

Postpartum Polymyositis Following Intrauterine Fetal Death

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ABSTRACT

Polymyositis (PM) is an uncommon inflammatory myopathy that affects striated muscles. It causes weakness of the limb girdles, neck, and pharyngeal muscles. We are presenting a case of PM which manifested after intrauterine death (IUD). The patient was referred to our hospital for breathing difficulty, 4 days after delivery of a dead fetus. Initially, she was treated in line of puerperal sepsis and peripartum cardiomyopathy. Patient's cardiopulmonary functions improved but she had persistent high-grade fever. Gross muscle weakness was found on day 5 of admission, involving all four limbs, predominantly in proximal muscles and she had dark colored urine. Laboratory tests revealed myoglobinuria, high serum creatine phosphokinase (CPK) levels, and high lactate dehydrogenase (LDH) levels. Polymyositis diagnosed on the basis of high CPK levels, magnetic resonance imaging (MRI) of cervical spine, electromyography (EMG), and muscle biopsy findings. We question, whether the PM could be pathogenically related to the pregnancy? Literature review of the previously reported cases of PM/dermatomyositis and our case report suggests that pregnancy can trigger the new onset of PM.

Keywords: Creatine phosphokinase, Polymyositis, Postpartum.

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INTRODUCTION

Polymyositis (PM) is an idiopathic inflammatory myopathy that targets predominantly skeletal muscles. It causes symmetrical proximal muscle weakness of limbs in particular while distal muscle weakness tends to be mild. Bulbar muscle involvement can be seen in 30% of patients. The exact etiology of PM is unknown; it is thought to be an immune-mediated syndrome associated with other autoimmune systemic diseases. Evidence supports T-cell (CD8(+)/T-mediated cytotoxic process.¹ Among the myositis-specific autoantibodies, antibodies against aminoacyl transfer ribonucleic acid (t-RNA) synthetases and anti-histidyl-tRNA synthetase (anti-Jo-1) antibodies are associated with PM, but the cause of their production remains unclear.² Environmental triggers like infectious viruses [human immunodeficiency virus (HIV), human T-lymphotropic virus-1 (HTLV-1), coxsackievirus B, hepatitis B, and influenza] have been implicated.

Polymyositis and pregnancy association has been reported few times, and most of them reported about the effect of myositis on pregnancy and high-risk pregnancy management in known myositis patients, but the pregnancy itself causing PM is not reported that widely.

CASE DESCRIPTION

A 20-year-old woman, primigravida, transferred to our hospital because of breathing difficulty 4 days after delivering a dead fetus at 24 weeks of gestation. She was intubated in emergency department in view of respiratory distress and shifted to critical care department. There were no significant past medical or genetic history of patient or her family. There was no history of complications like preeclampsia, gestational diabetes, or infections during pregnancy.

On examination, patients' Glasgow coma scale (GCS) was E4M6Vt. Her temperature was 101°F, pulse was 122 beats/minute, and blood pressure was 118/78 mm Hg. No rash was observed. Systemic examination revealed bilateral crepitations. Chest X-ray posteroanterior (PA) view showed bilateral infiltrates and echo assessment showed poor left ventricle (LV) systolic function and

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no right atrium (RA)/right ventricle (RV) dilatation. Patient was treated in line of pneumonia, sepsis, acute kidney injury (AKI), and peripartum cardiomyopathy. Patient's hemodynamics and cardiopulmonary-renal functions improved but she had persistent high-grade fever. Gross muscle weakness was found on day 5 of admission, involving all four limbs, predominantly in proximal muscles and she had dark colored urine. Laboratory tests revealed positive urine myoglobin, raised erythrocyte sedimentation rate (ESR) 47 mm/hour, raised serum creatine phosphokinase (CPK) levels 109,200 U/L, and raised lactate dehydrogenase (LDH) levels 460 U/L. Thyroid function tests were normal. Systemic lupus erythematosus (SLE) was ruled out based on clinical criteria, negative antinuclear antibody (ANA) and negative anti-dsDNA antibody. Initially, we thought of rhabdomyolysis as a differential diagnosis, as urine myoglobin was positive, CPK was high, but we did not find any major recovery with hemodialysis and other supportive measures. As weakness was more in proximal muscles, we decided to investigate her further.

Inflammatory myopathy was suspected in view of proximal muscle weakness and very high CPK levels. Magnetic resonance imaging (MRI) of cervical spine showed diffuse edema within the neck and shoulder girdle muscles on both sides. Electromyography (EMG) showed myopathic pattern, i.e., polyphasic motor unit

potential of decreased amplitude and duration recorded predominantly in proximal muscles of bilateral upper limbs and lower limbs (Fig. 1). Muscle biopsy from the left thigh muscle showed an inflammatory reaction as evident by endomysial infiltration with lymphocytes along with few necrotic muscle fibers (Fig. 2). Polymyositis diagnosed based on Bohan's criteria³ and she was started on high dose of intravenous (IV) steroids for first 3 days (methylprednisolone—1 g/day) then prednisolone 60 mg/day. Tracheostomy was performed on day 10 in view of prolonged ventilation. She responded well to the steroids and other supportive measures, her limbs power recovered progressively, and CPK levels decreased from more than 1 lac to less than 20 thousands by the end of second week).

