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LETTER TO THE EDITOR

Primary ciliary dyskinesia: a case of complete Kartagener's syndrome in a patient undergoing cesarean section

Dear Editor,

We recently anesthetized a patient with complete Kartagener's syndrome undergoing a cesarean section and would like to share our experience conducting this rare case.

Kartagener's syndrome is a subtype of primary ciliary dyskinesia, a rare genetic disorder and its pathophysiology involves the dysfunction or lack of arms of the ciliary protein dynein, responsible for the mechanics of ciliary movement, causing chronic respiratory symptoms. The triad bronchiectasis, rhinosinusitis, and situs inversus, with or without dextrocardia, which defines the syndrome as complete or incomplete, is classic for the diagnosis of the disease [1]. In women, ciliary dyskinesia of the fimbriae impairs the conduction of the oocyte along the uterine tubes, increasing the chance of infertility and ectopic pregnancy [2]. Due to the fertility problems in Kartagener's syndrome, a cesarean section is a very rare situation for anesthesiologists. In a recent published review of 99 cases, only two were in obstetric patients [3].

A 34-year-old patient with complete Kartagener's syndrome and ongoing planned pregnancy resulting from natural conception was schedule for an elective cesarean section, according to her will. She had previously undergone two surgical procedures under general anesthesia: sinus clearance due to chronic pansinusitis, and laparoscopic gastric fundoplication, with no cardiac or respiratory complications. She reported that her respiratory status did not worsen during pregnancy and the use of nasal formoterol and budesonide was maintained.

On physical examination, she presented abundant mucopurulent sputum, rhinorrhea with phonatory alteration, digital hippocratism, and moderate edema of the lower limbs. Lung auscultation revealed rhonchi and rales globally and bilaterally. On cardiac evaluation, she had regular heart rhythm and heart sounds were normal but better audible in the right hemithorax, with no murmurs. The 12-lead electrocardiogram showed sinus rhythm and P and T wave inversions in the DI lead. The echocardiogram showed the situs inversus with dextrocardia, mild mitral insufficiency, and an ejection fraction of 63%. The chest X-Ray showed the dextrocardia and relatively hyperinflated lungs (Fig. 1a). The computed tomographic scan showed situs inversus with dextrocardia, pulmonary consolidations, atelectasis, bronchiectasis, and areas of bilateral mucoid impaction (finger-in-glove sign) in the lung bases (Fig. 1b). Spirometry showed a mild mixed ventilatory disorder, with no alteration after the bronchodilator. The patient was informed of the risks inherent to any anesthetic procedure, as well as clarification about the possible anesthetic techniques. We opted for spinal anesthesia after the patient gave her informed consent.

According to obstetric recommendation, oral prednisolone, 5 mg per day, was added for five days prior to delivery. Elective cesarean section was carried out in the 39th week of pregnancy. Standard monitoring showed initial parameters within normal range and oxygen saturation equal to 98%, in room air. Two peripheral venous accesses were obtained, and a lactated Ringer's solution was administered. Antibiotic prophylaxis was performed with intravenous cefazolin, 2 g. The patient was also given intravenous hydrocortisone, 500 mg, omeprazole, 40 mg, and oxygen via a nasal catheter, 2 l min^{-1}.

The spinal anesthesia was performed with the patient in the sitting position with a 27G Quincke needle in the intervertebral space L_3–L_4, using 0.5% hyperbaric bupivacaine, 15 mg, and morphine, 50 μg. Due to the situs inversus, the uterus was displaced to the right for aortocaval decompression. During the procedure, overall, three doses of metaraminol, 0.5 mg, were administered to treat hypotension. Oxygen saturation remained between 98–100% and the heart rate was above 60 beats.min^{-1}. Pulmonary auscultation remained with rhonchi and bibasilar wheezing. Soon after the birth and clamping of the umbilical cord, oxytocin, 10 units, was administered intravenously. Intravenous ondansetron, 4 mg, dimenhydrinate, 30 mg, tramadol, 50 mg, and methamizole, 2 g, were also administered. The total volume of lactated Ringer's solution at the end of the procedure was 1500 ml. A female infant was born in very good conditions and the patient was discharged 48 hours after delivery with no complaints and no change in her breathing pattern.

