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BJAN-D-21-00072 - Case Report

Successful perioperative management of a primary pulmonary arterial angiosarcoma – Case report

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Abstract

Introduction: Primary pleomorphic pulmonary angiosarcomas are extremely rare tumors which could be easily mistaken for pulmonary emboli.

Background and findings: We describe the successful perioperative management of a patient with a pulmonary arterial mass which turned out to be a primary pulmonary angiosarcoma. The severe pulmonary hypertension was a particular challenge compounded with the site and adhesions of the tumor, and pulmonary hemorrhage. The procedure was successfully performed with strict hemodynamic control ensuring stable systemic and pulmonary arterial pressures using perioperative transesophageal echocardiography to continuously monitor cardiac function, along with other standard cardiac surgical monitors including depth of anesthesia monitoring.

Conclusion: Tight hemodynamic control, ensuring stable pulmonary arterial pressures using perioperative echocardiography, and thorough preparation with measures to reduce and prevent

increase in pulmonary arterial pressure along with close communication within the multi-disciplinary team are essential for successful management of patients with this pathology.

KEYWORDS: Pulmonary angiosarcoma; Pulmonary embolism; Cardiopulmonary bypass; Pulmonary hypertension; Case report

Introduction

Primary angiosarcomas are rare malignant tumors accounting for only 2% of all sarcomas.[1] Primary malignant cardiac and great vessel tumors are rarer with an incidence of only 0.001% to 0.03%, and a majority of them being rhabdomyosarcomas. Pleomorphic pulmonary arterial intimal angiosarcomas are much rarer and only about a few hundred cases have been reported since first described by Moritz Mandelstamm in 1923, with most of them diagnosed post-mortem. It could easily be mistaken for chronic pulmonary emboli.[2] The site and the adhesion of the tumor to the pulmonary artery and surrounding structures presents a particular challenge in its perioperative management.[3]

Report

Pre-operative period

A 72-year-old chronic smoker (weight 71 kg, height 1.68 m), with known hypertension, and dyslipidemia presented with worsening shortness of breath over the last month consisting of intermittent episodes of moderate shortness of breath with no other cardiac symptoms. There was no history of prolonged immobilization or long-distance travel nor any recent weight-loss.

On admission, the patient had a clear airway, respiratory rate of 20/minute, oxygen saturation of 94% on room air, heart rate of 75 beats per minute, and blood pressure 150/100 mmHg. He was well hydrated, warm with good peripheral perfusion, conscious and oriented, and normothermic. The hemogram was within normal limits with elevated D-dimer. The biochemistry profile was within normal limits, and the blood gas analysis demonstrated mild hypoxemia with compensated respiratory acidosis.

A transthoracic echocardiogram (TTE) performed on admission showed impaired right ventricular function with TAPSE 1.3 cm, mildly dilated right ventricle with thickened wall, severe tricuspid regurgitation with gradient 85 mmHg and estimated pulmonary arterial systolic pressure (PASP) of 100 mmHg, peak gradient across pulmonary trunk of 50 mmHg, and severely dilated right atrium. The left ventricle showed good systolic function with ejection fraction 60% and a cardiac output of 5.4 litres/minute and no diastolic dysfunction. The patient

underwent unsuccessful local thrombolysis with rTPA based on pulmonary angiogram (Fig. 1), and the subsequent echocardiogram performed demonstrated no improvement. A CT angiogram (Fig. 1) subsequently performed showed hemopericardium and pulmonary hemorrhage in the posterior segments of right middle and apical lobes. A decision to perform a pulmonary thromboendarterectomy was made by the multidisciplinary team involving cardiac surgeon, cardiac anesthesiologist and critical care team after 48 hours of stabilisation.

