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GAMMA-SARCOGLYCANOPATHY AND PREGNANCY: WHAT IS THE ANESTHETIC MANAGEMENT?

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ABSTRACT

Gamma-sarcoglycanopathy is a rare genetic limb-girdle muscular dystrophy that presents significant anesthetic and obstetric challenges during pregnancy.
We report the case of a 30-year-old patient at 39 weeks of gestation with generalized muscle weakness graded at 2/5 who was admitted in labor.
Due to her inability to deliver vaginally, a cesarean delivery was indicated and successfully performed under spinal anesthesia.
The anesthetic block utilized a mixture of 0,5% isobaric bupivacaine, fentanyl, and morphine, allowing for a complication-free extraction and stable recovery.
Regional anesthesia seems to be the best option for myopathic patients to avoid the inherent risks of general anesthesia, such as difficult intubation and rhabdomyolysis.
A comprehensive preoperative evaluation remains crucial to optimize maternal and fetal safety in the absence of standardized management guidelines.

KEYWORDS

Gamma-sarcoglycanopathy, Pregnancy , Anesthetic Management

MAIN ARTICLE

INTRODUCTION

Gamma-sarcoglycanopathy, or limb-girdle muscular dystrophy type 2C, is a rare genetic disease that affects the muscles. It manifests as a decrease in strength predominantly in the muscles of the pelvis (pelvic girdle) and shoulders (shoulder girdle) (1). It begins in childhood and progresses over the years. It is a severe and debilitating condition that requires medical support from the moment of diagnosis (2) (3).

We report the case of a 30-year-old female patient, followed for gamma-sarcoglycanopathy, who was admitted in labor at 39 weeks of gestation.

CASE PRESENTATION

We report the case of a 30-year-old female patient, followed for hereditary gamma-sarcoglycan muscular dystrophy with two similar cases among her siblings, who presented to the obstetric emergency department in labor at 39 weeks of gestation.

On clinical examination, we found a conscious patient who was hemodynamically and respiratory stable with a BP = 120/60 mmHg, HR = 90 bpm, and SpO2 = 98% on room air. Furthermore, the neurological examination revealed generalized muscle weakness graded at 2/5 in all four limbs.

The obstetric evaluation confirmed the presence of a progressing pregnancy estimated at 39 weeks of gestation. Following a multidisciplinary assessment involving obstetricians, anesthesiologists, and neurologists, a cesarean delivery was indicated. The decision was driven by the patient's severe pelvic girdle and abdominal muscle weakness (graded 2/5), which significantly compromises the generation of an effective Valsalva maneuver required during the active second stage of labor. Furthermore, a trial of labor was contraindicated due to the high risk of rapid maternal exhaustion, prolonged expulsive efforts, and potential secondary respiratory compromise

In the operating room, the patient was positioned and monitored. An 18G peripheral intravenous (IV) line was placed, followed by the administration of a 500 cc fluid bolus of 0.9% isotonic saline and antibiotic prophylaxis with 2 g of cefazolin. We administered spinal anesthesia with the patient in the lateral position, using a mixture of 10 mg of 0,5% isobaric bupivacaine, 25 µg of fentanyl, and 100 µg of morphine. The patient was then placed back in

the supine position (Figure 1). The delivery was performed without complications, and the patient remained under observation in the post-anesthesia care unit (PACU) for 2 hours, before being admitted to the intensive care unit (ICU) as a precaution. The patient was discharged from the hospital 2 days later.

DISCUSSION

Myopathies belong to the family of neuromuscular diseases involving the striated skeletal muscle, specifically the muscle fiber, either in its structure or its development. Two main categories of myopathies are distinguished: genetic myopathies and acquired myopathies. Genetic myopathies have an autosomal or X-linked mode of inheritance. These pathologies are caused by a qualitative and/or quantitative alteration of one of the constitutive proteins of the sarcolemma. These proteins contribute to the structure of the fiber. In the case of dystrophy, muscle fibers are destroyed. Among dystrophies, a distinction must be made between muscular dystrophies and myotonic dystrophies.

Limb-girdle muscular dystrophies have a very similar clinical presentation. In the case of gamma-sarcoglycanopathies, the clinical examination often reveals the existence of selectivity, early distal involvement, and muscle hypertrophy—particularly of the calves and tongue—as well as early contractures involving the spine and elbows (4).

Regarding the anesthetic evaluation, it remains identical for all myopathies: it quantifies the overall impact of the myopathy, anticipates potential complications, and determines the appropriate management for the patient. It relies on the assessment of the disease, preferably in coordination with the treating physician, to detect its impact on various systems: respiratory, cardiac, and nutritional.

