



Pregnancy in Dermatomyositis Complicated with Covid

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ABSTRACT

32-year-old primigravida, CATB2, Covid positive patient with acute onset of facial palsy, fever, sore throat and productive cough referred from a peripheral hospital came for caesarian section. Past history revealed occurrence of juvenile dermatomyositis for which she took steroids for one and a half year which was slowly weaned off. Prevalence of dermatomyositis is 1 to 8 in 100,000. She was started on steroids which was to be continued for a week after caesarian section. She was on paracetamol and cefuroxime on admission. Basic investigations were normal. Caesarian section was done in covid theater following covid protocols under subarachnoid anesthesia. Baby had an APGAR score of 9 and was covid negative. Mothers' recovery was slow. Facial deviation got corrected in 4 to 5 months. Ptosis persisted even after 9 months and is on treatment. Dermatomyositis is an uncommon inflammatory disease that affects mainly proximal muscle limiting day to day activities. But it can affect respiratory, cardiac and gastrointestinal systems, eyes and joints. Diagnosis is by muscle biopsy and an increase in muscle enzymes. Pregnancy during remission period is safe. But pregnancy can provoke the disease. Skin changes usually precedes muscle symptoms, though we could not notify any lesions. Lung involvement is prominent, and covid also carries the same amount of risk with respiratory system. Systemic glucocorticoids are the first line of treatment. Immunosuppressant drugs Azathioprine and methotrexate are added as second line of therapy, though these drugs are unsafe during pregnancy. Intravenous immunoglobulin, give good outcome during pregnancy. Plasmapheresis is also beneficial. Most of the pregnant patients end up in caesarian section. Regional anesthesia is the choice of anesthesia. Covid patients with respiratory infections, maintaining saturation above 90%, choice of anesthesia is usually regional anesthesia.

Key Words: Juvenile Dermatomyositis, COVID, Cesarian Section



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INTRODUCTION

Dermatomyositis is an inflammatory disease of the muscles. It is more common in women. Juvenile dermatomyositis can affect children at the age of 5 to 15 years. The disease has been widely treated with corticosteroids, intravenous immunoglobulin, plasmapheresis, and immunosuppressant drugs. The basic pathology is found to be vasculitis caused by immune complexes. The disease can go into remission or get precipitated during vulnerable situations. The diseased patients need careful evaluation of all systems before any anesthetic procedure whether symptomatic or not. Pregnancy during remission period goes unnoticed. Physiological changes during pregnancy can mask exacerbation of dermatomyositis. Pulmonary complications are more common in dermatomyositis and in covid. We had a 32-year-old primi posted for caesarean section with history of juvenile dermatomyositis, presenting with upper respiratory infection, wet lungs and facial palsy during covid period who was tested positive for covid.

CASE HISTORY

We had a 32-year-old female patient, 40 weeks of gestation admitted with fever, sore throat, productive cough, with acute onset of facial deviation posted for caesarean section, referred from a peripheral hospital during the peak time of covid. She was CATB2 covid positive and was admitted in covid ward. She was diagnosed to have dermatomyositis at the age of 15 years which was confirmed with muscle biopsy and treated with steroids twice daily and was gradually tapered off in one and half years and then stopped. She was asymptomatic and got married and conceived. On admission neurologist started her on prednisolone 16mgm bd which was to be continued for a week after caesarian section. She was started on Paracetamol and Cefuroxime. We couldn't elicit any history of skin lesions and on examination also we couldn't notice any. Though hemodynamically stable her upper respiratory status was poor. We decided to proceed for caesarian section under regional anesthesia in covid theater following covid protocol. We could successfully complete caesarian section under spinal anesthesia with 1.8ml 0.5% Bupivacaine [heavy]. She was given 1gm Methyl prednisolone per operatively considering her poor lung condition. The baby had an Apgar score of 9. Baby was tested

covid negative. Mother's recovery was very slow. Facial deviation got corrected after 4 to 5 months during postpartum period. Ptosis persisted even after 9 months for which she is under neuromedicine evaluation.

