Case Report

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A patient with Churg-Strauss syndrome who underwent endoscopic sinus surgery under general anesthesia
-A case report-

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There are many cause of cholinesterase deficiency, including drugs, liver disease, chronic anemia, malignant states, cardiac failure, severe acute infection, surgical shock, severe burn, collagen disease and vasculitis syndromes. Vasculitis syndromes are relatively rare, and among them, Churg-Strauss syndrome (CSS) is even rarer. We report here on a case of a patient with CSS who underwent endoscopic sinus surgery under general anesthesia. (Korean J Anesthesiol 2010; 59: 49-52)

Key Words: Cholinesterase deficiency, Churg-Strauss syndrome, Vasculitis syndrome.

Churg-Strauss syndrome (CSS) is an uncommon disease that is characterized by allergic rhinitis, asthma and eosinophilia, which is an altered blood count [1,2]. The involved organs are usually the lung and skin, although any organ system can be affected, including the cardiovascular system, the gastrointestinal system, the renal system and the central nervous system [3].

Immune complexes are probably involved as the cause of CSS, although the exact etiology of this syndrome is unknown [4]. The annual incidence rate of CSS per million of the population has been estimated to be between 1.3 and 6.8 [5,6].

Making the diagnosis is difficult because patients with CSS have various clinical symptoms and levels of disease progress. If treatment is delayed by a late diagnosis, then heart failure can be the cause of death. So, a rapid diagnosis and aggressive treatment are required to effectively treat patients with CSS.

Patients with CSS display cholinesterase deficiency, hypersensitivity of the airway and multiple organ dysfunctions. For this reason, anesthetic management of patients with CSS can be difficult. There are not many reports on performing anesthesia for patients with CSS. So, we report here on the anesthetic management of a patient with CSS and we also review the relevant medical literature.

Case Report

A 34-year-old woman (168 cm tall and weighing 55 kg) complained a productive cough, postnasal drip and maxillary

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pain. The patient was scheduled for endoscopic sinus surgery to treat her chronic sinusitis.

She had a history of bronchial asthma for the previous 2 years and was on brochodilators (salmeterol 1 puff once a day). She was diagnosed with CSS 1 year previously when she started complaining of gradually progressive numbness of the left 4th and 5th fingers, but the sensory tests were normal and the nerve conduction tests showed non-specific finding. Her differential WBC count showed peripheral eosinophilia and the antineutrophil cytoplasmic autoantibody (ANCA) was negative. The skin biopsy showed vasculitis with many eosinophils, neutrophils and nuclear debris in the dermis. The bronchoalveolar lavage (BAL) fluid showed an increased total cell count and the eosinophils were especially dominant. A diagnosis of CSS was then made. She was taking oral prednisolone 10 mg once a day.

The preoperative routine laboratory values were within the normal limits. Mild cough, sputum and rhinorrhea were present on the physical examination. The chest X-ray showed minimal patchy opacity in the left lower lung zone. The results of the pulmonary function tests were within the normal limits with a FEV_1 of 96%, a FVC of 80% and a FEV_1/FVC of 94% of the predicted values. Electrocardiography showed inferior myocardiac ischemia, but the findings of the echocardiogram were within the normal limits. Arterial blood gas analysis on room air showed a pH of 7.356, a PaO_2 of 76.2 mmHg, a $PaCO_2$ of 43.2 mmHg and a HCO_3 of 23.6 with a SaO_2 of 99.0%.

She was premedicated with intramuscular glycopyrrolate 0.2 mg before surgery and on the morning of surgery, along with overnight fasting. Oral prednisolone was given on the morning of surgery. All her medications for asthma were continued.

In the operating room, routine monitoring was used along with a neuromuscular transmission (TOF-Watchr, Organon, Ireland) monitor and a continuous arterial pressure monitor to determine the degree of muscle relaxation and hypotension due to deep anesthesia. General anesthesia was induced with propofol 120 mg, inhaled sevoflurane 3–4 vol% and remifentanil 0.2–0.3 ug/min/kg. The TOF was kept at 90–100% for 3 minutes, but after 3 minutes the blood pressure dropped to 70/50 mmHg. An intravenous injection of ephedrine (5 mg) was given. The blood pressure was stable and the TOF count was 2. Tracheal intubation was performed immediately. The anesthesia was maintained with sevoflurane 2–3 vol%, remifentanil 0.2–0.3 ug/min/kg, air and oxygen at FiO_2 0.5. Any muscle relaxant was not administered during surgery. The TOF was kept at 90–100% during surgery. No major problems occurred during surgery.

At the end of surgery, the remifentanil infusion and sevoflurane were stopped and any anticholinesterase agent was not used. The breathing sounds were clear and the TOF was kept at 100% before extubation, so the tracheal tube was taken out with the patient in a deep state of anesthesia. Oxygen was supplemented by a face mask till the patient recovered full consciousness and self respiration. Her postoperative period was uneventful.

Discussion

CSS was first described in 1951 by Dr. Jacob Churg and Dr. Lotte Strauss as a syndrome consisting of asthma, eosinophilia and additional vasculitis of multiple organ systems [1].

