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CASE REPORT

Anaesthetic Management of a Parturient with Mixed Connective Tissue Disorder

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Introduction

Mixed connective tissue disorders are a group of autoimmune disorders with overlapping clinical features of rheumatoid, systemic lupus erythematosus, polymyositis, and scleroderma. They are associated with the presence of high titers of anti-U1RNP, anti-La, and anti-Ra antibodies [1]. The underlying pathophysiology is non-inflammatory vasculopathy. The presentation of these disorders includes arthralgia, Raynaud's phenomenon, puffy hands, esophageal abnormalities, and cardiac problems such as arrhythmias, conduction block, and also proximal muscle weakness with impaired pulmonary function [1,2]. In Pregnancy, mixed connective tissue disorders have varied presentations and outcomes, from spontaneous abortion to normal/cesarean delivery. Lung involvement is more common and could result in either ILD or fibrosis, which would hamper the normal physiology of lung mechanics. The altered respiratory physiology in pregnant women along with pathological lung changes due to MTCD makes the management of such cases more challenging, especially under general anaesthesia.

Case Report

We report the successful management of a 32-year-old parturient, gravida 2, abortion 1, with mixed connective tissue disorder and early onset interstitial lung disease with oesophageal and vaginal candidiasis for emergency cesarean section at 37 weeks of gestation. Twelve years ago in 2009, she had complaints of swelling of toes and fingers and underwent medical treatment with tablet mycophenolate mofetil 1g once daily and tablet prednisolone 40 mg once daily. Her CT-Chest revealed fibrotic changes suggestive of ILD affecting both lungs in the lower lobes. She took medications until 2018 and later stopped taking drugs for 3 years. Later in 2021, she had an abortion so she was started on tablet hydroxychloroquine 200 mg once daily and resumed her pharmacotherapy. In the present pregnancy, she had a history of herpes zoster ophthalmicus and vaginal candidiasis in her 3rd month of gestation, which was medically managed. Also, she was diagnosed with hypothyroidism in her 2nd month of gestation and was started on tablet eltroxin 50 mcg once daily. She had no complaints of joint pain, breathlessness, or chest pain and was hemodynamically stable with normal laboratory investigations throughout her pregnancy. Workup for mixed connective tissue disorders showed that she was positive for Anti Ro and Anti La antibodies. At 37 weeks of gestation, labor was induced in view of oligohydramnios, and because of the incoordinate uterine contraction and fetal distress, she was planned for an emergency cesarean section. Fetal heart rate tracing revealed variable deceleration and bradycardia with a heart rate dropping up to 90 beats/min. Considering the severity of fetal distress, general anesthesia with rapid sequence induction and intubation (RSII) was administered to the patient. The patient was placed in the 30-degree head-up position on the operating table to minimise the compression of the gravid uterus

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on the lungs. After adequate preoxygenation, the patient was induced with propofol 100 mg i.v. and succinylcholine 100 mg i.v., and the trachea was intubated with 7 mm ID cuffed ETT, after which bronchospasm was observed which was confirmed with auscultation and ventilatory parameters showing maximum inspiratory pressure of 40 cmH_2O and sharkfin pattern on the capnographic waveform, and was managed with salbutamol MDI puffs, propofol boluses to deepen the plane of anaesthesia and adjusting ventilatory settings as per the requirement. Anesthesia was maintained with sevoflurane in O_2:Air (40:60) mixture with muscle relaxant, vecuronium. The remaining intraoperative course was uneventful with no adverse events. The baby was delivered promptly, was active, and healthy. The uterine contraction after delivery was satisfactory with oxytocin infusion and there was no evidence of postpartum haemorrhage. The patient was successfully extubated. Postoperatively, salbutamol nebulization and incentive spirometry was advised for alveolar recruitment. The patient was discharged after a week and was advised to resume her medications with an immunology workup.

Pregnancy in women with autoimmune disorders is not uncommon, and systemic effects vary from one individual to another. Most manifestations of autoimmune disorders would be suppressed during pregnancy due to their immunosuppressed state and the presence of the placental barrier. However, maternal complications include remission and exacerbations of the disease warranting treatment with corticosteroids and immunosuppressants, abortion, pre-eclampsia, HELLP syndrome, eclampsia, premature delivery, anemia, and maternal mortality due to renal failure and cardiopulmonary complications. In the fetus, vasculopathy could cause IUGR or might result in perinatal mortality. Pulmonary involvement in MTCD may be unnoticed and patients remain asymptomatic for long periods. The longstanding disease may eventually lead to ILD and fibrosis. Major manifestations include interstitial pneumonitis and fibrosis (20% to 65%), pleural effusion (50%), pleurisy (20%) and pulmonary hypertension (10% to 45%) [3]. Ground glass opacities with hyperattenuated regions in 78.2% of patients and ground glass opacity with mild fibrosis, with thickened non-septal or septal lines in 21.8% of patients are the most common findings in HRCT-chest [4]. These changes would result in desaturation, bronchospasm, and respiratory acidosis during anaesthesia administration. In these patients, reduction in the FRC resulting from the gravid uterus and ILD involving the lower lobes of both lungs leading to increased pulmonary artery pressure and bronchospasm resulting from airway manipulation could cause rapid desaturation due to decreased oxygen reserve, which should be given special consideration during general anesthesia.

Although neuraxial anaesthesia remains the preferred anesthetic technique in the treatment of pregnant women with MCTD, general anesthesia is unavoidable in certain emergency conditions. In such conditions, all necessary measures including the appropriate choice of anesthetic agents, measures to avoid hypoxia, hypercarbia, and respiratory acidosis by appropriate adjustment of ventilatory parameters must be taken into account to maintain hemodynamic stability with no or less damage to the pulmonary system. Desaturation after induction and bronchospasm should be anticipated and appropriately managed. The decision to extubate the patient should be based on the degree of underlying pulmonary pathology and recovery of pulmonary and neuromuscular function. Preoxygenation and induction in the head-up position, intra-operative alveolar recruitment maneuvers, and smooth induction and extubation would prevent desaturation and bronchospasm. These anticipated complications could occur even during the postoperative period. Also, post-operatively, these patients should be followed up for other pulmonary complications and managed appropriately.

Highlights

Mixed connective tissue disorders present in various forms in pregnancy and the outcome also varies from abortion to normal delivery. The multisystem involvement in the connective tissue disorders makes it difficult from the anaesthetists point of view to formulate appropriate plan of anesthesia in these patients. Herewith we report a successful management of a case of mixed connective tissue disorder with interstitial lung disease for emergency ceserean section. Due to the involvement of the lung, anaesthetic plan also varies in these patients, which has been discussed here.

Declaration of Conflicting Interest

Nil.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Acknowledgement

Nil.

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