Case Report

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Anesthesia Management in a Patient with Cystic Fibrosis (A Case Report)

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ABSTRACT

Cystic fibrosis (CF) is a hereditary disease resulting from a chlorine channel defect with autosomal recessive transmission, a structural and functional disorder in the transport of chlorine (Cl) through the plasma membrane in epithelial cells in organs such as the lungs, pancreas, liver, intestines, sweat glands, and epididymis. The most affected regions are the lungs and the gastrointestinal system. These cases are important for their perioperative respiratory complications. We present an anesthesia method conducted on a 16-year-old female CF case receiving general anesthesia.

Keywords: anesthesia, cystic fibrosis

ÖZET

Kistik fibrozisli bir hastada anestezi yönetimi (Olgu sunumu)

Kistik fibrozis (KF), otozomal resesif geçişli, epitel hücresindeki klor (Cl) kanalının defekti sonucunda ortaya çıkan kalıtımsal hastalıktır. Yapısal ve fonksiyonel bozukluk, akciğer, pankreas, karaciğer, bağırsaklar, ter bezleri ve epididim gibi organlarda Cl transportunun bozulmasına neden olur. En çok akciğerler ve gastrointestinal sistem etkilenmektedir. Bu olgular, perioperatif solunumsal komplikasyonlar yönünden özellik-lidir. Yazımızda genel anestezi uyguladığımız 16 yaşında kız KF olgusundaki anestezi yöntemini sunduk.

Anahtar kelimeler: anestezi, kistik fibrozis

Introduction

Cystic fibrosis (CF) is a common genetic disease. Its prevalence is one in 2,000-3,500 live births and its carrier rate is 1 in 25 (1). Its heredity mode is autosomal recessive; the responsible gene is 7q31.3. It can affect all exocrine epithelials and it manifests due to a chlorine channel defect called CF-Transmembrane Regulator (CFTR) in the epithelial cell membrane (2). The slowing water movement due to the defect of the exocrine channels necessary for normal anion flow leads to mucosa dehydration and ductus obstruction. Most affected parts are the lungs and the gastrointestinal system (3). There is a wide range of clinical findings that vary according to age group. Lung findings are the most common ones for all age groups, followed by gastrointestinal findings.

In patients with CF, since the most frequently affected organ is the lung, doctors need to be careful about perioperative and postoperative respiratory complications (4). In this report, we aim to discuss in the context of the literature a case that presented to our clinic for an adenoid vegetation and biopsy operation of a retention cyst in the left maxillary sinus performed under general anesthesia.

Case Report

The female patient was 16 years old, height 170 cm and weight 57 kg. Adenoid vegetation and biopsy operation for retention cyst in the left maxillary sinus was planned. In the patient's medical records it was noted that she had been followed due to CF since 2007 and had undergone a parathyroid operation under general anesthesia at age 11. The patient did not have respiratory system complaints.

In the nasal endoscopic examination, adenoid vegetation was detected but no nasal polyps were found. During the preoperative systemic examination, no pathological findings were observed.

Laboratory and electrocardiography findings were normal. In the respiratory function test (RFT), the results were

respectively: FEV1: 2.90 (expected 3.32); 87%, FVC: 3.49 (expected 3.74); 93%, FEV1/FVC 83.1 (expected 87.3); 95%. In the posteroanterior chest radiograph, increase in vascularity was found; in the thorax CT, an appearance in line with chronic atelectasis and early period bronchiectasis are found; in the cranial MRI, symmetrical soft tissue hardening at the nasopharynx level (adenoid vegetation?) and retention cyst in the left maxillary sinus were found.

The patient was considered to have an ASA score of II and a Mallampati score of I.

