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Mini Review

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Pregnant Women With Dermatomyositis or Polymyositis

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ABSTRACT

Dermatomyositis (DM) and polymyositis (PM) belong to the group of inflammatory muscle diseases characterized by inflammation of the muscles. These diseases are usually studied together as dermatomyositis/polymyositis (DM/PM). Regarding pregnant patients affected with DM/PM, important individual issues arise as: (1) Do female patients with DM/PM successfully complete pregnancy, giving birth to healthy infants or is there high risk of complications for both mother and fetus? (2) Is there a connection between activity of DM/PM and high risk of complications during gestation? (3) Does pregnancy increase the risk of DM/PM activation? (4) Does pregnancy increase the risk of DM/PM relapse during or right after gestation? After our attempt to answer these questions, we will refer to the treatment of the disease during pregnancy and the effect it could have on the completion of pregnancy.

KEYWORDS: Dermatomyositis; Polymyositis; Pregnancy; CPK; Immunoglobulin; Prednisolone.

ABBREVIATIONS: DM: Dermatomyositis; PM: polymyositis; CPK: Creatinine Phosphokinase.

INTRODUCTION

Dermatomyositis (DM) and polymyositis (PM) belong to the group of inflammatory muscle diseases characterized by inflammation of the muscles and represent immune-mediated syndromes secondary to defective cellular immunity with an incidence in the United States that ranges from 0.5-8.4 cases per million population and it is more common within the black population. Evidence support the idea of a *T*-cell–mediated cytotoxic process directed against unidentified muscle antigens with the factors triggering a *T*-cell–mediated process being still unclear. Due to common symptoms and laboratory tests (increased levels of muscle enzymes), DM and PM are usually studied together. The most common symptom of those diseases is symmetric muscle weakness, pain and tenderness. DM also appears to have skin manifestations (93%) not observed in PM, a characteristic that also makes it easier to diagnose (in adittion to the other diagnostic criteria that involve abnormal elctromyograph, elevated serum levels of CK and muscle biopsy).[1-4]

Regarding pregnant patients affected with DM/PM, important individual issues arise as: (A) Do female patients with DM/PM successfully complete pregnancy, giving birth to healthy infants or is there high risk of complications for both mother and fetus? (B) Is there a connection between activity of DM/PM and high risk of complications during gestation? (C) Does pregnancy increase the risk of DM/PM activation? (D) Does pregnancy increase the risk of DM/PM relapse during or right after gestation? After our attempt to answer these questions, we will refer to the treatment of the disease during pregnancy and the effect it could have on

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the completion of pregnancy.

DERMATOMYOSITIS AND POLYMYOSITIS DURING PREGNANCY

A. Do Female Patients with DM/PM Successfully Complete Pregnancy, Giving Birth to Healthy Infants or is there High Risk of Complications for Both Mother and Fetus?

Given the international literature, a wide number of studies have focused on female patients with DM/PM where the symptoms appeared during adolescence, teenage or adult life:

1) Regarding patients contracting the disease during adolescent or teenage years.

In 1992 Pinheiro Gda et al published a case report of a woman in her 37th week of gestation with adolescent DM who due to fetal distress underwent urgent cesarean section giving birth to a healthy infant. Both the mother and the child were healthy upon reexamination eight months later.

The progress of the first gestation of a 22-year-old woman with adolescent DM was first described by Madu et al[5] while she was being treated with low dosage corticosteroids (5 mg prednisolone every two days). The examination of the fetus in 20th week of gestation was normal. An ultrasound test with Doppler analysis was conducted in 34th, 36th and 37th week showing no abnormalities. The patient presented exacerbation of skin rash on the 39th week and labor induction with intravaginal prostaglandin infusion which led to spontaneous rupture of the membranes, causing childbirth. The infant, weighting 2790 gr, and the mother were both in good condition.

