American Journal of
Anesthesia & Clinical Research

Case Report

Anesthetic Management for Anterior Mediastinal Mass with Strong Family History of Malignant Hyperthermia

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Submitted: 05 June 2018; **Approved:** 10 July 2018; **Published:** 16 July 2018

Cite this article: Elmatite W, Upadhyay S, Alfahel W, Mireles R, Ramsdell R, et al. Anesthetic Management for Anterior Mediastinal Mass with Strong Family History of Malignant Hyperthermia. Am J Anesth Clin Res. 2018;4(1): 015-018.

ABSTRACT

Anesthesia for children with anterior mediastinal masses can present life-threatening and hemodynamic challenges in the perioperative period. We encountered a two-year-old child with a family history of malignant hyperthermia and a symptomatic anterior mediastinal mass due to significant airway compression. This case report describes an alternative technique of airway management by keeping the child breathing spontaneously without using inhalational anaesthetics and discusses other potential airway management techniques.

Keywords: General anesthesia; Anterior mediastinal mass; Malignant hyperthermia

INTRODUCTION

Anesthesia for children with anterior mediastinal masses is clinically challenging because of the risk of loss of airway leading to life threatening respiratory obstruction and hemodynamic instability. Compression of the major airways and cardiovascular obstruction is most likely to occur with the induction of general anesthesia [1]. Induction of anaesthesia is often accompanied with loss of muscle tone, which may lead to collapse of a mediastinal mass on the airway and major vessels, causing fatal airway obstruction and severe hemodynamic compromise [2-4]. With induction of anaesthesia, even a previously asymptomatic child may develop life threatening airway obstruction and haemodynamic collapse so alternative techniques such as local anesthesia or preoperative radiotherapy should be considered before general anesthesia [2-4]. Maintaining spontaneous respiration by using inhalational anaesthetics is a safe and popular approach for the airway can be secured and obstruction is bypassed. For patients who have a symptomatic anterior mediastinal mass, particularly those with greater than 50% tracheal compression, any mainstem bronchial compression, or FEV1 < 50% predicted, general anesthesia should be avoided (whether inhalational or consequently, keeping the patient spontaneously breathing during induction of anesthesia is essential. We present a case of anterior mediastinal mass with sign of airway obstruction along with strong family history of Malignant Hyperthermia (MH) which adds another challenge to keep the patient spontaneously breathing without using inhalational anesthesia which is a strong trigger of malignant hyperthermia [5].

CASE REPORT

A two-year-old male child weighing 17.4 Kg was referred for preoperative evaluation for ultrasound guided mediastinal mass and bone marrow biopsies under general anaesthesia. The child was admitted in Paediatric Intensive Care Unit (PICU) with acute onset respiratory distress associated with fever and cough. The child was on antibiotics and steroids to reduce airway edema. Other supportive respiratory care measures were also instituted including head elevation, supplemental oxygen administration, and bronchodilator therapy. The child's past medical history was unremarkable except for mild cough and low grade fever diagnosed as respiratory tract infection one week prior to admission and initially treated on an outpatient basis. On preoperative evaluation, the child had orthopnea, tachypnea with a respiratory rate of 50 per min, heart rate of 120 per min and blood pressure of 90/56 mmHg and oxygen saturation of 98% on 4 litre oxygen via face mask. His respiratory distress was partially relieved after giving steroids, but he still exhibited plethora of the face with engorged neck veins, suggestive of partial vena cava obstruction. A chest x-ray revealed a wide mediastinum (Figure 1A). Laboratory investigations were grossly normal except for elevated uric acid (7.3 mg/dl) (N = 2.0-5.5 mg/dl) and lactate dehydrogenase (LDH 554 unit/L) (N = 120-220 unit/L). A chest CT scan showed a large, anterior mediastinal mass extending to up to neck and compressing the airway, superior vena cava and left brachiocephalic vein with bilateral mild pleural effusions, but no pericardial effusion. (Figure 1 B) Echocardiography was normal. The hemato- oncologist decided to proceed with ultrasound-guided tumor biopsy and bone marrow biopsy. Based on the risk of general anesthesia in such cases we discussed with the hemato-oncologist the possibility of biopsy under local anesthesia but the patient was not co-operative at this time. Similarly, due to the concern of change of the pathological features of the biopsy preoperative radiotherapy was excluded.

