Turk J Anaesth Reanim 2015; 43: 205-8 DOI: 10.5152/TJAR.2015.94546

Case Report / Olgu Sunumu

Anaesthetic Management of a Patient with Synchronous Kartagener Syndrome and Biliary Atresia

Kartagener Sendromlu ve Bilier Atrezili Bir Olguda Anestezi Yönetimi

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Abstract / Özet

Kartagener syndrome is an autosomal recessive disorder characterized by primary ciliary dyskinesia accompanied by sinusitis, bronchiectasis, and situs inversus. Synchronous extrahepatic biliary atresia and Kartagener syndrome are very rare. During the preoperative preparation of patients with Kartagener syndrome, special attention is required for the respiratory and cardiovascular system. It is important to provide suitable anaesthetic management to avoid problems because of ciliary dysfunction in the perioperative period. Further, maintaining an effective pain control with regional anaesthetic methods reduces the risk of pulmonary complications. Infants with biliary atresia operated earlier have a higher chance of survival. Hepatic dysfunction and decrease in plasma proteins are important for the kinetics of drugs. In this presentation, the anaesthetic management of patients with synchronous Kartagener syndrome and biliary atresia, both of which are rare diseases, is evaluated.

Keywords: Kartagener syndrome, biliary atresia, anaesthesia, infant

Kartagener sendromu primer siliyer diskineziye eşlik eden sinüzit, bronşektazi ve situs inversus ile karakterize otozomal resesif geçen bir hastalıktır. Ekstrahepatik biliyer atrezi ile Kartagener sendromunun birlikteliği oldukça nadirdir. Kartagener sendromlu hastalar, preoperatif solunum ve kardiyovasküler sistem açısından özel olarak hazırlanmalıdır. Perioperatif dönemde uygun bir anestezi yönetimiyle, siliyer işlev bozukluğunun neden olabileceği problemlerden sakınılabilinir. Ayrıca rejyonal anestezi metotları ile etkili ağrı kontrolü pulmoner komplikasyon risklerini azaltmaktadır. Biliyer atrezili bebekler ne kadar erken ameliyat edilir ise hayatta kalma olasılıkları o kadar artmaktadır. Karaciğer işlev bozukluğu ve plazma protein miktarının azalması ilaçların farmakokinetiği açısından önemlidir. Her ikisi de oldukça nadir hastalıklar olan eş zamanlı Kartagener sendromlu ve biliyer atrezili olgumuzda anestezi yönetimi değerlendirilmiştir.

Anahtar Kelimeler: Kartagener sendromu, bilier atrezi, anestezi, infant

Introduction

Kartagener syndrome is an autosomal recessive disorder characterized by primary ciliary dyskinesia accompanied by sinusitis, bronchiectasis, and situs inversus. The prevalence of the syndrome is approximately 1:32000. Two main symptoms are chronic and repetitive respiratory tract infections and male infertility (1, 2). Preoperative respiratory and cardiovascular examination is crucial in patients with Kartagener syndrome. Our case was diagnosed with synchronous Kartagener syndrome, biliary atresia, and polysplenia while receiving treatment for infant respiratory distress syndrome. Here, we report the challenging anaesthetic management of this patient who had two consecutive biliary atresia surgeries.

Case Presentation

A 50-day-old, 3900 g male patient applied to our center complaining of bronchopneumonia and respiratory distress (Figure 1). Chest X-ray revealed dextrocardia and a right-sided stomach bubble (Figure 2). Upon further examination, he was diagnosed with Kartagener syndrome. During his respiratory treatment, jaundice, acholic stool, increased bilirubin, alkaline phosphatase, and transaminase levels were observed, and the patient was diagnosed with biliary atresia. The patient was planned to undergo hepatic portoenterostomy (Kasai procedure). Preoperative anaesthesia evaluation revealed apex beat on the right side. On auscultation, bilateral wheeze and right basal crackles were audible, with heart sounds best heard on the right side of the chest. The patient had a peripheral oxygen saturation of 93%-94% on spontaneous ventilation with room air.