Intra-operative period

Defibrillator pads were placed on each side of chest and a cardiac arrest trolley was kept ready as a precaution before beginning of anaesthesia as per the European Resuscitation Council (ERC) Advanced Life Support (ALS) guidelines.[4] Prostacyclin and nitric oxide were kept available to reduce pulmonary hypertension as necessary. Monitoring was established as per national standards with addition of Bispectral Index (BIS; Aspect Medical Systems Inc., Newton, MA) monitoring. After placement of a 16-gauge peripheral intravenous cannula and a 20-gauge left radial arterial cannula, 100% oxygen was administered with a face mask. Induction of anesthesia was performed using 0.1mg.kg^{-1} of Midazolam and 6 micrograms.kg^{-1} of Fentanyl titrated to clinical response and BIS response (target BIS 40 - 50), and 80 mg Rocuronium administered to facilitate intubation. A double lumen 37 Fr left sided tube couldn't be sited despite multiple attempts and hence, the patient was intubated with 8.0-mm tracheal tube with a plan to use a bronchial blocker to control the potential spillage of right pulmonary haemorrhage. The patient remained hemodynamically stable throughout induction of anesthesia. The left subclavian vein was cannulated with a 3-lumen central venous catheter and a second 14-gauge intravenous peripheral cannula was sited. Anaesthesia was maintained with Sevoflurane targeting a mean alveolar concentration (MAC) of 1.0 for age maintaining 100% oxygen delivery ventilating both lungs with ventilation settings of tidal volumes 490 mL (7 mL.kg^{-1}) on pressure-controlled ventilation volume guaranteed mode, at a rate of 12 breaths/minute, I:E ratio 1:2, peak pressure 18 cm H_2O and PEEP 5 cm H_2O. Further fentanyl boluses (up to 2.0 mg) and rocuronium were administered as required. Perioperative antibiotic prophylaxis was administered as per protocol. Perioperative transesophageal echocardiography (TEE) was performed confirming the findings of the TTE performed earlier.

A midline sternotomy and pericardiotomy were performed. After full heparinization with 220 mg (3 mg.kg^{-1}) of unfractionated heparin to achieve an activated clotting time of 452 seconds, aortic cannulation and right atrial 2 stage cannulation were performed, and a cardioplegia cannula was placed in the aortic root. Cardiopulmonary bypass (CPB) was rapidly

initiated and the entire procedure was performed without inducing cardiac standstill and aortic cross clamping, with the patient cooled to 24 degrees Celsius. The tumor principally extended in the right pulmonary artery which was clamped to prevent tumor migration. Anesthesia was maintained during CPB using Propofol infusion at 4 $mg.kg^{-1}.hour^{-1}$ (300 $mg.hr^{-1}$) and Sevoflurane 1-2% administered by the Perfusionist guided by BIS (target 40-50) and mean arterial pressure (MAP) (target 40-50 mmHg). The tumor was excised piece-meal to avoid damage to the pulmonary artery and valve leaflet to which it was adherent, and sent for histopathological evaluation. Teflon patch was applied to the posterior wall and right branch of the pulmonary artery. Dobutamine 3 $micrograms.kg^{-1}.min^{-1}$ infusion was commenced as the patient was being rewarmed to 36.5-degree Celsius. The patient was, subsequently, successfully weaned off cardiopulmonary bypass and decannulated. Noradrenaline infusion was started at 0.04 $micrograms.kg^{-1}.min^{-1}$ to maintain a MAP of 70-75 mmHg.

The total cardiopulmonary bypass time was 210 minutes. Protamine 270 mg was administered via peripheral cannula as a slow infusion over 20 minutes monitoring the right ventricle in mid-esophageal 4 chamber view on transesophageal echocardiography with final activated clotting time of 124 seconds (baseline 128 seconds). A pool of platelets and 4 units (approximately 1,000 mL) of fresh frozen plasma were administered. Perioperatively, a total of 1,500 mL of crystalloids was administered with a 600 mL blood loss and 1,050 mL urine output. A post CPB echocardiogram demonstrated a reduction in the tricuspid regurgitation from severe to moderate with PASP reduced to 50 mmHg from 100mmHg and peak gradient across pulmonary trunk from 50 mmHg to 24 mmHg. The patient was transferred with tracheal tube in situ, with lungs ventilated with portable ventilator with 100% oxygen, and full portable monitoring according to international standards to the critical care unit with infusions of Noradrenaline titrated to optimal hemodynamic parameters, and Propofol at 150 $mg.hr^{-1}$.

Post-operative ICU management

The patient was successfully weaned off mechanical ventilation and extubated around 12 hours postoperatively, and Noradrenaline weaned off over the next 48 hours. Pain was managed on the ICU with regular paracetamol and morphine PCA (patient-controlled analgesia) with adequate pain control. The patient was then discharged to the ward after further 12 hours without any significant multiorgan consequences and discharged home after further 5 days. A post-operative CT Angiogram (Fig. 2) was performed which was negative for any residual lesion. A further echocardiogram performed just prior to discharge demonstrated an improvement in right ventricular function (TAPSE 1.5 cm) and further reduction in PASP to

44.5 mmHg. The patient underwent 6 cycles of chemotherapy with Paclitaxel following the diagnosis of intimal fusiform/pleomorphic grade 3 pulmonary artery angiosarcoma. A PET scan performed did not show any metabolically active lesion. At the 3 months follow up, the patient had symptomatically improved and leading a moderately active life. However, the patient had a recurrence at the next follow up at 4 months and, unfortunately, died of a cardiac arrest following an acute pulmonary thrombosis on the recurrent tumour.

