

Case Report

Anaesthetic management of a patient with non compaction cardiomyopathy for implantable cardioverter defibrillator lead replacement

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ABSTRACT

Non compaction cardiomyopathy (NCM) is a rare, primary genetically derived cardiomyopathy with a variable clinical presentation ranging from absence of symptoms to congestive heart failure, systemic thromboembolism, arrhythmias and sudden cardiac death. Being an uncommon condition, the perioperative concerns in a patient with NCM have not been studied much. With increasing awareness and improved diagnostic tools including high resolution echocardiography and cardiac MRI, there has been an increase in the reporting of cases which stresses on the need for a complete understanding of this form of cardiomyopathy and its perioperative anaesthetic management. Authors report the case of a 24 years old female, with NCM who underwent prophylactic Implantable cardioverter defibrillator (ICD) insertion 5 years ago and was now posted for ICD removal and replacement in view of inappropriate ICD functioning.

Keywords: Arrhythmias, Heart failure, Implantable cardioverter defibrillator, Noncompaction cardiomyopathy, Sudden cardiac death

INTRODUCTION

Noncompaction cardiomyopathy (NCM) also known as spongy myocardium or persistent embryonic myocardium or left ventricular non compaction (LVNC) is considered to be a genetically heterogeneous form of cardiomyopathy with a familial and sporadic form.¹ The NCM is a rare disorder, and is thought to occur due to the failure of left ventricle (LV) compaction during the 5th to 8th week of embryogenesis resulting in two layers of myocardium, a compacted one and noncompacted layer.

The clinical presentation is variable. The patient may remain asymptomatic (diagnosed during a family screening) or may present with the typical complications

associated with NCM including heart failure, ventricular arrhythmias, and systemic embolic events which may occur at any age and may cause sudden death.²

CASE REPORT

Authors report the case of a 24year old female, with noncompaction cardiomyopathy (NCM) for ICD lead replacement under general anaesthesia on 02/10/2019. At the age of 18 years, the patient was diagnosed with NCM, when she underwent cardiac screening in view of sudden cardiac death (SCD) of two elder siblings. She was asymptomatic, and on screening was found to have a prolonged QT and NCM with normal LV function on transthoracic echocardiography (TTE). The TTE revealed

noncompaction of both left and right ventricle with prominent trabeculations in left ventricle (LV) and right ventricle (RV) apex and colour flow was seen within the recesses. She was found to have a fair LV function with ejection fraction of 50-55%. The coronaries were found to be normal on CT coronary angiography. Genetic testing was also done to confirm the diagnosis. She underwent prophylactic single chamber St. Jude ICD implantation through an infra-mammary incision as a primary prophylaxis in august 2014 in view of the risk of SCD. The device was implanted beneath the pectoral muscle and was programmed to VVI mode and was started on spironolactone and nicorandil.

Now, she presented with history of palpitations on and off 2-3 times in a week, which resolved on its own after sometime. On evaluation of ICD, it was found that several inappropriate ICD therapies in the form of shocks were delivered due to mis-sensing of the right ventricular ICD lead, due to fracture of the lead.

She was now posted for ICD removal and replacement in view of its inappropriate functioning. She had no other comorbidities. She did not give history suggestive of any associated neuromuscular disease.

There was a risk of avulsion of subclavian vein, tricuspid leaflet damage and cardiac tamponade during the removal of ICD which was explained to the patient and relatives. Authors planned general anaesthesia with real time TEE monitoring to assess cardiac function and to rule out any complication. Also, a cardiac surgeon and perfusionist were standby for the procedure and cardiopulmonary bypass (CPB) pump was primed and kept ready.

