

Anaesthesia in Poland syndrome: A case report

Poland sendromunda anestezi: Olgu Raporu

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ABSTRACT

Poland syndrome is a rare congenital anomaly. It includes in different anomalies such as musculo-skeletal system, heart, kidneys and lungs. This syndrome is characterized by thorax deformity most commonly. Ipsilateral hand anomalies are most important features and can be seen as syndactyly or ectrodactyly. Patients may have lung complications due to the use of muscle relaxants, which is used induction and maintenance of general anesthesia. In addition, these patients show increased risk of malignant hyperthermia during general anesthesia. Because of malignant hyperthermia risk, general anesthesia should be provided carefully. In this report, we discuss the anesthetic concerns related to the management of a 23-year-old female patient with right-sided Poland syndrome undergoing breast reconstruction surgery. *J Clin Exp Invest* 2014; 5 (4): 608-609

Key words: Poland syndrome, anesthetic management, congenital anomaly

INTRODUCTION

Poland syndrome is a congenital anomaly that is not seen common. The incidence of Poland syndrome is 1/ 30,000 to 1/50,000 live births [1]. Typical features of the syndrome include cardiovascular (dextrocardia, atrial septal defect), musculo skeletal (contralateral syndactyly, clubfoot, toe syndactyly, hemivertebrae and scoliosis), genitourinary (renal aplasia, hypospadias), gastrointestinal (situs inversus) and haematopoietic (spherocytosis and myelogenous leukaemia, lymphoma) abnormalities [2]. Patients with Poland syndrome often do not have functional defect. Patients can present cosmetic surgery such as breast reconstruction in anesthetic practice [2].

ÖZET

Poland sendromu nadir görülen bir konjenital anomalidir. Kas iskelet sistemi, kalp, böbrek ve akciğerlerle ilgili birçok anomaliyi içerir. Bu sendrom sıklıkla göğüs deformitesi ile karakterizedir. İpsilateral el anomalileri en önemli özelliği olup sindaktili ve ektrodaktili gibi el anomalileri de beraberinde görülebilir. Hastalarda genel anestezi induksiyonu ve idamesinde kas gevşetici kullanımına bağlı akciğer komplikasyonları gelişebilir. Ayrıca genel anestezi bu hastalarda malign hipertermi gelişme riskini artırır. Malign hipertermi ve solunumsal komplikasyon gelişebilmesi riski nedeniyle genel anestezi dikkatli bir şekilde uygulanmalıdır. Biz Poland sendromlu, meme rekonstrüksiyon cerrahisi uygulanan 23 yaşında bayan hastanın anestezi yönetimini sunmayı amaçladık.

Anahtar kelimeler: Poland sendromu, anestezi yönetimi, konjenital anomali

CASE

A 23 year-old female patient (height 161 cm; weight 49 kg) with right-sided Poland's syndrome was scheduled for breast reconstruction surgery. On physical examination before operation physical findings was a right-sided chest wall defect, absence of the pectoralis major and pectoralis minor, rudimentary development of 3rd rib and 2, 3, 4, 5 syndactyly of the fingers that were operated before (Figure 1). There were no abnormalities in other organ systems. Paradoxical respiration did not determined. It is normal for both heart and lung auscultation. Respiratory function tests revealed that there was no restriction. Also echocardiography was evaluated as normal too. Preoperatively with the approval of the patient we decided to apply general anaesthe-

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sia and she was informed about anaesthesia type. On the day of operation, the patient was taken to the operating table after midazolam premedication. The preoperative oxygen saturation was 96%, heart rate was 89 beats/min and arterial pressure was 123/65 mmHg. Propofol (2 mg/kg) and fentanyl (2 µg/kg) was used for induction of general anaesthesia. To endotracheal intubation rocuronium (0.6 mg kg⁻¹) was used. Anaesthesia was maintained with TIVA (Total intravenous anaesthesia) that is used propofol (6 mg/kg/h) and remifentanyl (0.25 µg/kg/min). We monitored the patient's body temperature and end-tidal CO₂. The total duration of anaesthesia was 4 hours. The patient was extubated in the operating room and transferred to recovery room. There were no complications experienced during operation and postoperative period.



Figure 1. Patient's thorax and limb anomalies

DISCUSSION

General anaesthesia can cause malignant hyperthermia in some congenital diseases such as Poland syndrome, Kabuki makeup syndrome [3], Arthrogryposis multiplex congenita [4] due to musculoskeletal abnormalities. Particularly succinylcholine and inhalational agents usage increase the risk of malignant hyperthermia. Because of this reason these agents should be contraindicated during general anaesthesia in patients with Poland syndrome [5]. In this current case report TIVA is preferred for maintenance of anaesthesia, rocuronium is used for muscle relaxation rather than succinylcholine and body temperature and end-tidal CO₂ was monitored because of malignant hyperthermia risk [6].

Patients with Poland syndrome can be asymptomatic or paradoxical breathing. It is possible that if there were no symptom at rest there would be paradoxical breathing under anaesthesia [2]. Therefore the patients should be ventilated via positive

pressure ventilation. In our case we used positive pressure ventilation and did not witness paradoxical breathing during anaesthesia and after extubation. The paradoxical respiration may cause hypoxia [7]. We think that the chest deformity was not severe in our case. Therefore paradoxical respiration did not occur.

A clinical presentation of Poland syndrome is situs inversus. Dextrocardia can be a part of situs inversus in some cases. Bavinck reported that dextrocardia was demonstrated 5.6% and left-sided defect was 9.6% in 144 patients. In this case, the defect was right-sided and dextrocardia was not observed [8].

In conclusion, musculo skeletal abnormalities as Poland syndrome are important in anaesthesia practice. This patient should be evaluated cautiously. The possible complications such as malignant hyperthermia and hypoxia should be considered. Also we should avoid succinylcholine and inhalational agents to prevent malignant hyperthermia, should use controlled respiration to prevent hypoxia.

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