Can J Anesth/J Can Anesth (2012) 59:1048–1051
DOI 10.1007/s12630-012-9780-y

CASE REPORTS/CASE SERIES

Case report: Giant cardiac malignancy in a nine-year-old female
Étude de cas: tumeur cardiaque géante chez une fillette de neuf ans

John P. Scott, MD · Todd J. Troshynski, MD · William R. Clarke, MD · Ronald K. Woods, MD, PhD · David R. Lal, MD · Richard J. Berens, MD · Richard L. Tower, MD

Received: 20 July 2012 / Accepted: 23 August 2012 / Published online: 31 August 2012

Abstract

Purpose *To describe the management of a giant cardiac malignancy initially diagnosed as an anterior mediastinal mass.*

Clinical features *A nine-year-old female with right facial swelling and chronic cough was diagnosed with a large right mediastinal mass. Intermittent ventricular and supraventricular arrhythmias were noted on admission electrocardiograms. Empiric corticosteroid and radiation therapy did not reduce the size of the tumour, and initial tissue biopsies were non-diagnostic. Due to worsening tamponade physiology and persistent arrhythmias, the patient was scheduled for tumour debulking with potential resection. Prior to surgery, a multidisciplinary team was assembled to delineate team member responsibilities and treatment algorithms.*

The procedure was performed under general anesthesia with spontaneous ventilation preserved during endotracheal intubation and invasive line placement. The team was prepared to provide extracorporeal mechanical support if needed. The child required inotropic and vasoactive medications after transitioning to positive pressure ventilation, but her hemodynamics improved with sternotomy. The lesion was identified as a malignant cardiac clear-cell tumour that was unresectable. Her sternum was left open, as attempted closure led to the re-creation of tamponade physiology. She underwent delayed sternal closure days later. After months of chemotherapy that resulted in significant tumour involution, she underwent successful surgical resection.

Conclusion *Giant primary cardiac tumours may present similarly to large anterior mediastinal masses. The care of patients with these lesions requires an understanding of the risks associated with mediastinal masses as well as those unique to cardiac tumours. A multidisciplinary approach is critical to providing safe and effective care throughout this process.*

Author contributions *John P. Scott, Todd J. Troshynski*, *William R. Clarke*, *Ronald K. Woods*, *David R. Lal*, *Richard J. Berens*, and *Richard L. Tower* participated in direct patient care, construction of the care plan, and writing and reviewing the manuscript.

J. P. Scott, MD (✉) · T. J. Troshynski, MD · W. R. Clarke, MD · R. J. Berens, MD
Section of Pediatric Anesthesia, Department of Anesthesiology, Medical College of Wisconsin, Children's Hospital of Wisconsin, 9000 W. Wisconsin Avenue, PO Box 1997/MS735, Milwaukee, WI 53201, USA
e-mail: Scottjake@mcw.edu

J. P. Scott, MD · R. J. Berens, MD
Section of Pediatric Critical Care, Department of Pediatrics, Medical College of Wisconsin, Children's Hospital of Wisconsin, Milwaukee, WI, USA

R. K. Woods, MD, PhD
Section of Pediatric Cardiothoracic Surgery, Department of Cardiothoracic Surgery, Medical College of Wisconsin, Children's Hospital of Wisconsin, Milwaukee, WI, USA

D. R. Lal, MD
Section of Pediatric Surgery, Department of Surgery, Medical College of Wisconsin, Children's Hospital of Wisconsin, Milwaukee, WI, USA

R. L. Tower, MD
Section of Pediatric Oncology, Department of Pediatric, Medical College of Wisconsin, Children's Hospital of Wisconsin, Milwaukee, WI, USA

Résumé

Objectif *Décrire la prise en charge d'une tumeur cardiaque géante initialement diagnostiquée comme étant une masse médiastinale antérieure.*

