

CASE REPORT

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Postpartum-onset dermatomyositis: case report and literature review

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Abstract Dermatomyositis (DM) is rare during the reproductive period, but when it does occur most reports have noted that it has an adverse effect on fetal outcome. Conversely, there is little information concerning the contribution of pregnancy to the development and course of DM. We describe here a patient with DM that developed after delivery of an infant and summarize previously documented cases of postpartum-onset DM. This case suggests that pregnancy could be a trigger for the development of DM.

Key words Delivery · Dermatomyositis (DM) · Pregnancy

Introduction

Pregnancy requires special consideration in women with coexisting chronic rheumatologic diseases: notable are the effect of the disease on pregnancy and the effect of pregnancy on the disease course. The relation between dermatomyositis (DM) and pregnancy has rarely been documented, and most of the cases that were reported were from the viewpoint of the management of high-risk pregnancies. In contrast, the contribution of pregnancy to the development and course of DM has not been addressed. Two areas of inquiry regarding pregnancy-related DM have been suggested¹: (1) disease activity aggravated during pregnancy that tends to improve after delivery; and (2) DM with onset or aggravation after delivery.

We describe here a 33-year-old woman with DM that developed after she delivered a healthy infant. We also summarize previously reported cases of DM with onset after delivery.

Case report

A 33-year-old woman was referred to our department with a 5-day history of dysphagia and a 10-day history of fever, fatigue, muscle pain of the upper and lower extremities, and gradually worsening proximal muscle weakness. These symptoms appeared 3 weeks after delivery of her second child, who was healthy. At the current admission her body temperature was 38.1°C, blood pressure 110/70 mmHg, pulse rate 75 beats/min, and respiration rate 17/min. On examination, the patient had an erythematous and violaceous rash over the eyelids. She also had an erythematous, scaling eruption over her metacarpophalangeal and proximal interphalangeal joints, and slight periungual erythema on the dorsum of the fingers (Fig. 1). She had proximal muscular weakness graded as 2/5 in accordance with the Medical Research Council's grading system for muscle strength. Her neurological examination was unremarkable except for the muscular involvement.

Laboratory tests on admission showed a white blood cell count of 3500/mm³, hemoglobin 10.9 g/dl, platelet count 160000/mm³, and elevated levels of muscle enzymes: creatine phosphokinase 893 U/l (normal 35–200 U/l); aldolase 20.1 U/l (normal 0.5–3.1 U/l); and myoglobin 314 ng/ml (normal <50 ng/ml). The erythrocyte sedimentation rate was 47 mm/h (normal 1–20 mm/h), and the C-reactive protein was 4.5 mg/l (normal 0–5 mg/l). The antinuclear antibody and anti-Jo-1 antibody tests were negative, as were those for other autoantibodies, including anti-SS-A antibody, antiribonuclear protein (RNP) antibody, anti-SS-B antibody, anti-ssDNA antibody, anti-dsDNA antibody, and anti-Sm antibody. The viral serology tests for influenza A and B, coxsackievirus, Epstein-Barr virus, herpes simplex virus, parainfluenza, adenovirus, echovirus, cytomegalovirus, measles, varicella-zoster, and human immunodeficiency virus were all negative. The serum levels of endocrine hormones such as follicle-stimulating hormone (FSH), luteinizing hormone (LH), estradiol (E2), and progesterone were within normal levels. Electromyography showed the features of inflammatory myopathy, including bizarre high-

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Fig. 1. Erythematous, scaling rash over the knuckles and dorsum of the hands

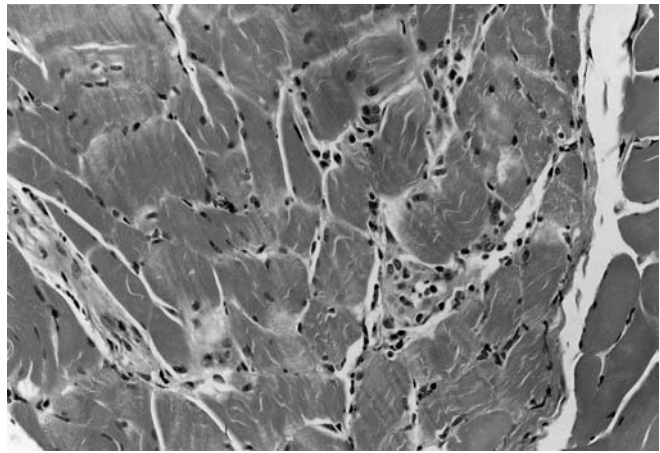


Fig. 2. Skeletal muscle biopsy showed the degeneration of myocytes, perifascicular atrophy, and interstitial and perivascular inflammatory cell infiltration

frequency discharges and repetitive fibrillations with a decrease in the amplitude and duration of the motor action potentials. A muscle biopsy from the thigh showed the degeneration of myocytes, perifascicular atrophy, and interstitial and perivascular inflammatory cell infiltration (Fig. 2).

