2%); cellcept (11.1%), cyclophosphamide (11.1%) and rituximab (11.1%) were used either in refractory forms or to reduce corticosteroid use [5] [14].

The outcome was favourable in 75% of cases, with 2 cases of complete remission (16.7%); 4 cases of stabilisation (33.3%) and 3 cases of relapse (25%) with a mortality rate of 25% caused by respiratory disorders and severe sepsis. Our results differ from those of Toujani [5]. The high mortality rate in our study could be explained by delayed or uncertain diagnosis, which allowed the disease to progress, making treatment more difficult and increasing the risk of complications.

5. Conclusion

Although rare, idiopathic inflammatory myopathies exist in Côte d'Ivoire and should not be overlooked. Despite intensive treatment with corticosteroids combined with immunosuppressants, these often debilitating conditions cause significant morbidity and mortality, clouding the prognosis for patients. This highlights the need for early diagnosis and the establishment of multidisciplinary consultation meetings for systemic diseases in general.

Contributions from Each Author

- ❖ Lead author: Konan N'guessan Michel: Designer, bibliographic review, working method, proofreading.
- ❖ Co-authors
- ✓ Abbe Jean-Fiacre Lidwine: Bibliographic review, data collection, data analysis, writing, translation, proofreading.
- ✓ Ouattara Rokia, Koffi Stéphane, Kouassi Laure, Yapa Stéphane, Lobah Yves: proofreading.
- ✓ U. Acko, Y. Binan: Proofreading and approval.

Conflicts of Interest

The authors report no conflicts of interest regarding funding sources or author affiliations.

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