ISSN (O):2395-2822; ISSN (P):2395-2814

Anaesthetic Management of a Case of Necrotising Fascitis with Uncorrected Tetralogy of Fallot with Scoliosis Posted for Debridement.

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Received: August 2019
Accepted: August 2019

ABSTRACT

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease, characterized by aortic overriding, right ventricular hypertrophy, pulmonary stenosis (PS) and ventricular septal defect (VSD). When left untreated, survival to adulthood is rare. There are few case reports of uncorrected TOF surviving to adulthood and then presenting for non-cardiac surgeries. We present a case of debridement of a necrotizing fasciitis under low dose spinal anaesthesia, in a patient of uncorrected TOF with scoliosis and cushingoid features.

Keywords: Tetralogy of Fallot, necrotizing fascitis, scoliosis, low dose spinal.

INTRODUCTION

Congenital heart disease (CHD) is a structural and functional heart disease, which is present at birth. Incidence of CHD is about 8–10/1000 live births worldwide and varies with modern diagnostics.[1] A study conducted between 2011 and 2014 showed the prevalence in India to be as high as 19.4/1000 live births. Common CHDs were Ventricular Septal Defect (VSD) (33%), Atrial Septal Defect (ASD) (19%) and Tetralogy of Fallot (TOF) (16%) in the age group of 0–5 years.[2] Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease, characterized by aortic overriding, right ventricular hypertrophy, pulmonary stenosis (PS) and ventricular septal defect (VSD). When left untreated, survival to adulthood is rare.[3] There are few case reports of uncorrected TOF surviving to adulthood and then presenting for non-cardiac surgeries.[3-5] We present a case of debridement of a necrotizing fasciitis under low dose spinal anaesthesia, in a patient of uncorrected TOF. Necrotising fasciitis, also known as necrotizing soft tissue injury is a progressive, often polymicrobial, bacterial infection of the fascia and surrounding soft tissue.[6] Early emergency surgical debridement is one of the key factors to minimize morbidity and mortality.[7]

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CASE REPORT

A 21-year-old male with cushingoid features got admitted to the hospital with the history of swelling of leg from last 15 days along with blackening and peeling of skin from past 3 days. He was diagnosed to have TOF at birth but did not present for follow up after the initial evaluation. His echocardiogram revealed the presence of TOF with a large conotruncal VSD of 2cms, overriding of the aorta and infundibular pulmonic stenosis. The right ventricular outflow tract (RVOT) gradient was 90mm Hg and had a right-to-left shunt. There was a history of an episode of a cyanotic spell at age of 3 months which was aborted spontaneously followed by multiple similar episodes for which no history of hospitalization was available. No history of seizures, altered sensorium or syncopal attacks. There was history of difficulty in breathing for which he was taking steroids from some quack. There was no history of diabetes mellitus. On admission, he had New York Heart Association class III dyspnea.

Examination revealed pallor, peripheral wasting, grade 4 clubbing in all fingers and toes [Figure 1] and blackening of left lower limb extending to the inguinal ligament. His heart rate was 103/min and blood pressure (BP) was 150/105mm Hg. His

oxygen saturation (SpO2) on room air was 80%. A parasternal heave was present along with a palpable systolic thrill in the left parasternal area. Auscultation revealed a harsh grade 4 ejection systolic murmur, more prominent in the left upper sternal border radiating to all the areas indicating right ventricular outflow tract (RVOT) obstruction. A pansystolic murmur could not be heard probably due to the large size of the VSD. EKG revealed Right Bundle Branch Block (RBBB), Right Ventricular Hypertrophy (RVH) and Right Axis Deviation (RAD) [Figure 3]. Blood investigations carried out included a complete blood picture which showed hemoglobin of 17.5 g/dL, a platelet count of 2.63 lakhs/mm3 and an INR of 1.21. Patient had active upper respiratory tract infection with harsh breath sounds. Chest X-ray revealed boot shaped heart and scoliosis [Figure 4]. Airway assessment showed a Malampati grading (MP) of 3 along with short neck and reduced thyromental distance.