In addition, the child's family history was strongly positive for Malignant Hyperthermia (MH), with both his father and grandfather having a documented history of MH.

In view of the airway obstruction by this anterior mediastinal mass along with strong positive family history of MH, we were unable to safely use inhalational anesthetic for induction. Therefore, we utilized IV ketamine and dexmedetomidine to maintain spontaneous breathing until the airway was secured. Before the procedure, the anesthesia machine was purged for 10 minutes at 10 L per min after removing the vaporiser and carbon dioxide absorber. A fresh breathing circuit and carbon dioxide canister were installed. Before anesthetic induction, a paediatric rigid bronchoscope was set up and an Extra Corporeal Membrane Oxygenator (ECMO) circuit was immediately available and primed in case of cardiorespiratory compromise requiring ECMO. ENT and vascular surgical colleagues were present during induction. The patient was kept in the semi-sitting position to maintain comfortable breathing. After applying

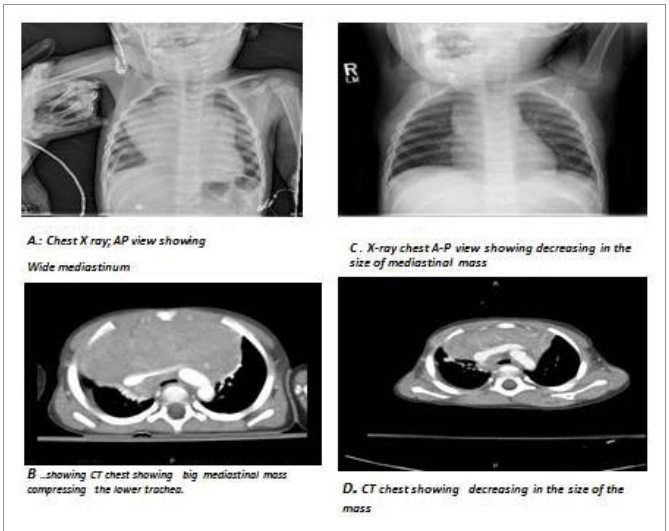

Figure 1: a. Chest X ray; AP view showing large mediastinal mass, b. showing CT chest showing big mediastinal mass compressing the lower trachea, c. X-ray chest A-P view showing decreasing in the size of mediastinal mass, d. CT chest showing decreasing in the size of the mediastinal mass.

standard monitoring that included a Non-Invasive Blood Pressure Cuff (NIBP), pulse oximetry (SPO2), End Tidal Carbon Di-Oxide (ETCO2) Electrocardiogram (ECG) and temperature. A bolus dose of Dexmedetomidine 1 mcg/ kg over 15 min) was started along with 100% supplemental oxygen. After completion of the Dexmedetomidine bolus, a small dose of ketamine 0.5 mg/ kg along with fentanyl 0.3 mcg were given before the start of the procedure. The interventional radiologist injected local anesthesia to decrease the dose of opiate required. We were also prepared to move the patient into the lateral or prone position, if necessary. The patient breathed spontaneously throughout the procedure via a mask with a mixture of oxygen and air to keep Spo2 above 95%. Ultrasound guided mediastinal mass biopsy was performed with the patent in semi erect position and then a bone marrow biopsy was obtained from the iliac bone with the patient in lateral position. Both procedures were completed uneventfully within 20 min and paracetamol 250 mg as rectal suppository was given for post procedural analgesia. The patient awakened and was transported to the PICU for observation.

