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# Dermatomyositis and Pregnancy: A Case Report with Review of the Literature

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**Abstract** **Case Report**

Dermatomyositis (DM) is a systemic autoimmune disease, characterised mainly by non-infectious inflammatory involvement of the muscles and skin. The association of dermatomyositis with pregnancy is rarely described in the literature. We present a case of a patient with dermatomyositis who presented with clinical symptoms typical of dermatomyositis, confirmed by investigations. The patient was put on corticosteroids and Azathiopyrin, an ultrasound scan under treatment showed a pregnancy followed by an abortion shortly after the ultrasound scan.

**Key words:** Dermatomyositis; pregnancy; Senegal.



## INTRODUCTION

Dermatomyositis (DM) is an idiopathic systemic disease with both skin and muscle symptoms [1, 2]. The diagnosis is based on criteria that are still relevant today. The occurrence of pregnancy during dermatomyositis is rarely described in the literature. We report the case of a patient followed in the Rheumatology Department for dermatomyositis under background treatment in whom an ultrasound scan for amenorrhoea revealed a pregnancy of 6 weeks and 4 days ultrasound age followed by an abortion shortly after the ultrasound scan.

## OBSERVATION OF THE PATIENT

This is a 19 year old Senegalese woman, married for one year, nulligest, nulliparous, with a last menstrual period (LMP) date of more than one month, with no previous pathological history, the history of the disease goes back to about six months marked by the progressive onset of muscular pains, located in the pelvic and cervical girdles, of proximal topography, bilateral and symmetrical, fixed and non-irradiating, of variable intensity, of inflammatory schedule (2 nocturnal awakenings, morning rollover of more than 30 minutes), without any triggering or calming factor. Associated signs included odynophagia and false routes when the food bolus passed, and general signs of physical asthenia. The clinical examination revealed pain provoked by pressure on the muscular lodges of the pelvic and scapular girdles, an abolished idiomuscular reflex with preservation of the osteo-tendinous reflexes (ROT), positive Stool and Scarf signs. We also noted hairless skin, diffuse poikyloderma lesions (Figure 2, Figure 3) located in the photo-exposed areas (face, décolleté and anterior face of the elbows), bilateral periorbital erythema (Figure 1) and a diffuse manicure sign (Figure 4). Biological findings included : AST 47 IU/l (N<40) ; CPK 427 IU/l (N=2.6) ; LDH 469 IU/l (N=210-425). A myogenic syndrome was observed on EMG. Thus, the diagnosis of DM was made on the basis of the diagnostic criteria (Bohan and Peter criteria : 4/5 ; Hoogendijk (2004) : 3/6 ; Troyanov (2005) : 3/6). The following pre-therapeutic tests were performed : Creatininemia, Azotemia, Glycemia, Uric acid, HBS Ag, anti-HBc antibody, SRV are normal. Our patient was put on Prednisone : 1 mg/kg/d ; Imurel 50 mg : 100 mg/d ; Kaleorid : 1 tablet per day ; Troycal : 1 tablet per day ; Omepral 20 mg : 1 tablet per day at bedtime. An ultrasound was ordered and showed a pregnancy of 6 days + 4 days followed by an abortion shortly after the ultrasound (abortion reported by the patient).

## REVIEW OF THE LITERATURE

From an epidemiological point of view, in America, the sex ratio is 2F/1H, DM occurs at any age, but most often between the ages of 40 and 60, and is three times more frequent in the black population than in the Caucasian population [3]. In France DM affects 2F/1H, it can occur at any age, but appears between 50 and 60 years of age [4]. In Dakar, Senegal, a

retrospective study of the records of patients hospitalised for DM in the adult medical services of the main hospital in Dakar (HPD) from January 2001 to December 2007, based on 21 cases collected [5]:

- All black Africans, 71% had dermatomyositis and 29% polymyositis (PM).
- The sex ratio was 9F/6H in DM, 4F/2H in PM.
- The mean age of the patients was 52 years (26-80 years). There was no juvenile form.
- Frequency of 3 cases on average per year in Senegal with a preferential involvement of women, especially around the age of 40.

Diagnostically, the Reasons for consultation (RM) were dominated by myalgias and proximal motor deficits [6], as illustrated by the table below.

**Table-1: RM of DM in the elderly and DM in the young in a group of patients seen in the internal medicine department of the CHU Hèdi Chaker, Tunisia [6].**

| | **Young adult N = 40** | **Elderly N = 4** |
|---|---|---|
| Gender | 9H/31F | 1H/3F |
| Myalgias n (%) | 87,5 | 50 |
| Proximal muscle deficit n (%) | 87,5 | 75 |
| Laryngopharyngeal involvement n (%) | 52,5 | 50 |
| Digestive signs n (%) | 50 | 50 |
| Joint signs n (%) | 97,5 | 25 |
| Lung signs n (%) | 17,5 | 25 |
| Gender | 5 | 25 |

On a retrospective study of 67 cases of DM conducted in the internal medicine department of the CHU Hèdi Chaker of SFAX (Tunisia) ; over a period from 1979 to 2007 [7]:

- Before the onset of DM, the fertility rate was 4.4 pregnancies per patient
- After the diagnosis of DM this figure is 2
- This decrease in fertility rate in these patients is explained by the late onset of DM, with a higher frequency after the age of 40.

