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Case report

Anaesthetic management of a patient diagnosed with amyotrophic lateral sclerosis taken to caesarean section: Case report☆

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ABSTRACT

Introduction: In the obstetric population with amyotrophic lateral sclerosis, caesarean delivery may be indicated in order to avoid greater maternal respiratory distress induced by the physiologic changes at the end of gestation and the progressive muscle weakness caused by the disease. Optimal anaesthetic management is required in order to guarantee the ideal conditions for surgery and to preserve patient respiratory function.
Objective: To describe the anaesthetic management of a patient diagnosed with amyotrophic lateral sclerosis scheduled for caesarean section.
Methods and results: We describe the case of a 38-week pregnant woman diagnosed with amyotrophic lateral sclerosis who underwent caesarean section under sequential combined spinal-epidural anaesthesia. In addition, a review of the literature was conducted.
Conclusions: Combined spinal-epidural anaesthesia was a good option as anaesthetic technique for caesarean section in this patient with amyotrophic lateral sclerosis.

Anestesia para cesarea en paciente con esclerosis lateral amiotrofica: reporte de caso

Palabras clave:
Esclerosis lateral amiotrófica
Enfermedad de la neurona motora
Cesárea
Anestesia
Informe de Caso

RESUMEN

Introducción: En las gestantes con diagnóstico de esclerosis lateral amiotrofica puede estar indicado parto por cesárea para evitar una mayor dificultad respiratoria materna debido a los cambios al final de la gestación, y a la debilidad muscular progresiva propia de la enfermedad, motivo por el cual es necesario un manejo anestésico que garantice las condiciones idóneas para la cirugía y preserve la función respiratoria.

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Objetivo: Describir el manejo anestésico de una paciente con diagnóstico de Esclerosis lateral amiotrofica programada para cesárea

Métodos y resultados: Mostramos el caso de una gestante de 38 semanas con diagnóstico de Esclerosis lateral amiotrofica a quien se le realizó una cesárea, y cuya técnica anestésica empleada fue CSE (combinada espinal-epidural) secuencial. Además, se realizó una revisión de la literatura al respecto.

Conclusiones: La anestesia regional CSE (combinada espinal-epidural) secuencial fue una buena opción como técnica anestésica en esta paciente con esclerosis lateral amiotrofica para cesárea.

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive neurological disorder that mainly affects male patients between the fifth and sixth decades of life, with an incidence of 1:50 000.[1] It is characterised by degeneration of higher and lower motor neurons because of degenerative changes in lateral corticospinal tracts of the spinal cord and, consequently, muscle denervation.[2]

ALS, or Lou-Gehrig disease, is extremely rare in pregnant women. Only some 12 cases have been reported of pregnant women with ALS since 1993. It is important to highlight that respiratory demands during pregnancy may result in acute respiratory failure in patients with ALS. For this reason, it is recommended to establish baseline respiratory function early on in pregnancy and ensure regular follow-up. In some cases, vaginal delivery may be expected because the perineum is not affected and uterine dynamics is not altered.[3] However, physiological changes that become more intense towards the end of pregnancy, such as reduced residual functional capacity resulting from lower residual volume due to diaphragm displacement by the pregnant uterus, may worsen the mother's respiratory status secondary to progressive muscle weakness inherent to the disease. This could lead to the indication of early caesarean section.

There are few recommendations in the literature regarding anaesthetic management of women in childbirth with ALS. Epidural analgesia and anaesthesia have been used in these patients with no evidence of worsening of their neurological condition following the procedure. Progressive loss of innervation leads to muscle atrophy and hypersensitivity of acetylcholine nicotinic receptors. These patients are more sensitive to the use of non-depolarising neuromuscular relaxants[4] and, furthermore, the use of depolarising muscle relaxants is associated with neuromyotonia, rhabdomyolysis and severe hyperkalemia.[5] Because of all these considerations, a pregnant woman with ALS is a true challenge when it comes to peri-operative management and critical care.

Case description

A 38-year-old primigravida 38 weeks pregnant diagnosed with ALS some 10 years before was scheduled for category 4 caesarean section by obstetric indication. Upon neurological assessment, the patient presented with progressive muscle weakness, severe dysarthria, dysphagia, preserved superior function, fasciculations in the anterior region of the chest and the tongue, and is emotionally labile.

In terms of muscle function, the patient presented with quadriparesis (muscle strength in upper limbs 2/5 and 2/5 in the lower limbs), hypotonia, generalised laxity, hyperreflexia and positive bilateral Babinsky sign. In terms of pulmonary function, the chest X-ray showed evidence of moderate fibroemphysema and there was severe restriction on spirometry, leading to a diagnosis of mild respiratory failure. There was absence of cardiovascular symptoms, grade 2 cardiological risk (Goldman score), no functional capacity assessment was made, and there was no evidence of murmurs or cyanosis. Anaesthetic assessment: ASA III, body weight 50 kg, height 155 cm, BMI 20.83, mouth opening 3 cm, thyromental distance greater than 6 cm, modified Mallampati grade 3, preserved cervical mobility and mandatory decubitus. Vital signs: heart rate 86 bpm, arterial blood pressure 134/92 mmHg, respiratory rate 20 rpm, oxygen saturation 95% in room air, and no fever.

The patient and husband were explained the anaesthetic technique, and after discussing the risks and benefits, a sequential combined spinal-edpidural (CSE) was performed.

Before anaesthesia administration, the patient received ranitidine 50 mg IV, metoclopramide 10 mg IV and dexamethasone 4 mg IV. Basic ASA monitoring was instituted and advanced airway equipment and aspiration tube were prepared. With the patient in lateral decubitus and following asepsis and antisepsis, the epidural space was identified at the level of L3–L4, 4 cm from the skin; using a N° 18 Tuohy needle and the technique of intermittent loss of resistance, the epidural space was reached with no complications. A N° 27 Whitacre spinal needle was then inserted through the Tuohy needle until CSF flow was ascertained, and hyperbaric bupivacaine 6 mg plus fentanyl 10 μg were injected. Finally, a N° 20 multi-orifice epidural catheter was put in place and fixed 9 cm from the skin.

The catheter aspiration test was negative and the decision was made not to use a test dose; sensory anaesthetic level was obtained up to T7. With the patient in supine decubitus and 15° uterine lateralisation, 0.25% isobaric bupivacaine in 10 ml was administered through the epidural catheter 5 min after the intrathecal injection, achieving sensory block of T5 after 5 min.