The patient was given methylprednisolone 100mg/day IV. Her muscle strength gradually recovered, and the muscle enzyme levels began to decrease during the second week of the methylprednisolone treatment. She was then treated with prednisolone 50mg daily and azathioprine 100mg/day, with a decrease in the prednisolone dose to 15mg/day over 8 weeks. There were no laboratory abnormalities or muscle weakness with this treatment. After 3 months follow-up, relief of signs and symptoms of the disease were noted.

Discussion

Dermatomyositis is a systemic, autoimmune disease characterized by a nonsuppurative inflammation of striated muscle and the presence of characteristic skin lesions. Muscle weakness and inflammatory infiltrates in the skeletal muscles are its principal clinical and histologic characteristics.² The incidence of the disease is approximately 5 cases per million population.³ Because there are two peaks in the incidence of DM in women (age groups 10–15 and 45–60 years), only 14% of the cases occur during the reproductive period.⁴ Hence there have been few cases of DM reported that complicated pregnancy, and little is known about the relation between DM and pregnancy.

Pregnancy requires special consideration in women with coexisting chronic rheumatologic diseases such as systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), systemic scleroderma, Sjögren's syndrome, DM, and polymyositis. Fetal and maternal complications of pregnancy were reviewed in women who have underlying rheumatologic diseases.⁵ The relation between DM and pregnancy could

be studied from two viewpoints: the effect of the disease on pregnancy (fetal complications) and the effect of pregnancy on the course of the maternal rheumatologic disease.

Until now, the association of DM and pregnancy has rarely been documented, and most of the cases reported were from the viewpoint of the effect of DM on the pregnancy and the fetal outcome. Most reports believed that DM has an adverse effect on fetal outcome. Among 31 fetal outcomes, 10 (32%) ended with fetal death or abortion, 8 (26%) deliveries were premature, and 13 (42%) of the pregnancies resulted in healthy babies.⁶ These relations seem to parallel maternal disease activity. Stillbirth or neonatal death complicated pregnancies in 7 of 15 patients with active disease in one study, and in 5 of the 15 patients the baby was born prematurely.⁶ Although fetal complications may be overestimated owing to the possibility that only the complicated cases were reported, ideally for women with DM pregnancy should be planned during disease remission, with contraception carefully recommended otherwise. The patients should be followed carefully during pregnancy for clinical and laboratory signs of disease exacerbation.

The contribution of pregnancy to the development and course of DM has not yet been addressed. The inflammatory myopathies rarely complicate pregnancy: only a few cases of DM associated with pregnancy have been reported, and only some of these cases have developed during pregnancy. Bauer et al.⁷ conjectured that the fetus and its complement of foreign antigens might be involved in the development of DM during pregnancy. The production of antibodies to fetal (paternal) HLA antigens has been confirmed. Such immunological changes in the mother may be potentially relevant to the development of autoimmune disease.⁸ More recently, it has been shown that fetal cells can persist after delivery.⁹ The hypotheses suggesting the role for viral infections in the development of DM, and a decrease in the humoral response to certain viral antigen during pregnancy, have been suggested to provide the link between DM and pregnancy.¹⁰ Persistent fetal

Table 1. Cases of dermatomyositis with a postpartum onset

Patient	Age	Onset	Therapy	Disease evolution	No. of delivery	Delivery	Fetal outcome	Reference
1	NK	15 Days after delivery	NK	NK	Second	Abortion	Death	12
2	30	^a	No	Worsened gradually ^b	Second	Full term	Alive	13
3	33	5 Days after delivery	Steroid	Improved after steroid therapy	First	Full term	Alive	1
4	33	3 Weeks after delivery	Steroid	Improved after steroid therapy	Second	Full term	Alive	Present case

NK, not known

^aThe patient gave a history of progressive limb weakness shortly after the birth of her child

^bSix weeks after the next delivery (at 35 years of age), severe exacerbation occurred, and the patient died

microchimerism in patients with systemic sclerosis has been suggested to be one of the causes of the increased susceptibility to the disease in parous women.¹¹ Thus, microchimerism may have caused susceptibility to DM or triggered subclinical DM during the pregnancy.

We report a patient with DM that developed after delivery of a healthy, full-term infant. Until now, postpartum onset of DM has been rare, with only three cases having been reported^{1,12,13} (Table 1). DM developed within 1 month after delivery in all cases. One case was associated with abortion, and the remaining three cases developed after delivery of healthy, full-term babies. As DM with onset during pregnancy tends to improve after delivery, the contribution of pregnancy in cases with postpartum onset seems to be different.

