JARSS 2019;27(1):66-69
doi: 10.5222/jarss.2019.77487

Successful Anesthetic Management of a 14 Year Old Boy Undergoing Tricuspid Valvular Surgery for Carcinoid Heart Disease

Aynur Camkıran Fırat
Murat Özkan
İlkay Erdoğan
Pınar Zeyneloğlu

Karsinoid Kalp Hastalığı Nedeniyle Triküspid Kapak Replasmanı Yapılan 14 Yaşındaki Erkek Çocuğunda Başarılı Anestezi Yönetimi

ABSTRACT

Objective: *We present our strategy for successful management of pediatric patients with carcinoid heart disease when weaning from cardiopulmonary bypass (CPB) during tricuspid valve surgery.*
Case Presentation: *The patient was a 14-year-old boy who had been diagnosed with a grade IV neuroendocrine tumor. The patient was premedicated with oral midazolam, hydroxyzine, somatostatin analogue (Octreotide®), and pheniramine maleate. Octreotide® and serotonin 5-hydroxytryptamine receptor antagonist were also added during the rewarming period. Dopamine was continued as an infusion (5 μg kg^{-1} min^{-1}) for the first day. Bioprosthetic valve placement and transannular right ventricular outflow tract patch augmentation were performed. He was extubated within 20 hours and discharged from the hospital at postoperative 6 day.*
Discussion: *Thanks to the premedication of this case and the addition of somatostatin analogue, pheniramine maleate and methylprednisolone to the pump reservoir and avoiding agents such as atracurium, morphine and meperidine, which may cause histamine release, the surgery was accomplished have been accomplished without development of carcinoid crisis.*

Keywords: *Carcinoid heart disease, carcinoid crisis, cardiopulmonary bypass, anesthesia, paraneoplastic syndrome*

ÖZ

Amaç: *Bu olgu sunumunda karsinoid kalp tutulumu nedeniyle triküspid kapak replasmanı yapılan çocuk hastada kardiyopulmoner baypastan (KPB) ayrılma sırasında uygulanan strateji anlatılmaktadır.*
Olgu Sunumu: *On dört yaşında olan hastamızın evre IV karaciğer metastazı olan nöroendokrin tümörü vardı. Hastaya premedikasyon amacıyla peroral verilen midazolam ve hidroksizinin yanına somatostatin analogu (Octreotid®) ve feniramin maleat ilave edildi. Octreotid® ve 5-hidroksitriptamin reseptör antagonisti aynı zamanda pompa rezervuarına da yapıldı ve ısınma döneminde doz yinelendi. Triküspid kapak yerine biyolojik kapak yerleştirildi ve pulmoner çıkıma krosanüler yama koyuldu. Postoperatif ilk gün 5 μkg^{-1} dk^{-1} dopamin infüzyonu devam etti. Cerrahi sonrası 20. saatte ekstübe edilen hasta, postoperatif 6. gün taburcu oldu.*
Tartışma: *Bu olgunun premedikasyonuna ve pompa rezervuarına somatostatin analogu, feniramin maleat ve metilprednizolon ilave edilmesi ve histamin salımınına neden olabilecek atrakuryum, morfin ve meperidin gibi ajanlardan kaçınılması ile karsinoid kriz gelişmeden cerrahinin tamamlanması sağlanmıştır.*

Anahtar kelimeler: *Karsinoid kalp hastalığı, karsinoid kriz, kardiyopulmoner baypas, anestezi, paraneoplastik sendrom*

Alındığı tarih: 23.11.2018
Kabul tarihi: 26.12.2018
Yayın tarihi: 31.01.2019

Aynur Camkıran Fırat
Başkent Üniversitesi Tıp Fakültesi,
Anesteziyoloji Anabilim Dalı,
Ankara, Türkiye
acamkiran@gmail.com
ORCİD: 0000-0003-1470-7501

M. Özkan 0000-0003-3991-8479
Başkent Üniversitesi Tıp Fakültesi,
Kalp Damar Cerrahisi Anabilim Dalı,
Ankara, Türkiye

İ. Erdoğan 0000-0003-0964-1528
Başkent Üniversitesi Tıp Fakültesi,
Pediyatrik Kardiyoloji Anabilim Dalı,
Ankara, Türkiye

P. Zeyneloğlu 0000-0003-2312-9942
Başkent Üniversitesi Tıp Fakültesi,
Anesteziyoloji Anabilim Dalı,
Ankara, Türkiye

INTRODUCTION

Carcinoid tumors derive from neuroendocrine cells that are capable of metastasis, and these slow-growing neoplasms cause the release of vasoactive amines, such as serotonin, histamine and quinine peptides [(1)]. These bioactive substances are not metabolized in the liver, and they enter the systemic circulation directly and bring about hemodynamic instability.