Caractéristiques cliniques *Un diagnostic de volumineuse masse médiastinale droite a été fait chez une fillette âgée de neuf ans, présentant une enflure de la partie droite du visage*

et une toux chronique. Des épisodes intermittents d'arythmie ventriculaire et supraventriculaire ont été notés sur les électrocardiogrammes à l'admission. Un traitement empirique par corticostéroïdes et radiothérapie n'a pas réduit la taille de la tumeur. Les biopsies tissulaires initiales n'avaient pas permis le diagnostic. Compte tenu de l'aggravation de la symptomatologie de tamponnade et des troubles du rythme persistants, une réduction de taille de la tumeur, avec éventuelle résection, a été programmée. Une équipe multidisciplinaire a été réunie avant la chirurgie pour définir les responsabilités des membres de l'équipe et les algorithmes thérapeutiques.

L'intervention a été réalisée sous anesthésie générale et la ventilation spontanée a été préservée pendant l'intubation endotrachéale et la mise en place des tubulures invasives. L'équipe était prête, le cas échéant, à fournir un soutien mécanique extracorporel. La patiente a nécessité des médicaments inotropes et vasoactifs après le passage à une ventilation à pression positive, mais son état hémodynamique s'est amélioré avec la sternotomie. La lésion a été identifiée comme étant une tumeur maligne à cellules claires du cœur impossible à réséquer. Son sternum a été laissé ouvert, car la tentative de fermeture entraînait la réapparition de la symptomatologie de tamponnade. La fermeture sternale a été retardée et réalisée plusieurs jours plus tard. Après plusieurs mois de chimiothérapie, la tumeur a présenté une involution significative; il a alors été possible de réséquer chirurgicalement la tumeur.

Conclusion *Les tumeurs géantes cardiaques primitives peuvent ressembler à de grandes masses médiastinales antérieures. La prise en charge de patients présentant de telles lésions nécessite la connaissance des risques associés aux masses médiastinales, ainsi que les risques spécifiques aux tumeurs cardiaques. Une approche multidisciplinaire est essentielle pour assurer des soins sécuritaires et efficaces tout au long de ce processus.*

Space occupying lesions of the mediastinum may impose a significant risk for cardiorespiratory compromise. Typically, these scenarios are associated with anterior mediastinal masses. Large cardiac tumours located within the middle mediastinal space pose similar and additional challenges. We present the management of a giant primary cardiac malignancy in a nine-year-old female who was initially diagnosed with an anterior mediastinal mass. Informed parental consent was obtained for publication of this case report.

Case report

The patient presented primarily for new onset right-sided facial swelling, which, with further history, was associated with a six-month history of cough, wheezing, and dyspnea on exertion. The initial chest *x-ray* revealed a large right-sided thoracic lesion (Fig. 1), and a chest computed tomography scan showed a large mass in the right chest with compression of the right atrium, right ventricle, aortic root, superior vena cava, inferior vena cava, right main pulmonary artery, right pulmonary veins, and right mainstem bronchus (Fig. 2). Transthoracic echocardiogram (TTE) confirmed these findings and showed a Doppler pattern consistent with tamponade physiology. The results of these studies were thought to be consistent with an anterior mediastinal mass, such as a thymoma, lymphoma, or a germ cell tumour.

Admission electrocardiograms revealed dysrhythmias, including non-sustained ventricular and supraventricular tachycardias (Fig. 3). The patient, who noted no difference in her condition, initially tolerated these. Antiarrhythmic therapy was started with minimal improvement. Given the size of the tumour and the anticipated risks associated with sedation, empiric corticosteroid and radiation therapy were administered to decrease the size of the tumour before tissue diagnosis could be made. This strategy led to minimal tumour involution with no change in chest *x-ray* or biomarkers of tumour lysis syndrome.

In order to make a tissue diagnosis, needle tissue biopsy, pleural tap, and bone marrow aspiration were performed. For these procedures, the patient received a balanced anesthetic consisting of intravenous midazolam, inhaled sevoflurane, and supplemental local anesthetic while spontaneous ventilation was maintained with the patient in the semi-fowler's position. There were no complications.

