Can J Anesth/J Can Anesth (2009) 56:604–608
DOI 10.1007/s12630-009-9118-6

CASE REPORTS/CASE SERIES

Hemodynamic collapse under anesthesia in a patient with pulmonary artery sarcoma

Collapsus hémodynamique sous anesthésie chez une patiente souffrant d'un sarcome de l'artère pulmonaire

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Received: 5 February 2009 / Accepted: 8 May 2009 / Published online: 23 May 2009

Abstract

Purpose *To describe a case of acute right ventricular dysfunction and hemodynamic collapse under general anesthesia in a patient undergoing resection of a pulmonary artery sarcoma.*

Clinical features *A 67-yr-old woman with a presumptive diagnosis of pulmonary embolism presented to the hospital with progressive shortness of breath and syncope despite therapeutic anticoagulation. An echocardiogram revealed a large mass in the main pulmonary artery and a right ventricular systolic pressure of 105 mmHg. She was referred to our centre for urgent surgical management. One hour following induction of general anesthesia, the patient sustained sudden hemodynamic collapse. Transesophageal echocardiography revealed massive right ventricular dilation, effectively producing tamponade of the left ventricle. Urgent pericardiotomy was performed with immediate hemodynamic recovery. After initiation of cardiopulmonary bypass, the patient was found to have a large pulmonary artery sarcoma involving the pulmonary valve. Following successful resection with grafting and pulmonary valve replacement, the patient's trachea was extubated the following day, and she was discharged from hospital 1 week later in satisfactory condition.*

Conclusions *Pulmonary artery sarcomas pose rare and unique challenges to the anesthesiologist. Given the high perioperative mortality, careful monitoring for catastrophic acute cor pulmonale is crucial. Urgent pericardiotomy or cardiopulmonary bypass for sudden hemodynamic collapse may be life-saving components of intraoperative management.*

Résumé

Objectif *Décrire un cas de dysfonction aiguë du ventricule droit et de collapsus hémodynamique sous anesthésie générale chez une patiente subissant une intervention de résection d'un sarcome de l'artère pulmonaire.*

Éléments cliniques *Une femme de 67 ans chez qui le diagnostic présomptif d'une embolie pulmonaire avait été posé s'est présentée à l'hôpital avec une histoire de dyspnée progressive et de syncope malgré un traitement anticoagulant. Une échocardiographie a révélé la présence d'une importante masse dans l'artère pulmonaire principale et une pression systolique du ventricule droit de 105 mmHg. Elle a été référée à notre centre pour une prise en charge chirurgicale d'urgence. Une heure après l'induction de l'anesthésie générale, la patiente a soudainement subi un collapsus hémodynamique. L'échocardiographie transoesophagienne a révélé une dilatation massive du ventricule droit provoquant la tamponnade du ventricule gauche. Un drainage péricardique d'urgence a été réalisé, résultant en une récupération hémodynamique immédiate. Après l'amorçage de la circulation extra-corporelle, il a été découvert que la patiente souffrait d'un important sarcome de l'artère pulmonaire affectant la valve pulmonaire. Après une résection réussie accompagnée d'un pontage et du remplacement de la valve pulmonaire,*

Electronic supplementary material The online version of this article (doi:10.1007/s12630-009-9118-6) contains supplementary material, which is available to authorized users.

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l'extubation trachéale de la patiente a eu lieu le lendemain; elle a reçu son congé de l'hôpital une semaine plus tard dans un état satisfaisant.

Conclusion *Pour l'anesthésiologiste, les sarcomes de l'artère pulmonaire sont une source de défis rares et singuliers. En raison de la mortalité périopératoire élevée liée à cette condition, un monitorage attentif surveillant l'occurrence d'un cœur pulmonaire aigu catastrophique est essentiel. Un drainage péricardique ou une circulation extra-corporelle d'urgence pour traiter un collapsus hémodynamique soudain pourraient constituer des éléments salvateurs de la prise en charge péropératoire.*

Pulmonary artery tumours are exceedingly rare and associated with a poor prognosis. Their management often includes surgical intervention requiring anesthesia. With surgical resection, mean survival time after diagnosis increases from 1.5 months to 10 months, although perioperative mortality may be as high as 22%.[1] Due to similarities in clinical presentation, patients often are misdiagnosed with pulmonary thromboembolism, with the correct diagnosis revealed intraoperatively.[2] Whereas hemodynamic collapse under anesthesia has been reported in patients undergoing pulmonary embolectomy,[3] specific anesthetic complications associated with pulmonary artery tumours, including acute right ventricular (RV) outflow tract obstruction, have not been described. We report a case of hemodynamic collapse under anesthesia resulting from acute RV outflow tract obstruction and left ventricular tamponade in a patient with a large pulmonary artery sarcoma. In accordance with institutional guidelines, written consent was obtained from the patient prior to publication of this report.