The clinical manifestations of CSS usually have three stages [7]. The first phase is called the "allergic" phase and this is characterized by allergic inflammation of the nose, the skin and the lung. People are often diagnosed with late onset asthma during this phase. The second phase is called the "hypereosinophilic" phase, which means that there are too many eosinophils in the body. This phase is characterized by inflammation of the esophagus, stomach or intestine. The third phase is the "systemic vasculitis" phase. During this phase there is inflammation and damage of the blood vessels; the blood vessels in different parts of the body can be damaged. During this phase, the patients with CSS may suffer from fever, weight loss and a lack of energy. These 3 phases are not necessarily contiguous.

The American College of Rheumatology (ACR) has offered criteria that must be fulfilled in order to classify a patient as having CSS [8]. In order to be classified as a CSS patient, a patient should have at least 4 of the 6 ACR criteria for CSS to be diagnosed with a sensitivity of 85% and a specificity of 99.7%. These criteria are 1) asthma, 2) eosinophilia [>10% on the differential WBC count], 3) mononeuropathy, 4) transient pulmonary infiltrates on chest X-rays, 5) paranasal sinus abnormalities and 6) a biopsy containing a blood vessel with extravascular eosinophils. Our patient's asthma, peripheral blood eosinophilia, the skin biopsy with an infiltration of eosinophils and the chronic sinusitis fulfilled the ACR diagnostic criteria.

CSS commonly responds to prednisone. High doses of oral prednisone are initially used in an attempt to get the disease into remission as quickly as possibly (e.g., oral prednisone 40–60 mg/day). After the first month or so, this high dose of prednisone is slowly tapered down over the ensuing months [9]. Other immunosuppressive drugs such as azathioprine, cellcept, cyclophosphamid or methotrexate may be used in addition to prednisone. High doses of intravenous steroids, and commonly methylprednisolone, maybe useful for those patients with severe disease or for those who are unresponsive to oral prednisone combined with other immunosuppressive medications.

The French Vasculitis Study Group has identified five prognostic factors, the so-called five-factor score (FFS),

for patients with necrotising vasculitis, including CSS [10]. These are (1) elevated serum creatinine levels (>1.58 mg/dl), (2) proteinuria (>1 g per day), (3) gastrointestinal tract involvement, (4) cardiomyopathy and (5) central nervous system involvement. The presence of one or more factors is correlated with a 5-year mortality rate ranging from 25.9–46%, whereas an FFS of 0 is correlated with a 5-year mortality rate of 11.9% [10]. The prognosis of patients with CSS is not clearly known, but a 1 year survival rate of 90% and 5 year survival rate of 62% have been reported after treatment with corticosteroid. A significant reduction in mortality has been reported after treatment [11].

Most patients with CSS have asthma. So, pulmonary function tests are essential as part of the preoperative evaluation of CSS patients to assess the risk of perioperative and postoperative pulmonary complications. The patient in this case had been experiencing asthma for the previous 2 years, but her respiratory symptoms were not severe and they were well-controlled with brochodilators (salmeterol 1 puff once a day), and the pulmonary function tests were within the normal limits.

Surgery requires that a patient have nothing to eat or drink for about 6 hours before anesthesia, with an exception being medication. Patient with CSS and who have been taking corticosteroid for a long time should take their usual medication up until the time of surgery. Adrenal insufficiency could develop, and CSS patients may require perioperative supplemental corticosteroids [12]. The patient in this case took oral prednisolone 10 mg on the morning of surgery and there was no adverse event during the perioperative period. So, we did not medicate her with additional corticosteroid.

For our case, the anesthesia management during the perioperative period was chosen to minimize tracheal activity as the patient had asthma. Propofol reduces the intubation-induced bronchoconstrictive response [13]. Propofol leads to bronchodilation in patients with chronic obstructive pulmonary disease. The volatile anesthetic agents are bronchodilators that relax the airway smooth muscle and decrease the resistance of the respiratory system. Some opioids can release histamine, and so they can cause bronchospasm. But fentanyl and its analogues have an antihistaminergic action and these may be more effective than morphine for patients with asthma. Furthermore, Groeben reported that the suppression of the cough reflex and the deepening of level of anesthesia level that are achieved after opioid administration can be helpful in asthmatic patients [14]. The anesthetic management was successful in this case by using propofol, remifentanil and sevoflurane to induce and maintain anesthesia without respiratory complication.

Taylor and colleagues [15] reported on two patients with CSS who were found to have decreased cholinesterase activity after abnormal sensitivity to suxamethonium was suspected. So, we did not use muscle relaxant as there was no need for muscle relaxation for surgery and our patient was adequately anesthetized using a volatile anesthetic agent and opioid. A deep state of anesthesia was induced and maintained. This also reduced any bronchial hyper-reactivity, yet hypotension can occur because of deep anesthesia, so a continuous arterial pressure monitor was used to detect hypotension early. The use of an anticholinesterase agent to reverse muscle relaxation can cause bradycardia, increased secretion and brochial hyper-reactivity, so we did not use an anticholinesterase agent.

In conclusion, this case was successfully managed using general anesthesia following careful preoperative evaluation and precise perioperative management. The patients with CSS may require intensive care during the course of their disease. Muscle relaxant should be used with caution during surgery, and successful anesthetic management can be achieved.

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