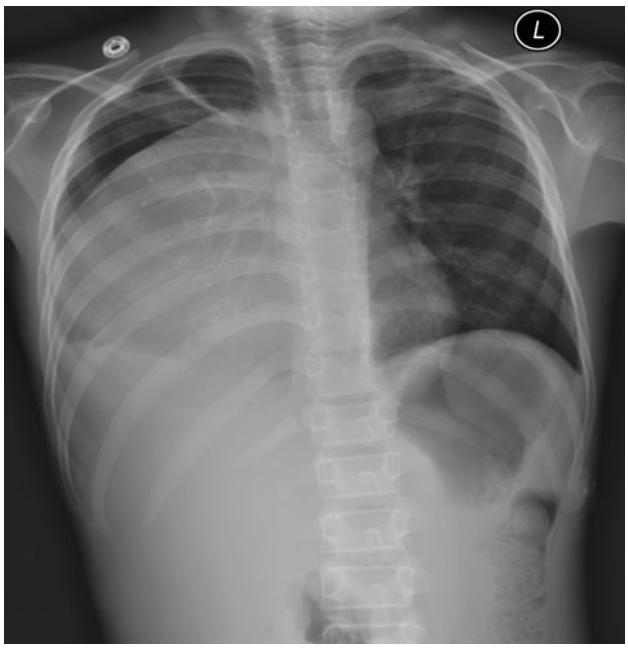

Fig. 1 Anteroposterior chest *x-ray* showing a large right-sided chest mass

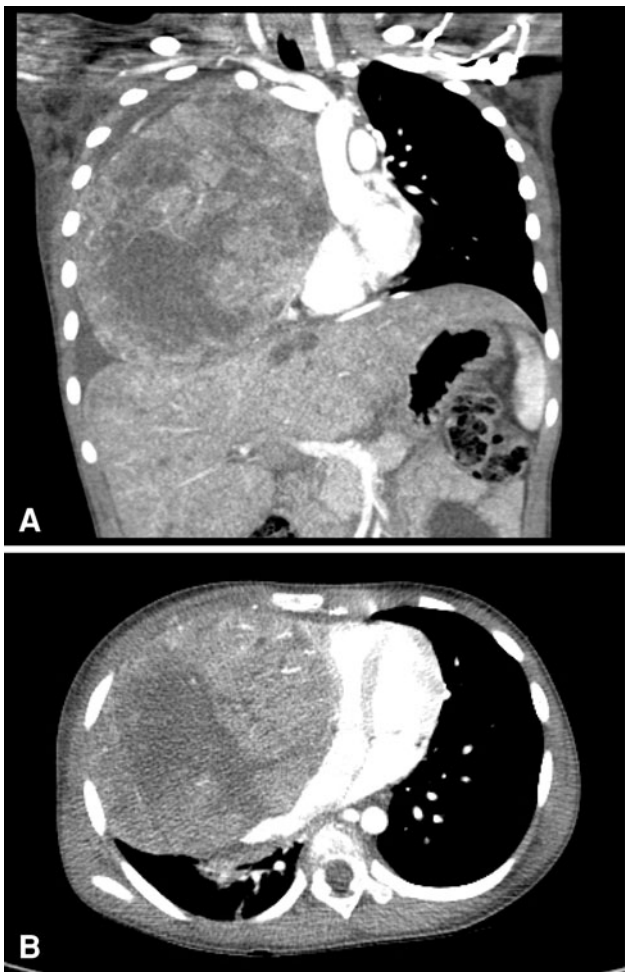

Fig. 2 Coronal (A) and axial (B) views of a computed tomography scan of the chest showing significant compression of the heart and mediastinal structures