Carcinoid syndrome is a paraneoplastic syndrome that includes heart failure. The classical presentation of carcinoid heart disease is right heart failure, which results from a right-sided valvular (tricuspid and pulmonic) lesion caused by fibrous reaction [(2)]. Catecholamines and histamine-releasing drugs may trigger a carcinoid crisis. This can occur during induction of anesthesia, and it is characterized by severe flushing, cardiac arrhythmias and bronchoconstriction [(3)].

The most important causes of perioperative mortality in this patient group are carcinoid crisis and right ventricular heart failure. There is no clear consensus about medical therapies for weaning these patients from cardiopulmonary bypass (CPB) or during anesthesia. Here we present our strategy for successful management of pediatric patients with carcinoid heart disease when weaning from CPB during tricuspid valve surgery.

CASE PRESENTATION

The patient was a 14-year-old boy (23 kg, 146 cm) who had been diagnosed with a grade IV neuroendocrine tumor (well-differentiated, non-responsive to chemotherapy and Ki-67 proliferative index 1%) and liver metastases. He had right ventricular dysfunction due to carcinoid heart disease and presented with shortness of breath. Transthoracic echocardiography revealed severe pulmonary stenosis, dysplastic tricuspid valve, severe tricuspid regurgitation (4/4), loss of leaflet coaptation, abnormal interventricular septal motion, left deviation of the interventricular septum, and heart failure (Fig. 1). The tricuspid valve annulus measured 28 mm^2. Abdominal ultrasonography showed hepatomegaly and splenomegaly, with multiple images suggesting necrotic sites in the liver: an 8.4x5.2x4.5 cm area in the left lobe and a 6.3x5.6x5.7 cm area in the right lobe.

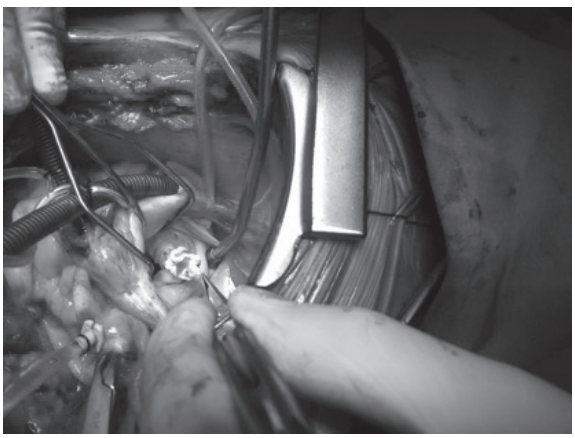

Figure 1. Dysplastic tricuspid valve.

The patient was transported to our center on the day before surgery, so we performed only echocardiography additionally. During preoperative period his medications included a mammalian-target-of-rapamycin inhibitor (Everolimus® 5 mg day^{-1} orally) and a somatostatin analogue (Octreotide® 100 µg b.i.d. subcutaneously). Before the surgery the patient fasted for 6 hours and he was premedicated with oral midazolam 0.5 mg kg^{-1}, hydroxyzine 1 mg kg^{-1}, Octreotide® 100 µg, and pheniramine maleate (Avil®, 45.5 mg). Anesthesia was performed using a combination of high-dose narcotics (fentanyl 50 mg kg^{-1} bolus, 10 mg kg^{-1}; infusion for one hour), a benzodiazepine (midazolam), an inhalation agent (sevoflurane), and a neuromuscular blocker (vecuronium). An indwelling radial artery catheter was placed and standard monitoring was carried out as recommended by the American Society of Anesthesiologists. A central venous catheter was inserted through the right internal jugular vein and a transesophageal echocardiographic (TOE) probe was inserted to monitor valvular and ventricular function, and to reveal any intraoperative hemodynamic problems, such as excessive fluid administration.

Methylprednisolone (Prednol® 10 mg kg^{-1}) was added to the prime solution. Octreotide® 100 µg and serotonin 5-hydroxytryptamine 3 receptor antagonist (Emetril® 1.2 mg) were also added during the rewarming period. Once CPB was achieved, the right, and left ventricular pressures were 37 mmHg and 74 mmHg, respectively. CPB time was 117 minutes. Arterial blood gas analyzer was used to measure blood-glucose concentration during operation. Our hemodynamic parameters were stable, we didn't use bolus vasoactive medication during the operation. We used only dopamine infusion. Dopamine was then initiated at 7 µg kg^{-1} min^{-1}, and was continued as an infusion (5 µg kg^{-1} min^{-1}) for the first postoperative day and 4 µg kg^{-1} min^{-1} for the second day. Bioprosthetic valve placement and transannular right ventricular outflow tract patch augmentation were performed. Two units erythrocyte suspension and

two units fresh frozen plasma were delivered during operation.

Once the operation was completed, the patient was transferred to the intensive care unit. In the postoperative period, TOE revealed severe pulmonary regurgitation and minor pleural effusion. He was extubated within 20 hours and discharged from the hospital on postoperative 6th day. The patient's parents read this case description and consented to its publication.