She was liberated from mechanical ventilator and decannulated on day 19 of admission, but her bulbar weakness persisted resulting in intermittent cough, intolerance to oral feeds, and retained secretions. Videolaryngoscopy was performed which confirmed dysphagia due to cricopharyngeal muscle discordance, because of that we continued to feed her through Ryles tube. She improved gradually; her CPK levels decreased to 1066 U/L. She was able to walk without support, started to take oral feeds, and was discharged on tapering dose of oral steroids. Patient was followed up after 2 weeks then monthly. Prednisone dose at 60 mg/day given for 6 weeks, then the steroid dose was tapered every week by 5 mg for 8 weeks and then tapered off by 2.5 mg every week. She was counseled to avoid to get conceive for a year at least.

DISCUSSION

Pregnancy has been involved in eruption of various autoimmune diseases like SLE⁴ and associated with bad consequences in diffuse scleroderma patients.⁵ Diseases like myasthenia gravis⁶ and multiple sclerosis⁷ have been diagnosed or relapsed in postpartum period. Autoimmune disease may be erupted because of the change in immune function or as a repercussion of passing of fetal cells into the maternal circulation during pregnancy.⁸ Bianchi et al. suggested that fetal cells can remain in maternal circulation following delivery.⁹ Fetal microchimerism can increase the vulnerability to systemic scleroderma in postpartum.¹⁰ Katz reported aggravation of herpes gestationis in postpartum due to drop in progesterone levels; these maternal hormonal changes can trigger autoimmune disease.¹¹ These changes in immune system and maternal hormones may justify the onset of PM in postpartum. Pinheiro et al. suggested that reduction in the humoral response to certain viral antigens explains the association between pregnancy and dermatomyositis (DM),¹² Gutierrez et al. studied 18 women with PM/DM to find the potential effect of pregnancy on the disease and the effect of disease on pregnancy. They observed three patients with prior quiescent disease, which aggravated during pregnancy, their observations allowed them to think that pregnancy as a trigger factor for the onset and exacerbation of PM/DM.¹³

Silva et al. reported 15 patients (9 DM and 6 PM) who developed the disease in antepartum and 7 patients (4 DM and 3 PM) who