On his 90th day, the patient was taken to the operation room for the Kasai procedure. Electrocardiogram (ECG), non-invasive blood pressure, and peripheral oxygen saturation was monitored (Datex Ohmeda S/5 Avance). Anaesthesia was

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Received / Geliş Tarihi : 21.04.2014
Accepted / Kabul Tarihi : 24.09.2014
Available Online Date / Çevrimiçi Yayın Tarihi : 16.02.2015

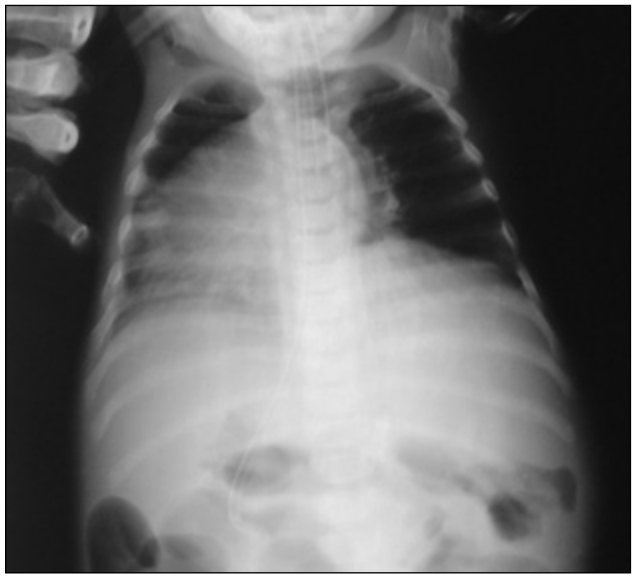

Figure 1. Infiltration of the right lower-middle lobe

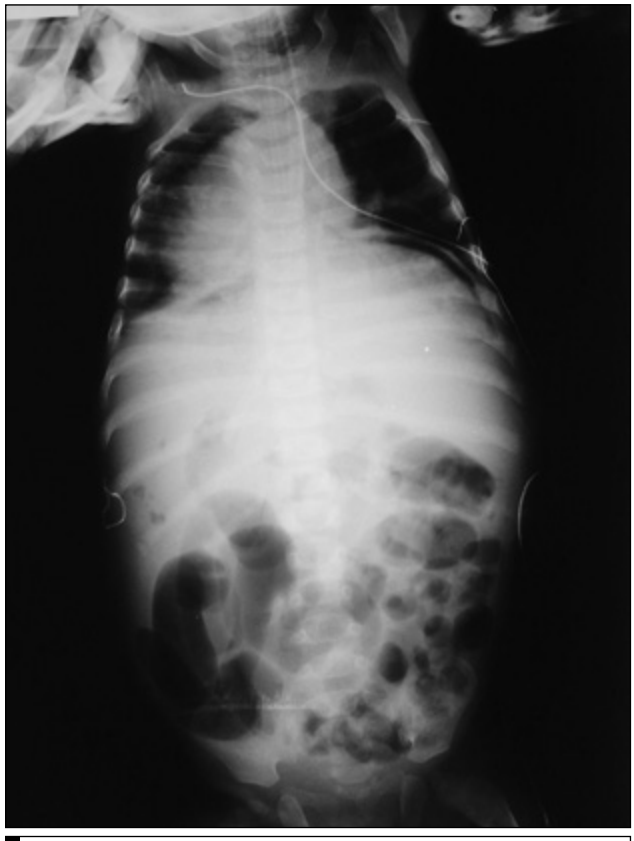

Figure 2. Chest X-ray showing dextrocardia

induced with 8% sevoflurane in 100% O_2 and IV access was performed from the dorsum of the right hand. Further, 0.002 mg kg^{-1} IV fentanyl was administered, and intubation was performed with 0.5 mg kg^{-1} IV atracurium. Anaesthesia was maintained with 2% sevoflurane in 40% O_2-air mixture and 0.02-0.05 mcg kg^{-1} min^{-1} remifentanyl infusions.