The fertility problems that occur in primary ciliary dyskinesia and Kartagener's syndrome prevent most patients from successfully getting pregnant, which accounts for the

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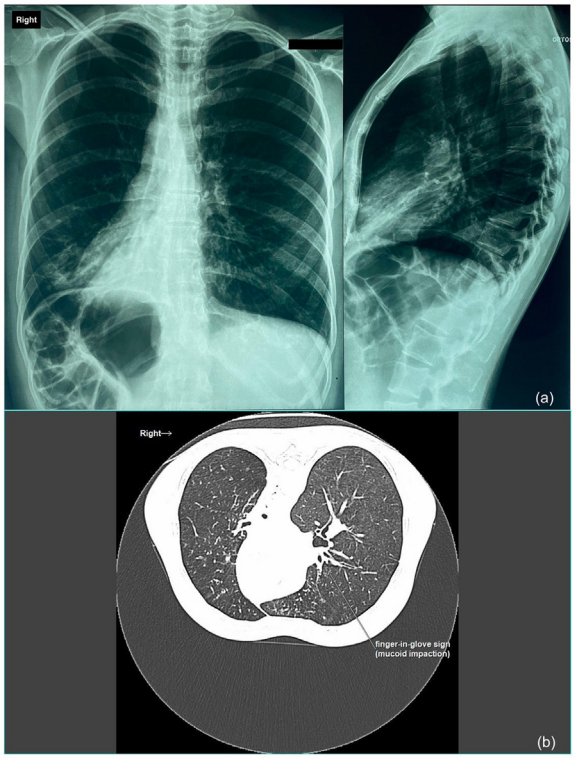

Figure 1 X-ray and CT Kartagener.

scarcity of studies on anesthetic procedures in obstetrics, particularly in patients with respiratory dysfunction and severe heart disease. As for the most advised anesthetic recommendation, it is suggested the one with the least airway manipulation associated with the most efficient postoperative analgesia. In light of this, the use of regional anesthesia becomes a priority over general anesthesia whenever possible [4]. In view of the frequent lung impairment, some measures can be taken to prevent respiratory complications involving bronchoaspiration prophylaxis, the use of antibiotics and bronchodilators, and respiratory physiotherapy [5].

Several physiological alterations resulting from pregnancy are relevant to the anesthesiologist, such as reduced functional residual capacity, supine hypotension, and greater cephalic dispersion of local anesthetics in the subarachnoid space. Considering these alterations and the possible reduced cardiovascular and pulmonary functions due to the syndrome, the effects of the neuraxial anesthesia may be exacerbated. Thoracic and lumbar sympathetic blockade, mainly at higher spinal levels, aggravates hypotension by decreasing peripheral vascular resistance and preload, the latter already reduced by aortocaval compression. Moreover, dorsal decubitus and uterine compression further limit diaphragmatic contraction and lung function. If any invasive airway intervention is required, bronchial reactivity may be accentuated in the airway already compromised by the disease and result in upper and lower airway obstruction.

We provided adequate hemodynamic control, with volume replacement and use of vasopressor, besides adequate uterine displacement (situs inversus) and no invasive airway interventions were required to maintain blood oxygenation.

Despite the lack of literature, apparently, there is no contraindication for labor in Kartagener's syndrome. It basically will depend on the patient's cardiovascular and respiratory status and if these conditions are compatible with the physiological overload seen in the progressive stages of the labor.

Kartagener's syndrome comprises several challenges and the support to these patients should include preanesthetic evaluation and care directed to the main possible aspects of perioperative decompensation of the disease, especially respiratory function. Neuraxial anesthesia seems to be a good alternative for patients with Kartagener's syndrome undergoing cesarean section, a very rare situation for these patients.

Conflicts of interest

The authors declare no conflicts of interest.

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