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Case Report

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Total Intravenous Anesthesia for Immobile Cilia Syndrome. About a Case

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Abstract

Immotile cilia syndrome is an autosomal recessive disorder which is characterized by ciliary dysfunction and decreased mucociliary clearance. This ciliary function is a mechanism whose primary pulmonary defense alteration can be a problem during inhalational anesthesia, as these cause decreased ciliary movement. However during total intravenous anesthesia this normal ciliary movement can be preserved using remifentanil and propofol. A case of immotile cilia syndrome was handled with complete intravenous anesthesia for surgery protocol type ovarian handling post-operative pain epidural catheter is presented.

Keywords: Immobile Cilia Syndrome, Mucociliary Clearance, Total Intravenous Anesthesia.

Introduction

The immobile cilia syndrome also called Kartagener syndrome or primary ciliary dyskinesia is an autosomal recessive heterogeneous genetic disorder located on chromosome 9p21-p13 characterized by ciliary dysfunction, decreased mucociliary clearance resulting in chronic bronchitis that leads to bronchiectasis, chronic rhinosinusitis, chronic otitis media, situs inversus (50% of cases) and male infertility. The first cases were described at the beginning of 1900, where there was a triad characterized by chronic sinusitis, bronchiectasis and situs inversus. Then it was evidenced that these patients had immobility of the cilia and this is where this term is associated with the name of the syndrome, later it is discovered that the cilia are not immobile but describe a rigid, uncoordinated or inefficient rhythm modifying this syndrome to dyskinesia primary ciliary which constitutes the term currently accepted [1,2]. The incidence of primary ciliary dyskinesia in the white population is 1 per 40,000. Mutations in 08 genes have been identified up to now (DNAI1, DNAH5, DNAH11, DNAI2, KTU, RSPH9, RSPH4A and TXNDC3). The primary defect is the dysfunction of the structure of the axoneme, which is constituted by multiple proteins so that the Disorder is very heterogeneous. Of these proteins, the dynein is the most studied whose most common alteration is the loss of the external arms [1,3,4].

The most important anesthetic consideration in the immobile cilia syndrome is the evaluation of possible cardiovascular and pulmonary complications [5].

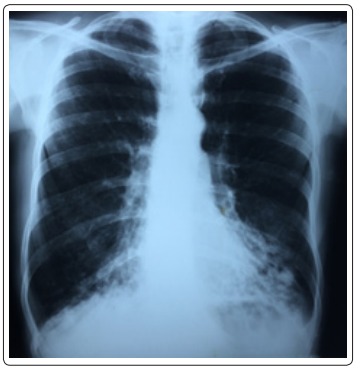
Clinical Case

This is a 50-year-old female patient with a diagnosis of immobile cilia syndrome for 30 years who goes to the general surgery service at Central Hospital of Maracay, Aragua State-Venezuela, for presenting diffuse abdominal pain and performing clinical and paraclinical evaluation. Cystic LOE is evidenced in retro vesical pelvic excavation of probable adnexal etiology, so they request for laparotomy type ovarian cancer diagnosis protocol with extemporaneous biopsy.

Preoperative evaluation: Personal Background: Diagnostic of immobile cilium in 1988 through electron microscopy and spectrophotometry. Surgical history: thyroidectomy in 2011 and hysterectomy in 2010 both with general anesthesia without complications.

To the physical examination: Weight: 40 kgs TA: 100 / 70mmHg FC: 92 lpm FV: 18rpm. Airway evaluation: patient with prominent maxilla. Thorax: symmetric hypoexpansible respiratory noises present with sibilant aggregates in the left hemithorax, predominantly basal.

Paraclinical: HB: 12.4g / dl HTO: 40.8% LEU: 12,000 / mm3 PTL: 329,000 / mm3 PT: 1.08 PTT: 0 sec GLYCEMIA: 84 mg / dl Urea: 18 mg / dl Creatinine: 0.8 MG / DL HIV: Negative VDRL: Not reactive TSH: 3.2mUI / ml T3L 2.53 pg / ml T4L 0.87 ng / dl.



X-RAYS OF THORAX: Figure 1: PA chest x-ray showing bilateral basal infiltrate with left predominance of inflammatory type

Evaluation by endocrinology: patient without hormone replacement since February 2016, currently euthyroid without surgical contraindication

Evaluation by Pulmonology: Patient who attends in January 2016 with cough with whitish expectoration and respiratory difficulty at moderate efforts. Physical examination showed wheezing predominantly at the left basal level and radiological findings of bilateral basal infiltration indicated treatment with: Levofloxacin, Formoterol + Budesonide and respiratory physiotherapy. At the end of treatment and revaluation not contraindicated for surgical intervention and suggests: Nebulotherapy with short-acting beta 2 before and after surgery, cortico intravenous steroids and respiratory physiotherapy.