On the day of surgery, the baseline heart rate (HR) was noted to be 81 bpm, the blood pressure (BP) was 104/70mmhg and 98% spo2 on room air. All emergency drugs and equipments including defibrillator were kept ready. Anaesthesia was induced with propofol, midazolam and fentanyl. Muscle relaxation was achieved using atracurium to aid tracheal intubation. Right radial artery was cannulated for continuous blood pressure monitoring and TEE probe was inserted. Central venous access was not taken, as femoral vein access was taken by the cardiologist and could be used during any emergency. Anaesthesia was maintained with 50% oxygen and air mixture with sevoflurane at MAC of 0.8-1. Intraoperative monitoring included ECG, spO2, invasive blood pressure (IBP), EtCO2, temperature, TEE, bispectral index (BIS), neuromuscular monitoring (TOF) and urine output. The real time TEE demonstrated increased trabeculation in both LV and RV (LV>RV) with normal ventricular function as shown in Figure 1.

There were no significant hemodynamic changes during the procedure. The ICD was removed under fluoroscopy guidance as shown in Figure 2 with real time TEE confirming no damage to tricuspid valve leaflets or tamponade/ pericardial effusion. The new ICD was

inserted through subclavian vein via an infraclavicular incision and placed in right ventricle (RV) and its position was confirmed under fluoroscopy. It was set on VVI mode. At the end of the procedure, the patient was extubated after adequate reversal and was shifted to ICU for post-operative monitoring. The postoperative course was uneventful and she was discharged on the 7th post operative day and had regular follow up in cardiology department.

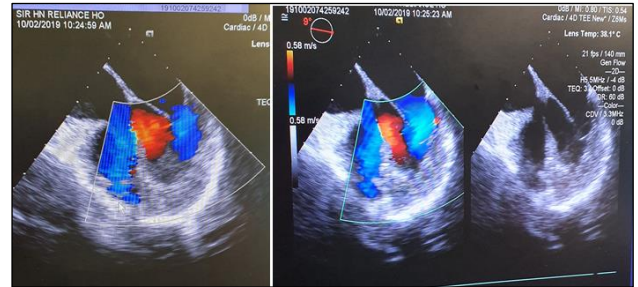


Figure 1: Intraoperative TEE monitoring showing hypertrabeculation in both left and right ventricle with intertrabecular recess in right ventricle communicating with the ventricular cavity.

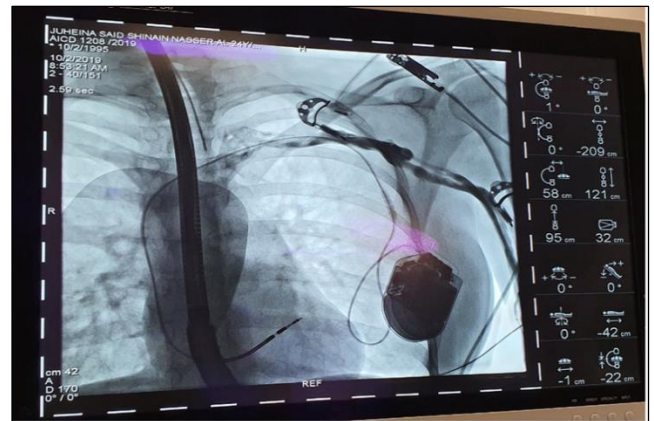


Figure 2: ICD replacement under fluoroscopy guidance with real time TEE to rule out any complications.

DISCUSSION

Noncompaction cardiomyopathy is a rare disorder, and is thought to occur due to the failure of left ventricle (LV) compaction during the 5th to 8th week of embryogenesis resulting in two layers of myocardium, a compacted one and noncompacted layer. It is characterized by the prominent trabeculations with deep intertrabecular recesses in continuity with the ventricular cavity, which are filled with blood from the ventricle and have no communication with the coronary system.³ A decrease in the coronary flow reserve measured by PET-CT is observed in most segments that show ventricular wall motion abnormalities.⁴ Different diagnostic criteria have been described to diagnose NCM over time, but the Jenni criteria is the most frequently used.⁵⁻⁷ Echocardiography

is the first line diagnostic modality with Cardiac magnetic resonance being the best for diagnostic confirmation.¹

Being an uncommon condition, the perioperative concerns and anaesthesia considerations have not been studied in detail. With increasing awareness and improved diagnostic tools including high resolution echocardiography and cardiac MRI, there has been an increase in the reporting of cases which stresses on the need for a complete understanding of this form of cardiomyopathy and its perioperative anaesthetic management.

