CASE REPORT OPEN ACCESS

Giant Abdominal Neuroblastoma in an Infant with Dilated Cardiomyopathy: An Intraoperative Challenge

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ABSTRACT

Neuroblastomas are cancers that originate from neuroblasts and are found most commonly in infants. An association of neuroblastoma with dilated cardiomyopathy in an infant is rare and we report the successful perioperative management of a five-month-old baby with giant abdominal neuroblastoma and severe left ventricular systolic dysfunction. Our patient had a cardiac arrest during bone marrow aspiration and complexity of anaesthesia was determined by the large tumour mass compromising the respiration, increased intraabdominal pressure and cardiomyopathy with severe left ventricular dysfunction owing to effects of both chemotherapy and post cardiorespiratory arrest. Care of such patients is complicated by the fact that they have differing anaesthesia management goals, aiming at maintaining the patient's hemodynamic variables of preload, heart rate, contractility and afterload, while also simultaneously maintaining respiratory parameters.

Key words: Anaesthetic management; Dilated cardiomyopathy; Neuroblastoma, Severe left ventricular systolic dysfunction

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INTRODUCTION

Neuroblastoma is the most common extra-cranial tumour in children and accounts for 6-8% of paediatric tumours [1]. An association of neuroblastoma with dilated cardiomyopathy in infants is rare. The overall incidence of paediatric cardiomyopathy (CM) is 4.8 per 100 000 infants. [5] Our case was unique as besides the post-successful cardiopulmonary-resuscitation and chemotherapy induced myocardial depression, anaesthetic management included maintenance of normal diastolic pressures to optimize coronary perfusion, maintenance of preload the and avoiding tachycardia . All measures were taken to avoid increase in pulmonary vascular resistance i.e. avoiding high airway pressures, hypercarbia, or hypoxia, which, in the presence of a low cardiac output, may lead to a rapid hemodynamic deterioration.

CASE REPORT

A 6-month-old girl weighing 5kg was admitted with breathing difficulty, failure to thrive, progressive weight loss and abdominal distension. Laboratory testing revealed α-feto protein level 18913.32 ng/ml, hemoglobin 9.0g/dl, platelet count 70,000/microliter, INR 1.9. Trucut biopsy of the left renal mass and bone marrow aspirate (BMA) was suggestive of neuroblastoma. Contrast-

enhanced computed-tomography scan of abdomen revealed heterogeneously enhancing mass lesion measuring 9.3 x 10.8 x 8.5 cm in the left suprarenal location encasing vessels with extension. The child received OPEC regimen of chemotherapy (4-cycles). Since child had no response to OPEC regimen, second line doxorubicin chemotherapy 40 mgm^{-2} was added for 2 cycles. She had suffered a cardiorespiratory arrest during bone marrow aspiration after receiving midazolam and revived after 5cycles of cardiopulmonary resuscitation (CPR), mechanically ventilated and extubated uneventfully after 48 hours.

Post arrest there was no focal neurological deficit and Computed Tomography scan of brain revealed no focal lesion. However her echocardiography suggested dilated cardiomyopathy, severe systolic dysfunction (ejection fraction 20–25%), global hypokinesia and moderate mitral regurgitation. Her chest X ray was suggestive of diffuse haze in right lung middle and lower zone. Written informed high risk consent was taken from relatives and post-operative ventilator was arranged in paediatric intensive care unit owing to nature of surgery and child's general condition. After attaching standard basic monitors (heart rate, non-invasive blood pressure, and peripheral oxygen saturation and end tidal carbon dioxide), pre-oxygenation was done with 100% oxygen and rapid sequence induction was done with intravenous Fentanyl (6µg), Thiopentone sodium (15mg) and Suxamethonium chloride (6mg). Tracheal intubation was performed uneventfully and she was put on pressure-controlled ventilation mode to maintain normocapnia. A central venous line was inserted by surgeons by open technique and Central Venous Pressure was maintained at 10–12 cmH20. Maintenance of anesthesia was done with Sevoflurane and fentanyl boluses. Epidural catheter was not placed owing to thrombocytopenia and coagulopathy. Intraoperative ABG was within normal limits (pH 7.42, pO2 245, pCO2 34, and HCO3 25).