2) Regarding female patients whose disease was first diagnosed with DM in adult life the conclusions are contradictory with reports advocating good pregnancy outcome, or correlation between disease activity and adverse outcome, or even initiation of the disease during pregnancy or after childbirth.[4-10]

In most cases the gestation was successfully completed with only a small number of complications. In 1984, a group of 18 women with DM/PM was studied by Gutierrez et al.[8] Ten pregnancies involving 7 women were monitored. Concerning 4 of these women the start of the gestation coincided with the beginning of the disease while with the other 3, the disease was dormant and an outbreak sometime during pregnancy occurred. Fifty-five percent of these cases ended in fetal death whilst 50% of the remaining gestation was premature terminated.

In contrast, in 1992 Ohno et al[9] presented two women with active DM who became pregnant, giving birth without complications to healthy infants.

In the study of Missumi et al[10] in 2015 regarding the progress of gestation during active DM/PM, from a total of 98 women (60 women with DM and 38 with PM) 78 pregnancies were monitored and studied between June 2011 and June 2012. The study resulted in good pregnancy outcomes (except 1 intrauterine reactivation, 1 diabetes mellitus, 1 hypertension, 1 disease reactivation, 1 hypothyroidism and 2 fetal losses) with patients developing dermatomyositis during pregnancy and 4 during the post-partum period, with good control after corticoid and immunosuppressive therapy. Therefore, the authors concluded that pregnant patients do not seem to be connected to worse prognosis or high risk for either the mother or the fetus, while the disease, either during pregnancy or the postpartum, had good outcome after therapy.

B. Is There a Connection Between Activity of DM/PM and High Risk of Complications During Gestation?

A research of the international literature proves a possible correlation between connective tissue diseases and complications during gestation. In 1998, Skomsvoll et al[11] presented a study were the births recorded in the medical birth registry of Norway in the period 1967-1995 were studied for obstetrical complications and interventions at delivery. The authors reported 72 pre-mature births, 30 pre-eclampsia cases, 42 low birth weight, reporting a possible connection between the outcomes and active disease.

Other studies also reported a more specific association between the activity of the DM/PM and the increased risk of complications during pregnancy.[12-14] However, complications of pregnancy in women whose disease was in remission were also reported[15] as well as lack of correlation between disease activity and progression of pregnancy.[8,16] Even though the results seem more satisfying than worrying, more studies need to be conducted to reach safe conclusions considering the connection of complications during pregnancy to DM/PM.

C. Does Pregnancy Increase the Risk of DM/PM Activation?

In 2014 Pinal-Fernandez et al[17] reported 102 gestations out of 51 women with inflammatory muscle disease (41 with DM and 10 with PM) diagnosed during 1983-2013. From these, 14 pregnancies regarding 8 women took place during activity of the disease. Seven women showed clinical improvement whilst exacerbation of the disease occurred in 5 pregnancies with 2 women. In other women, the disease remained inactive during pregnancy. No myopathy in any pregnancy was observed. Thus, the researchers concluded that pregnant patients with DM/PM are not accompanied by an unfavorable prognosis for the mother or the fetus, and that approximately on 50% of the patients the disease showed improvement during pregnancy.

D. Does Pregnancy Increase the Risk of DM/PM Relapse During or Right After Gestation?

The research of Missumi et al[11] in 2015 resulted in 2 patients developing the disease during pregnancy and 4 women (2 with

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DM and 2 with PM) after childbirth.

Similarly, Park et al[18] reports the appearance of a typical DM's skin rash and intense muscle weakness in the 12th week of pregnancy in a woman 22 years of age while Kofteridis et al[19] presented a patient with acute appearance of DM combined with rabdomyolysis in the 14th week of pregnancy resulting in miscarriage of the fetus. Also, Ohno et al[9] described a woman with the appearance of DM in the third trimester of pregnancy. In the second pregnancy, the possible outbreak of the DM was avoided with oral use of 0.3 mg/kg body weight prednisolone per day. Both pregnancies were successfully completed with the mother and the newborn developing no health problems.

The case of a 30 year old female that developed muscular pain and muscle weakness combined with facial, elbow and knee skin rash, while pregnant with triplets (confirmed by ultrasound) in 8th week of gestation was presented by Tojyo et al.[20] The laboratory tests revealed increased creative phosphokinase levels (CPK serum) and anti-Jo-1 antibodies. A skin biopsy showed edema in the superficial layer of the dermis and aqueous alteration in the stratum basale of the epidermis. Diagnosis of DM was then confirmed and oral treatment with 80 mg of pre-dnisolone daily without observation improved the symptoms. All the embryos died. Their death was linked to DM.