In children, a variety of congenital cardiac anomalies have been reported together with the finding of non-compacted left ventricular segments, including atrial and ventricular septal defects, aortic coarctation and aortic stenosis.⁸ In a study by the Vienna group, neuromuscular diseases were detected in upto 65% patients with NCM including myotonic dystrophy and was found to be a predictor of adverse prognosis.⁹

The clinical presentation is variable. The patient may remain asymptomatic (diagnosed during a family screening) or may present with the typical complications associated with NCM including heart failure, ventricular arrhythmias, and systemic embolic events which may occur at any age and may cause sudden death. In our case, the patient was diagnosed on a routine family screening in view of SCD in family and had prolonged QT interval on ECG. In a study, by Rosa et al, 34 cases of NCM were identified over 15 years and it was observed that the clinical presentation at the time of diagnosis included dyspnoea in 79% patients, heart failure in 35%, chest pain in 26% and chronic atrial fibrillation in 26% patients. Real time TEE is a useful monitor to assess cardiac function and thromboembolism during general anaesthesia in these patients. Non-invasive cardiac output monitoring devices is another alternative when TEE is not suitable.

Also, patients with NCM are susceptible to develop severe arrhythmias. Any factors that increase sympathetic tone should be avoided and anaesthetic drugs which can provoke arrhythmias should be used with caution. In view of associated neuromuscular diseases, the dose of non-depolarizing neuromuscular blocking agents should be titrated under continuous monitoring of TOF.

CONCLUSION

The anaesthetic implications in a patient with NCM includes assessing the degree of myocardial reserve, risk of arrhythmias, heart failure, thromboembolism and screening for neuromuscular disease in adults and associated congenital heart disease in children. The risk of potential hyperpyrexia or hyperkalaemia with

anaesthesia in certain neuromuscular diseases should also be borne in mind. Attention to any muscular weakness at the time of tracheal extubation is required and neuromuscular blocking agents should be used with caution under continuous monitoring of TOF.

In the best of our knowledge, this is a rare case report and highlights the perioperative concerns in these patients.

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REFERENCES

1. Frentzou GA, Drinkhill MJ, Turner NA, Ball SG, Ainscough JF. Non-compaction cardiomyopathy. *Heart* 2013 May 1;99(2):A138.
2. Oechslin EN, Jost CH, Rojas JR, Kaufmann PA, Jenni R. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. *J Am Coll Cardiol*. 2000 Aug 1;36(2):493-500.
3. Rosa LV, Salemi VM, Alexandre LM, Mady C. Noncompaction cardiomyopathy: a current view. *Arq Bras Cardiol*. 2011 Jul;97(1):e13-9.
4. Jenni R, Wyss CA, Oechslin EN, Kaufmann PA. Isolated ventricular noncompaction is associated with coronary microcirculatory dysfunction. *J Am Coll Cardiol*. 2002;39(3):450-4.
5. Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. *Heart*. 2001;86(6):666-71.
6. Petersen SE, Selvanayagam JB, Wiesmann F, Robson MD, Francis JM, Anderson RH, et al. Left ventricular non-compaction: insights from cardiovascular magnetic resonance imaging. *J Am College of Cardiol*. 2005 Jul 5;46(1):101-5.
7. Kohli SK, Pantazis AA, Shah JS, Adeyemi B, Jackson G, McKenna WJ, et al. Diagnosis of left-ventricular non-compaction in patients with left-ventricular systolic dysfunction: time for a reappraisal of diagnostic criteria?. *Eur Heart J*. 2008 Jan 1;29(1):89-95.
8. Pantazis AA, Elliott PM. Left ventricular noncompaction. *Curr Opin Cardiol*. 2009;24(3):209-13.
9. Maron BJ, Towbin JA, Thiene G. Contemporary definitions and classification of the cardiomyopathies. *Circulation* 2006;113:1807-16.

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