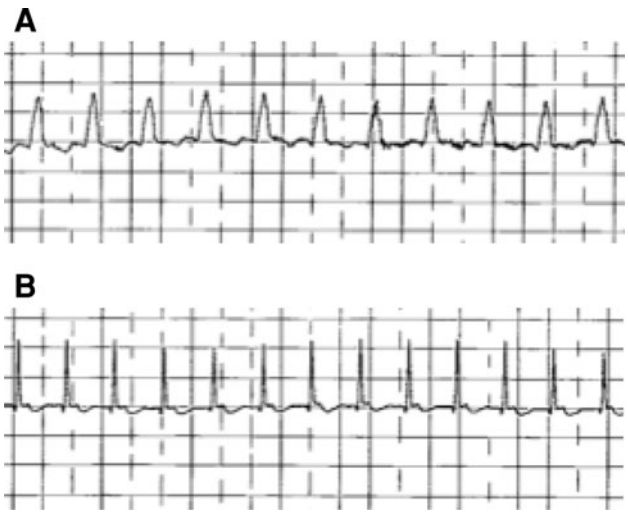

Fig. 3 (A) Rhythm strip illustrating monomorphic ventricular tachycardia. (B) Supraventricular tachycardia

Unfortunately, these studies were non-diagnostic. The patient continued to have dysrhythmias despite medical management and exhibited worsening tamponade physiology. Thus, after a multidisciplinary care conference, including oncology, intensive care, cardiothoracic surgery, general surgery, and anesthesiology, the patient and her family agreed to mediastinal exploration for tumour debulking and potential resection. Prior to this procedure, the multidisciplinary team met to designate team member responsibilities and to formulate procedural plans and treatment algorithms in order to maximize efficiency and reduce error.

The patient was transferred to the operating room with monitoring, including electrocardiogram, pulse oximetry, noninvasive blood pressure, and two-site (cerebral and somatic) near-infrared spectroscopy. She was placed in the semi-fowler's position. Prior to induction, her oropharynx was topicalized with Cetocaine® spray. Anesthesia was induced with intravenous midazolam and inhaled sevoflurane while maintaining spontaneous ventilation. An arterial line was placed after induction. With spontaneous ventilation preserved, nasal fibreoptic intubation was performed, followed by placement of a left subclavian central venous line. Anesthesia was maintained with sevoflurane and fentanyl as the cardiovascular surgical team dissected out the innominate artery and femoral vein for potential cannulation in the event that extracorporeal mechanical support was required. This procedure was performed prior to the introduction of positive pressure ventilation due to a concern for potential circulatory collapse during the transition from negative to positive pressure ventilation.

Positive pressure ventilation was methodically introduced as the anesthesiologist manually assisted with ventilation. A neuromuscular blocking agent was administered only after controlled positive pressure ventilation was successfully implemented. During this transition, epinephrine and norepinephrine infusions were initiated to maintain adequate cardiac output and perfusion. Surgery commenced and the patient's hemodynamics improved during sternotomy as tamponade was relieved. The pericardium was opened, but debulking of the tumour was aborted as no tissue plane was identifiable between the mass and the heart. The lesion appeared to be a primary cardiac tumour. Additional tissue biopsies confirmed the diagnosis of a malignant cardiac clear-cell tumour with features of myogenic differentiation. The sternum was left open, as attempted closure led to the re-creation of tamponade physiology. The child remained intubated and mechanically ventilated post-operatively, and underwent delayed sternal closure at a later date. She was extubated thereafter and eventually discharged. She received months of chemotherapy resulting in significant tumour involution. She has

since undergone successful surgical resection and cryoablation of the mass and is doing well at this time.

Discussion

The presenting signs and symptoms of both benign and malignant primary cardiac tumours are diverse and vary according to tumour size and location.[1-3] Patients with small tumours may be asymptomatic at diagnosis. However, large tumours may be associated with profound derangements in cardiovascular and respiratory physiology. Direct myocardial invasion may result in dysrhythmias, valvular insufficiency, dynamic outflow tract obstruction, congestive heart failure, and cardiac tamponade.[3,4] Vascular obstruction may lead to superior vena cava syndrome, hepatic congestion, ascites, and lower extremity edema.[1-3,5] Respiratory symptoms, such as cough, stridor, or wheezing may occur secondary to external tracheobronchial compression.