Figure 1: showing clubbing of hands

Figure 2: showing clubbing of toes

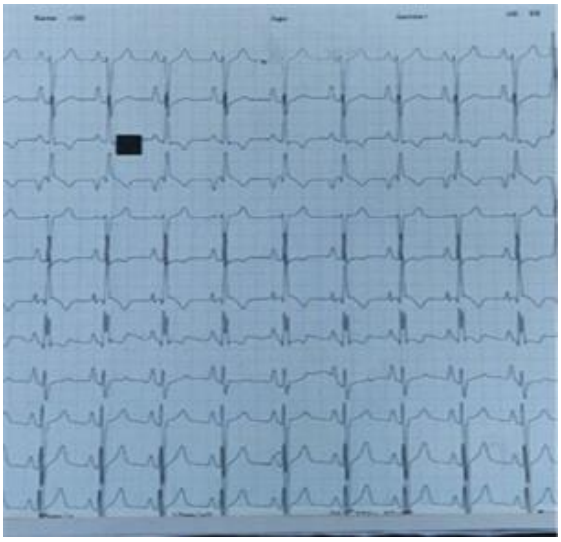

Figure 3: ECG showing RAD, RVH and RBBB

Figure 4: boot shaped heart and scoliosis on chest x-ray

In view of the difficult airway and risk of respiratory infection, we planned a subarachnoid block with low-dose local anesthetic and opioid combination since the coagulation profile was normal. High risk consent was obtained. Defibrillator, difficult airway cart and emergency drugs were kept ready. Phenylephrine was loaded. Standard monitoring was started. A 16-gauge and 18-gauge intravenous cannulation was performed. All intravenous lines were de-aired. Fluid loading of 500ml was done before giving subarachnoid block. Injection vancomycin 600mg was infused over one hour for as prophylaxis for infective endocarditis. After this, background infusion of phenylephrine at 0.75mcg/kg/minute was started which was titrated as per the hemodynamic parameters. Subarachnoid block (SAB) was performed under all aseptic conditions with a 25-gauge Quincke needle using 1.0 ml of injection bupivacaine hyperbaric 0.5% along with 10 µg of injection fentanyl. 4 min after

institution of the spinal anesthetic, a sensory level of T-12 to pin prick was achieved and the debridement was started. The sensory level was re-assessed and found to be at T-10, about 10 min after the subarachnoid block. Intra-operatively, there were no episodes of hypotension or concomitant desaturation.

Post-operatively, the patient was shifted to a high dependency unit, positioned propped up and administered oxygen with FiO2 of 60% via a venturi mask.

DISCUSSION

The natural history of unoperated TOF is variable and depends largely on the degree of obstruction to pulmonary blood flow. One third of untreated infants die in the first year of life. These infants have the most severe obstruction to pulmonary blood flow. Fifty one percent of infants die by the age 3 years and 76 percent by the age of 10 years. Risk of sudden death remains thereafter.[8]

Prolonged survival with uncorrected TOF is associated with a well-developed left ventricle, mild to moderate PS with adaptations such as systemic-pulmonary collaterals, persistent patent ductus arteriosus or systemic hypertension.[4] The longevity of our patient is explained by a large bidirectional VSD.

Right to left shunting leads to poor pulmonary perfusion resulting in chronic hypoxemia and cyanosis.[5] Chronic hypoxemia is associated with polycythemia and abnormal hemostasis, both of which are of concern to the anesthesiologist. As the hematocrit increases, blood viscosity increases dramatically. Hyperviscosity is associated with thromboses of intracranial veins and sinuses, sometimes resulting in stroke. Children under the age of 5 years are at highest risk. Adults with cyanotic CHD appear to be at decreased risk for thrombotic intracranial accidents, even with hematocrits >65%, but instead suffer from a propensity toward intracranial bleeding. The etiology of this age-related difference is unknown. Reported hemostatic abnormalities include thrombocytopenia, platelet dysfunction, hypofibrinogenemia, accelerated fibrinolysis and factor deficiencies.[9]

The perioperative goals are (i) ensure adequate hydration. Patients who are markedly polycythemic should not be allowed to become dehydrated. The duration of the preoperative fasting should be minimal for age, or an intravenous infusion should be started to prevent dehydration. (ii) Care should be taken to prevent infusion of bubbles in patients with right-to-left shunt (iii) maintain systemic arterial blood pressure (iv) minimize additional resistance to pulmonary blood flow and (v) avoid sudden increases in systemic oxygen demand (crying, struggling, inadequate level of anesthesia).[9]

According to latest guidelines of European Society of Cardiology, infective endocarditis prophylaxis is recommended for uncorrected TOF.[10] Vancomycin is the suggested antibiotic for prophylaxis when patient is posted for debridement of skin and subcutaneous tissue.[11] Adequate hydration ameliorates the increased blood viscosity, sludging and thromboembolism.[12] Hypovolemia can exacerbate the right ventricular outflow tract obstruction if the patient has infundibular stenosis. Also in necrotizing fasciitis, the intravascular volume is depleted, thereby necessitating the need for preoperative fluid resuscitation. Therefore a fluid bolus was given before subarachnoid block.

Maintaining higher systemic vascular resistance (SVR) relative to pulmonary vascular resistance (PVR) minimises right-to-left shunting. Hypoxia, hypercarbia and acidosis can cause significant increases in PVR. Hyperventilation without PEEP helps to decrease PVR.

These patients are vulnerable to hypercyanotic spells perioperatively. An intraoperative spell is treated with fluid bolus, deepening the anaesthesia plane, fentanyl, phenylephrine and hyperventilation with 100% oxygen to decrease PVR.

Scoliosis significantly alters the geometry of the chest, and ultimately three-dimensional range of motion of both the thoracic cage and spine during breathing. In addition, the deformity places respiratory muscles at biomechanical disadvantage ultimately leading to decreased compliance of chest wall and a restrictive syndrome that may affect the respiratory function.[13]

The presence of thoracic scoliosis, difficult airway and an active respiratory tract infection could increase the likelihood of respiratory complications and hemodynamic fluctuations had general anesthesia been planned. Hence, we planned a subarachnoid block with low-dose local anesthetic and opioid combination since the coagulation profile was normal. Another important postoperative consideration in these patients is the blunting of the chemoreceptor response to hypoxia. Profound hypoxia can occur without eliciting the normal response of increased ventilation, particularly when respiratory depressants have been given.