DISCUSSION

Dermatomyositis is an uncommon inflammatory muscle disease characterized by symmetric muscle weakness and distinctive skin lesions. It is more common in females and affect both children and adults [1]. Juvenile dermatomyositis affect children between 5 to 15 years and prevalence is 2 to 3 /million [2]. In adults it affects at age between 40 to 60 years and prevalence is 1 to 8/100,000. Estimated prevalence of pregnancy with dermatomyositis is 11/100,000 [3]. Relative risk of obstetric complication is calculated to be 7.6 and fetal risk is 2.7 after onset of myositis [4]. Etiology includes genetic factors, environmental factors, various drugs, high intensity radiation exposure in women and immunological factors.

PATHOPHYSIOLOGY

Activation of compliment factor and formation of neoantigens and membrane attack complex deposits on vascular walls resulting in inflammation of blood vessels [2, 5 & 6]. Perifascicular atrophy around periphery of fascicles is the hallmark of dermatomyositis on muscle biopsy. Presence of antisynthetase antibody is most specific and elevated muscle enzymes helps in confirmation of diagnosis. Electromyography, Xray and MRI assist in confirmation of diagnosis.

CLINICAL SYMPTOMS

Muscle weakness is the most common presenting symptom which can be subacute in onset and progressive. Proximal muscles are affected which restricts movements. In severe cases dysphonia and dysphagia occurs. Dermatomyositis affects respiratory system (interstitial lung disease) [1], cardiac muscles (hypertension, arrhythmia), gastrointestinal tract (aspiration) and small joints (restricted airway). Dermatomyositis can affect fertility [1].

PREGNANCY AND DERMATOMYOSITIS

Onset of dermatomyositis during pregnancy is rare. Dermatomyositis seems to remain inactive during pregnancy in most patients [3]. Initial symptoms like fatigue are easily overlooked [7, 8]. Conception after onset of disease and new onset of dermatomyositis during first trimester carry a very high risk of spontaneous abortions [1, 2]. Population based studies showed an increase in hypertensive disorders [1, 9], eclampsia, eclampsia and antepartum haemorrhage [7, 10 & 11]. If the disease flare up during pregnancy IUGR, prematurity and foetal loss can occur [1, 3, 12 & 13]. Foetal prognosis depends on maternal disease level [14, 15]. Mortality during first trimester is 83%. 17% relapse during pregnancy [16] Acute onset of dermatomyositis during 2nd trimester can lead to rhabdomyolysis, and myoglobinuria causing spontaneous abortions [17]. Such patients can land up with renal failure.

Uterus is unaffected by myositis [8]. Dermatomyositis during remission period is not a threat to pregnancy [2, 18 & 19]. Pregnancy can trigger autoimmune diseases due to hormonal changes [7] and cellular immunity is decreased during pregnancy which will make them more prone for infections. In India, poor foetal outcome and spontaneous abortions are reported.

Corticosteroids are the first line of treatment for dermatomyositis during pregnancy [20] and are relatively safe [16] [21]. Prednisolone does not cross placenta fully. Most of it goes to maternal side. Prednisolone can affect growth of the baby by interfering with collagen formation [18]. Corticosteroids for more than 6 months can produce myopathy, secondary diabetes osteoporosis, cushingoid features, and make them more susceptible to infections. Intravenous immunoglobulin [20] is 60 to 70% efficient. Plasmapheresis is needed in some [20]. Antineoplastic drugs like methotrexate, azathioprim and others are not safe [22, & 3]. But used in resistant cases in combination with steroids and IViG.

Delayed gastric emptying, reduced gastroesophageal sphincter activity, straightening of esophageal gastric angle during pregnancy and increase gastric acid volume stimulated by placental gastrin make pregnant patients more prone for gastric regurgitation and pulmonary aspiration. Dermatomyositis can affect muscles of oropharynx and upper oesophagus. So, chances of pulmonary aspiration is very high [23].

Nonspecific ST-T wave changes, left axis deviation, enlargement of cardiac chambers are all part of physiological changes during pregnancy. Inflammation of cardiac muscles during pregnancy in dermatomyositis can cause hypertension, congestive cardiac failure and arrhythmias. The presentation may be subclinical and always do an echocardiogram [24].

