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Anesthetic Management of a Patient with Poland's syndrome: A Case Report

Young Eun Jun, Yun Do Jung, Lee Kyeong Kang and Kyu Nam Kim

ABSTRACT

Poland syndrome is a rare congenital disease characterized by various degrees of chest wall defects, such as deficits in the small thoracic muscle and defects in the neuromuscular region of the small thoracic muscle. In addition, this condition may present with loss or weak development of the papilla and breasts and can be accompanied by brachydactyly, syndactyly, and limb defects in the hands and feet on the same side. Poland syndrome also may cause various deformities in the heart, lungs, and kidneys. These lung deformities can produce hypoventilation and hypoxia as the function of the respiratory muscle decreases, leading to increased pulmonary complications postoperatively. Since these patients also have an increased risk of malignant hyperthermia, careful attention is required in the selection of drugs for inducing general anesthesia. We would like to report an experience of general anesthesia for breast reconstruction in a 20-year-old female patient with Poland syndrome.

Keywords: General anesthesia, Malignant hyperthermia, Poland syndrome.

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K. N. Kim*
Hanyang University Hospital, Seoul, Republic of Korea.
(e-mail: vesicle100@naver.com)
Y. E. Jun
Hanyang University Hospital, Seoul, Republic of Korea.
(e-mail: youngeun5870@gmail.com)
Y. D. Jung
Hanyang University Hospital, Seoul, Republic of Korea.
(e-mail: fcfrankfurty@gmail.com)
L. K. Kang
Hanyang University Hospital, Seoul, Republic of Korea.
(e-mail: valen527@naver.com)

****Corresponding Author***

I. Introduction

Poland syndrome is a rare congenital anomaly characterized by the absence of part of one of the major chest muscles, breasts, or shoulder muscles and rib cage abnormalities [1] and can lead to asymmetric lung sizes. Many people with Poland syndrome also have hand abnormalities on the affected side like brachydactyly, syndactyly, and vestigial fingers. The incidence of Poland syndrome ranges from 1 in 7,000 to 1 in 100,000, and males are affected more frequently than females by a 2:1 to 3:1 ratio [2]. There is a higher preponderance of patients with abnormalities in the right thoracic and upper limbs than on the left. [1] Poland syndrome is a nongenetic congenital disorder with a low (<1%) risk of reoccurrence in the same family [2].

The anesthetic management of patients with Poland syndrome is complicated by the risk of hypoxia that can result from any existing lung deformities. In addition, structural deformities can hinder airway management, especially in children. Poland syndrome also carries a risk of malignant hyperthermia (MH) [3]. Herein, we describe the anesthetic management of a patient with the rare Poland syndrome throughout the perioperative period.

II. Case Presentation

A 20-year-old female patient with right-side Poland syndrome was scheduled for breast reconstruction surgery at our facility. At the time of surgery, she was 162 cm tall and weighed 72 kg. She had a history of multiple surgeries for hand sclerodactyly relaxation without any complications. Preoperative chest computed tomography (CT) scanning revealed hypoplasia of the left pectoralis major and latissimus dorsi muscle, asymmetry of the breasts, and no abnormalities of the ribs (Fig. 1A and 1B). No abnormalities of the lungs were visible on chest radiography (Fig. 2). Although the patient reported symptoms of dyspnea and dyspnea on exertion, there was no paradoxical respiration. She had normal neck movements and a Mallampati score of II. Preoperative laboratory testing revealed hemoglobin: 12.6 g/dL, hematocrit: 38.4%, white blood cell count: 8000 mm3, platelet count: 357,000 mm3, prothrombin time: 10.8 sec (109%), international normalized ratio: 0.95, platelet adhesion-aggregation time: 137 sec, and activated partial thromboplastin time: 35 sec.

On the day of surgery, the patient was taken to the operating room after administration of intramuscular midazolam 2.5 mg and atropine 0.5 mg as premedication. Routine monitoring was established, including non-invasive

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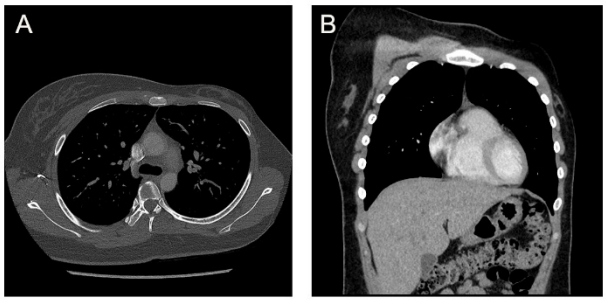

Fig. 1. Chest computed tomographic images of a patient with Poland syndrome showing asymmetry of the breasts and hypoplasia of the left pectoralis major and the latissimus dorsi muscle. (A) The axial view and (B) the coronal viewMagnetization as a function of applied field.

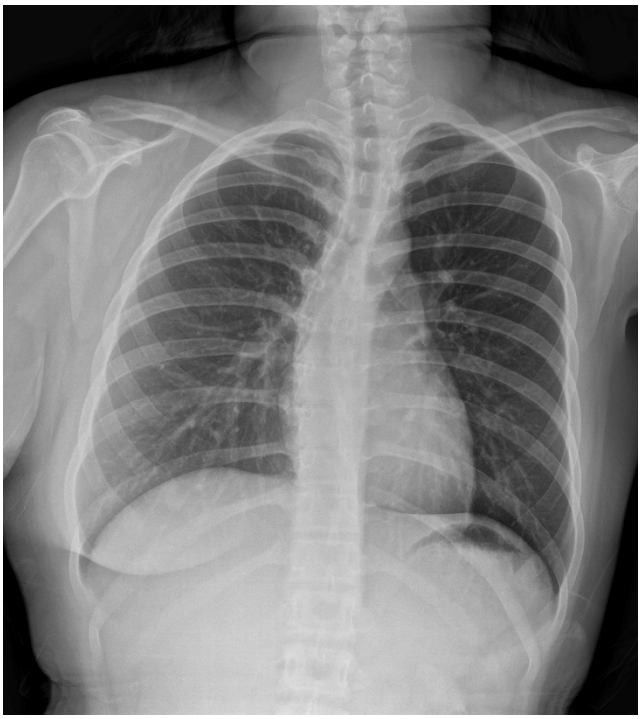

Fig. 2. Preoperative chest radiography showing no active lung lesion.