The child remained hemodynamically stable, until the surgeons lifted huge tumor mass after extensive dissection, there was sudden fall in ETCO2 followed by drop in heart rate to 30/min. The surgeons were immediately asked to stop the surgery and flood the field with saline. Aspiration of air was attempted via central line, subsequently followed by episode of pulseless electrical activity (PEA), slow rate which reverted with two cycles of CPR and adrenaline 100µg. Immediate post arrest ABG was suggestive of acidosis (pH 7.1) however subsequent trends of ABG were within normal limits. After completion of surgery, infant was ventilated, owing to intraoperative hemodynamic instability. She was extubated uneventfully after 96 hours. Tumour was excised completely as suggested by surgeons and specimen was sent for histopathological examination which was suggestive of neuroblastoma. She did not receive any post-operative chemotherapy owing to poor general condition and was discharged home after 25 days of hospital stay.

DISCUSSION

Neuroblastoma is most common cancer in infants arising from neuroblasts of sympathetic nervous system [1]. Rapid Sequence Induction is necessary as huge abdominal mass increases intra-abdominal pressure, thereby risk of aspiration and appropriate adjustment in the dose of induction agent was made to avoid undue hypotension [2]. Also the large abdominal mass compromises respiration by compressive effects, cephalad displacement of diaphragm, decreased lung compliance leading to impaired ventilation-perfusion ratio.

The effect on the cardiovascular system depends upon underlying cardiomyopathy, intravascular volume status, intra-abdominal mass, mode of mechanical ventilation, surgical manipulation and anaesthetic agents used [3]. Chemotherapeutic agents, like cisplatin, doxorubicin and daunorubicin, are associated with cardiotoxicity which can manifest as myocardial depression, ischemia, myocarditis, endomyocardial fibrosis or cardiac conduction defects [4]. Our patient had received multiple cycles of chemotherapy and sustained a cardiorespiratory arrest during Bone Marrow Aspiration which probably added a further ischemic insult to the myocardium. Nugent et al. reported annual incidence of dilated cardiomyopathy as 4.76 per 100,000 infants in Australia [5].

Dilated cardiomyopathy is defined as primary myocardial disease characterised by left ventricular or biventricular dilatation with decreased ventricular contractility. Probable causes of DCMP in our case could be Ischaemic (post cardiac arrest), Chemotherapy induced (Doxorubicin) and association with neuroblastoma, which however is least possible. Anaesthetic principles in an infant with DCMP include maintaining normal blood pressure, avoiding tachycardia, avoiding myocardial depression, adequate preload and avoiding increase in afterload in the presence of increase in left ventricular diastolic pressure. Other aim is to maintain myocardial contractility and maintaining balance between systemic pulmonary vascular resistances. All fac-

tors such as hypoxia, hypercarbia, and increase in airway pressure are to be avoided s they increase the pulmonary vascular resistance [3]. Inotropic support may be required and titrated to desirable hemodynamic targets. We chose fentanyl and titrated dose of sevoflurane to provide maximum hemodynamic stability during maintenance of anaesthesia. Unfortunately our child had an episode of sudden fall in ETCO2, severe bradycardia and PEA during lifting of tumor. The diagnosis of an air embolism came to mind as many vessels were being handled. However any sudden fall in cardiac output (Functional hypovolemia due to decompression of abdominal organs and vessels by lifting of the tumor) could be another reason for sudden fall in ETCO2. Lifting of the tumor could have led to embolization of a thrombus in the IVC due to stasis of blood in the inferior vena cava due to compressive effect. Lifting of the tumor may have also led to an acute kinking of the IVC with a precipitous and sudden fall in venous return leading to a functional hypovolemia and PEA.

To conclude, children with large abdominal malignant mass with severe systolic dysfunction are a challenge to the anaesthesiologist and can be meticulously assessed during preoperative check-up, optimized by medical management, and managed by formulating perioperative anaesthetic plan with prompt diagnosis and management of complications. This case report highlights the unique presentation of neuroblastoma in an infant with dilated cardiomyopathy and perioperative challenges in dealing with such patients.

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