Also with general anesthesia any hypoxia, hypercarbia, acidosis or hypothermia resulting therefrom as also the effects of intermittent positive pressure ventilation, could decrease pulmonary blood flow, by increasing pulmonary vascular resistance (PVR). In a patient with an active lower respiratory tract infection, the potential for triggering undesirable airway sequelae, such as a bronchospastic episode during airway manipulation is always there. Any such episode in a cyanotic patient might result in a degree of hypoxemia difficult to manage. Furthermore, a decrease in SVR could also result from the effects of the anesthetic

drugs administered as part of a general anesthetic regimen.

Hence, we opted for a spinal anesthetic with a combination of "low-dose" bupivacaine (5mg) along with fentanyl (10 μg) to minimize the subsequent fall in SVR. A spinal anesthetic in this case, also allowed minimal alterations in ventilation perfusion relationships and did not result in any increase in PVR. Phenylephrine is the vasopressor of choice in this case to counteract any episode of hypotension following SAB as it is pure α1 agonist and maintains BP by increasing SVR without causing tachycardia. Moreover, phenylephrine-induced rise in BP is associated with increased pulmonary blood flow and improved oxygenation in patients with TOF.[14]

CONCLUSION

With the above mentioned strategy of volume loading and low dose phenylephrine infusion, we could manage the right to left shunting effectively, and we did not have any episodes of hypotension and desaturation, resulting from worsening of the shunt.

REFERENCES

1. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol 2002;39:1890-900.
2. Bhardwaj R, Rai SK, Yadav AK, Lakhotia S, Agrawal D, Kumar A, et al. Epidemiology of congenital heart disease in India. Congenit Heart Dis 2015;10:437-46.
3. Thomas SH, Bass P, Pambakian H, Marigold JH. Cyanotic tetralogy of Fallot in a 77 year old man. Postgrad Med J 1987;63:361-2.
4. Sousa P, Santos W, Marques N, Cordeiro P, Ferrinha R, Pereira S, et al. A 72-year-old woman with an uncorrected tetralogy of Fallot presenting with possible pulmonary endocarditis: A case report. J Med Case Rep 2013;7:150.
5. Solanki SL, Jain A, Singh A, Sharma A. Low-dose sequential combined-spinal epidural anesthesia for Cesarean section in patient with uncorrected tetrology of Fallot. Saudi J Anaesth 2011;5:320-2.
6. Tsoa H. Arthritis and musculoskeletal and skin disease. In: Freedberg IM, Eisen AZ, Wolf K, Goldsmith LA, Katz SI, Fitzpatrick TB, eds. Fitzpatrick's Dermatology in General Medicine. 5th ed. New York, NY: McGraw-Hill; 1999:2213-2231.
7. Chapnick EK, Abter EI. Necrotizing soft-tissue infections. Infect Dis Clin North Am. 1996;10:835-855.
8. Lell WA, Pearce FB. Tetralogy of Fallot. In. Lake CL, Booker PD (Eds). Pediatric Cardiac Anaesthesia. 4th ed. Philadelphia: Lippincott Williams & Wilkins; 2005. p 344-356.
9. Frankville D. Children and Adults with Congenital heart disease. In. Lake CL, Booker PD (Eds). Pediatric Cardiac Anaesthesia. 4th ed. Philadelphia: Lippincott Williams & Wilkins; 2005. p 601-632.
10. Wilson W, Taubert KA, Gewitz M, Lockhart PB, Baddour LM, Levison M, et al. American Heart Association. Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. Circulation. 2007;116:1736-1754.
11. Rossi AM, Mariwalla K. Prophylactic and empiric use of antibiotics in dermatologic surgery: a review of the literature and practical considerations. Dermatol Surg. 2012; 38: 1898-921.
12. Goyal R, Singh S, Bangi A, Singh SK. Case series: Dexmedetomidine and ketamine for anesthesia in patients with uncorrected congenital cyanotic heart disease presenting for non-cardiac surgery. J Anaesthesiol Clin Pharmacol 2013;29:543-6.
13. Qiabi M, Chagnon K, Rakovich G. Scoliosis and bronchial obstruction. Can Respir J. 2015 Jul-Aug; 22(4): 206–208.
14. Tanaka K, Kitahata H, Kawahito S, Nozaki J, Tomiyama Y, Oshita S. Phenylephrine increases pulmonary blood flow in children with tetralogy of Fallot. Can J Anaesth 2003;50:926-9.

How to cite this article: Popli S, Gupta P, Verma A, Juneja A, Das N, Arya P. Anaesthetic Management of a Case of Necrotising Fascitis with Uncorrected Tetralogy of Fallot with Scoliosis Posted for Debridement. Ann. Int. Med. Den. Res. 2019; 5(5):AN23-AN26.

Source of Support: Nil, **Conflict of Interest:** None declared