

Spinal anesthesia in Kartagener's syndrome

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ABSTRACT

Kartagener's syndrome, an autosomal recessive disorder is a combination of dextrocardia (situs inversus), bronchiectasis and sinusitis. We report a 22-year-old woman with this syndrome scheduled for appendectomy. Spinal anesthesia was preferred for the patient with this rare disorder due to the relative advantages of the regional technique over general anesthesia.

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Kartagener's syndrome (KGS) is a rare inherited disorder including dextrocardia (situs inversus), bronchiectasis, and chronic paranasal sinusitis. It is transmitted as an autosomal recessive trait. The clinical features of this syndrome are productive cough, respiratory tract infections, sinusitis, otitis media and infertility.¹ Bronchiectasis and chronic infections may result in end-stage pulmonary disease.² The implications of the syndrome for anesthesia are related to the respiratory system and increased predisposition to infections. Here, we aimed to report the anesthetic management of a patient with KGS and polycystic ovarian syndrome (PCOS) who underwent surgery for acute appendicitis.

Case Report. A 22-year-old woman with acute appendicitis was scheduled for appendectomy. The patient was a known case of KGS with ciliary dyskinesia, bronchiectasis, and chronic sinusitis. Concomitant disease included PCOS. She had once suffered from deviation of the nasal septum, had undergone septorhinoplasty 2 years ago, and was hospitalized for the following 3 weeks for the treatment of the pneumonia that had begun in the early postoperative period. Pre-anesthetic evaluation

revealed PCOS and a history of bronchitis. She had coughing, wheezing and dyspnea. Her breathing rate was 26 per minute. Chest examination revealed a decrease in the breathing sounds and bilateral basal crepitations. Chest radiography confirmed bilateral infiltrations in the middle and lower zones of the lungs (**Figure 1**). Other routine biochemical and blood investigations were normal. Spinal anesthesia was planned. The patient was preloaded with lactated Ringer's solution. Monitoring of continuous ECG, non-invasive blood pressure, and pulse oximetry was established. The patient was placed in the sitting position. Spinal anesthesia was administered by bupivacaine 0.5% 2 ml plus fentanyl 20 µg through the L3-L4 interspinous space. The patient was positioned supine, and 6 L.min⁻¹ oxygen was delivered through a nasal oxygen cannula. No prophylactic antibiotic was administered as amoxicillin 1 gr intravenously was given just before the surgery as a prophylaxis for pneumonia. Ephedrine 10 mg was administered intravenously for the prophylaxis of hypotension. Fentanyl 1 µg kg⁻¹ and midazolam 4.5 mg was administered intravenously during the surgery for sedation. The maximum sensory level achieved was T8. The surgery lasted for 50 minutes, and the

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Figure 1 - Chest radiography showing bilateral infiltrations in the middle and lower zones of the lungs.

hemodynamic parameters were stable during this time. Postoperatively, diclofenac was given twice a day for analgesia and the patient was discharged from hospital on the fourth postoperative day.

Discussion. Kartagener's syndrome is a variant of immotile cilia syndrome, and is known as primary ciliary dyskinesia. It is characterized by the presence of chronic sinusitis, bronchiectasis, and dextrocardia (situs inversus). It is a result of an ultrastructural defect of the cilia, specifically a defect in the synthesis of dynein protein.² This abnormality results in ciliary immobility with impaired mucociliary clearance in the airway and sinuses, and reduced motility of sperm. Clinical symptoms of KGS include productive cough, chronic or recurrent respiratory tract infections, sinusitis, otitis media, bronchiectasis, and male infertility.^{3,4} The main anesthetic considerations are related to the respiratory system. These include preoperative respiratory tract infections, obstructive pulmonary function pattern, and increased risk of postoperative pulmonary infections causing increased morbidity. Considering the risk of general anesthesia including atelectasis, prevention of coughing reflex, increase in airway secretions, and aspiration resulting in perioperative respiratory tract infections, we preferred regional anesthesia for our patient. By using regional anesthesia in KGS, it is possible to protect respiratory muscle functions. In addition, keeping the level of blockade at T6-8 protects pulmonary functions. However, asepsis during the regional anesthesia procedure should be stressed as these patients have abnormal neutrophil chemotaxis.⁵ Therefore, we did not prefer combined

spinal epidural or epidural anesthesia to avoid catheter placement. Patients with KGS usually undergo surgery for pulmonary disorders, sinusitis, otitis, and cardiac disorders. There are very few reports of anesthesia in such patients. In some patients with KGS, general anesthesia was used, however for some of them regional anesthesia was preferred.^{1,5-7} Reidy et al,¹ reported postoperative respiratory tract infection following general anesthesia for diagnostic laparoscopy in a patient with KGS. In our patient, the procedure did not last too long and postoperative pain could be managed without the epidural route. In the view of these factors, and since her last procedure had been complicated by pneumonia, the spinal anesthetic technique was the best choice for our patient wherever feasible. Preoperative evaluation of pulmonary and cardiac anatomy and functions is essential in these patients.² Fifty percent of these patients have situs inversus. Dextrocardia is usually not associated with cardiac anomalies when it occurs with situs inversus. If dextrocardia occurs with situs solitus or situs ambiguous, the incidence of congenital cardiac diseases increases up to $\geq 90\%$.⁸ These cardiac malformations include ventricular inversion, single ventricle, pulmonary stenosis, abnormal atrioventricular valves and anomalies of systemic and pulmonary venous return.⁹ Our patient had no dextrocardia according to ECG, chest radiography, and auscultation. Considering the risks of general anesthesia, including potential hepatic dysfunction with exposure of inhalation anesthetics, perioperative respiratory tract infection, and abnormal neutrophil chemotaxis, we considered regional anesthesia as the best technique for our patient. The major advantage of the regional technique is that it has no influence on respiratory muscle functions intraoperatively and postoperatively, also it aids clearance of airway secretions effectively. There are very few reports about sufficiency of epidural anesthesia in KGS.^{1,7} We did not apply epidural anesthesia because of risk of epidural catheter infection as these patients have abnormal neutrophil chemotaxis. In addition, risks of elevation of blockade level appear in epidural analgesia. Early ambulation helps to prevent retention of respiratory tract secretions in these patients. Postoperative analgesia should be managed to contribute in early ambulation that will help the clearance of the secretions of the airway. The pulmonary functions may be depressed by opioid analgesics including morphine, meperidine or fentanyl. Optimal postoperative analgesia with avoidance of excessive sedation is needed in such patients. Diclofenac can effectively be used for this purpose. Therefore, in our patient, postoperative

analgesia was achieved with 75 mg diclofenac sodium twice daily intramuscularly with a Visual Analog Score of less than 40 mm. The postoperative period was uneventful, and the patient was discharged from hospital on the fourth postoperative day.

As a conclusion, preoperative evaluation of pulmonary and cardiac functions and anatomy is essential in patients with KGS. Regional anesthetic techniques should be considered in these patients because of the potential risks of general anesthesia. In addition, the analgesic drugs with no adverse effects on pulmonary function should be preferred for the management of postoperative pain.

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