Discussion

Primary pulmonary artery angiosarcomas are very rare tumors with very poor prognosis even in the absence of metastasis. Less than half of the patients diagnosed survive the first year despite treatment.[3] Pulmonary artery sarcomas usually originate from the intimal cells and histologically consist of poorly differentiated mesenchymal cells, with fibroblastic and myofibroblastic components. Thus, according to the French Federation of Cancer Centers Sarcoma Group (FNCLCC) grading they are high grade tumors, which was the case with our patient. Due to the extremely rare occurrence of the tumour the literature on the medical and perioperative including anaesthetic management is very limited.

Given the severe pulmonary hypertension (PASP 100 mmHg) and a physical partial obstruction in the pulmonary artery, along with hemopericardium and right pulmonary hemorrhage, it is prudent to take precautions to reduce the risk of cardiac arrest and also put measures in place to readily treat such an occurrence as per international and ERC ALS guidelines.[4] Lung isolation strategy should be planned to reduce the possibility of spilling over of hemorrhage from the right to the left lung and for the possibility of performing a pneumonectomy. However, the evidence for pneumonectomy for a complete eradication is inconclusive and could result in worsening of pulmonary hypertension.[5]

Induction of anesthesia should be closely titrated to avoid haemodynamic instability and 100% oxygen administered throughout the procedure before initiation of CPB to avoid worsening of pulmonary hypertension. Measures to control acute fluctuations in pulmonary arterial pressures should be in place. The use of BIS or cerebral oximetry is recommended to titrate the depth and ensure cerebral perfusion, respectively. The maintenance of hypothermia and infusion of Propofol provides cerebral protection during the procedure thus helping to reduce adverse postoperative cerebral consequences. Early support with inotropes and/or vasoconstrictors helps to ensure hemodynamic stability thus maintaining good tissue and organ flow as well as perfusion in the post CPB period. The use of perioperative TEE is paramount

in such cases as a perioperative monitor in detecting acute chamber and valve failures due to tumor embolism, the status of the repair, and damage to the surrounding tissues.

Conflicts of interest

The authors declare no conflicts of interest.

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Figure 1 – Pre-operative Pulmonary angiogram and CT Angiogram showing pulmonary artery mass.

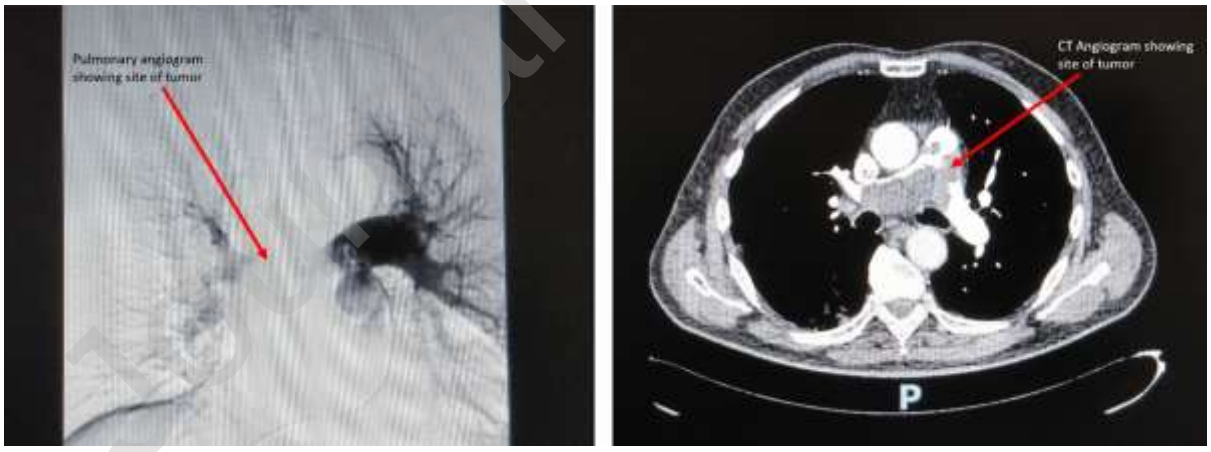

Figure 2 – Post-operative CT Angiogram demonstrating absence of residual tumor.