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CLINICAL INFORMATION

Laparoscopic cholecystectomy under continuous spinal anesthesia in a patient with Steinert's disease

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Abstract Steinert's disease is an intrinsic disorder of the muscle with multisystem manifestations. Myotonia may affect any muscle group, is elicited by several factors and drugs used in general anesthesia like hypnotics, sedatives and opioids. Although some authors recommend the use of regional anesthesia or combined anesthesia with low doses of opioids, the safest anesthetic technique still has to be established.

We performed a continuous spinal anesthesia in a patient with Steinert's disease undergoing laparoscopic cholecystectomy using 10 mg of bupivacaine 0.5% and provided ventilatory support in the perioperative period. Continuous spinal anesthesia was safely used in Steinert's disease patients but is not described for laparoscopic cholecystectomy. We reported a continuous spinal anesthesia as an appropriate technique for laparoscopic cholecystectomy and particularly valuable in Steinert's disease patients.

PALAVRAS-CHAVE
Raquianestesia contínua;
Doença de Steinert;
Colecistectomia laparoscópica

Colecistectomia laparoscópica sob raquianestesia contínua em paciente com doença de Steinert

Resumo A doença de Steinert é uma desordem intrínseca do músculo com manifestações multissistêmicas. A miotonia pode afetar qualquer grupo muscular e é provocada por vários fatores e medicamentos usados em anestesia geral, como hipnóticos, sedativos e opiáceos. Embora alguns autores recomendem o uso de anestesia regional ou anestesia combinada com opiáceos em doses baixas, a técnica anestésica mais segura ainda precisa ser estabelecida.

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Administramos raquianestesia contínua em um paciente com doença de Steinert submetido à colecistectomia laparoscópica, com 10 mg de bupivacaína a 0,5%, e fornecemos suporte ventilatório no período perioperatório. A raquianestesia contínua foi usada com segurança em pacientes com doença de Steinert, mas não foi relatada em colecistectomia laparoscópica. Relatamos a raquianestesia contínua como uma técnica adequada para a colecistectomia laparoscópica e particularmente valiosa em pacientes com doença de Steinert.

Introduction

Steinert's disease (SD) is an intrinsic disorder of the muscle with multisystem manifestations. Inheritance is autosomal dominant and a prevalence of about 3–5 in 100,000.[1] Patients usually present between ages of 15 and 35 years with weakness of grip, impaired foot dorsiflexion, cataracts and infertility. Muscular weakness is usually found in the face, neck and distal muscles group contributing to characteristic facial appearance. Myotonia may affect any muscle group and is elicited by drugs, pain, cold, shivering, surgical manipulations and electric scalpel among other factors.[2]

Extramuscular involvement is almost invariable with cardiac (conducting system and cardiac muscle), ventilatory (respiratory muscles weakness, central abnormalities), gastrointestinal (dysphagia, reduction in the rating of gastric emptying) and endocrine affection (hypothyroidism, primary gonadal failure, diabetes mellitus).

The perioperative assessment of these patients should target the extramuscular manifestations of the disease which may be life-threatening. The conduct of anesthesia poses particular problems which include the increase of sensitivity to several drugs used during general anesthesia. The patients with this disease have an increase of the sensibility to hypnotics and sedatives which can cause apnea even with small doses.[3] Inhalation agents may further risk the already compromised cardiovascular and respiratory systems, while postoperative shivering may precipitate myotonic crisis.[4] Depolarizing relaxants should be avoided because they can trigger myotonic crisis and difficulty in ventilation and intubation.[5] Non-depolarizing neuromuscular blocking agents usually evoke a normal response but if muscle wasting exists a prolonged response may occur.[4]

The postoperative complications usually result in pulmonary and cardiac dysfunction and pharyngeal muscle weakness. Regarding this knowledge regional anesthesia is considered the best option in these patients, although the safest anesthetic technique still has to be established.

We present a continuous spinal anesthesia (CSA) in SD patient for laparoscopic cholecystectomy (LC).

Case description

A 35-year-old Caucasian female, with SD was scheduled for elective LC.

She is being followed in the pneumology department and presented restrictive pulmonary disease with mild ventilatory impact and indication for nocturnal BIPAP support that she does not adhere. She is currently not under any medication. Previous cervical cesarean under combined spinal epidural anesthesia was without complications. Physical examination revealed facial atony, mild prognatism and short neck.

