Thoracic Anesthetic Management of a Patient with Kartagener’s Syndrome

Kartagener Sendromlu Hastada Torasik Anestezik Yaklaşım

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Kartagener’s syndrome (KGS) is an autosomal recessive disorder characterized by the triad of bronchiectasis, sinusitis and situs inversus. In this report we present a patient with KGS and bronchiectasis, who underwent pulmonary resection under thoracic epidural anesthesia (TEA) combined with general anesthesia (GA). Lung isolation techniques vary with respect to the patient and anesthesiologist in this rare disorder. We placed a left sided double lumen tube (DLT) with the bronchial lumen on the right for left -sided middle and lower lobectomy in this case. The appropriate anesthetic approaches and advantages of the TEA combined with sevoflurane in patients with KGS undergoing thoracic surgery are discussed.

Key Words: Kartagener syndrome, anesthesia, thoracic surgery; sevoflurane

Introduction

Kartagener’s syndrome (KGS) is an autosomal recessive disorder, characterized by the triad of bronchiectasis, sinusitis and situs inversus (1). Functional and ultrastructural abnormalities of respiratory cilia have been described in patients with the congenital respiratory disease known as primary ciliary dyskinesia (PCD). Approximately 50% of patients with PCD exhibit situs inversus, which indicates the KGS (2, 3). Defects in the mucociliary transport predisposes the patient to recurrent pulmonary and upper respiratory tract infections resulting in bronchiectasis (1, 3).

Patients with KGS are usually presented for thoracic surgery due to bronchiectasis. The most important anesthetic considerations are related to the pulmonary function, anatomic changes and increased incidence of respiratory infection in the perioperative period. There are few articles reporting the anesthetic management of patients undergoing thoracic surgery due to KGS. This report presents a patient with KGS and bronchiectasis, who was scheduled for pulmonary resection under thoracic epidural anesthesia (TEA) combined with general anesthesia (GA).

Case Report

A 29-year-old female patient (40 kg, 160 cm) with KGS was admitted to the hospital with repeated pulmonary infections and suppurative productive cough since her childhood. There was no clubbing on examination, and the apex beat was on the right side. Auscultation of the heart was normal, and there were crepitations in the basalmost left lung field. Hemoglobin level was 12.8 g/dL and peripheral leukocyte count was 15x10^9/L with neutrophils 74.3%. Other routine laboratory investigations were within the normal ranges. The patient had received antibiotics and chest physiotherapy preoperatively. Chest X-ray revealed dextrocardia and a right-sided stomach bubble. Electrocardiograph (ECG) showed sinus rhythm and inversion of the P wave in lead I, with a deep Q wave and inverted T wave. Computed tomographic (CT) scan of the thorax revealed situs inversus, reversed large vessels and bilateral bronchiectasis. Bronchiectasis was seen especially in the right middle lobe (Figure 1) and left lower and middle lobes (Figure 2). Computed tomography also confirmed the presence of an anatomical right lung in the left hemithorax. Echocardiography revealed a structurally normal heart with an ejection fraction (EF) of 63%. Pulmonary function tests (PFT) showed a moderate restrictive pattern. Results of arterial blood gas (ABG) analysis in room air showed pCO₂: 31.6 mmHg, pO₂: 69.3 mmHg, pH: 7.450, SO₂: 94.5%. The patient was scheduled for left thoracotomy, middle and lower lobectomy under TEA combined with GA.
After premedication with 0.07 mg/kg im midazolam, pulse oximetry, ECG and noninvasive blood pressure monitoring were established. ECG electrodes were placed in reverse positions. An epidural catheter was inserted at T 7-8 level in the sitting position. An epidural test dose of 3 mL lidocaine 2% with 1: 200,000 epinephrine was injected through the catheter, and a negative response to the test dose was confirmed. Colloid infusion at a rate of 15 mL/kg/h was applied prior to an epidural dose of 0.1 mL/kg bupivacaine (0.5%). Propofol, alfentanil and vecuronium were used for anesthesia induction and 1.5% sevoflurane was used for maintenance. The right radial artery and left jugular vein were both cannulated (Figure 3). After induction of anesthesia, bupivacaine (0.375%) infusion was initiated at a rate of 0.1 mL/kg/h through the epidural route.