Spirometry: mixed type severe obstructive alteration

Anesthetic management: ranitidine 50 mg, metoclopramide 10 mg and hydrocortisone 500 mg were administered as pre-anesthetic impact medication. Standard monitoring was placed in the operating room: TA: 110/70 mmHg FC: 60 lpm SatO2: 100%. Patient is placed in left lateral decubitus previous asepsis and antisepsis is infiltrated L2-L3 is approached epidural space with 16G tuohy needle. Picking and positive Nessi signs are placed 16G epidural catheter. The patient is repositioned in dorsal decubitus and after denitrogenization for 05 minutes induction is performed with: Lidocaine 40 mg, Propofol 100 mg, Rocuronium 40 mg and remifentanil 20 mcg. After administration of 40 mg of rocuronium, 60 seconds are expected and laryngoscopy with Macintosh sheet number 3 orotracheal intubation with tube 6.5 mm (ID) without any complication in the first attempt. The position of the endotracheal tube was confirmed by the auscultation of respiratory sounds and evidence of capnography wave. The ventilatory parameters VT were kept: 7-10 ml / kp I: E 1: 1.5 FV: 10-12 rpm with FiO_2: 0.6 (air 1.5 l / min and O_2 1.5 l / min).

Maintenance is carried out with infusion with propofol previously prepared at a concentration of 10mg / cc, dosing 1 mg / kp / hour and remifentanil at a concentration of 10 mcg / cc, dosing 0.15 mcg / kp / min. The concentration of remifentail and propofol is adjusted keeping the bispectral index between 40 - 60. The surgery lasted 90 minutes, it was decided to perform left oophorectomy by extemporaneous biopsy because it was negative. There was no need to use a new dose of neuromuscular blocker, 30 minutes before the end of surgery, a mixture (fentanyl + bupivacaine) was placed via the epidural catheter. At the end of the surgery, the infusions stop. It is reverted with neostigmine 0.5 mg + atropine 0.25 mg. The patient is extubated after aspiration of secretions in the oropharynx and when this presents a clinical TOF> 0.9. He moved to the post anesthetic care unit where he stayed for 45 minutes, during which he started respiratory physiotherapy. The mixture was placed every 6 hours and the catheter was removed at 24 hours. Patient was discharged 48 hours after surgery without complications.

Discussion

The immobile cilia syndrome is caused by ciliary dysfunction that results from a defect in the ultra structure of the same, which causes an alteration in mucociliary clearance which constitutes an important defense mechanism of the airways [5]. Anesthetic considerations are varied can be used the conductive or general anesthesia and the choice of one or the other depends on the lung function integrity, as well as the type of surgery [5]. In view of having an adequate preoperative preparation, since there is a bibliography that sustains that to administer general anesthesia in this type of patients in addition to pulmonary function it is necessary to consider respiratory physiotherapy, antibiotic prophylaxis [6]. By fulfilling these parameters and by the type of surgery this anesthetic technique is decided. Inhalation anesthetics decrease the frequency of ciliary movements [7,8]. There are studies that show that the decrease in these ciliary movements is greater with the use of Sevoflurane and remifentanil than with the use of propofol and remifentanil and others that report a marked decrease in motility with isoflurane than with propofol and alfentanil [3,7]. It was shown that intravenous anesthetics have no impact on ciliary mobility and propofol stimulates ciliary mobility through the release of nitric oxide from vascular endothelial cells, which in turn causes the release of guanosine monophosphate cyclical whose role is important in the regulation of ciliary mobility [3,4,9]. Because the use of long-acting opioids for pain management is associated with respiratory depression and can lead to decreased thoracic expansion and ineffective coughing which leads to atelectasis, hypoxemia and even nosocomial infection, the use of ultra-short-acting opioids is recommended [5]. It has been shown that with the use of remifentanil there is a marked decrease in ciliary mobility which has an important repercussion in patients with immobile cilia syndrome or Kartagener [10]. The propofol was chosen for reasons previously described with remifentanil, which was used at low doses, due to the pharmacokinetic characteristics it offers, such as its short latency time and short elimination route, although taking into account that pain management with this it becomes somewhat ineffective.

Although there are studies that report that the laryngeal mask produces a lower impact on mucociliary activity than the endotracheal tube, it was decided to use the latter because it is considered safer due to the type of surgery [11,12].

An epidural catheter was placed as its use had shown good results in the postoperative period for pain management, although there are studies that conclude that the quality of analgesia is not related to post-operative respiratory complications [13,14]. It was decided to place an epidural catheter prior to induction, which was maintained for 24 hours, with which the postoperative pain was effectively managed, thus decreasing the pulmonary repercussions already described that the pain itself could cause.

In conclusion when surgery is proposed for patients with primary ciliary dyskinesia, it is best to prepare the patient adequately to safeguard pulmonary and cardiac function and cause the least type of repercussions in this area when deciding the anesthetic technique according to the requirements of the procedure and the general conditions of the patient.

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