Journal of Anesthesia and Clinical Research

Paolo et al., J Anesth Clin Res 2018, 9:11
DOI: 10.4172/2155-6148.1000864

Case Report Open Access

Emergency Caesarean Section in Systemic Sclerosis: Multidisciplinary Management

Murabito Paolo[1], La Rosa Valeria[1], Scarpinati Giuseppe[2], Bonsignore Christian[1], Tringali Eleonora[1], Minardi Carmelo[1*], Pitino Sofia[1] and Astuto Marinella[1]

[1]*Department of Anesthesia and Intensive Care, Policlinico University Hospital, G Rodolico, Catania, Italy*

[2]*Department of Anesthesia and Intensive Care, Santo Bambino Hospital, Catania, Italy*

[*]**Corresponding author:** Minardi Carmelo, Department of Anesthesia and Intensive Care, Policlinico University Hospital, G Rodolico, Catania, Italy, Tel: +393398643162; E-mail: minardi.carmelo@virgilio.it

Received date: November 10, 2018; **Accepted date:** November 21, 2018; **Published date:** November 28, 2018

Copyright: ©2018 Paolo M, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Systemic sclerosis (Ss) is a rare autoimmune disease, it is associated with abnormal deposition of extracellular collagen in visceral organs and skin. Another feature of Systemic sclerosis is vascular dysfunction, causing distal amputation secondary to multiple Raynaud's Phenomenon. Ss it is an anaesthetic challenge from different point of views, patients usually show poor oral aperture and consequently difficult airway management. Moreover they may even be affected by restrictive pulmonary disease, representing an obstacle for adequate mechanical ventilation. Ss it is also associated with cardiovascular and respiratory comorbidities. Management of systemic sclerosis involved many different specialties such as: paediatrician and gynecologist who gave 32 weeks of pregnancy as an acceptable moment to perform caesarean delivery if needed. Also thoracic surgeon was engaged to help us in managing difficult airways by flexible bronchoscopy if general anesthesia would have been necessary. Patients affected by Ss, are usually young woman, so we should expect to manage pregnancy in this rare pathology also because the immunosuppressant therapies are more effective now than in the past. In this case report, a pregnant women affected by Ss was managed even in emergency situation. In order to reduce mistake and to manage any possible condition such as elective caesarean section or emergency one, we decided to adopt an anesthesiologic protocol to manage our patient. Our protocol established three attempt for each anesthesia technique, first line anesthesia was epidural anesthesia, then spinal anesthesia and as last chance general anesthesia. As a result of non-reassuring fetal heart rate, patient underwent an emergency caesarean section, we had to perform spinal anesthesia because it was difficult to place epidural catheter. Patient was stable after spinal anesthesia and surgery, so our case report strongly recommends to plan carefully anesthesia, in this case spinal anesthesia is a viable solution for women with Scleroderma undergoing caesarean section.

Keywords: Morquio syndrome; Spinal surgery

Case Report

Systemic Sclerosis (Ss) is a rare autoimmune disease, characterized by deposition of extracellular collagen in multiple organs. Its etiopathogenesis is associated to vascular dysfunction and inflammation that ultimately causes connective tissue fibrosis. Etiology of Ss is not fully elucidated yet, some evidence suggest that, as many autoimmune disease, many factors may be involved in developing Ss. Human Leukocyte antigens (HLA-DR1,2,3,4,5 and HLA DQA2) are frequently involved as well as anti-nuclear antibodies, anticentromere and antinucleolar antibodies [1]. Usually Raynaud's phenomenon is the initial symptom and precedes skin changes by less than 1 year in cases of diffuse, cutaneous Ss [2]. Abnormal collagen deposition cause skin tightening that can lead to microstomia. Pulmonary involvement is secondary to diffuse fibrosis, smooth muscle hypertrophy of pulmonary vessels resulting in pulmonary hypertension and ultimately to right heart failure. Pulmonary function tests are consistent with restrictive changes, demonstrating decreased compliance, vital capacity, and diffusion capacity. Another cardiac involvement is associated to myocardial fibrosis resulting ventricular hypertrophy and diastolic dysfunction, and cardiac conduction defects, furthermore, coronary vessels become sclerotic and are prone to vasospasm.