Of interest is the case of a 27-year-old woman described by Pasrija et al.[21] During the third trimester of pregnancy, the patient showed a sudden lilac rash and Gorton papules on the dorsal surface of the upper hands, weakness of the proximal muscles and palpitations combined with high creatinine phosphokinase (CPK) serum levels. Electromyography in the left deltoid muscle was conducted, showing findings compatible with myopathy while biopsy of the right deltoid muscle revealed the presence of lymphocytic inflammatory infiltrates and necrotic muscle fibers. The patient started administration of 8 mg dexamethasone intramuscularly twice daily combined with hydroxychloroquine. The muscle symptoms retreated immediately but not the skin rash. The patient experienced episodes of dyspnea and intense palpitation. In the 33rd week of pregnancy and echocardiogram revealed generalized hypokinesia on the wall of the left ventricle and 45% ejection fraction. The Doppler examination in the 36th week showed high resistance index (resistance index=0,6) in the left uterine artery. In the 37th week a healthy infant weighing 2.500 g by spontaneous vaginal delivery followed.

Relapse of DM shortly after birth or miscarriage has also been reported. In the study of Vancsa et al[12] one out of 9 patients with DM/PM, developed DM during the 3rd trimester of gestation.

In the study of Kaddour et al[14] in 9 pregnancies with an active DM/PM, an outbreak was reported in one woman 10 days after childbirth.

The Kanoh et al[22] also report the case of a female who developed DM after childbirth of a healthy infant. A similar case was described by Lee et al.[23]

The Yassaee et al[24] described in 2009 the case of a 38 year old woman that revealed cutaneous symptoms of DM 4 days after spontaneous abortion (facial erythema, Gorton papules on both hands and elbows and telangiectasias) but without muscle participation or increase of CPK levels. The skin biopsy was compatible with DM.

Treatment during Gestation

In 2015 Ochiai et al[25] reported a case of a patient with rheumatoid arthritis and interstitial pneumopathy that developed amyotrophic dermatomyositis during pregnancy, treated with pre-dnisolone that improved skin symptoms and interstitial pneumopathy as well as giving birth to a healthy infant in 35th week.

In 2007, Williams et al[26] described a case of a young first time pregnant woman where DM appeared in the first weeks of pregnancy. The administration of intravenous immunoglobulin (1 g/kg month body weight for two consecutive days) was accompanied by a decline of symptoms in conjunction with the reduction of CPK levels. The pregnancy was completed normally for both mother and fetus (healthy infant weighing 3.657, 5 g).

Likewise, in 2008 described a case of a 31-year-old woman that developed fever, muscle weakness, skin rash (periorbital and Gorton papules) as far as joint pain and CPK increase. Prednisolone was administered (60 mg/day) orally with regression of clinical events and reducing serum levels CPK. This dosage was progressively reduced to 35 mg/day. The pregnancy evolved satisfactorily but the ultrasound scan showed intrauterine growth restriction.

In the 35th week cesarean section was conducted. The infant had lower body weight (1502 g), but was in good condition. In contrast, the mother lost consciousness during spinal anesthesia and had to be intubated. The extubation was difficult because of the presence of weakness of the respiratory muscles. Intravenous immunoglobulin was administered at 20 g/day for 5 consecutive days in combination with 30 mg/day pre-dnisolone orally. The skin rash and muscle weakness subsided, the serum CPK levels decreased significantly but without full restoration of the respiratory muscles. Rehabilitation program for the respiratory muscles followed, leading to a reduction of the partial pressure of carbon dioxide ($PaCO_2$) and successful extubation attempt. The patient left the hospital in satisfactory condition only treated with low doses of pre-dnizolone.[27]

CONCLUSION

Dermatomyositis (DM) and Polymiositis (PM) are inflammatory myopathies of unknown etiology but with common symptoms. When the symptoms occur during pregnancy, the disease represents a challenge for both the patient and the obstetrician in order to follow therapy with no effect for the mother and

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especially the fetus. Most evidence support that the appropriate treatment with immunosuppressants allows a normal pregnancy without major problems and with no further risk for post-partum relapse. However, follow-up studies need to confirm such claims and to provide clear evidence on acute exacerbation during pregnancy.

