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 epidymis. 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)

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...
respiratory system should be used. Chest physiotherapy may also help eliminate secretions. In appropriate operation areas, regional anesthesia and analgesic methods may help in

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 adenoectomy, endotracheal intubation was required. Besides, enabling ventilation via ETT offers an advantage in this operation. Since the operation was short and included an adenoectomy, 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

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

References


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<th>Contribution Categories</th>
<th>Name of Author</th>
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<tbody>
<tr>
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