Case report

A 67-yr-old previously healthy woman (weight, 72 kg; height, 165 cm) presented to hospital with shortness of breath on exertion and syncope following a vacation in Mexico. She was diagnosed with pulmonary embolism based on radiographic findings and discharged home on therapeutic anticoagulation with coumadin (international normalized ratio, 2–3). One month later, she returned to the hospital with severe shortness of breath and syncope despite adequate anticoagulation. Lower extremity Doppler ultrasonography showed no evidence of deep venous thrombosis, and a pulmonary ventilation-perfusion scan was normal. A computed tomographic scan of the chest, however, showed a large filling defect measuring 3.8 × 2.5 cm in the RV outflow tract involving the pulmonary valve (see Fig. 1). Filling defects also were noted

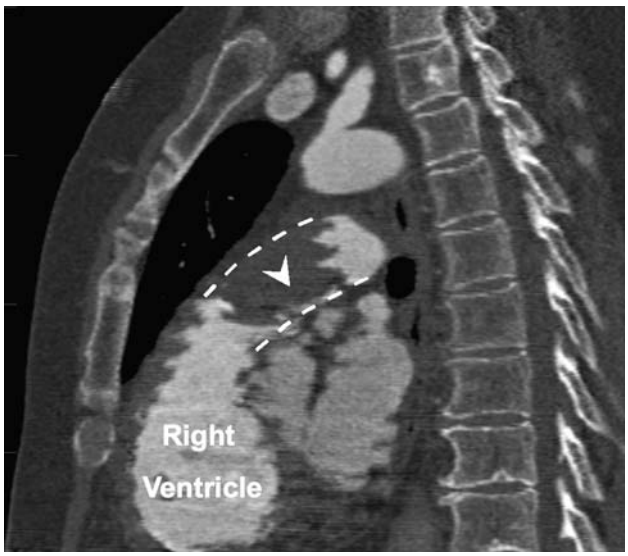

Fig. 1 Computerized tomography scan of the chest. Sagittal view of the patient's chest demonstrates a large filling defect measuring 3.8 × 2.5 cm in the right ventricular outflow tract (*arrow*), producing almost complete occlusion

in the proximal left pulmonary artery and the right apical segment. A diagnosis of pulmonary embolism was made, and the patient was referred to our centre for urgent pulmonary embolectomy. On arrival, the patient had blood pressure of 116/63 mmHg, heart rate of 80 beats · min^{-1}, respiratory rate of 20 · min^{-1}, and oxygen saturation on supplemental oxygen (2 L · min^{-1} by nasal prongs) of 96%. Whereas she was in no distress at rest, the patient experienced syncopal symptoms with minimal exertion. Cardiovascular and respiratory physical examination was unremarkable with the exception of a right-sided heave. The remainder of the physical examination was noncontributory.

A preoperative electrocardiogram showed normal sinus rhythm with right axis deviation and a classic S1Q3T3 pattern of RV strain. A chest radiograph was normal. An echocardiogram revealed a large echogenic mass in the main pulmonary artery (Video available as supplementary material). The gradient across the mass was 80 mmHg, and the RV systolic pressure was 105 mmHg. There was moderate global RV systolic dysfunction and abnormal septal motion. The left ventricle and the remainder of the echocardiographic examination were normal. A preoperative coronary angiogram revealed minimal coronary artery disease. Laboratory investigations were significant for slight elevations in alanine aminotransferase (54 U · L^{-1}; normal range, 5–45 U · L^{-1}) and lactate dehydrogenase (260 U · L^{-1}; normal range, 115–230 U · L^{-1}). The complete blood count, electrolytes, and creatinine were normal. Arterial blood gas analysis resulted in the following: pH, 7.47; $PaCO_2$, 37 mmHg; PaO_2, 81 mmHg.