The onset of cutaneous manifestations may precede myositis by several months or years, essentially an orbital erythema in spectacles pathognomonic of DM, Gottron's papules are present in 30% of cases, a manicure sign, other cutaneous manifestations are possible such as calcinosis, vasculitis, and Raynaud's phenomenon [8]. A motor deficit affects the striated musculature bilaterally, symmetrically and non-selectively [8] : proximal muscles (scapular and pelvic girdles and cervical muscles) ; myalgias, observed in 25-70% of myositis cases ; distal muscles, late noted in 25-30% of cases ; involvement of the pharyngeal musculature 25-30% of cases Joint manifestations, 15 to 30%. Cardiac involvement is observed in 10 to 15% of myositis ; the most frequent cardiac abnormalities are rhythm disorders, coronary or intramyocardial vasculitis, inflammatory myocarditis and pericarditis ; pulmonary manifestations occur in 15 to 45% ; they are observed during the course of the anti-synthetase syndrome ; they are rare in DM [8].

The paraclinical literature reports that the sedimentation rate (SR) is moderately elevated in 50-60% of patients [9]. Muscle enzymes are elevated in 75-85% of DM [9]. Isolation of the MM iso-enzymes from muscle or MB from heart, CPK (creatine phosphokinase) does not differentiate between myocardial involvement (regenerating muscle fibres secrete the MB isoenzyme) [9]. Rheumatoid factors (RF) are positive in 20% of DM. Antinuclear and anti-cytoplasmic antibodies (AAN) present in 30-50% : antibodies (Ac) directed against non-specific muscle proteins, and antibodies found in autoimmune conditions (anti-RNP, anti-PM-Scl, anti-SSA and anti-SSB Ac, anti-Ku Ac). NAAs directed against a 220 KD protein of the nuclear complex [9] : DM-specific anti-Mi-1 and anti-Mi-2 antibodies in 5 to 10% of highly corticosensitive classical DMs and anti-synthetase antibodies which are rare in DM. The electromyogram (EMG) can show very suggestive abnormalities in the clinically affected territories [9]. It shows two important negative signs: the absence of a neurogenic aspect and the normality of nerve conduction velocities (NCV). On magnetic resonance imaging (MRI), the distribution of DM lesions, which are characterised by inflammation without fatty infiltration, is similar to that of PM; other abnormalities may be noted, such as the existence of subcutaneous calcifications or fluid collections [9]. Surgical proximal muscle biopsy is used to confirm the diagnosis [9]:

- Myocytes are the site of ischaemic lesions where major histocompatibility complex (MHC) class I expression is predominant.
- The sensitivity and specificity of MHC class I expression in myocytes are 78% and 95% respectively in DM.
- Other tissues than muscle are involved : skin, kidney, lung, heart, digestive system.

An association between DM and tumour pathology is found in 20 to 30% of cases [10, 11]. It is more frequent after the age of 40. DM precedes the onset of cancer in 70% of cases. The average delay

between the two conditions is usually less than 1 year. Breast, uterine and ovarian cancers in women, and bronchial, prostate and digestive epithelial tumours in men predominate [4]. A drug or retroviral trigger seems to be observed in DM. Interstitial lung disease with a poor prognosis can complicate amyopathic DM (AMD), or DM without myositis [12, 13].

In the course of the disease, the onset of DM during pregnancy is a rare event. 17% of DM that started before pregnancy relapses during pregnancy [14]. When DM relapses or starts during pregnancy, the prognosis is poor with fetal death in 50% of cases [14]. Prior to the era of corticosteroid therapy, myositis was a particularly severe condition with spontaneous survival rates of less than 40% [15]. In the absence of underlying tumour pathology, adult myositis now has a relatively favourable prognosis, with 5-year survival rates of around 90% [15]. Functional sequelae are observed in 30-50% of cases [15].

