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Case Report

Anesthetic management of a patient with Hermansky-Pudlak syndrome undergoing video-assisted bullectomy☆

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Abstract The Hermansky-Pudlak syndrome (HPS) is a rare set of disorders characterized by oculocutaneous albinism, bleeding diathesis, and pulmonary fibrosis, with the latter 2 conditions presenting major challenges in anesthetic management. We report a 53-year-old woman with pulmonary fibrosis secondary to HPS who underwent video-assisted bullectomy to treat recurrent pneumothorax. Preoperative bleeding time and platelet count were within normal limits, but the surgeons had difficulty with continuous oozing from the incision site; the surgical blood loss was 270 mL, which was a relatively large amount for this surgery. Because of her restrictive lung disease, the patient's tidal volume was only 250 mL under pressure-controlled ventilation, with a peak inspiratory pressure of 30 cm H_2O and a positive end-expiratory pressure of 5 cm H_2O. She also had postoperative respiratory insufficiency, with a partial pressure of arterial CO_2 of 112 mm Hg and a pH of 7.08 on arterial blood gas analysis. Then, the patient needed mechanical ventilation for 4 days. In conclusion, patients with HPS require strict respiratory management to support their restrictive pulmonary dysfunction, and, also, we should consider preventive management for hemostasis and adequate analgesia to reduce the patient's work of breathing.

1. Introduction

The Hermansky-Pudlak syndrome (HPS), first described by Hermansky and Pudlak [1] in 1959, is a type of oculocutaneous albinism associated with bleeding diathesis and pulmonary fibrosis. Hermansky-Pudlak syndrome is rare, with a worldwide prevalence of 1 in 500,000 to 1 in 1,000,000. Although anesthetic management is difficult in these patients, owing to bleeding diathesis and restrictive pulmonary dysfunction, few reports focused on this aspect have been published. We describe the anesthetic management of a patient with HPS who underwent video-assisted bullectomy. Our case is the first report describing anesthetic management of pulmonary thoracic surgery in a patient with HPS.

2. Case report

A 53-year-old woman (height, 156 cm; weight, 35 kg; and American Society of Anesthesiologists physical status III) with

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pulmonary fibrosis secondary to HPS was scheduled to undergo video-assisted bullectomy. She had albinism from birth and had experienced subcutaneous hemorrhage since childhood. She was diagnosed with pulmonary fibrosis at the age of 48 years. The patient was diagnosed with HPS at the age of 50 years, when genetic testing and screening were performed because of the bleeding diathesis and albinism. Although prednisolone and pirfenidone, a pyridine molecule with antiinflammatory and antifibrotic activities, were administered, the patient's pulmonary fibrosis gradually progressed, and home oxygen therapy had been initiated 3 years before surgery. During the next 2 years, she had 2 incidents of pneumothorax that were successfully treated by chest tube drainage. Her vital capacity had decreased to 42%, suggesting advanced restrictive lung disease, and she was registered as a candidate for lung transplantation.

During this follow-up period, the patient presented to the emergency department with severe dyspnea and was diagnosed with a recurrent right pneumothorax, and she did not respond well to chest tube drainage, as air leakage continued, and the lung failed to reinflate (Fig. 1). A video-assisted bullectomy under general anesthesia was planned. Preoperative chest computed tomography showed pulmonary fibrosis (Fig. 2). The preoperative platelet count and bleeding time were within normal limits, and we did not perform further examinations such as platelet aggregation tests.

The patient received no premedication. On arrival in the operating room, her baseline oxygen saturation was 90% with oxygen delivery at 2 L/min via a nasal cannula. Preoxygenation was performed with 100% oxygen for 5 minutes. After induction of anesthesia with propofol and rocuronium, we initiated mask ventilation with 100% oxygen. Although mask ventilation was difficult because of the patient's decreased pulmonary compliance, we were able to maintain an oxygen saturation of 94%-96%. Intubation was performed uneventfully