Pre-operative evaluation should focus on skin tightening of face and oral aperture, microstomia and neck mobility should be assessed [3]. Microstomia may require fiber-optic intubation or elective tracheostomy when needed. Cardiac and pulmonary function should be assessed in order to evaluate cardiovascular function and pulmonary involvement. It is also advisable to perform pulmonary functional test to assess restrictive changes. Other anaesthetic issues are operatory room or temperature, it is mandatory to maintain OR temperature above 21°C and intravenous fluids should be warmed to avoid vasoconstriction and Raynaud phenomenon [4]. Intravenous access may be difficult due to skin thickening. There are no specific contraindications about use of anaesthetic but drugs with extensive renal metabolism should be avoided if possible, in patient with advanced kidney failure. Pregnancy may affect or deteriorate pre-existing condition lowering 10 year survival rate of scleroderma patients [5]. During pregnancy, gravid uterus may accentuate symptoms of gastroesophageal reflux, a pre-existent condition in Ss. Moreover, limitation of diaphragmatic excursion may be exacerbate pre-pregnancy dyspnea. Women affected by systemic sclerosis are at greater risk of cardiac and pulmonary problems as well as renal ones. Scleroderma renal crisis rate may be increased during pregnancy, differential diagnosis should exclude pre-eclampsia. Ace inhibitor therapy, reduced incidence of scleroderma crisis but it is not recommended because when administered during first trimester of pregnancy, congenital malformations increase [6]. During second and

third trimester, differentiation between pre-eclampsia or renal crisis is difficult because seizure or elevated transaminases may be present. If possible, measurement of plasma renine may rule out pre-eclampsia in which renine levels are low or normal. If SRC is indistinguishable from pre-clampsia, trial of ACE inhibitor is recommended.

Our patient was 36 years old and she was diagnosed with Ss at 16 years old, after multiple Raynaud phenomenon causing severe digital ulcers. Other physical feature was typical Mauskopf Facies characterized by multiple telangiectasias, microstomia, thin lips and pinched nose; she also had Mallampati IV secondary to poor oral aperture. In addition, patient showed severe sclerodactyly and digital ankylosis in both hands and elbow ankylosis.

Patient had a rapid progression and severity of the disease, so she underwent to an autologous transplant, an off label treatment for Ss [7].After autologous transplant, her clinical situation remained stable, HRCT did not show pulmonary fibrosis, echocardiography did not show any sign of pulmonary hypertension or diastolic dysfunction.

The patient was poorly compliant to immunosuppressant therapy, because she decided to stop cyclosporine without any medical advice to get pregnant. Nevertheless systemic sclerosis did not worse, probably due to immunomodulation during pregnancy [8].

She was admitted to hospital at 26 week of gestational for oligohydramnios and intrauterine growth retard (IUGR). During her hospital stay, conservative therapy with bed rest, hydration and periodic laboratory tests were performed.

Consultant paediatrician and obstetrician considered 32 week as an acceptable gestational age to perform potential caesarean section if clinical condition worsened. Perioperative management involved elective ultrasound-guided Central Venous Catheter placement in Left Internal Jugular vein due to skin thickness.

During her hospital stay, patient developed gestational hypertension, and then showed pre-eclamptic signs such as worsening proteinuria, peripheral edema and refractory hypertension to multimodal therapy.

Epidural anesthesia was considered as first line treatment option for caesarean section, spinal anesthesia would have been a second line option and general anesthesia with fiberoptic intubation would have been last extreme choice but was planned [9].