DISCUSSION

Perioperative management of patients with carcinoid heart disease who require cardiac surgery is challenging because of the multiple processes involved, including carcinoid activity, heart failure, and CPB. All of these can cause perioperative complications, including death. Reported perioperative mortality rates for this patient group range from 7% to 16% [4]. Several different strategies have been described for perioperative anesthesia management in these cases, and most of them involve Octreotide® administration.

Our patient had been admitted to another hospital for severe tricuspid regurgitation, and he was transferred to our hospital only 1 day before the surgery. While urinary 5-hydroxyindoleacetic acid is known to be a good marker for carcinoid tumor activity, we were unable to test for this due to lack of time. Our patient's right heart failure was associated with severe symptomatic functional limitations.

Carcinoid crisis is a life-threatening form of carcinoid syndrome that can be provoked pharmacologically through administration of thiopental, atracurium, succinylcholine, meperidine or morphine. Emotional stress can also cause carcinoid crisis. Besides, patients who undergo cardiac surgery always receive vasoactive medications, especially once CPB has been achieved, and these drugs can also trigger such an event. Given that Octreotide® acts on somatostatin receptors to decrease the secretion of vasoactive amines and peptides from carcinoid tumor cells, this agent is very effective at preventing carcinoid crises [3,5].

Pericardial and pleural effusions and wound-healing complications reported for a mammalian-target-of-rapamycin inhibitor after cardiac transplantation, but we didn't see these complications [6]. A prolonged QT interval has been observed with Octreotide use [7], but we didn't see this complication.

Both carcinoid activity and cardiac disease cause hemodynamic instability. Medical therapies may resolve or relieve symptoms of carcinoid syndrome. Hemodynamic instability in patients with carcinoid heart disease should be treated with a somatostatin analogue, an antihistamine (pheniramine maleate), methylprednisolone and a serotonin 5-hydroxytryptamine3 receptor antagonist. Our pediatric patient was hemodynamically stable throughout his surgical procedure. It can be difficult to wean any patient from CPB without vasoactive drugs. In our case of pediatric carcinoid heart disease, we chose to administer dopamine during CPB weaning and we added Octreotide® and Emetril® during rewarming. In addition, we monitored cardiac function via TOE to regulate dopamine administration and assess fluid needs.

CONCLUSION

This case report highlights the importance of careful anesthetic management for preventing carcinoid crisis in pediatric patients. Appropriate premedication with midazolam and hydroxyzine is recommended, as this can prevent emotional stress, induction can be carried out safely using a combination of fentanyl, midazolam and vecuronium. Drugs that cause histamine release should be avoided and weaning from CPB should be done with care, cardiac function should be monitored via TOE, and vasoactive agent administration must be timed carefully to avoid hemodynamic instability.

REFERENCES

1. Patel C, Mathur M, Escarcega RO, et al. Carcinoid heart disease: Current understanding and future directions. Am Heart J. 2014;167:789-95. https://doi.org/10.1016/j.ahj.2014.03.018
2. Toby N, Weingarten TN, Abel MD, et al. Intraoperative Management of Patients with Carcinoid Heart Disease Having Valvular Surgery: A Review of One Hundred Consecutive Cases. Anesth Analg. 2007;105:1192-9. https://doi.org/10.1213/01.ane.0000284704.57806.0b

3. Dobson R, Burgess MI, Pritchard DM, et al. The clinical presentation and management of carcinoid heart disease. International Journal of Cardiology. 2014;173:29-32.
https://doi.org/10.1016/j.ijcard.2014.02.037
4. Suphathamwit A, Dhir A, Dobkowski W, et al. Successful hepatectomy using venovenous bypass in a patient with carcinoid heart disease and severe tricuspid regurgitation. J Cardiothorac Vasc Anesth. 2016;30:446-51.
https://doi.org/10.1053/j.jvca.2015.05.061
5. Castillo JG, Filsoufi F, Adams DH, et al. Management of patients undergoing multivalvular surgery for carcinoid heart disease: the role of the anaesthetist. Br J Anaesth. 2008;101:618-26.
https://doi.org/10.1093/bja/aen237
6. Bouzas-Mosquera A, Crespo-Leiro MG, Paniagua MJ, et al. Adverse effects of mammalian target of rapamycin inhibitors during the postoperative period after cardiac transplantation. Trans Proc. 2008;40:3027-30.
https://doi.org/10.1016/j.transproceed.2008.09.021
7. Celik N, Cinaz P, Emeksiz HC, et al. Octreotide-Induced Long QT Syndrome in a Child with Congenital Hyperinsulinemia and a Novel Missense Mutation (p. Met115Val) in the ABCC8 Gene. Horm Res Paediatr. 2013;80:299-303.
https://doi.org/10.1159/000354666