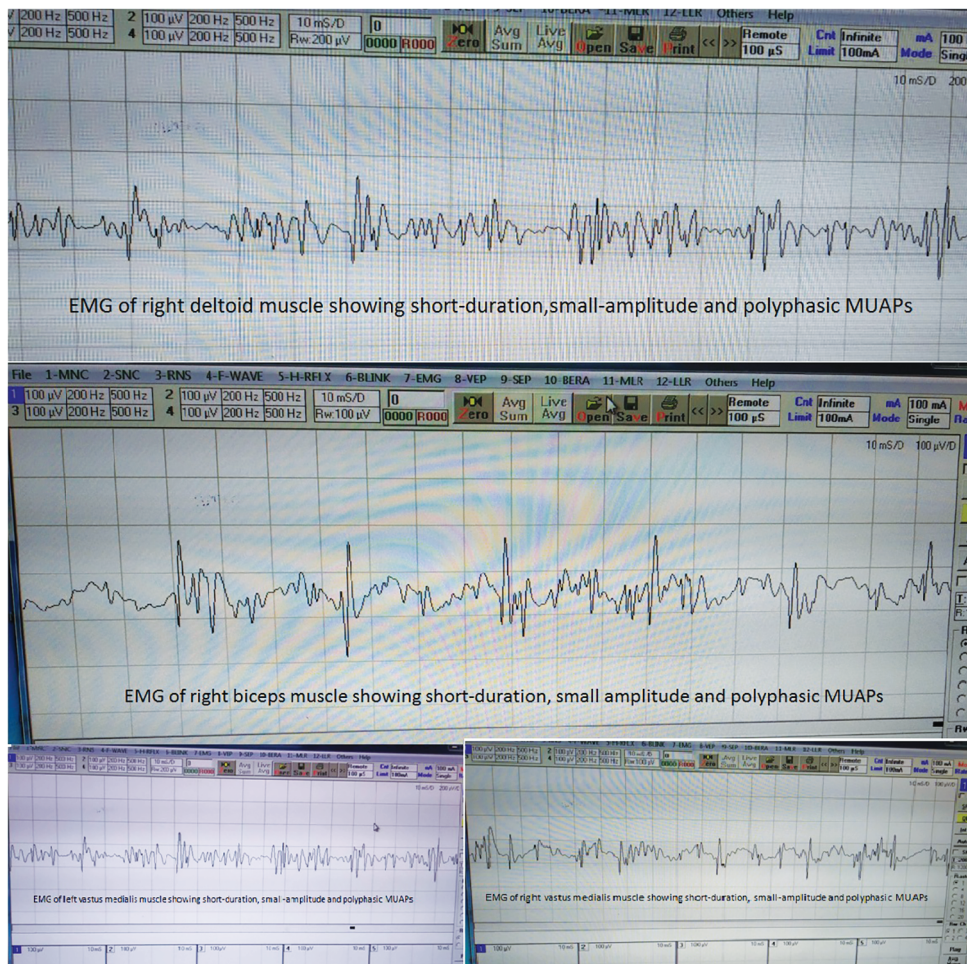


Fig. 1: EMG shows myopathic pattern i.e., polyphasic motor unit potential of decreased amplitude and duration

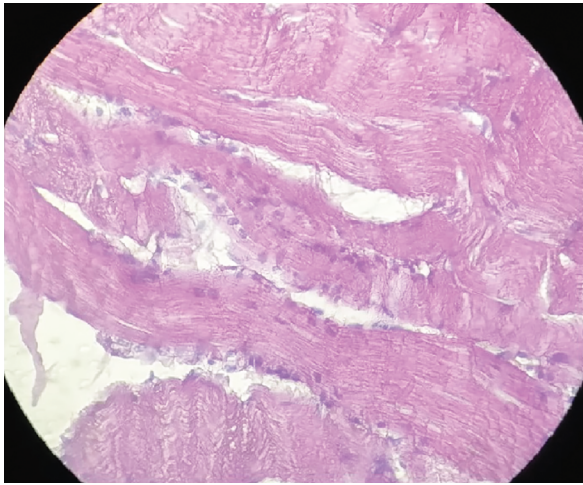


Fig. 2: Muscle fibers showing increased internal nuclei with endomyosial lymphocytes along with degeneration of fibers

developed the disease in postpartum. They concluded that severity of maternal disease speculate the fetal prognosis. Severe the myopathy, greater the chance of fetal loss.^{14,15} Misumi et al. reported four cases of myositis (two DM and two PM) developed in postpartum. All patients responded well to steroid and other immunosuppressant drugs.¹⁶

Other case reports which have reported myositis after child birth or abortion, suggesting that the fetal antigens or maternal hormonal changes precipitate disease activity during pregnancy.^{17–22}

Takei et al. reported a similar case of postpartum PM following intrauterine fetal death. They suggested PM should be considered as differential diagnosis of puerperal fever.¹⁷ Noelyn et al. reported the case of PM in a pregnancy complicated by stillbirth and placental massive perivillous fibrinoid deposition (MPFD). They suggested the possibility of role of the placenta in autoimmune disease.¹⁸ Pappapetropoulos et al. reported a case of PM with three pregnancies.

They concluded that the incidence of PM during one pregnancy does not guarantee its recurrence in next pregnancy. They suggested that women should be counseled to avoid to get conceive during the active disease and regular follow-up is essential even if pregnancy occurs in remission.¹⁹

Kanoh et al. reported a case of DM following delivery of a healthy baby. They suggested that pregnancy can elicit the onset of DM.

They classified pregnancy-related DM into two types; type I—disease aggravated in pregnancy and tends to improve following delivery, type II—has postpartum onset.²⁰ Lee and Yoo reported a case of postpartum DM and discussed earlier reported cases. They concluded that pregnancy can elicit the onset of DM.²¹ Yassaee et al. reported a case of amyopathic dermatomyositis (ADM) following a spontaneous abortion and progressed 2 years later to classic DM following the delivery of a healthy baby.²²

We are reporting a case of PM which manifested following the delivery of dead fetus. The disease process could have started peripartum but it manifested first in postpartum. Postpartum PM is rare, with only few reported cases.

Though our patient's initial presentation was more suggestive of pneumonia, sepsis, and peripartum cardiomyopathy, but after few days she developed myoglobinuria and had gross symmetrical proximal predominant muscle weakness. On further evaluation, definitive diagnosis of PM was established. She had excellent response to corticosteroids, as her limb muscle weakness improved in 2nd week but dysphagia which was reported in 30% of patients persisted till 7th week.

CONCLUSION

This case of PM which manifested following delivery indicates the possibility of new onset PM during postpartum period and it should be considered as differential diagnosis of puerperal fever. Literature review of the previously reported cases of PM/DM and our case report suggests that pregnancy can trigger the new onset of PM.

CONSENT

Written informed consent was acquired from the patient prior to writing this case report.

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