During pregnancy diaphragm is elevated. Functional Residual Capacity is reduced by 20%. Closing volume will be above functional residual capacity which makes the alveoli more prone for atelectasis and hypoxia. Intrapulmonary shunts increase towards term. Tidal volume decreases and respiratory rate increases. In dermatomyositis 50% of patients show evidence of pulmonary diseases like pneumonitis, alveolitis and bronchopneumonia. Progressive weakening of thoracic and intercostal muscles provides a restrictive pattern. Exertional dyspnoea, exercise intolerance and nonproductive cough will be there. Reduced chest movements, bilateral crackles and patchy infiltration of both lungs are

seen. After 28 weeks, of pregnancy supine hypotension syndrome sets in which can compromise placental blood supply and affects the foetus.

GIT perforations, anaemia, intercurrent infections during pregnancy are also associated with dermatomyositis. Extreme muscle weakness may demand assisted delivery and most of them ended up in caesarian section [24].

Covid virus can enter through transcribal route, axonal transport, transsynaptic transfer, haematogenous or lymphatic route [25]. Covid virus can affect cranial nerves: Olfactory, trigeminal, vagus and facial nerves [26, 27, 25, 4, 28, 29, 30, 31 & 32]. Isolated facial nerve palsy is the most common acute mononeuropathy reported [30]. Bell's palsy has been reported in some patients. In pregnant patients' frequency of constitutional symptoms were the same. Sars Covid infections during pregnancy is a risk factor for eclampsia. There is a twofold increase in still birth reported [31]. Spontaneous abortions, preterm delivery and foetal death are reported [33]. During pregnancy physiological and immunomodulatory changes exacerbate the presentation. Most common method of delivery is by caesarian section. Covid virus can alter development of foetal brain predisposing them for neurodevelopmental disorders. [6%] [32] Interleukin 1 and TNF alfa is found in pregnant patients with covid. Most common treatment is with antiviral drugs, antibiotics, corticosteroids, intravenous immunoglobulin and oxygen therapy.

ANAESTHETIC PROBLEMS

- 1) Preop history evaluation with onset of disease, confirmation of diagnosis and treatment history is important.
- 2) 50% of patients with dermatomyositis will have lung involvement. Interstitial lung disease, bronchopneumonia and alveolitis will be present [22]. Respiratory assessment with clinical evaluation and accessory evaluation with the help of PFT is valuable. Can ask for Pulmonologist advice and management preoperatively.
- 3) Diffusion capacity may be low [34]. Increase in intra pulmonary shunt towards term, decrease in FRC, alveolar collapse with a higher closing volume during pregnancy make them more prone for rapid desaturation.
- 4) Cardiac involvement may be subclinical. An echocardiogram should be advised along with ECG. Evaluation of exercise tolerance ideally should be done, but may not be practical during pregnancy. Myocardial fibrosis, CCF, cardiac arrhythmias can occur. [35, 34].
- 5) Delayed gastric emptying, loss of gastroesophageal sphincter activity, straightening of gastro esophageal angle during pregnancy and increase in gastric acid volume stimulated from placental gastrin make pregnant patients highly prone for acid aspiration. Dermatomyositis can affect muscles of oropharynx and upper oesophagus if the disease is active. This will be added up to the risk of aspiration under GA.
- 6) Joint involvement can affect air way assessment. Mouth opening, neck extension and flexion should be meticulously done.
- 7) Rhabdomyolysis and muscle necrosis can release potassium. So, level of potassium is important [14, 36].
- 8) Long term steroid therapy can induce hyperglycemia. Blood sugars should be normalized.
- 9) Myopathy, associated with dermatomyositis and secondary myopathy due to long term steroid therapy can affect duration of nondepolarizing muscle relaxant. Consider short acting muscle relaxants and opioids if there is a need for general anaesthesia.
- 10) Some patients may be on anticholinesterases therapy for severe muscle weakness. Consider delayed awakening and recovery.
- 11) Prepare and obtain consent for elective postop ventilation if the situation demands general anaesthesia.
- 12) Consider anaemia and intercurrent infections. Crossmatch and arrange blood and consider antibiotics.
- 13) Conception after onset of disease can have hypertension, preeclampsia and eclampsia who will need proper management with intravenous antihypertensive drugs like magnesium sulphate, labetalol etc. Nondepolarizing muscle relaxants should be used with caution.
- 14) Neonatal experts should be present as most of these babies are IUGR or preterm.
- 15) Regional anaesthesia is always safe for caesarian sections.
- 16) General anaesthesia requires adequate preoxygenation, antiapnoea prophylaxis, sellicks maneuver, short acting muscle relaxants and opioids and an informed consent for elective postop ventilation is mandatory.

