

Case Report

Scleroderma, The Prone Position and Profound Hypotension: A Case Report

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Abstract:

Purpose: *This case report describes a case of profound hypotension in a patient with a diagnosis of systemic sclerosis (scleroderma).*

Clinical Features: *A 59 year old Japanese diagnosed with scleroderma was scheduled for self bone marrow harvest and monocyte plantation to the affected skin in his hands. The patient suffered from cardiovascular and respiratory comorbidities and was taking oral prostacyclin. The hypotension correlated with the onset and offset of the prone position during the anesthesia.*

Conclusions: *This case is an example of profound hypotension of a patient with a diagnosis of scleroderma in the prone position. The diagnoses of both hypotension and the cause of hypotension were delayed. The etiology of the patient's hypotension was thought to be mostly caused by reduced venous return to the heart due to relative hypovolemia by vasospastic phenomena with scleroderma and improper positioning leading to IVC compression in the prone position. This case indicates that preoperative adequate volume replacement and proper positioning are imperative to maintain the circulatory status during anesthesia in the prone position for a patient diagnosed as scleroderma.*

(Key words : hypotension, complication, anesthesia, scleroderma, prone position)

Case Report

A 59 year old Japanese man who had been diagnosed with scleroderma 12 years ago was booked for a procedure as part of a clinical trial which involved bone marrow harvest and subsequent monocyte injections into the hands for treatment of intractable ischemic lesions of the skin. His respiratory symptoms included a chronic dry cough and dyspnea with walking one block. He had Raynaud's phenomenon, pulmonary hypertension, pulmonary fibrosis and gastroesophageal reflux. His medications included oral prostacyclin, theophylline, prednisone 5mg/day (taken on day of surgery), Gastrom (a gastric cytoprotective agent) and vitamin E. He denied the use of cyclosporin or ACE inhibitors.

The patient was 63kg and 163cm tall. Preoperatively, he was in mild respiratory distress, slightly diaphoretic and flushed after minimal exertion. His fingers demonstrated a fusiform

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appearance with extreme pallor, small ulcerations and no clubbing. Telangiectasias were noted on his chest. On auscultation, there were fine crackles in the lower left base and in the lower one half of the right lung. Heart sounds were of normal intensity with no abnormal splitting of S2 ; P2 (pulmonic valve) was not louder than A2 (aortic valve). There were no murmurs or gallop rhythms. The JVP was 2cm above the sternal angle. The airway exam demonstrated a small oral aperture and Mallampatti 3 score.

Pre-operative blood work including : CBC (Hb was 12.5g/dl), electrolytes, glucose, calcium, liver function tests and coagulation studies were normal. The creatinine was 0.84mg/dl. A 12-lead ECG showed sinus rhythm at 85 beats per minute (bpm) with a right bundle branch block. There was no evidence of right atrial enlargement. Chest x-ray demonstrated basal reticular patterns bilaterally with greater involvement on the right. Echocardiogram revealed normal biventricular function, estimated systolic pulmonary pressures of 50mmHg and a small pericardial effusion. A thallium scintigram demonstrated one region of fixed reduced uptake in the inferior wall. A chest CT scan was consistent with right middle and lower lobe fibrosis. Pulmonary function tests revealed an FEV₁ 2.79l (89%), FVC3.20 (86%), and a DLCO of 49%.

Pre-induction vital signs were blood pressure (BP) 134/88mmHg by a non invasive blood pressure (NIBP) monitor, heart rate (HR) 84bpm, and pulse oximetry (SpO₂) 98%. The patient was treated with 100% oxygen and induced with propofol 90mg, vecuronium 6mg, and fentanyl 100 µg with cricoid pressure. The airway was uneventfully secured by direct laryngoscopy and an 8.0mm endotracheal tube. Post intubation vital signs were BP 130/70mmHg, HR 75bpm, SpO₂ 100%, end tidal CO₂ (EtCO₂) 39mmHg and peak ventilation pressures were 25 mmHg. Sevoflurane, air and oxygen were administered for maintenance. Arterial catheterization was not recommended because of the increased risk of severe ischemia to patient's hands.

The patient was turned prone after the intubation. After re-application of NIBP cuff, the NIBP monitor showed blood pressure 130/120mmHg at first measurement and 45/30mmHg at the second measurement. Repeat measurements were approximately 45/30mmHg. The cuff was noted to deflate slowly during measurement and the suspicion of kinking or obstruction of the NIBP cord was considered. The cuff was removed and reapplied with the cord visibly straight, then applied to the other arm, and then a second cuff was applied, with all methods revealing a BP of 45/30mmHg. The other vital signs at this time were: HR 55bpm, SpO₂ 100% and EtCO₂ 38mmHg. The ECG showed normal sinus rhythm with no indication of ischemia. End tidal sevoflurane was 1.6%. The carotid pulse was difficult to palpate due to the patient's prone position with the head turned but the temporal artery was easily palpable. Radial pulses and pedal pulses were not palpable, but brachial and popliteal pulses were easily felt. Perfusion of the fingers was deemed normal by a capillary refill time of 2 seconds and a warm temperature.

