Cholecystectomy of a patient with Churg–Strauss syndrome
— A case report —

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Churg-Strauss syndrome is an allergic granulomatous angiitis and the organ most commonly involved in this condition is the lung. However, this syndrome also affects the skin, cardiovascular system, kidney, peripheral nervous system and gastrointestinal system. Cardiac involvement is a rare complication but can lead to rapid-onset heart failure as the result of specific cardiomyopathy. Pericardial effusion may also occur. Acalculous cholecystitis is also a rare complication of Churg-Strauss syndrome. Here, we present a case of a patient with Churg-Strauss syndrome and severe heart failure scheduled for cholecystectomy due to acalculous cholecystitis.

The patient had mild asthma symptoms, peripheral neuritis in both legs, and severe heart failure. During the preoperative period, steroids, β2 agonists, diuretics, and antihypertensive drugs were administered. During anesthesia we attempted to prevent compromising the patient’s cardiac and pulmonary functions. The surgery was completed successfully, and the patient was discharged without any complications.

Key Words: Asthma, Cholecystectomy, Churg-Strauss syndrome, Heart failure.

Churg-Strauss syndrome (CSS) is a rare necrotizing systemic vasculitis occurring in estimated two to four patients per million [1]. According to the criteria of the American College of Rheumatology, the diagnosis of CSS is confirmed when four of the six following signs are present. Asthma, eosinophilia >10%, neuropathy, pulmonary infiltrates, paranasal sinus abnormality, and extravascular eosinophil infiltration on biopsy finding. CSS is often accompanied by cardiologic features that suggests poor prognosis, usually in the form of rapid-onset heart failure [2]. Acalculous cholecystitis has been considered to be rarely involved in CSS [3]. This case highlights the anesthetic management of cholecystectomy in a patient with the CSS with heart failure.