CONCLUSION

Dermatomyositis with pregnancy and covid infection coming for caesarian section is very rare. Pregnancy during remission period is like any other normal pregnancy. Dermatomyositis with pregnancy if symptomatic can be treated with steroid and intravenous immunoglobulin. The outcome recorded is good. Mothers can have restricted movements, restrictive disease of lung with poor pulmonary functions. Pregnant patients are more prone to aspirate when gastrointestinal system is also affected. Dermatomyositis makes pregnant patients to become diabetic, hypertensive and hypothyroid. Most of these patients need an early termination of pregnancy and most of them ended up in caesarian section [37]. We could successfully manage the caesarian section under regional anaesthesia. Our patients recovery was slow and her ptosis persisted even after 9months. The baby is healthy and active.

REFERENCES

1. Arthritis, Semin Rheum. 2018 Jun. (2017). Pregnancy outcomes in adult patients with dermatomyositis and polymyositis. Kathleen D kostad, MD, PhD, Fiorentino, MD, PhD, [. . .] and Lorinda Chung. *Seminars in Arthritis and Rheumatism*, 47(6), 865–869 Published online. <https://doi.org/10.1016/j.semiarthrit.2017.11.005>

2. Madu, A. E., Edwin Omih, E. B., & Lindow, S. W. (2013). Juvenile dermatomyositis in pregnancy. Case reports in obstetrics and gynecology. Volume. Article iD890107. <https://doi.org/10.1155/2013/890107>. Published
3. Akalin, T., Akkaya, H., Büke, B., & Koçak, İ. (2016). A Case of New -Onset dermatomyositis in second trimester of pregnancy: A case report and review of literature. *Case reports in obstetrics and gynecology. Case Reports in Obstetrics and Gynecology/OpenAccess*. Volume 2016/ArticleID6430156, 2016, 6430156. <https://doi.org/10.1155/2016/6430156>
4. Paliwal, V., & Garg, R. and Nidhi Teyan. (2020). Neuromuscular presentation in Covid !9. *Neuro sci*. 41[11]. Published online 2020 Sept 15, 7 491599, 3039–3056. <https://doi.org/10.10072-020-04708-8.pmicd:pmc>, PubMed: 32935156
5. Christine. S. Rinder Diseases related to immune system dysfunction. *Stoelting Anaesthesia and coexisting Diseases* (5th ed).Page 603.
6. Bradely, W. G., & Maria Salam, A. *Harrisons principle of internal medicine*. 10th edition Page 2184.
7. Zhong zhiqiang, Fuanlin, Jing Yang, Feng Chun Zhang. (2017). Pregnancy in polymyositis or dermatomyositis; Retrospective results from a teritary centre in China. *BJ of Rheumatology* 56, issue8. Page 1272–1275, <https://doi:10.1093/rheumatology/k ex 070>.
8. Dourmishev Assen, L. A., & Dourmishev, L. *Dermatomyositis. Advances in recognition, Understanding and management* (pp. 103–106).
9. Sirajum Munira, M. D., & Christopher-Stine, L. MD. Pregnancy in myositis and scleroderma. M. P.H. <https://doi.org/10.1016/j.bpobgyn.2019.10.004>
10. Missumi, L. S., Dzousa, F. H. C., Queiroz Andrade, J., & Shinyo, S. K. (2015). Pregnancy outcomes in dermatomyositis and polymyositis. [Revista Braseleira de Rheumatologia ElsevierREV BRAS]. *RHEUMATOL*, 55(2). 95-102. www.rheumatologia.com.br.
11. Akalin, T., Akkaya, H., Buke, B., & Kocak, I. (2016). A case of New Onset dermatomyositis in 2nd trimester of pregnancy: A case report and review of literature. published 10th July Case report/open access. Volume/ArticleID6430156. <https://doi.org/10.1155/2016/6430156>. Case reports in Obstetrics and Gynecology