Following the intubation, 8F nasogastric tube was inserted and the oesophagus temperature was monitored. Invasive blood pressure and central venous pressure were monitored after radial artery cannulation and left subclavian central venous catheterization. Epidural catheter was inserted from T10/11 level and bupivacaine was injected with a dose of 2 mg kg^{-1} with 5 mL 0.9% NaCl solution through the catheter in two doses.

During the perioperative period, the patient required frequent endotracheal aspirations; auscultation revealed wheezing and bronchodilator therapy was administered. Further, tracheal lavage was applied with (10^{-5}) adrenaline solution to lessen the symptoms. During surgical exploration, polysplenia and micronodular structured liver with no biliary tract were observed, and hepatic portoenterostomy (Kasai procedure) was performed. Venous blood and arterial blood gas samples, electrolytes and glucose levels were checked four times in the perioperative period. The patient did not require blood replacement during the operation. The surgery lasted 360 min and was completed without any complications. Total infusion volume was 230 mL 2% dextrose 0.45% NaCl solution and 70 mL fresh frozen plasma (FFP). The urine output during the operation was 25 mL. Because the respiratory and metabolic values in the arterial blood sample taken at the end of the surgery were appropriate for extubation, the patient was successfully extubated with 0.01 mg kg^{-1} atropine and 0.02 mg kg^{-1} neostigmine and was sent to the paediatric surgery intensive care unit. Postoperative analgesia treatment was maintained with an epidural infusion for two days. He was discharged to the ward in the postoperative 3rd day.

After discharge from the hospital, the patient applied to paediatric surgery clinics intermittently, complaining with ascendant cholangitis and respiratory distress.

When he was 11 months old (5100 g), he was admitted to paediatric surgery clinic with increased bilirubin levels and dilatation in the hepatic biliary tract. Reoperation was decided. In the preoperative examination, bilateral crepitus in the lung auscultation, flaring of the nostrils, and increased expiration time were observed. Peripheral oxygen saturation was 80%-82% in room air. Anaesthesia was induced with 5 mg kg^{-1} thiopental and 0.002 mg kg^{-1} IV fentanyl. Intubation was performed with IV 0.5 mg kg^{-1} atracurium. Anaesthesia was maintained with 2% sevoflurane in 40% O_2-air mixture and 0.02-0.05 mcg kg^{-1} min^{-1} remifentanyl infusion. In addition to standard monitoring, invasive arterial and central venous blood pressures, core temperatures were monitored. Epidural catheterization was not planned because of elevated international normalized ratio for prothrombin time.

Intermittent tracheal lavage was required, owing to the tracheal mucoid secretions that blocked the endotracheal tube. Arterial blood gas samples, blood glucose, and electrolyte levels were monitored. Chiba procedure was performed. İntravenous 350 mL 2% dextrose, 0.45% NaCl, 150 mL erythrocyte suspension, and 150 mL FFP were administered throughout the operation. The duration of the surgery was 7