A Doppler ultrasound was obtained and revealed return of flow to the patient's right radial artery only when the NIBP cuff pressure was below 50mmHg. The duration of the prone position at this time was 15 minutes.

Intravenous Ephedrine 4mg was given and the BP and HR increased to 70/35mmHg and 68 bpm respectively. Ringer's lactate 500ml was given as a bolus and pre-operatively donated autologous packed red blood cells 200ml was also given with the first dose of ephedrine. The

increases in BP and HR lasted for 20 minutes and one repeat dose of 4mg was given with similar results.

The external jugular vein was noted to be variably filled and fluctuated with positive pressure ventilation. Ventilation pressures were 22-24mmHg and there was bilateral air entry with no evidence of bronchospasm on auscultation. There was no flushing at the intravenous site or on the arms, face or back.

Total bone marrow aspiration volume after 20 minutes prone was 300ml with a goal to reach 600ml. There was no other blood loss.

Patient positioning was checked and there was no external compression on the shoulders or arms that could have accounted for an aorto-radial pressure gradient. The patient's hips and abdomen were checked and the patient's hips were not on the hip bolsters. After 55 minutes in the prone position, the procedure was complete and the patient was returned to the supine position. As monitors were being re-applied, both radial and popliteal and posterior tibial pulses were noted to be strong and easily palpable. The blood pressure *by the NIBP monitor* was 117/65mmHg and the heart rate was 90bpm. Impression marks on the patient's abdomen from padding on the operating table were as prominent as the markings on the patient's chest. The patient remained anesthetised (supine) during the processing of the bone marrow and local infiltration of monocytes to his hands. Vital signs remained stable during these times and emergence was uneventful. Postoperative recovery was uneventful.

Discussion :

The anesthetic considerations of scleroderma are significant and excellent reviews are available^{1,2}. The airway can be difficult to intubate due to a decrease in size of the oral aperture. Pulmonary fibrosis is the most prominent respiratory feature and there is a risk of aspiration due to esophageal reflux. Esophageal dysmotility is found in 75% of patients³. The pathogenesis involves a vasculopathy that results in tissue ischemia and progressive enteric neuropathy that disturbs gut peristalsis leading to gastroparesis³. Cardiac involvement includes pericardial effusions but pericarditis is not common. Pericardial disease is usually clinically silent and benign but the incidence is as high as 40% by echocardiography and 50% in autopsy series⁴. Conduction abnormalities and arrhythmias can occur as a consequence of patchy myocardial fibrosis. Raynaud's phenomenon is common and arterial catheterization can theoretically trigger vasospasm. Volume depletion and hypertension may be present as consequences of vasoconstriction. Pulmonary hypertension is frequently seen in association with pulmonary fibrosis, but can be present in the absence of pulmonary fibrosis. Different therapies are being evaluated for pulmonary hypertension, one of which includes administration of prostacyclin via oral (Beraprost), subcutaneous (Treprostinil), inhaled (Iloprost), and intravenous infusion (Epoprostenol)². Endothelin receptor antagonists (Bosentan) have also shown promising hemodynamic results in patients with scleroderma and pulmonary hypertension⁵ and have been used in systemic sclerosis. Renal compromise is associated with scleroderma and ACE inhibitors are thought to play a role in slowing the progression of renal dysfunction. Finally, contractors due to dermatological involvement can increase risk of pressure sores

and peripheral neurological damage.

Important features of this case include : the delayed diagnosis of hypotension, the aetiology of hypotension, and the aesthetic considerations of scleroderma.

The diagnosis of hypotension was delayed. The reason for the delay was primarily two-fold : assumption of monitor error and the presence of concurrent clinical findings that did not support profound hypotension. The reasons for the presumption of monitor error were : 1) the presence of an initial measurement of 130/120mmHg followed by a measurement of 45/30 mmHg ; 2) what appeared to be a slowly deflating BP cuff and ; 3) the improbability of such a low blood pressure. Only after the Doppler ultrasound was the diagnosis of *right radial artery* hypotension accepted. At this point we noticed that the NIBP monitor was working properly. The features of the case that were considered to be non-supportive of profound hypotension included : the absence of tachycardia, the presence of adequate perfusion of the hands, the presence of a popliteal pulse, the absence of hypoxia, the absence of signs of ischemia on the ECG, and the absence of a decreased ETCO₂.