After standard monitoring, continuous spinal block was performed without any pre-medication, in right lateral decubitus, at L2–L3 level with paramedian approach, using a spinocath kit B.Braun® (24 G, 29 G) and 20 mcg of fentanyl with 1 ml of normal saline solution were administered. Having the patient already in prone position, 5 mg of hyperbaric bupivacaine were administered through the catheter with 2 ml of normal saline solution. The sensory level after 5 min was T12 and subsequently a top-up with 5 mg of isobaric bupivacaine with 1 ml of normal saline allowed a spreading through T7 level. T4 level was achieved 5 min later.

Analgesia included 80 mcg of fentanyl before the incision, acetaminophen 1 g and 40 mg of parecoxib during the procedure. Referred left shoulder pain was effectively controlled with diaphragmatic instillation of 2% lidocaine. After 40 min a spinal perfusion of ropivacaine 0.1% at 2 ml/h was started and maintained during 24 h.

Non-invasive ventilatory (NIV) support was instituted 12 h before surgery until 24 h after surgery with BIPAP S/T (Spontaneous/Timed) 6/14 cmH2O and cough assist device. Spirometry and blood gasometry were assessed during perioperative period (Tables 1 and 2). Maximal intra-abdominal pressure (IAP) was 10 mmHg and at the end of the surgery abdominal was 8 mmHg. Minimal blood oxygen saturation was 92%

No myotonic crises were triggered during the procedure.

After 3 h in the Post Anesthesia Care Unit (PACU) completed motor block was completely reversed and the patient referred no pain or dyspnea.

At 24 h the spinal catheter was removed without complications and she was discharged home without noticed intercorrences.

Discussion

CSA allowed a sensory block suitable for the surgery and minimal respiratory impairment. CSA was chosen concerning co-morbidities of the patient, suitability to the procedure and the already described complications of general anesthesia in this context. In fact, Cope et al.[6] considered that regional anesthesia is the best option in these patients because trigger drugs of myotonic crisis are not used. March et al.[7] recommended regional anesthesia or combined general and regional anesthesia with restricted use of opioids, because these patients have higher risk of respiratory depression. CSA has already been used with success in high risk patients for abdominal surgery[8] including

Table 1 Perioperative spirometric evaluation.

	Baseline	CSA + NIV	Maximal IAP + NIV	PACU + NIV
FVC (L)	2.60	2.24	2.00	2.59
FEV 1 (L)	1.99	1.79	1.65	1.99
FEV 1/FVC (%)	76.50	79.88	82.58	78.35

FVC, functional vital capacity; FEV 1, forced expiratory volume in 1 s; NIV, non-invasive ventilation; IAP, intra-abdominal pressure; PACU, Post-anesthesia care unit.

Table 2 Perioperative blood gasometric evaluation.

	Baseline	Intraoperative + NIV	PACU + NIV
pH	7.550	7.587	7.552
pCO_2 (mmHg)	24.5	24.5	22.2
pO_2 (mmHg)	116.0	115.1	117.8
HCO_3 (mmol/L)	22.9	22.8	22.7
$EtCO_2$ (mmHg)	23.7	23.6	23.5
Sat O_2 (%)	98	98	98

pO_2 and pCO_2, blood partial pressure of oxygen and carbon dioxide; $EtCO_2$, end tidal carbon dioxide; Sat O_2, oxygen saturation; HCO_3, bicarbonate ion.

LC.[9–13] Also, Verdaguer et al.[14] described a case of CSA in a patient with SD proposed for hysterectomy. But to our knowledge this is the first case of LC under CSA in a patient with Steinert's disease. Bennun et al.,[15] reported a significant decrease in mean postoperative vital capacity (from 965 to 349 mL) from the preoperative value during continuous propofol anesthesia. In our patient there was no decrease in postoperative vital capacity comparing with preoperative values. According to the spirometric evaluation there was not a significant impairment of the mechanics of ventilation even after full establishment of the sensitive block or after the pneumoperitoneum. Furthermore, seriated blood gas analysis validated the contribution of perioperative non-invasive ventilatory support.

Conclusion

Postoperative pain control was effective and opioids were avoided. No spinal headache was noticed which could be explained by the removal of the catheter only after 24 h.[16] In conclusion, we reported a CSA as an appropriate technique for LC and with particular value in SD patients.

Conflicts of interest

The authors declare no conflicts of interest.

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