AUTHORS CONTRIBUTIONS

All authors participated in writing the paper and approved the final version of the manuscript.

CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

COMPLIANCE WITH ETHICAL STANDARDS

This article does not contain any studies with human participants or animals performed by any of the authors.

REFERENCES

1. Bohan A, Peter JB, Bowman RL, Pearson CM. Computer-assisted analysis of 153 patients with polymyositis and dermatomyositis. *Medicine (Baltimore)*. 1977; 56(4): 255-286. Web site. http://journals.lww.com/md-journal/Citation/1977/07000/A_Computer_Assisted_Analysis_of_153_Patients_With.1.aspx. Accessed March 14, 2017.

2. Tymms KE, Webb J. Dermatopolymyositis and other connective tissue diseases: A review of 105 cases. *J Rheumatol*. 1985; 12(6): 1140-1148. Web site. http://europepmc.org/abstract/med/4093921. Accessed March 14, 2017.

3. Gerami P, Schope JM, McDonald L, Walling HW, Sontheimer RD. A systematic review of adult-onset clinically amyopathic dermatomyositis (dermatomyositis siné myositis): A missing link within the spectrum of the idiopathic inflammatory myopathies. *J Am Acad Dermatol*. 2006; 54(4): 597-613. doi: 10.1016/j.jaad.2005.10.041

4. Oddis CV, Conte CG, Steen VD, Medsger Jr TA. Incidence of polymyositis-dermatomyositis: A 20-year study of hospital diagnosed cases in Allegheny County, PA 1963-1982. *J Rheumatol*. 1990; 17(10): 1329-1334. Web site. http://europepmc.org/abstract/med/2254890. Accessed March 14, 2017.

5. Madu AE, Omih E, Baguley E, Lindow SW. Juvenile dermatomyositis in pregnancy. *Case Rep Obstet Gynecol*. 2013; 2013. doi: 10.1155/2013/890107

6. Ishii N, Ono H, Kawaguchi T, Nakajima H. Dermatomyositis and pregnancy. *Dermatologica*. 1991; 183(2): 146-149. doi: 10.1159/000247657

7. Papapetropoulos T, Kanellakopoulou N, Tsibri E, Paschalis C. Polymyositis and pregnancy: Report of a case with three pregnancies. *J Neurol Neurosurg Psychiatry*. 1998; 64(3): 406-408. doi: 10.1136/jnnp.64.3.406

8. Gutierrez G, Dagnino R, Mintz G. Polymyositis/dermatomyositis and pregnancy. *Arthritis Rheumatism*. 1984; 27(3): 291-294. doi: 10.1002/art.1780270307

9. Ohno T, Imai A, Tamaya T. Successful outcome of pregnancies complicated with dermatomyositis. Case reports. *Gynecol Obstet Invest*. 1992; 33(3): 187-189. doi: 10.1159/000294879

10. Missumi LS, Souza FH, Andrade JQ, Shinjo SK. Pregnancy outcomes in dermatomyositis and polymyositis patients. *Rev Bras Reumatol*. 2015; 55(2): 95-102. doi: 10.1016/j.rbr.2014.10.001

11. Skomsvoll JF, Østensen M, Irgens LM, Baste V. Obstetrical and neonatal outcome in pregnant patients with rheumatic disease. *Scand J Rheumatol*. 1998; 27(Supp107): 109-112. doi: 10.1080/03009742.1998.11720781

12. Vancsa A, Ponyi A, Constantin T, Zenher M, Dankó K. Pregnancy outcome in idiopathic inflammatory myopathy. *Rheumatol Int*. 2007; 27(5): 435-439. doi: 10.1007/s00296-006-0239-8

13. Chopra S, Suri V, Bagga R, Thami MR, Sharma A, Bambery P. Autoimmune inflammatory myopathy in pregnancy. *Medscape J Med*. 2008; 10(1): 17. Web site. http://www.medscape.com/viewarticle/567525_2. Accessed March 14, 2017.