Many factors could have contributed to hypotension in this patient, including : pulmonary hypertension, the presence of a pericardial effusion, an abnormal thallium scintigram, long term use of oral prostacyclin, chronic steroid use, and vasospastic state of scleroderma. In this case, there was a clear association with the onset and offset of hypotension and the prone position.

It is assumed that our patient was mildly hypovolemic as a consequence of vasospastic property of scleroderma itself and preoperative fasting. Volume depletion may be enhanced due to vasodilating action of sevoflurane in the prone positioned patient with the compression or obliterate the IVC, as is seen in an improperly. If left ventricular preload was impaired due to pericardial tamponade, acute pulmonary hypertension, pulmonary embolus, pneumothorax, or undiagnosed intrathoracic mass causing cardiac compression, then the jugular venous pressure would have been elevated and the external jugular vein would have been distended, which it was not.

It is unlikely that acute ventricular dysfunction would have been the major contributor to hypotension in the absence of anesthetic overdose. Patients with scleroderma can develop patchy replacement of myocardium with fibrosis, resulting in atrial or ventricular dysrhythmias¹, and fibrosis is a probable explanation for the fixed defect on scintigraphy.

Chronic steroid use is associated with hemodynamic instability when supplemental steroid coverage is not provided. A study evaluating hemodynamic responses to major surgery in 12 patients receiving 7.5mg of prednisone daily (including the day of surgery) for many months and with documented adrenal suppression did not find any difference in hemodynamics in the placebo group vs the supplemental steroid group⁶.

Anaphylaxis could have been the cause of hypotension, but there was no bronchospasm, flushing or urticaria seen. However, the lack of these findings does not rule out anaphylaxis. The interaction of general anesthesia and orally administered prostacyclin could be another potential source of low afterload as a cause for the hypotension but a literature search failed to find any reports on this type of interaction.

Vagal reflex also could induce hypotension during the position change. However, vagal reflex

rarely lasts as long as 55 minutes after the intravenous administration of the ephedrine.

In summary, the diagnoses of both hypotension and the cause of hypotension were delayed in this case due to multiple factors. The etiology of the patient's hypotension was thought to be mostly caused by reduced venous return to the heart due to relative hypovolemia by vasospastic phenomena with scleroderma and preoperative fasting, and improper positioning leading to IVC compression in the prone position. This case indicates that preoperative adequate volume replacement and proper positioning in the prone position are important during anesthesia for a patient diagnosed as scleroderma.

References :

- 1) Cossio M, et al. Life-threatening complications of systemic sclerosis. *Critical Care Clinics*. 18 : 819-39, 2002
- 2) Blaise G, et al. Pulmonary arterial hypertension : pathophysiology and anesthetic approach. *Anesthesiology*. 99 : 1415-32, 2003
- 3) Sjogren RW. Gastrointestinal features of scleroderma. *Curr Opin Rheumatol*. 8 : 569-75, 1996
- 4) Gowda RM, et al. Scleroderma pericardial disease presented with a large pericardial effusion-a case report. *Angiology*. 52 : 59-62, 2001.
- 5) Channick RN, et al. Effects of the dual endothelin-receptor antagonist bosentan in patients with pulmonary hypertension : A randomised placebo-controlled study. *Lancet*. 358 : 1119-23, 2001.
- 6) Glowniak JV. A double-blind study of perioperative steroid requirements in secondary adrenal insufficiency. *Surgery*. 121 : 123-9, 1997

症例報告：強皮症，腹臥位と難治性低血圧

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要 約

目的：全身性硬化症（強皮症）の患者において高度の低血圧をきたした症例を報告する。臨床経過：59歳，男性。全身性硬化症による下肢の虚血性病変に対し，骨髄よりの単球分離採取術が予定された。循環器系及び呼吸器系の合併症を有しており，経口プロスタサイクリンを内服中であった。プロポフォールで導入後，腹臥位にて手術を行った。腹臥位中，難治性の低血圧が発生した。手術終了後，仰臥位に戻すと血圧は回復した。結論：本症例は低血圧の診断及び低血圧の原因の発見が共に遅れる結果となっ

た。本症例の低血圧の原因としては，全身性硬化症による血管収縮による血管床減少状態に麻酔薬による血管拡張と腹臥位での下大静脈の圧迫による静脈還流の低下が最も考えられた。全身性硬化症患者の麻酔においては，血管収縮により循環血液量が減少していることを考慮して十分な補液後に慎重な体位変換が必要であると考えられた。

（キーワード：低血圧，合併症，麻酔，強皮症，腹臥位）