12. Tuccnardi, A., Czuzoj-Shulman, N., & Halim, A. Abenhaim(2015). Retrospective study using data from healthcare cost and utilization project, Nationwide in patient sample from 1999–2015. *American Journal of Obstetrics and Gynec.ology*. <https://www.ajoj.org.articlepdf>.
13. Sayuri Missumi, L., Desouza, F. H. C., & Andrade, J. (2015). 55[2]. *Samuel Katsuyuki Shinyo Pregnancy Outcomes in Dermatomyositis and Polymyositis*, November 7, 95–102. <https://doi.org/10.1016/j.rbr.2014.10.001.Epub2014>
14. Silva, C. A. SMSultanand. A. Isenburg(2003). Pregnancy outcome in adult onset idiopathic inflammatory myopathy. *Rheumatology*, vol 42, 10 pp. 1168–1172.
15. Chopra, S., Suri, V., & Bhagya, R. MRThami, A. Sharma, P(2008). Bambery Autoimmune inflammatory myopathy in pregnancy. *Medscape Journal of Medicine*, vol.no,no, 1 [Article], 17.
16. Awatef, K., Salim, G., & Zahra, M.F. (2016). A rare case of dermatomyositis revealed during pregnancy with good outcome. *Pan African Medical Journal*, 23, 117. <https://doi.org/10.11604/pamj.2016.23.117.9198.Pan Afr Med.2016>; 23:117. Published online 2016 March 24. doi:10.11604/pamj:2016.23.117.9198
17. Joshi, D., Kumar, N., & Rai, A. (2009). Dermatomyositis presenting with rhabdomyolysis and acute renal failure; an uncommon manifestation. *Annals of Indian Academy of Neurology*, 12(1), 45–47. <https://doi.org/10.4103/0972-2327.48853>
18. Madu, A. E. (1990). Omih E Baguley E. Lindow S.W. Juvenile dermatomyositis in pregnancy 20 years study of hospital diagnosed alleghency county P A1963-1982. *Journal of Rheumatology*, 17(10). 1329-1334. website <https://Europe.pmc.org/abstract/med/2254890>. Accessed March 14, 2017.
19. Wahl, M. Amy-Labbe and Mariam Davidson(2010). Pregnancy and childbirth with neuromuscular diseases, July–September, 2010 issue of MDAs Quest Magazine.
20. Abasse, A. A., Moustaphe, N., Mohammed, D.L., & Hassen, A(2021). Dermatomyositis and pregnancy. A case report with review of literature. *Rheumatology department. Chu Artiside le Dante Dakar Senegal*. DOi:10.36347/sasjm. Vol 7105.006SASJM.75 194-198.
21. Zacharia, F. N., Nikolaos PhD, G. G., & Kontomanolis, N. Emmanuel, PhD. (2017). Patients with dermatomyositis/polymyositis during pregnancy. *Gynecology and obstretic research*. Volume4: Issue 1Article Ref #1000gj4138.
22. Mayu, S., Isyojima, S., Miura, Y., Nishimi, S., Tokunaga, M. H.T., Takahashi, R., & Miwa, K. K.Y. (2019). Polymyositis-dermatomyositis and interstitial lung disease in pregnant woman successfully treated with cyclosporin and tapered steroid therapy. *Case Reports in Rheumatology*. Volume 2019/Article ID 4914631. <https://doi.org/10.1155/2019/4914631.Published>
23. Williams, L., Chang, P. Y., Park, E., & Gorsen, K. (2007). Successful treatment of dermatomyositis during pregnancy with intravenous immunoglobulin monotherapy. *Obstreticsand Gynecology*, 109(2 Pt. 2), 561–563. <https://doi.org/10.1097/01.AOG.0000253244.45837.7c.Pubmed>
24. Kolstad, K. D. MD PhD, & Fiorentino, D. MD PhD,[...]and Lorinda Chung, MD, MS. (2018). Pregnancy outcome in adult patients with dermatomyositis and polymyositis. Author manuscript; 47 (6). *Seminars in Arthritis and Rheumatism*, 865–869. Published online 2017Nov20.doi:10.1016/j.semiarthrit.2017.11005.
25. Martins, A. S. P., & Losa, F. J. F. [...] and Mercedes Garcia Gasalla. Facial nerve as neurologic manifestation of Covid 19. *Journal of Global Infectious Diseases Walters Kluwer-Medknow Publications*.