A left-sided double lumen tube (DLT), (35 Fr Mallinkrodt) with the bronchial lumen facing the right was inserted into the anatomical left main bronchus in the right hemithorax. After advancement through the vocal cords, the left DLT was turned 90 degrees to the right and advanced into the right-sided bronchus. Following blind insertion, the tube placement was checked by auscultation and flexible fiberoptic bronchoscope. During the surgery, the left lung was selectively collapsed without difficulty. A volume-controlled mode was used with the anesthesia machine. Tidal volume was set to 8 and 10 mL/kg, for one and two lung ventilations respectively. Respiratory rate was adjusted to maintain the end tidal CO₂ at 30-35 mm-Hg. Arterial blood gas taken at the 20th minute of double lung ventilation was as follows; pCO₂ : 42 mmHg, pO₂ : 330 mmHg, pH: 7.325, SO₂ : 99.5%. During one lung ventilation with 50% oxygen and air mixture, ABG analysis revealed a pCO₂ : 43.5 mmHg, pO₂ : 120 mm-Hg, pH: 7.321, SO₂ : 98.3%. Peak pressures were measured as 28 and 22 cmH₂O during one and two lung ventilations respectively.

The patient’s trachea was extubated in the operating room without any complication after completion of left sided bilobectomy. In ABG analysis; pCO₂ : 38.5 mmHg, pO₂ : 100 mmHg, pH: 7.325 and SO₂ was 97% while she was breathing O₂ 2 L/min via nasal cannula on postoperative 30th minute.

The patient was followed in intensive care unit on the first postoperative day. Postthoracotomy pain control was achieved by patient controlled epidural analgesia. She was discharged uneventfully on the 6th postoperative day.

**Discussion**

Preoperative evaluation of pulmonary and cardiovascular systems is essential for preparing an anesthesia plan in patients with KGS. Complete situs inversus represents a full mirror image of the normal anatomy of the thoracic and abdominal viscera (4). In dextrocardia with situs inversus, there are no congenital septal or valvular cardiac defects in 90-95% of cases (5). The position of ECG leads should be the mirror image of normal in these patients. The large vessels and thoracic duct are likely to be reversed. Cannulation of the left internal jugular vein is analogous to cannulation of the right internal jugular vein in normal patients. It should be performed in order to avoid damaging the thoracic duct and to ensure direct access to the right atrium (6).
Proper lung isolation is important in KGS patients undergoing lobectomy because it protects the dependent lung from the spillage of suppurative secretions. Pulmonary inversion should be kept in mind while choosing a DLT. Anesthetic approach and choice of DLT in patients of KGS undergoing thoracic surgery were rarely emphasized in the literature (Table 1).

Ho et al. (7) performed fiberoptic bronchoscopy during operation due to bowel adenocarcinoma of a patient with KGS. They suggested that if these patient require one lung ventilation, usage of left-sided double lumen tube with the bronchial lumen on the right was necessary. Habibi et al. (8) reported that it was the suitable approach for a patient with KGS who require a DLT. We also placed a left-sided DLT with the bronchial lumen facing the right for left-sided middle and lower lobectomy. Moreover, correct DLT placement in the right-side was verified with bronchoscopy. Lung isolation and one-lung ventilation were uneventful. Reidy et al. (9) reported left middle lobectomy and successful management of general anesthesia and in their patient, the trachea was intubated with a right-sided DLT. Sahajananda et al. (6) reported an 8-year-old child with KGS who underwent right lobectomy using a modified single lumen tube. Bould et al. (10) have reported anesthetic management of an adolescent with KGS undergoing thoracic surgery. They reported that the patient, who had been intubated with a single lumen tube, rapidly developed severe desaturation due to viscous secretions. Eldawlatly et al. (11) presented anesthetic management during lobectomy in a 12-yr-old child with KGS. These authors had used an univent tube to achieve lung isolation. The main anesthetic consideration is related to increased risk of perioperative respiratory infections when managing patients with KGS (Table 1). Preoperative respiratory infection, if present, should be treated and precautions should be taken to prevent postoperative infection. Postoperative chest physiotherapy and early mobilization help to clear airway secretions and reduce the risk of infection (9, 10). Epidural analgesia should be considered in thoracotomy patients with PCD for early mobilization and uneventful discharge (10).