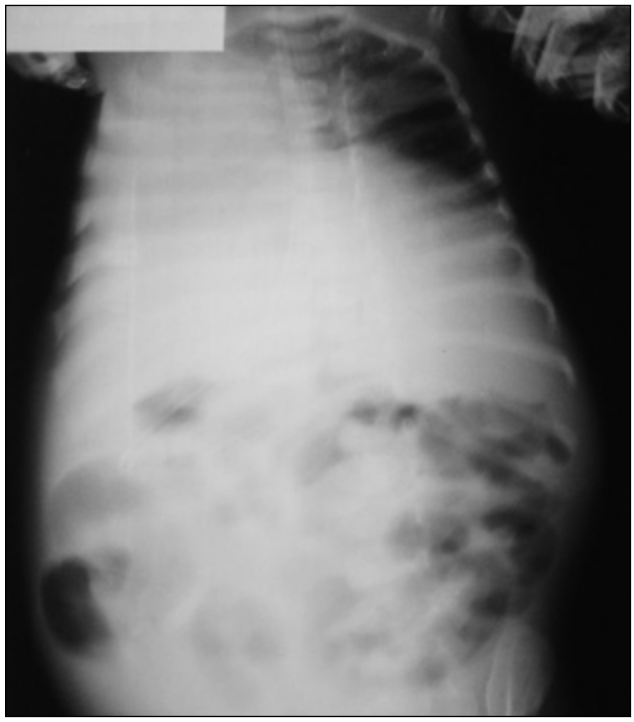

Figure 3. After the development of the right lung atelectasis

hours, and total diuresis was 40 mL. At the end of the surgery, the patient was taken to the paediatric surgery intensive care unit without extubation. On the 2nd day of the ventilator therapy, pneumonia developed. Atelectasis was observed in the right lung on the 4th day (Figure 3), which was treated with aspiration through a rigid bronchoscopy (Figure 4). On the 6th day of the operation, the patient was extubated. On the 7th day, he was taken to the ward and was discharged from the hospital on the 10th day of the operation.

Discussion

The anaesthetic management of our case included major surgery for the liver pathology in a patient with important respiratory problems.

Biliary atresia is an obliterative cholangiopathy that is fatal when not treated. Moreover, Kasai reported that infants operated before the 60th day of birth had a better outcome; hence, an early diagnosis is crucial (3). Diagnosis in our patient was late because respiratory problems dominated the situation. He underwent Kasai procedure when he was 90 days old.

In the anaesthetic management of biliary atresia, hepatic failure, hypoglycaemia, decrease in the plasma proteins, and coagulation defects should be considered. The vitamin K replacement, preparation of FFP, and evaluation of haemogram and coagulation factors should be considered during the preoperative preparation. Hypoglycaemia should be considered, and intravenous fluids should contain dextrose (3). Protection from hypothermia is an important issue for infants who are undergoing major surgery. The operating room should be kept warm. Warmed intravenous fluids, rinsing solutions,

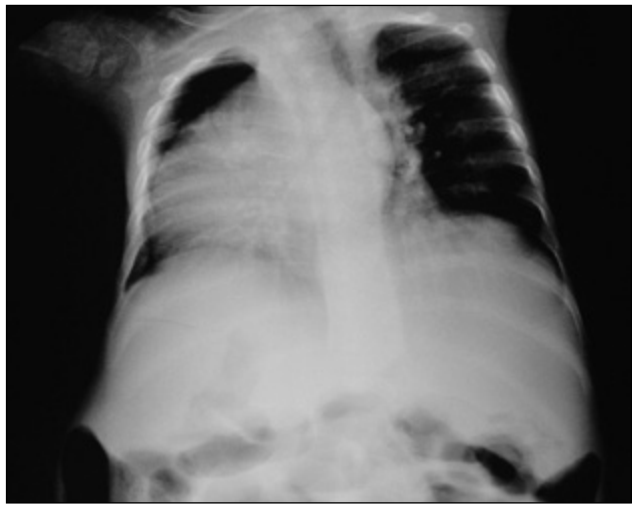

Figure 4. Right lung after bronchoscopy

and hot air blowers or warming mattresses should be used to maintain the patients' temperature.

Productive cough, sinusitis, repetitive respiratory tract infections, otitis media, and infertility are observed in Kartagener's syndrome (1). Physiopathology is associated with the dysfunction of dynein arms in the cilia structure, damaging the coordinated movements of the cilia of the epithelial cells and mucociliary activity. This dysfunction triggers mucus retention, chronic respiratory system infections, which would eventually lead to bronchiectasis (4). The disease can be diagnosed by the frequent respiratory infections during infancy (5).