Some consideration must be given to the possible contribution of pregnancy to the development of DM. First, it is sometimes difficult to specify the time of DM onset. The initial symptoms of DM during pregnancy could be nonspecific (e.g., slight fatigue, which is common during pregnancy, and periungual erythema on the backs of the fingers, which could easily be overlooked). Second, DM can overlap other connective tissue diseases, such as SLE, which is one of the common disorders associated with DM.¹⁴ It was well known that the disease activity of SLE can be affected by pregnancy, but the elements related to SLE could not be involved in the postpartum development of DM in our patient. The susceptibility to DM or subclinical DM triggered during pregnancy can be provoked by another factor, such as the puerperium or a viral infection. The factor responsible for immediate postpartum onset in our patient may be hormonal. Analogous to a possible mechanism of postpartum exacerbation of herpes, a dramatic fall in the progesterone level, a hormone with immunosuppressive effects, may allow expression of a suppressed autoimmune disease entity.¹⁵

Because the viral serology results were all negative in our patients, we excluded a viral factor leading to an immediate postpartum onset of dermatomyositis. The serum levels of such hormones as FSH, LH, E2, and progesterone were all within normal levels. However, these women during the puerperium showed large hormonal variations due to lactation and menstruation. Accordingly, a correlation between these hormonal levels and the postpartum onset of DM could not be excluded despite the fact that the levels were quite normal.

The relation between DM and pregnancy should be considered in terms of fetal and maternal complications. The former is commonly considered a high-risk pregnancy. The contribution of pregnancy to the development and course of DM was not addressed. Two subtypes of pregnancy-induced DM may exist: in one type, which is more common, disease activity occurs during pregnancy; the other type appears during the postpartum period. Whether postpartum-onset DM is a subtype of DM is not known.

Conclusions

We described a 33-year-old woman who developed DM after delivering a healthy infant. Because postpartum onset of DM has rarely been reported, we must consider the possibility of accidental onset after delivery, although we believe that pregnancy might have been a trigger in the development of DM or aggravated DM in this patient. Longer follow-up and further evaluation are necessary for a better understanding of postpartum-onset DM.

References

- 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;141:897-900.
- Dalakas MC. Polymyositis, dermatomyositis and inclusion-body myositis. *N Engl J Med* 1991;325:1487-98.
- Harris A, Webley M, Usherwood M, Burge S. Dermatomyositis presenting in pregnancy. *Br J Dermatol* 1995;133:783-5.
- Oddis CV, Medsger TA. Current management of polymyositis and dermatomyositis. *Drugs* 1989;37:382-90.
- Mary JJ. Obstetric complications and rheumatic disease. *Rheum Dis Clin North Am* 1997;23:169-82.
- Ishii N, Ono H, Kawaguchi T, Nakajima H. Dermatomyositis and pregnancy: case report and review of the literature. *Dermatologica* 1991;183:146-9.
- Bauer KA, Siegler M, Lindheimer MA. Polymyositis complicating pregnancy. *Arch Intern Med* 1979;139:449.
- Nelson JL. Pregnancy immunology and autoimmune disease. *J Reprod Med* 1998;43:335-40.
- Bianchi DW, Zickwolf GK, Weil GJ, Sylvester S, DeMaria MA. Male fetal progenitor cells persist in maternal blood for as long as 27 years postpartum. *Proc Natl Acad Sci USA* 1996;23:93:705-8.
- Pinheiro GdR, Goldenberg J, Atra E, Pereira RB, Camano L, Schmidt B. Juvenile dermatomyositis and pregnancy: report and literature review. *J Rheumatol* 1992;19:1798-801.

11. Nelson JL, Furst DE, Maloney S, Gooley T, Evans PC, Smith A, et al. Microchimerism and HLA compatible relationships of pregnancy in scleroderma. *Lancet* 1998;351:559-62.
12. Gutierrez G, Dagnino R, Mintz G. Polymyositis/dermatomyositis and pregnancy. *Arthritis Rheum* 1984;27:291-4.
13. England MJ, Perimann T, Veriava Y. Dermatomyositis in pregnancy: a case report. *J Reprod Med* 1986;31:633-6.
14. Caro I. Dermatomyositis as a systemic disease. *Med Clin North Am* 1989;73:1181-92.
15. Katz SI. Herpes gestationis. In: Fitzpatrick TB, Eisen AZ, Wolff K, et al., editors. *Dermatology in general medicine*. 4th ed. New York: McGraw-Hill; 1993. p. 626-9.