14. Iba-Ba J, Mayi-Tsonga S, Ibouili Bignoumba R, et al. Dermatomyositis and pregnancy: A case in Gabon. *Med Trop (Mars)*. 2009; 69(5): 503-505.

15. Kaddour N, Snoussi M, Marzouk S, et al. Pregnancy in dermatomyositis and polymyositis. *Tunis Med*. 2012; 90(8-9): 606-612. Web site. http://europepmc.org/abstract/med/22987374. Accessed March 14, 2017.

16. Silva CA, Sultan SM, Isenberg DA. Pregnancy outcome in adult-onset idiopathic inflammatory myopathy. *Rheumatology (Oxford)*. 2003; 42(10): 1168-1172. doi: 10.1093/rheumatology/keg318

17. Pinal-Fernandez I, Selva-O'Callaghan A, Fernandez-Codina A, et al. Pregnancy in adult-onset idiopathic inflammatory myopathy: Report from a cohort of myositis patients from a single center. *Semin Arthritis Rheum*. 2014; 44(2): 234-240. doi: 10.1016/j.semarthrit.2014.05.004

18. Park IW, Suh YJ, Han JH, et al. Dermatomyositis developing in the first trimester of pregnancy. *Korean J Intern Med*. 2003; 18(3): 196-198. doi: 10.3904/kjim.2003.18.3.196

ISSN 2377-1542 http://dx.doi.org/10.17140/GOROJ-4-138

19. Kofteridis DP, Malliotakis PI, Sotsiou F, Vardakis NK, Vamvakas LN, Emmanouel DS. Acute onset of dermatomyositis presenting in pregnancy with rhabdomyolysis and fetal loss. *Scand J Rheumatol*. 1999; 28(3): 192-194.

20. Tojyo K, Sekijima Y, Hattori T, et al. A patient who developed dermatomyositis during the 1st trimester of gestation and improved after abortion. *Rinsho Shinkeigaku*. 2001; 41(9): 635-638. Web site. http://europepmc.org/abstract/med/11968753. Accessed March 14, 2017.

21. Pasrija S, Rana R, Sardana K, Trivedi SS. A case of autoimmune myopathy in pregnancy. *Indian J Med Sci*. 2005; 59(3): 109-112. Web site. https://tspace.library.utoronto.ca/handle/1807/22595. Accessed March 14, 2017.

22. Kanoh H, Izumi T, Seishima M, Nojiri M, Ichiki Y, Kitajima Y. A case of dermatomyositis that developed after delivery: The involvement of pregnancy in the induction of dermatomyositis. *Br J Dermatol*. 1999; 141(5): 897-900. doi: 10.1046/j.1365-2133.1999.03165.x

23. Lee SI, Yoo WH. Postpartum-onset dermatomyositis: Case report and literature review. *Mod Rheumatol*. 2002; 12(4): 362-365. doi: 10.1007/s101650200066

24. Yassaee M, Kovarik CL, Werth VP. Pregnancy-associated dermatomyositis**.** *Arch Dermatol*. 2009; 145(8): 952-953.

25. Ochiai M, Sato E, Tanaka E, et al. Successful delivery in a patient with clinically amyopathic dermatomyositis during pregnancy despite first-trimester acute exacerbation of interstitial lung disease. *Mod Rheumatol*. 2015; 27(2): 1-5. doi: 10.3109/14397595.2014.975906

26. Williams L, Chang PY, Park E, Gorson KC, Bayer-Zwirello L. Successful treatment of dermatomyositis during pregnancy with intravenous immunoglobulin monotherapy. *Obstet Gynecol*. 2007; 109: 561-563. doi: 10.1097/01.AOG.0000253244.45837.7c

27. Nozaki Y, Ikoma S, Funauchi M, Kinoshita K. Respiratory muscle weakness with dermatomyositis during pregnancy: Successful treatment with intravenous immunoglobulin therapy. *J Rheumatol*. 2008; 35(11): 2289. doi: 10.3899/jrheum.080389