Anesthetic considerations vary significantly among different neuromuscular disorders. While susceptibility to classic Malignant Hyperthermia (MH) is strongly associated with specific channelopathies (e.g., RYR1 mutations), patients with structural myopathies like gamma-sarcoglycanopathy face a distinct, life-threatening risk: Anesthesia-Induced Rhabdomyolysis (AIR). Because gamma-sarcoglycanopathy involves a deficiency in the sarcoglycan complex, the muscle sarcolemma is highly fragile. Exposure to depolarizing neuromuscular blockers (such as succinylcholine) or halogenated volatile anesthetics can trigger massive muscle breakdown. This results in acute, severe hyperkalemia and subsequent cardiac arrest, a cascade that can clinically mimic MH but has a fundamentally different pathophysiology. Consequently, these agents are strictly contraindicated in this patient population. If general

anesthesia is required, a Total Intravenous Anesthesia (TIVA) approach using propofol is the gold standard.

There are no definitive rules for choosing between regional and general anesthesia in patients with myopathies. In the event of general anesthesia, the agents that can be safely used for induction in a patient with confirmed or suspected myopathy include thiopental, propofol, and ketamine. Maintenance of anesthesia should be achieved using a continuous propofol infusion. Halogenated anesthetic agents are contraindicated due to the risk of myocardial depression, rhabdomyolysis, and hyperkalemia. They can also induce shivering upon awakening, which may increase the risk of myotonia (5).

In certain situations, regional anesthesia appears to optimize patient management and should be considered whenever possible to avoid intubation difficulties and reduce the risk of vasoplegia associated with general anesthesia (6). The combination of both techniques (general anesthesia and ultrasound-guided regional anesthesia) is possible, including the use of central neuraxial blocks (7).

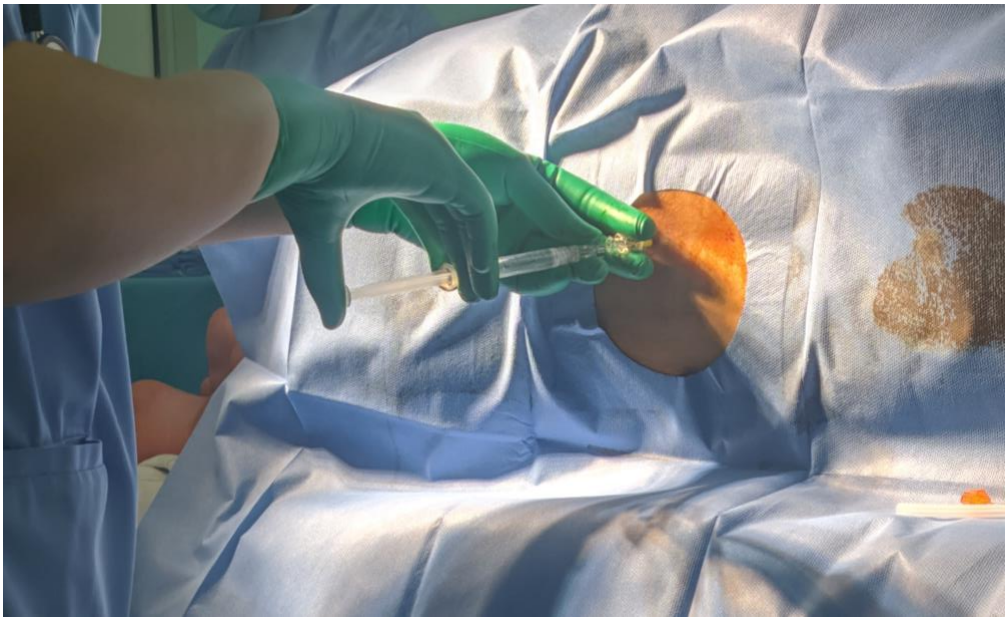
In the case of our patient, we opted for regional anesthesia, a technique that allows the delivery to be performed with minimal risk to both the mother and the baby.

CONCLUSION

The significant clinical and nosological variability of myopathies requires a comprehensive preoperative evaluation of these patients in order to optimize their anesthesia and reduce intraoperative risks. The anesthetic management of these patients constitutes a challenge for any anesthesiologist, particularly in the presence of superimposed physiological conditions. In the absence of clear guidelines for the management of these types of patients, respecting contraindications and reviewing the literature remain the only ways to safely manage them.

FIGURES:

Figure 1: Patient positioned in the lateral decubitus position during the performance of spinal anesthesia.



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