CASE REPORT

A 22-yr-old female weighing 52 kg was admitted complaining of right upper quadrant (RUQ) pain. In the past history, she had been suffering from asthma and paranasal sinusitis for 2 years. She had been taking β2 agonist bronchodilator inhaler with intermittent oral steroid, and antibiotics. On admission, asthma symptom was well-controlled, but on the physical examination, severe RUQ pain, both legs edema, numbness, paresthesia and multiple erythematous macules were detected. To relieve her neuropathic pain symptom gabapentin 300 mg and amitriptyline 10 mg tid were prescribed. On the skin biopsy, eosinophilic infiltration and c-anti-neutrophil cytoplasmic antibody (c-ANCA) was positive. On hematologic examination, leukocytosis (17,700/mm3) and eosinophilia (50% of differential count) were detected. In the chest X-ray, multiple patchy infiltrates were observed (Fig. 1). On her abdominal ultrasonography and computed tomography, gall bladder wall thickening and edema were shown but no gall bladder stone was detected. Endoscopic retrograde cholangiopancreatography (ERCP) was tried but her symptom did not improve, then the percutaneous pig-tail catheter was inserted to drain her bile. Two weeks later she developed bile peritonitis and urgent chol-
ecystectomy was scheduled. In the preoperative pulmonary function test, forced vital capacity (FVC): 2.72 L, forced expiratory volume of 1 second (FEV₁): 2.34 L, FEV₁/FVC: 80%, mild restrictive pattern was shown. Arterial blood gas analysis in the room air showed pH: 7.488, PaCO₂: 42.2 mmHg, PaO₂: 96.8 mmHg, SaO₂: 99.7%. Oral methylprednisolone 40 mg and salbutamol 1 g inhalation were continuously administered to the patient. In the transthoracic echocardiography, severe left ventricular systolic function with global hypokinesia and mild pericardial effusion were observed and ejection fraction was 30%. Therefore, spironolactone 25 mg, ramipril 10 mg, bisoprolol hemifumarate 2.5 mg were administered for 4 weeks. On three days before the operation, transthoracic echocardiography was rechecked and it showed normal left ventricular systolic function without regional wall motion abnormalities and ejection fraction was 59%. On three days before the operation, oral methylprednisolone was replaced to hydrocortisone 100 mg i.v tid and it was repeated until the operation day. Salbutamol inhalation therapy was also maintained until the operation day. In preoperative CBC, hemoglobin 11.0 g/dl, hematocrit 36.0, white blood cell 14,330/mm³, and eosinophil: 2% were reported. In the preoperative chest X-ray, there was no active lung lesion (Fig. 2). In the morning of operation day, the patient was premedicated with glycopyrrolate 0.2 mg i.m. and midazolam 3 mg i.m., and hydrocortisone 100 mg i.v. She took the last oral medications with a 30 ml of water before leaving the ward. An epidural catheter was placed at the T₈–T₉ intervertebral space after 2% lidocaine 3 ml infiltration for postoperative patient-controlled epidural analgesia (PCEA). Epidural test dose (2% lidocaine 3 ml, epinephrine 15 μg) was injected. Anesthesia was induced by propofol 80 mg i.v., rocuronium 30 mg i.v., and 100% O₂ inhalation. Cobra perilyngeal airway, size 3 (CobraPLA™, Engineered Medical System, Inc., USA) is inserted blindly instead of endotracheal tube to minimize airway irritation. Five-lead electrocardiogram, non-invasive BP, SpO₂, EtCO₂, spirometry, airway pressure, body temperature, central venous pressure and continuous arterial pressure were monitored by S/S™ Anesthesia Delivery Unit (Datex-Ohmeda, Finland). Anesthesia was maintained with 50% N₂O in O₂ with 1−1.2% isoflurane, and remifentanil infusion (5−25 μg/hr) to avoid cardiovascular instability. During the intraoperative period peak airway pressure was maintained between 18−20 cmH₂O and hemodynamic variables and arterial blood gas levels were stable. The patient was provided with postoperative PCEA by injection of 8 ml of 0.375% ropivacaine and 50 μg fentanyl just before the completion of the surgery. Patient-controlled analgesia pump was connected to epidural catheter containing 0.25% levobupivacaine 150 ml and fentanyl 500 μg. We established the infusion rate: 3.0 ml, bolus dose : 1.5 ml, and lock-out time : 20 min. Neumuscular
Churg-Strauss syndrome (CSS) is an uncommon disease characterized by necrotizing vasculitis of multiple organ systems. The main symptom is asthma. Many patients show asthma that may precede systemic vasculitis by many years. About half of the patients with CSS show non-fixed, patchy pulmonary infiltrates on chest radiography and these are often associated with fever, cough and dyspnea. This patient showed mild fever and leukocytosis at admission, but infection sign was not detected in blood culture or sputum examination. In preoperative evaluation of the patient with CSS, pulmonary function tests are necessary for assessing a patient’s risk for developing perioperative pulmonary complications. The patient had been suffered from asthma for 2 years but fortunately symptom was not severe and well-controlled with corticosteroid and β2-agonist inhalation. Combined treatment with corticosteroids and a β2-adrenergic agonist can improve preoperative lung function and decrease the incidence of wheezing following endotracheal intubation [4].

CSS patients with cardiac involvement have more severe disease and a poorer prognosis [2]. Heart involvement was reported as a major cause of death. They include eosinophilic endomyocarditis, coronary vasculitis, coronary dissection, congestive heart failure, cardiac conduction abnormalities, arrhythmia, and pericardial effusion. Cardiomyopathy occurs in 13−40% of all cases and can lead to acute onset severe cardiac failure. Therapeutic responses to the corticosteroid are variable. Fortunately, this patient responded dramatically to medication.

CSS often develops abdominal pain. The causes are gastric lesions, colonic ulcers, pancreatitis and bowel perforations. Cholecystitis has been rarely reported but it is related to eosinophilic infiltration of gall bladder and surrounding vessels [3].

We used CobraPLA™ instead of endotracheal tube to lessen airway irritation. When randomly selected 52 nonasthmatic patients to receive either an endotracheal tube or a laryngeal mask airway under general anesthesia, airway resistance was lower in patients receiving laryngeal mask airways than in those submitted to endotracheal intubation [5]. Both the LMA-Classic and CobraPLA™ consist of a tube with a distal cuff, inflated when positioned in the patient’s hypopharynx, to achieve a seal, thus allowing mechanical ventilation of the lungs without leaks or aspiration. However, the CobraPLA™ has a larger internal tube diameter and a larger and more proximally positioned circumferential cuff than LMA-Classic. The CobraPLA has been evaluated and found to be an adequate alternative to the LMA-Classic in terms of insertion and recovery characteristics [6]. Higher sealing pressures have been reported with the CobraPLA™ when compared with the LMA-Classic during controlled ventilation [6]. Therefore we chose CobraPLA™ as a ventilating device instead of LMA-Classic that might give safer airway protection from aspiration in controlled ventilation.