Diagnosis of cardiac tumours is typically confirmed with TTE, [5] and a computed tomography scan should be performed to assess for compression of other vital structures within the patient's neck and chest. Early diagnosis of malignant primary cardiac tumours is critical, as these lesions tend to respond poorly to chemotherapy and adjuvant therapies.[5-7] Complete surgical excision tends to be the best curative measure.[5,8] Unfortunately, malignant primary cardiac tumours have high mortality rates, as advanced disease is present at diagnosis in most cases.[5-8]

The anesthetic considerations for patients with large cardiac tumours relate to risks associated with mediastinal masses and risks unique to cardiac tumours. As with anterior mediastinal masses, large cardiac tumours create significant mass effect on vital cardiovascular and respiratory structures, increasing the risk of cardiorespiratory compromise with the induction of anesthesia. Positioning patients in the semi-fowler's or right lateral decubitus position helps to minimize extrinsic airway or vascular compression.[9] Maintenance of spontaneous ventilation during induction of anesthesia is recommended, as the greatest risk of compromise occurs during the transition from negative pressure to positive pressure ventilation.[9] In instances of cardiorespiratory collapse, rigid bronchoscopy and/or extracorporeal mechanical support may be necessary resuscitative interventions.[9]

The anesthesiologist must be cognizant of risks unique to primary cardiac tumours. As with our patient, primary cardiac tumours are a substrate for generation of malignant arrhythmia, which may be medically refractory and increase the risk of sudden cardiac death.[4] Antiarrhythmic medications and defibrillation should be readily available.[4] Preload augmentation is essential to mitigate dynamic outflow tract obstruction or tamponade. Chronotropic agents should be used judiciously as these may be arrhythmogenic and may potentially worsen dynamic outflow tract obstruction. Finally, when the tumour extends into the endocardium itself, it may embolize spontaneously or during dissection.[6] In these instances, the use of transesophageal echocardiography (TEE) and/or transcranial Doppler may be useful supplemental monitors.

In this example, a giant cardiac malignancy was initially diagnosed and treated as a large anterior mediastinal mass; however, the lesion proved to be something more insidious, as was suggested by the persistent dysrhythmias. The anesthetic considerations of this rare lesion are more diverse and complex than those of an isolated anterior mediastinal mass. Given the significant anesthetic and surgical risks, a multidisciplinary approach is essential to the creation of a safe and effective treatment plan for any such patient.

Competing interests None declared.

References

1. *Becker AE*. Primary pediatric heart tumors in the pediatric age group: a review of salient pathological features relevant for clinicians. Pediatr Cardiol 2000; 21: 317-23.
2. *Vander Salm TJ*. Unusual primary tumors of the heart. Semin Thorac Cardiovasc Surg 2000; 12: 89-100.
3. *Maraj S*, *Pressman GS*, *Figueredo VM*. Primary cardiac tumors. Int J Cardiol 2009; 133: 152-6.
4. *Myers KA*, *Wong KK*, *Tipple M*, *Sanatani S*. Benign cardiac tumours, malignant arrhythmias. Can J Cardiol 2010; 26: e58-61.
5. *Verspoor FG*, *van Swieten HA*. Delays in diagnostics and treatment of cardiac tumors are unacceptable. Int J Cardiol 2011; 147: 157-8.
6. *Centofanti P*, *Di Rosa E*, *Deorsola L*, *et al*. Primary cardiac tumors: early and late results of surgical treatment in 91 patients. Ann Thorac Surg 1999; 68: 1236-41.
7. *Perchinsky MJ*, *Lichtenstein SV*, *Tyers GF*. Primary cardiac tumors: forty years' experience with 71 patients. Cancer 1997; 79: 1809-15.
8. *Simpson L*, *Kumar SK*, *Okuno SH*, *et al*. Malignant primary cardiac tumors: review of a single institution experience. Cancer 2008; 112: 2440-6.
9. *Hammer GB*. Anaesthetic management for the child with a mediastinal mass. Paediatr Anaesth 2004; 14: 95-7.