The biopsy result confirmed T -cell lymphoblastic non -Hodgkin lymphoma. Two days later, the child was again rescheduled for lumbar puncture for intrathecal chemotherapy and Mediport (central vascular access insertion) under General anesthesia. A repeat preoperative evaluation showed significant improvement of respiratory symptoms and a repeat chest x-ray and CT scan demonstrated decreased size of the mediastinal mass (Figure 1C,1D). The same anaesthetic technique with intravenous dexmedetomidine and ketamine was used for both lumbar puncture and placement of USG guided left subclavian central line placement. Both procedures were uneventful. After the completion of the procedures, the child was shifted back safely to the PICU for further post anaesthesia care.

DISCUSSION

This patient presented with the rare combination of a symptomatic anterior mediastinal mass and a strong family history of malignant hyperthermia. Our primary anesthetic concern was the potential for airway obstruction as well as hemodynamic instability during general anesthesia, as the patient was symptomatic, with progressive dyspnea and had signs of airway obstruction. This was supported with the CT finding of compression of lower part and reduction of tracheal diameter over than 50%.

The risk of major complications during the perioperative period is high in patient with symptoms of airway/ vascular compression; however, this cannot be completely ruled out in asymptomatic patients [4,6]. All patients should be thoroughly evaluated by radiologic imaging (X ray, CT; pulmonary function test may be useful in adult cooperative patient but its use is limited in paediatrics age group as it require higher patient cooperation. Airway complications are more common in children because of smaller airway diameter, decreased functional reserve and high metabolic rate [7].

There are limited case reports on successful use of a dexmedetomidne – ketamine combination for diagnostic or therapeutic procedures in paediatric patients with anterior mediastinal masses [8-10] Maintaining spontaneous respiration is a safe and popular approach. In this case, we were not able to utilize inhalational anaesthetics due to cardiovascular and respiratory compromise related to the anterior mediastinal mass and the family history of malignant hyperthermia. Therefore, we chose to use a combination of dexmedetomidine and ketamine which not only provided effective sedation, but also good analgesia with maintenance of spontaneous breathing and haemodynamic stability [11-13].

The optimal combination dose of dexmedetomidine and ketamine has not been defined yet. The use of local anesthesia and awake fiberoptic intubation is not possible in uncooperative pediatric patients. Therefore, in this case we utilized mask ventilation with intravenous agents that do not cause respiratory compromise. Given this strategy, it was of the utmost importance to ensure that advanced airway management, surgical airway management, and ECMO were all immediately available.

Appropriate patient positioning associated with best respiratory function should be selected during the preoperative period and the same position should be adopted during anesthesia. The supine position may aggravate the respiratory dysfunction and hemodynamic instability due to SVC obstruction. Children with large mediastinal masses causing obstructive symptoms may benefit from preoperative treatment to shrink the tumor and surrounding deems which helps in restoring cardio-respiratory stability. Steroids or radiation may be utilized, however both may compromise the eventual histologic and molecular diagnosis, particularly in cases of lymphoma. Preprocedural preparation of equipment and presence of experts in case of any life threatening event is crucial for safe salvage. The anesthesiologist should be ready to change the position to a lateral one or even prone to improve ventilation further. In cases of loss of airway or complete cardiovascular collapse, institution of Cardio-Pulmonary Bypass (CPB) or Extra Corporeal Membrane Oxygenation (ECMO) may be required. Rigid bronchoscopy can be performed successfully to bypass the tracheal compression by tumor in cases of lost airway, potentially before resorting to ECMO.

CONCLUSION

Maintenance of spontaneous ventilation is a safe and time tested technique in patients with symptomatic anterior mediastinal masses which have potential major cardiorespiratory complications. An intravenous dexmedetomidine- ketamine combination can be safe and effective as total intravenous anaesthetics when there is contraindication to inhalation anaesthetic.

ACKNOWLEDGMENT

The authors thank and acknowledge Dr. Jerrold leman, the professor of anesthesia. John R. Oishei Children's Hospital Buffalo, New York, USA. For his guide during management of this case.

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