At 30 week gestational age, emergency caesarean section was performed for non-reassuring fetal heart rate (NRFHR). Standard monitoring was used throughout surgery including: NIBP, SpO_2 and ECG. Invasive blood pressure was not monitored because arterial access is associated with increased rate of Raynaud phenomenon in Ss. An experienced anaesthetist performed 3 attempts of epidural anesthesia in different interspaces. Epidural space was identified in each attempt but it was difficult to advance epidural catheter.

Spinal anesthesia with Hyperbaric Bupivacaine 0.5% 12 mg and Fentanyl 20 μg was performed after 3 attempts. Patient had great haemodynamic stability throughout caesarean section, moreover surgery was uneventful and a good response to oxytocin was reported. A female baby (1450 gm) was born with Apgar score 8 and 9 at 1 and 5 min. Post-operative period was uneventful, we emphasize a prolonged motor block that regressed after 4 h, we hypnotize a slow anaesthetic reabsorption due to microvascular fibrosis.

Conclusion

In conclusion Ss is a rare disease, nevertheless anaesthetist may encounter pregnant women in delivery room or obstetric ward because immunosuppressive therapy are more efficacy than in the past. It is important to assess severity of disease before undergoing any kind of surgery, as mentioned before, airway management and intravenous access may be challenging. Cardiac and pulmonary involvements, even in mild form are always present, so their function should be assessed. As reported, patient developed hypertension, it is advisable to make a differential diagnosis between scleroderma renal crisis and pre-eclampsia, signs and symptomps are mostly identical, in case of scleroderma renal crisis, and plasmatic renin would be high. Plasmatic renin is not always available so ACE-inh trial should be performed, considering pros and cons of these drugs in pregnancy.

The aim of our case report is to bring an example of spinal anesthesia in pregnant patient with severe form of scleroderma. Even if epidural anesthesia should be the first line treatment in obstetric surgery, spinal anesthesia, both in elective and in emergency situation, may be a good choice considering that epidural space may be fibrotic or distorted by abnormal fibrosis.

References

1. Arnett FC, Howard RF, Tan F, Moulds JM, Bias WB, et al. (1996) Increased prevalence of systemic sclerosis in a native american tribe in oklahoma. Association with an amerindian HLA haplotype. Arthritis Rheum 39: 1362-1370.
2. Belch JJ (1991) Raynaud's phenomenon: its relevance to scleroderma. Ann Rheum Dis 4: 839-845.
3. Dorotta IR, Schubert A (2002) Multiple sclerosis and anesthetic implications. Curr Opin Anaesthesiol 15: 365-370.
4. Hasan O, Jessar M, Ashar M, Noordin S, Ahmad T (2017) Systemic sclerosis: Clinical manifestations, anesthetic and orthopedic considerations in a patient. Int J Surg Case Rep 42: 24-28.
5. Steen VD, Medsger TA (1999) Fertility and pregnancy outcome in women with systemic sclerosis. Arthritis Rheum 42: 763-768.
6. Cooper WO, Hernandez-Diaz S, Arbogast PG, Dudley JA, Dyer S, et al. (2006) Major Congenital Malformations after First-Trimester Exposure to ACE Inhibitors. N Engl J Med 354: 2443-2451.
7. Sullivan KM, Goldmuntz EA, Keyes-Elstein L, McSweeney PA, Pinckney A, et al. (2018) Myeloablative Autologous Stem-Cell Transplantation for Severe Scleroderma. N Engl J Med 378: 35-47.
8. Piccinni M-P, Lombardelli L, Logiodice F, Kullolli O, Parronchi P, et al. (2016) How pregnancy can affect autoimmune diseases progression? Clin Mol Allergy 14: 11.
9. Sanfilippo F, Conticello C, Santonocito C, Minardi C, Palermo F, et al. (2016) Remifentanil and worse patient-reported outcomes regarding postoperative pain management after thyroidectomy. J Clin Anesth 31: 27-33.