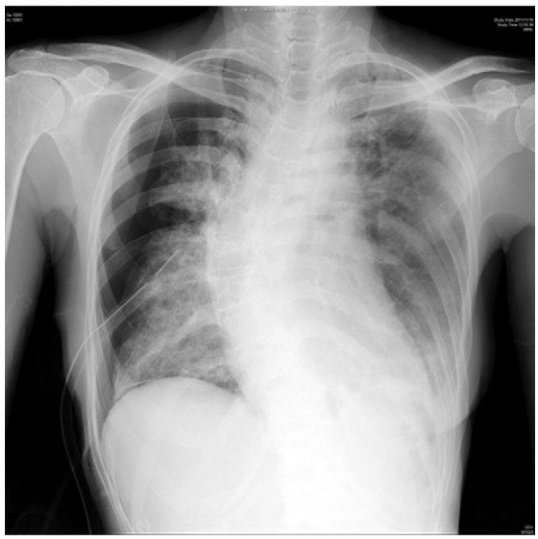

Fig. 1 Preoperative chest x-ray shows diffuse reticulonodular shadows in both lower lung fields and significant scoliosis. Pneumothorax was not improved by pleural cavity drainage.

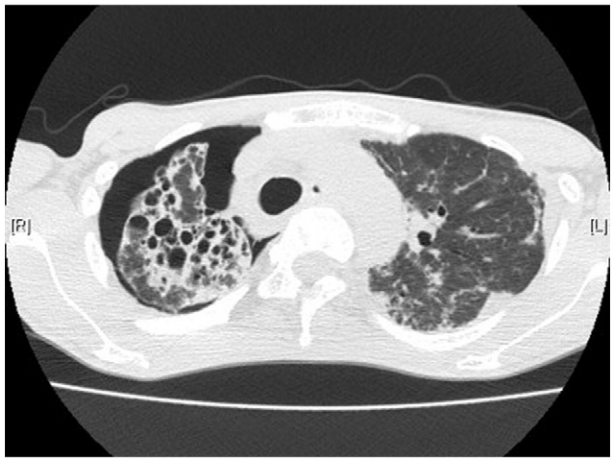

Fig. 2 Preoperative chest computed tomography at the level of the tracheal carina shows extensive pulmonary fibrosis with right pneumothorax.

using a 35F catheter left-sided Bronchocath double-lumen tube (Covidien, Tokyo, Japan). We confirmed the position of the endotracheal tube by fiberoptic bronchoscopy. The patient's tidal volume was only 250 mL under pressure-controlled ventilation, with a peak inspiratory pressure of 30 cm H_2O and a positive end-expiratory pressure of 5 cm H_2O. End-tidal CO_2 was as high as 70 mm Hg. We did not attempt one-lung ventilation.

Anesthesia was maintained by continuous intravenous infusion of remifentanil (0.1-0.3 μg/kg per minute), inhalation of sevoflurane (1.5%) in 100% oxygen, and intermittent intravenous injections of fentanyl and rocuronium. The total dose of intraoperative fentanyl was 250 μg. To maintain oxygen saturation, we did not decrease the fraction of inspired oxygen, and the intraoperative oxygen saturation was maintained between 90% and 100%. Video-assisted bullectomy for multiple bullae in the right lung was performed with the patient in the left lateral decubitus position. Because of pleural adhesions and the bleeding diathesis, which caused the surgeons difficulties in controlling continuous oozing from the surgical incision site, the procedure lasted for 130 minutes, and intraoperative blood loss and urine output were 270 mL and 200 mL, respectively. The patient received 2400 mL of intraoperative fluid (68 mL/kg). Intraoperative hypotension was corrected by fluid administration and intermittent intravenous infusion of phenylephrine, but the patient did not require continuous intravenous infusion of catecholamine.

At completion of the surgery, we switched the ventilator mode to spontaneous ventilation with continuous positive airway pressure and pressure support. However, patient-ventilator asynchrony developed, and the patient's breathing was insufficient even after reversal of neuromuscular blockade by sugammadex. She had a low tidal volume and tachypnea. The arterial blood gas analysis showed a partial pressure of arterial CO_2 of 112 mm Hg with a pH of 7.08, and we resumed mandatory ventilation to prevent progression of acute respiratory acidosis. We could not obtain sufficient tidal