In the operating theatre, indwelling radial arterial and large-bore intravenous (*iv*) access was obtained prior to induction of general anesthesia. The patient was induced in a titrated fashion with midazolam 2 mg *iv,* ketamine 50 mg *iv,* propofol 30 mg *iv,* a remifentanil infusion (0.06 $\mu g \cdot kg^{-1} \cdot min^{-1}$), and rocuronium 50 mg *iv*. After induction, a central venous sheath was inserted into the right internal jugular vein, and a pulmonary artery catheter was inserted into the superior vena cava. A milrinone bolus of 2 mg *iv* was administered over 15 min via the central venous catheter, and an infusion was started at 0.25 $\mu g \cdot kg^{-1} \cdot min^{-1}$. A transesophageal echocardiography (TEE) probe was inserted and used for monitoring throughout the case. General anesthesia was maintained with sevoflurane (end-tidal concentration, 0.6–0.9%, 50% oxygen in air) and remifentanil (0.06–0.12 $\mu g \cdot kg^{-1} \cdot min^{-1}$).

During the first hour following induction, the patient received three boluses of phenylephrine 0.1 mg *iv* to maintain the mean arterial pressure >60 mmHg. The surgeons performed the midline sternotomy 60 min following induction, at which point a total of 600 mL of normal saline had been administered. A sudden, fourth decrease in blood pressure occurred during sternal retraction, unresponsive to phenylephrine 0.2 mg *iv*. The arterial blood pressure decreased to 45/33 mmHg and a bolus of epinephrine 60 μg *iv* was administered without effect. The hypotensive episode was accompanied by a sudden decrease in end-tidal carbon dioxide tension from 35 to 13 mmHg and an increase in central venous pressure from 17 to 28 mmHg. The patient's heart rate remained stable at 80-85 beats $\cdot min^{-1}$. In preparation for urgent cardiopulmonary bypass, 30,000 units of heparin *iv* were administered. Meanwhile, TEE revealed a massively dilated and distended RV with significant interventricular septal shifting, effectively tamponading the left ventricle. A pericardiotomy was performed in preparation for cardiopulmonary bypass. Following opening of the pericardium, the arterial blood pressure immediately rose to 199/154 mmHg with a sinus tachycardia of 125–130 beats $\cdot min^{-1.}$ Improvement of septal shifting and LV tamponade were seen on TEE. Cardiopulmonary bypass was initiated shortly thereafter with no further complications.

The intraoperative frozen section was consistent with a diagnosis of sarcoma in the pulmonary artery invading the pulmonary valve. The patient underwent resection of the main pulmonary artery with grafting and a bioprosthetic pulmonary valve replacement. She was weaned from cardiopulmonary bypass easily and was transferred to the post-cardiac surgery intensive care unit where she received milrinone and norepinephrine infusions of 0.35 $\mu g \cdot kg^{-1} \cdot min^{-1}$ and 4 $\mu g \cdot min^{-1}$, respectively. The patient's trachea was extubated the following morning, and she was discharged home in good condition 7 days later. Final pathological examination of the intraoperative specimen revealed a high-grade sarcoma with myofibroblastic-type differentiation and positive resection margins. The patient was referred for further medical evaluation and management of her tumour.

Discussion

Pulmonary artery tumours are rare, with few cases reported in the literature.[4,5] They are histologically classified as undifferentiated sarcomas; further subclassification is not thought to be useful clinically or prognostically.[2,6] Pulmonary artery sarcomas commonly arise from the pulmonary trunk, often spreading to the main and proximal pulmonary arteries and involving the pulmonary valve.[1,7] These masses may produce luminal obstruction locally and also can dislodge and produce emboli to distal pulmonary vessels. Progressive obstruction of the pulmonary artery results in RV hypertension, which may ultimately precipitate RV failure, syncope, or sudden death.[5,8] Although right outflow tract obstruction is commonly fixed, a "to-and-from" movement caused by the contracting heart may result in dynamic obstruction.[9,10] Symptoms typically include dyspnea, cough, chest pain, syncope, and palpitations, and the disease is commonly misdiagnosed as a pulmonary embolism.[2] With recent advances in diagnostic imaging, such as high-resolution computed tomography, magnetic resonance imaging, and echocardiography, these tumours may be diagnosed more frequently and present to the operating room for resection.[2]