26. Chen, Xiangliang, Laurent, S., & Warnke, C. (2021). Systematic review of neurological symptoms and complications of covid 19. 20 July 2020. *Journal of Neurology*, 268, 392–402.
27. Lashrya, Noleu, T., & Mukherji, S. S., and Nictel. Mejia Post acute neurological consequences of Covid 19 an unequal burden.
28. Spudich, S., & Nath, A.. (2022). Nervous system consequences of Covid 19. *Science*. Doi.10.1126/Science.abm2052, 375(6578), 267–269. <https://doi.org/10.1126/science.abm2052>
29. Magalhaes, Joao E., and Pedro Augusto Sampaio-Rocha-Filtio(2022). Pregnancy and neurologic complication in pregnancy. A scoping View. Seaud 2022 July. *Acta Neurologica*, 146(1), 6–23. Published online. April 7. doi10.1111/ane.13621.PMCID:PMC911489.PMID:35388457.
30. Wang, Ruting, & Wu, Z. (2022). Chenyang Deleterious effect of nervous system in offspring following maternal SARS COVID 2 infection during Covid 19 pandemic. *Translational Psychiatry*, 12, article no.:232.
31. Bohania, N., & Ish, P.[...] and Karthikeyan P Ayyengar. (2021). Cranial Neuropathy in Covid 19 infection. A case series and review of literature. *Infez Med. Infezioni in Medicina*, 29(4), 609–613. Published online 2021 Dec10. doi.10.53854/liim-2904-15.PMICD:PMC8805475.PMID:35146371.
32. Mamidi, S. Rahul sony, Pawan Dhull, Sindhu Singh. J(2022). Muthukrishnan Postcovid Neurological Complications; Acasa series and review of literature. *Case Series Year 2022/Volume 24/Issue 2/page195-199*. April-1, 132–121. <https://doi.org/10.4103/jmms.jmms>
33. Berron-Ruiz, A. L., Sauer-Ramirez, R., & Johnson-Ponce, J. (2010); 24[3]. Dermatomyositis and pregnancy. *Perinatología y Reproducción Humana*, 3, 175–181.
34. Reidwaldman, & Madeline, E. Dewwane, JunLu, 82(2), 283–296. doi. <https://doi.org/10.1016/J.Jaad.2019.05.105>, PubMed: PMID;31279813Doi10;1016/J.Jaad.2019.05.105Dermatomyositis; Diagnosis and treatment JAMA cad Dematol 2020 Feb.
35. Stoelting MD. Stephen F Dierdorf and J Scott Walton(...) Anaesthesia for patients with rare and co-existing diseases. *Anaesthesia5thedition* Paul G Barash Bruce F Cullen MD Robert K, 521–525. Clinical
36. Shupak, B. S., Patel, R. C., Patel, C., & Calkins, J. M. (1992). Neuromuscular blockade in patients with active dermatomyositis. *Anesthesiology*, 77(5), 1031–1033. <https://doi.org/10.1097/00000542-199211000-00028>. Paul G Barash, Bruce F Cullen, Robert K Stoelting 2011 Chapter 25. Rare and Coexisting Diseases
37. Lara. C. Pullen Phd(2019). Pregnancy outcomes in females with idiopathic inflammatory myositis April 8, 2019 *Rheumatologist*. <http://www.rheumatologist.org->