On the day of surgery, the patient was taken to the operating room after administration of intramuscular midazolam 2.5 mg and atropine 0.5 mg as premedication. Routine monitoring was established, including non-invasive blood blood pressure (NBP), heart rate (HR), pulse oximetry (SpO2), electrocardiography (ECG), and invasive blood pressure (IBP) monitoring. Her initial NBP, HR, and SpO2 readings were 135/80 mmHg, 80 beats per min, and 98%, respectively.

General anesthesia was induced with total intravenous anesthesia (TIVA) and target-controlled infusions (TCIs) using an Orchestra® Base Primea TCI pump (Fresenius Kabi). Propofol and remifentanil were infused at target concentrations of 4.0 μg/mL and 3.0 ng/mL, respectively. Tracheal intubation was safely performed using the McGRATH™ video laryngoscope after injection of 40 mg of rocuronium. Maintenance of anesthesia was performed with propofol (3-4 mcg/mL) and remifentanil (1.5-3.5 ng/mL). The Bispectral Index score was measured at 40-60, and SpO2 was maintained between 98–100%.

There were no specific adverse events noted during the surgery. There was no increase in expiratory CO2 level. In addition, there were no hyperthermia episodes or any other signs of MH. The total anesthetic time was 2 hours 30 minutes, and the estimated blood loss was 50 mL. After completion of the operation, 15f0 mg of sugammadex was used to reverse the muscle relaxation. After extubation, the patient was transferred to the post-anesthesia care unit and discharged without any complications.

III. Discussion

The most common clinical feature of Poland syndrome is the absence of the sternocostal head of the pectoralis major muscle; the absence of the entire pectoralis minor muscle is also occasionally observed [1]. Despite these characteristics, most of these patients demonstrate no functional impairments [1]. Other deformities that may accompany a defect in the thoracic muscle include abnormal ribs, abnormal scapulae, abnormal upper arms, abnormal radial bones, brachydactyly, syndactyly, scoliosis, atrial septal defects, diaphragmatic hernias, renal agenesis, hepatobiliary defects, inguinal hernias, and hematologic disorders like acute leukemia [4], [5]. Fortunately, no abnormalities other than hypoplasia of the left pectoralis major and the latissimus dorsi muscle were observed in this patient.

In some Poland syndrome patients, chest deformities cause paradoxical chest wall motion. There is typically no paradoxical movement in the resting state, but paradoxical chest wall motion may occur during anesthesia. Therefore, even if a simple operation is performed, positive pressure ventilation should be applied when sedation or anesthesia is required. In addition, during the sixth week of gestation when the upper limb bud is in a stage of development, the embryonic blood supply is interrupted, which causes hypoplasia of the ipsilateral subclavian artery or its branches and explains the etiology of Poland syndrome. As a result, the subclavian artery size is decreased (<50% in diameter), and its blood flow velocity is lowered [1]. Therefore, when it is necessary to insert a central venous catheter, it should be placed on the unaffected side; similarly, it is safe to perform a brachial plexus block on the unaffected side [6].

In general anesthesia in patients with Poland syndrome, those with thoracic deformities (especially children) may have a difficult airway to intubate. Therefore, appropriate anesthesia management should be carefully planned through evaluation of chest movement related to breathing and, radiologic imaging to assess tracheal deviation or stenosis preoperatively. Although the patient in this case had some defects in the chest wall, the lung on that same side was normal, and there were no functional abnormalities related to lung function. Therefore, problems associated with lung function during anesthesia were predicted to be minor, and anesthesia management was planned with a specific focus for anticipating the occurrence of MH [3].

All inhalation anesthetics (e.g., halothane, ether, desflurane, sevoflurane, isoflurane) except nitrous oxide have been reported as MH triggers, and the depolarizing neuromuscular blocking agent succinylcholine is associated with adverse events graded as ‘ ‘very likely’’ or ‘‘almost certainly’’ to induce MH [7]. All anesthetic agents other than volatile anesthetic agents and depolarizing neuromuscular blocking agents are considered safe from an

MH point of view [8]. In addition to these volatile agents, Poland syndrome itself also increases the risk of MH [1], [3]. Therefore, TIVA is preferred for maintenance of general anesthesia using propofol and remifentanil. Additionally, monitoring of the body temperature and end-tidal CO2 are necessary to check for the occurrence of MH.

In conclusion, patients with Poland syndrome require a detailed physical examination because chest wall defects and their accompanying malformations can occur. It is also necessary to identify the associated deformities in advance and prepare for associated problems. Clinicians should be fully aware of the risk of lung complications and MH during anesthesia for patients with Poland syndrome.

IV. Conclusion

In conclusion, patients with Poland syndrome require a detailed physical examination because chest wall defects and their accompanying malformations can occur. It is also necessary to identify the associated deformities in advance and prepare for associated problems. Clinicians should be fully aware of the risk of lung complications and MH during anesthesia for patients with Poland syndrome.

Conflict of Interest

Authors declare that they do not have any conflict of interest.

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