In the morning of the operation, after 8 hours of fasting, the patient was taken into the operating room. A peripheral venule was cannulated in the left hand dorsally and hydration was initiated with 2 ml/kg/h isotonic NaCl. Preoperatively, 25 mg ranitine and 2 mg ondansetrone were administered intravenously. Monitoring was made by ECG, non-invasive blood pressure, peripheral oxygen saturation (SpO_2), and body temperature probe. Heart rate was 78 times per minute, blood pressure was 110/70 mmHg, SpO_2 was 99%. After 3 minutes of preoxygenation with 100% oxygen, anesthesia induction was administered with 2 mg/kg propofol, 1 µg/kg fentanyl and 0.6 mg/kg rocuronium. Endotracheal intubation was made with a 7 mm cuffed polyvinyl endotracheal tube (EET). The location of the EET was verified by auscultation and capnograph. Anesthesia maintenance was provided by 2% sevoflurane and 50:50 oxygen/air. Volume-controlled ventilation was given with an end tidal CO_2 value of 32-35 mmHg. The patient's postoperative heart rate, blood pressure values and SpO_2 values were normal. For postoperative analgesia, 750 mg paracetamol + 60 mg tramadol were administered. The operation was completed without any problem in 40 minutes. The lungs were sufficiently ventilated via the ETT. When spontaneous respiratory action began, the effect of the muscle relaxant was terminated by neostigmine 0.05 mg/kg and atropine 0.01 mg/kg. When the patient's airway reflexes returned completely, she was extubated. The patient was taken to the recovery room and received postural drainage and stream therapy. The patient was kept in the recovery room for nearly two hours and then sent to the ward. In the postoperative period, no complications were observed. The patient was discharged without any problems after 24 hours.

Written Informed Consent was taken from the patient's relatives.

Discussion

In CF patients, bronchial obstruction and stasis lead to chronic infection, inflammation, fibrosis, bronchiectasis, and cystic dilatation. When the disease progresses, the damage in the lung increases; ventilation is disordered, hypoxia, pulmonary hypertension, and as a result cor pulmonale and right ventricle failure develop. Nasal and paranasal mucosa retention result in nasal polyposis and chronic sinusitis (3). In respiratory system evaluation, respiratory function tests (RFT) should be made and a chest radiograph should be taken (4). Especially in patients with hypoxemia, postoperative ventilation is needed. In patients with right ventricle hypertrophy and cor pulmonale, ECG and echocardiography should be used. In our case, we have found that there was an appearance in line with chronic atelectasis in the thorax CT, and early period bronchiectasis was found. However, although FEV1 was 2.90 (expected 3.32) 87% in RFT, no respiratory complaint was present. Respiration system examination was normal. Fully monitoring is recommended during the operation (4). However, since the operation was short, monitoring tools were kept limited.

In specific patients, instead of using muscle relaxant, application of a laryngeal mask (LMA) may be preferred. If endotracheal intubation is needed, tracheal ventilation may be conducted during the operation. Since nasal polyps are frequent, nasal intubation needs to be performed carefully (4). Although LMA use is recommended in patients with CF, in our case, since the operation was for adenoidectomy, endotracheal intubation was required. Besides, enabling ventilation via ETT offers an advantage in this operation. Since the operation was short and included an adenoidectomy, regional anesthesia and analgesia were not used.

If a patient with CF receives general anesthesia, medications with short effect should be used. Hypothermia is to be avoided. In the postoperative period, analgesics that do not depress the respiratory system should be used. Chest physiotherapy may also help eliminate secretions. In appropriate operation areas, regional anesthesia and analgesic methods may help in postoperative pain therapy (5). Patients with CF continue to be at risk for respiratory depression, pneumothorax, pneumonia, atelectasis, and airway obstruction (6). After major surgery, an intensive care unit may be the most appropriate setting for close monitoring, continued intravenous hydration, airway management, and chest physiotherapy (6).

In conclusion, in these patients all interventions have to

focus on preventing respiratory complications in the postoperative period during and after administration of anesthesia. In the preoperative period, detailed medical records should be taken and physical examination and laboratory findings should be checked. Appropriate anesthesia method and medications should be used and monitoring should be conducted during the operation. The lungs should be ventilated intermittently during the operation, which will facilitate sufficient liquid entrance and removal.

Contribution Categories	Name of Author
Follow up of the case	H.Y., S.N.S., N.K.
Literature review	N.K., K.E., A.S., S.N.S.
Manuscript writing	H.Y., N.K., K.E., A.S.
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