Therapy [16, 17, 18, 19] is based on High-dose corticosteroid therapy (1 mg/kg/d of prednisone) is the first-line treatment, active in more than 70% of DM. Clinical efficacy is slow, with delayed improvements possible up to 3 months. These doses should be maintained until all clinical signs have regressed and muscle enzyme levels have been significantly reduced or normalised. A slow taper may be undertaken, limiting to 10% of the prescribed dose every 10 days, based on motor recovery and muscle enzyme levels. This taper should be continued until the minimum effective dose is maintained. A clinical relapse should always prompt discussion of an additional myopathy, particularly cortisone. Boluses of methylprednisolone, preceding oral corticotherapy, even if they are frequently used in clinical practice, have never been proven to be absolutely useful. In the case of primary or secondary resistance, intolerance or dependence on corticosteroids, various therapeutic alternatives are proposed:

- Immunosuppressive drugs are currently the most commonly used second-line drugs, notably azathioprine and methotrexate. Azathioprine seems to be of interest in the treatment of DM, where several publications in open studies report efficacy in 50% of cases. The effectiveness of methotrexate is noted, in published series, in 50 to 70% of cases.
- Several studies have shown the value of ciclosporin in 50-70% of corticoresistant myositis, even as a first-line treatment.
- The value of intravenous immunoglobulin (IV IG) in corticoresistant myositis has been reported. Their efficacy is estimated at 60-70% of DM. IV Ig is currently proposed as an alternative to immunosuppressive drugs, or in case of failure of the latter. Their tolerance is excellent but their efficacy seems to be lower in the first line.
- Plasmapheresis may be indicated in acute, severe and intractable myositis, in systematic association with an immunosuppressive agent or IV Ig to avoid any rebound on stopping.
- Hydroxychloroquine may be useful in skin lesions of DM, but has no effect on muscle manifestations.
- If swallowing problems occur, oral feeding should be discontinued, enteral or parenteral feeding should be started, and the patient should be monitored in the intensive care unit.
- In the prevention of inhalation pneumonitis, physiotherapy and occupational therapy are essential in the management of DM.

Azathioprine has been reported to decrease the effectiveness of intrauterine contraceptive devices. However, it is recommended to use alternative or additional contraceptive methods. This could explain the occurrence of pregnancy in our patient. Intrauterine growth restriction, abortion and preterm delivery have been reported with azathioprine-prednisolone. This combination seems to be the cause of the abortion in our patient even though the discovery of the pregnancy was incidental and the abortion reported by the patient.

## CONCLUSION

We report a case of dermatomyositis associated with a pregnancy of fortuitous discovery, in order to appreciate the seriousness of this association whose fetal mortality rate depends on the activity of the DM but also on the azathioprine-prednisolone association. The basis of early and adapted therapeutic management is based on corticosteroid therapy but the perspective remains the use of biotherapies (anti CD20).

**Fig-1: Bilateral periorbital erythema**

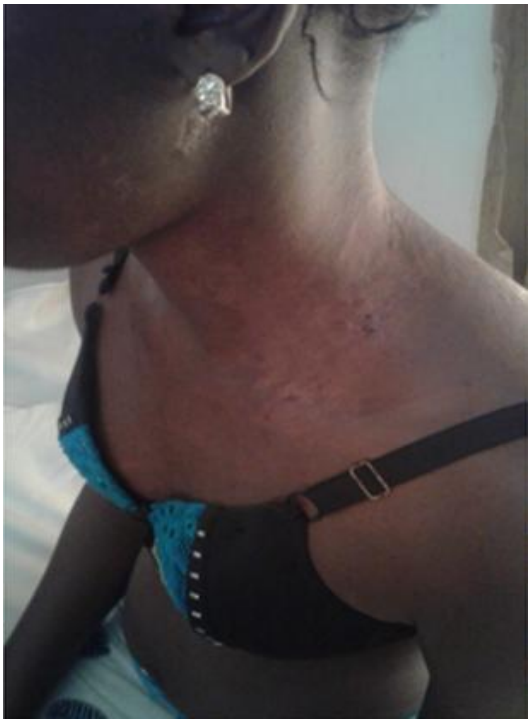

**Fig-2: Diffuse poikyloderma lesions**

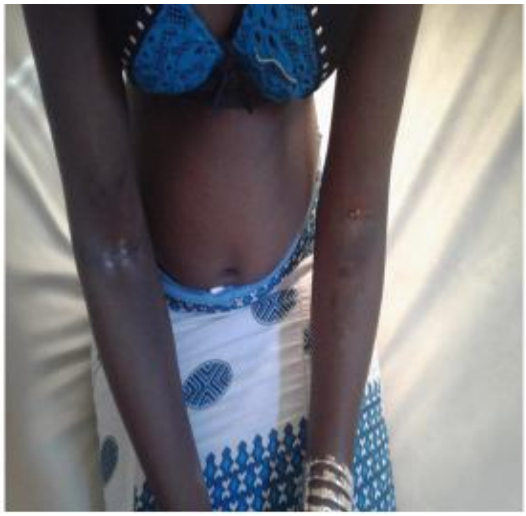

**Fig-3: Diffuse poikyloderma lesions**

**Fig-4: Sign of the diffuse manicure**

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