Propofol, a widely used short-acting intravenous anesthetic, has been associated with less bronchoconstriction during anesthetic induction than other anesthetic agents [7]. *In vitro* data suggest that propofol has a direct airway smooth muscle-relaxing property [8]. We used propofol as a induction agent because propofol could be a better choice than thiopental in asthmatic patients and had been reported to be safe in asthmatic patients in spite of the reports to induce histamine release in healthy volunteers [9]. Inhalational agents possess bronchodilatory effects, decrease airway responsiveness, and attenuate histamine-induced bronchospasm. The mechanism is thought to be β-adrenergic receptor stimulation leading to increased intracellular cyclic-AMP. Increased cAMP may bind free calcium within bronchial myoplasm and cause relaxation by negative feedback. It may impede antigen-antibody mediated enzyme production and the release of histamine from leukocytes as well. For all these reasons, volatile agents such as halothane and isoflurane have been recommended for general anesthetic techniques in patients with obstructive airway diseases for many years, and they are even helpful to treat status asthmaticus. An exception should be made for desflurane, which can lead to increased secretions, coughing, laryngospasm, and bronchospasm. There are some controversial results about the effect on respiratory resistance of sevoflurane. In comparison of bronchodilating effect of halothane, sevoflurane and isoflurane in non-asthmatic patients, sevoflurane was superior to halothane and isoflurane [10]. On the other hand, sevoflurane increased respiratory resistance in mild to moderate asthmatic children with endotracheal intubation that was not seen in non-asthmatic children [11]. In spite of this, no clinically adverse event was observed. Analysis of the respiratory mechanics and lung histology in rats anesthesized with sevoflurane showed that sevoflurane...
evoked stiffness of lung tissue and increased mechanical in-homogeneities [12]. Takala et al. measured pulmonary inflammatory mediators in bronchoalveolar lavage fluid after sevoflurane anaesthesia in pigs and reported that sevoflurane increased pulmonary leukotriene C4, NO3−, and NO2− production, suggesting an inflammatory response [13].

Although opioids can release histamine, they are considered safe for airway reactive patients. Fentanyl and its analogues are frequently used in the induction of anesthesia, and they can lead to thorax rigidity that can be misinterpreted as bronchospasm. With slow injection, this effect is hardly observed. Moreover, the suppression of the cough reflex and the deepening of anesthesia level achieved after opioid administration can be helpful in asthmatic patients. We used remifentanil infusion as adjuvant for deepening of anesthesia and minimizing airway reaction, and hemodynamic stability during operation.

We used rocuronium for the effective muscle relaxation and controlled ventilation. Among muscle relaxant, depending on which type of muscarinic receptor is stimulated, the effect on bronchial tone and reactivity can be expected. It has been shown that muscle relaxants which affect M2 receptors more than M3 receptors (gallamin, pipecuronium, rapacuronium) can cause and enhance bronchoconstriction [14]. Otherwise, muscle relaxants which seem to bind M3 receptors more or at least the same way as M2 receptors do not induce bronchospasm. Among those, vecuronium, rocuronium, cisatracurium, and pancuronium are considered safe. In addition to these direct effects on muscarinic receptors, atracurium and mivacurium dose dependently release histamine and have been identified as triggers of bronchoconstriction and should be used carefully in asthmatic patients. Furthermore, the reversal of muscle relaxation at the end of surgery should be avoided since neostigmine and physostigmine cause bradycardia, increased secretion, and bronchial hyper-reactivity. For this purpose, doses of muscle relaxants should be titrated so as to be worn off at the end of surgery. We provided thoracic epidural patient controlled analgesia with fentanyl and local anesthetics mixture, that could decrease the incidence of bronchospasm after tracheal intubation. Anesthesiology 1999; 100: 1052-7.