Prognosis is usually grim, with many patients dying a short time after diagnosis with signs of decompensated right heart failure.[1,5] Surgery remains the most effective intervention and includes pneumonectomy, local excision, and endarterectomy,[11] with isolated reports of surgical cure.[4] Mean survival times have ranged from 10 to 18 months with resection,[1,12] and chemotherapy and radiation therapy show modest response rates but no advantage in overall survival.[2]

Perioperative mortality during attempted resection of these tumours is significant. Pulmonary hypertension, RV hypertension, and RV failure are well-documented risk factors for perioperative morbidity and mortality during both cardiac[13,14] and non-cardiac surgery,[15] and, according to some authors, are the most common clinical manifestations of pulmonary artery sarcoma.[6] Based on the literature up to 1990, Kruger *et al.*[1] reported an early mortality rate after tumour resection of 22%. Chhaya *et al.*[7] recently described a case series of six patients who were scheduled to undergo pulmonary embolectomy and were subsequently found to have a pulmonary artery sarcoma intraoperatively. Three of the six patients died perioperatively; however, specific details were not provided.

Furthermore, in a large series of patients undergoing general anesthesia for emergent pulmonary embolectomy, which is associated with a similar pathophysiology, the incidence of hemodynamic collapse was 19%.[3] The authors concluded that these patients should be prepared and draped prior to induction of general anesthesia, with cardiopulmonary bypass immediately available.

Acute RV failure is precipitated by impaired contractility, volume overload, and pressure overload.[16] Acute *cor pulmonale*, clinically defined as sudden pressure overload and failure of the RV,[17] is the most likely etiology of our patient's acute RV failure and subsequent hemodynamic collapse. Sternal retraction and manipulation of the heart may have precipitated acute obstruction of the RV outflow tract by the tumour. The relatively stable hemodynamic profile during the initial hour after induction followed by a sudden decompensation supports this hypothesis. The patient's echocardiographic findings revealed left ventricular compression by the enlarged RV, as both cavities are enclosed by a stiff pericardial compartment. Animal studies have highlighted the phenomenon of ventricular interdependence, demonstrating shift of the interventricular septum during acute RV failure and impaired left ventricular filling that resolves with opening of the pericardium.[18] The patient's RV dysfunction may have been exacerbated by myocardial ischemia from hypotensive episodes, myocardial depressant effects of the anesthetic agents, and volume overload, although these factors were likely less important given the sudden onset of hypotension and immediate and dramatic resolution following pericardiotomy.

Treatment options in patients with pulmonary artery sarcoma and an acutely decompensated right ventricle are limited. The usual strategies for pharmacological RV afterload reduction are ineffective given the fixed proximal obstruction. Contractility can be optimized through inotropic agents and prevention of ischemia.[19] The importance of maintaining adequate RV perfusion pressure has been highlighted in animal studies showing reversibility of acute RV failure with improvement in coronary perfusion pressure. These studies suggest that maintaining systemic pressure may be a key factor in managing RV failure.[20] In this patient, septal shifting and LV tamponade seen with TEE were immediately relieved by opening the pericardium and restoring cardiac output. The inotropic effects of epinephrine likely contributed to an improvement in RV contractility and produced the subsequent tachycardia and hypertension. Anesthesia for pulmonary artery tumour resection must therefore focus on optimization of RV perfusion and contractility as well as early recognition of acute RV outflow tract obstruction. Preparation and cannulation for femoral-femoral bypass prior to induction of anesthesia should be considered given the potential need for emergency cardiopulmonary bypass.

In conclusion, the patient with a pulmonary artery sarcoma poses a rare and unique challenge to the anesthesiologist. Right ventricular hypertension and dysfunction are well-established risk factors for perioperative mortality, and both are closely associated with pulmonary artery tumours. The high risk of hemodynamic collapse and death is supported by the pulmonary embolectomy literature as well as the limited data available on pulmonary artery tumour resections. Our case highlights the potential risk of acute perioperative RV outflow obstruction in patients with this disease. Careful monitoring of RV performance and optimization of RV perfusion and contractility is warranted. The anesthesiologist should be aware of the potential benefits of urgent pericardiotomy and preparation for emergency cardiopulmonary bypass.

Acknowledgements We gratefully acknowledge Dr. Pari Tiwari for his kind assistance with the CT image and Dr. Chris Thompson for his invaluable help with the echocardiography clip.

Conflicts of interest None declared.

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