The co-occurrence of Kartagener syndrome and extrahepatic biliary atresia is a very rare event (4). In our case, in addition to biliary atresia, polysplenia accompanied the syndrome. Dextrocardia is a component of Kartagener syndrome. The ECG electrodes and defibrillator paddles should be placed in the mirror image of their normal positions. Large vessels and ductus thoracicus are also in the reverse position, and using the left side for central venous catheterization is safer (1). In both operations, we used the left side for the central venous catheterization and had not encountered any problems. Furthermore, an increased possibility of the advancing of the endobronchial tube to the left lung should not be ignored because of situs invertus. Respiratory physiotherapy, postural drainage, antibiotherapy, and bronchodilators in the preoperative period can help in the preparation of the patient. As a result of cilia dysfunction, perioperative pulmonary risk increases and if appropriate, regional anaesthesia should be preferred over general anaesthesia (2, 6). Anticholinergic drugs, inhaled medical gases, high O_2 concentrations, and anaesthetic agents can affect the already decreased mucociliary functions. The addition of epidural anaesthesia in general will decrease the use of anaesthetic agents that increase cilia dysfunction and would decrease the risk of pulmonary complications by achieving an effective pain control in the postoperative period (7). In our case, using epidural anaesthesia in the

first operation enabled the patient to have a more comfortable perioperative and postoperative period. If respiratory distress occurs as a result of viscous mucus plugs, bronchoscopic aspiration can be beneficial. Viscous secretions are reported to cause sudden hypoxia by blocking the main branches of the bronchus (6). Antisepsis during any intervention, mainly for epidural, central venous catheterization, and aspiration of the endobronchial tube is vital because of the abnormal neutrophil chemotaxis in these patients (8). Although all possible precautions are taken, infections, respiratory infections can occur and increase morbidity and hospital stay.

Conclusion

The challenge in the anaesthetic management of Kartagener syndrome is taking precautions against the pulmonary complications resulting from cilia dysfunction and bronchiectasis, removing the mucoid secretions with effective aspirations and maintaining oxygenation throughout the operation, avoiding the use of drugs that cause respiratory dysfunction, using regional blocks for pain treatment if possible, and securing postoperative respiratory support. The perioperative management of biliary atresia should consider possible hepatic failure, hypoglycemia, decrease in plasma proteins, and coagulation defects.

The anaesthetic management for each anomaly should be individually assessed if the patient has multiple congenital anomalies.

Informed Consent: Written informed consent was obtained from patient's parent who participated in this case.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - G.K.; Design - P.K., A.Ç.T., G.K.; Supervision - G.K., F.A.; Funding - G.K., F.A.; Materials - P.K, Ş.E.E.; Data Collection and/or Processing - P.K., A.Ç.T.; Analysis and/or Interpretation - G.K., A.Ç.T., P.K., Ş.E.E.; Literature Review - P.K., Ş.E.E.; Writer - P.K.; Critical Review - G.K., A.Ç.T., F.A.

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

Financial Disclosure: The authors declared that this study has received no financial support.

Hasta Onamı: Yazılı hasta onamı bu olguya katılan hastanın ebeveynlerinden alınmıştır.

Hakem Değerlendirmesi: Dış bağımsız.

Yazar Katkıları: Fikir - G.K.; Tasarım - P.K., A.Ç.T., G.K.; Denetleme - G.K., F.A.; Kaynaklar - G.K., F.A.; Malzemeler - P.K, Ş.E.E.; Veri toplanması ve/veya işlemesi - P.K., A.Ç.T.; Analiz ve/veya yorum - G.K., A.Ç.T., P.K., Ş.E.E.; Literatür taraması - P.K., Ş.E.E.; Yazıyı yazan - P.K.; Eleştirel İnceleme - G.K., A.Ç.T., F.A.

Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemişlerdir.

Finansal Destek: Yazarlar bu çalışma için finansal destek almadıklarını beyan etmişlerdir.

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