Anesthetic regimens used for thoracic surgery include total intravenous anesthesia (TIVA), general anesthesia with volatile anesthetics, and TEA combined with GA. They have different effects on hypoxic pulmonary vasoconstriction (HPV), pulmonary and systemic hemodynamics, and the incidence of hypoxemia during one-lung ventilation. TIVA has been compared to inhalation anesthesia in multiple studies with respect to oxygenation and shunt fraction during one-lung ventilation. Although it is accepted that volatile anesthetics inhibit HPV and may promote hypoxemia in a dose-dependent manner during one-lung ventilation, it was showed that TEA combined with GA did not impair arterial oxygenation to the same extent as TIVA, which might be a result of the changes in cardiac output. Therefore, patients with pre-existing cardiopulmonary disease and impaired oxygenation before one-lung ventilation might benefit from TEA combined with GA (12).

Patients with impaired ciliary function, such as KGS, are predisposed to perioperative pulmonary complications such as arterial hypoxemia, atelectasis, and pulmonary infection. It is important to maintain mucociliary transport in order to prevent respiratory complications. Matsuura et al. (13) reported that sevoflurane has a direct cilioinhibitory effect, but its effect is much weaker than that of halothane and isoflurane. They found that exposures to 2%-0.25% sevoflurane did not change ciliary beat frequency significantly. Casati et al. (14) showed that combining epidural anesthesia with GA could decrease volatile agent requirement. We also used a combination of TEA and GA to decrease sevoflurane requirement, and used 1.5% sevoflurane in the present case. Epidural anesthesia combined with GA reduces intraoperative stress and improves perioperative immune function (15). In addition, epidural anesthesia combined with GA significantly improves postoperative lung function and reduces the risk of pulmonary complications (12). Therefore, we preferred TEA combined with GA in this patient who had increased risk of perioperative respiratory complication.

**Conclusion**

We reported a case with KGS receiving a left-sided DLT with the bronchial lumen facing the right-side, into the anatomical left main bronchus in right hemithorax.

**Table 1. The review of the literature in anesthetic management of thoracic surgery in patients with KGS**

<table>
<thead>
<tr>
<th>Ref no.</th>
<th>Author/year</th>
<th>Lung isolation technique</th>
<th>Operation side and type</th>
<th>Anesthetic management/ volatile agent</th>
</tr>
</thead>
<tbody>
<tr>
<td>8</td>
<td>Habibi/1997</td>
<td>Left sided DLT</td>
<td>Left thoracotomy, biopsy and excision of pulmonary nodules</td>
<td>General anesthesia/ not reported</td>
</tr>
<tr>
<td>9</td>
<td>Reidy/2000</td>
<td>Right sided DLT</td>
<td>Left thoracotomy, Lobectomy</td>
<td>General anesthesia/ Isoflurane</td>
</tr>
<tr>
<td>6</td>
<td>Sahajananda/2003</td>
<td>Modified single lumen tube</td>
<td>Right thoracotomy, Lobectomy</td>
<td>General anesthesia/ Isoflurane</td>
</tr>
<tr>
<td>10</td>
<td>Dylan Bould/2006</td>
<td>Single lumen tube</td>
<td>Left thoracotomy, Suture of apical bullous</td>
<td>General anesthesia/ Isoflurane</td>
</tr>
<tr>
<td>11</td>
<td>Eldawlatly/2007</td>
<td>Univent tube</td>
<td>Right thoracotomy, Lobectomy</td>
<td>General anesthesia/ Sevoflurane</td>
</tr>
<tr>
<td>Our patient</td>
<td>Left-sided DLT (inserted into anatomical left main bronchus in right hemithorax)</td>
<td>Left thoracotomy, bilobectomy</td>
<td>Combined general and thoracic epidural anesthesia/ Sevoflurane</td>
<td></td>
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KGS: Kartagener Syndrome, DLT: Double lumen tube
bronchus in the right hemithorax. We considered that TEA combined with sevoflurane is the appropriate anesthetic choice in KGS patients undergoing thoracic surgery.

Conflict of Interest
No conflict of interest was declared by the authors.

References