Regional anesthesia as a safe option in patient with limb girdle muscular dystrophy undergoing total abdominal hysterectomy: A case report and case review

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1 INTRODUCTION

Limb-girdle muscular dystrophies (LGMDs) comprise a group of rare neuromuscular disorders, which primarily involve pelvic and shoulder girdle muscles1 and may progress to involve the heart and lungs.2 General anesthesia with volatile anesthetics and succinylcholine and the increased risk of rhabdomyolysis and malignant hyperthermia (MH) make this a very challenging prospect for anesthesiologists.1 With very few case reports on anesthetic management of patients with LGMDs, we aimed to present our hospital experience about a patient with LGMD undergoing total abdominal hysterectomy under regional anesthesia.

2 CASE REPORT

A 46-year-old female was admitted to the hospital with diagnosis of abnormal uterine bleeding secondary to fibroid and left complex ovarian cyst and was planned for total abdominal hysterectomy. She was diagnosed with LGMD with muscle biopsy (Figure 1) 8 years back and had symptoms predominantly affecting her lower limbs.

She had complaints of urinary incontinence. She had no complaints of difficulty in swallowing. On examination, there was weakness in bilateral upper and lower limbs with intact muscle tone and sensation. Nerve conduction study was normal, and electromyography of the lower limb muscles showed myogenic pattern.

Detailed pre-anesthetic workup was done. All the routine blood investigations were within normal limits. Echocardiography (Figure 2) was normal, and pulmonary function test (Figure 3) revealed moderate obstructive pattern.

She was explained about all the findings of the pre-anesthetic checkup and the risks associated with anesthetizing her, after which informed written consent was taken. Considering the high risk of rhabdomyolysis and malignant hyperthermia and the unavailability of...
dantrolene, we decided to proceed with regional anesthe-
sia with combined spinal and epidural anesthesia (CSE).

Operating room was prepared ensuring breathing
circuits were replaced with a new one and the vaporiz-
ers were removed from the anesthesia workstation. The
anesthesia machine was flushed with high flow oxygen.
Necessary backup for total intravenous anesthesia (TIVA)
was made ready in case of failed CSE. Patient was shifted
to operating room, monitors were attached, and baseline
vitals were recorded. Under all septic precaution in sitting
position, epidural catheter was inserted at L2-L3 inter-
space and fixed at 9 cm, and subarachnoid block was given
midline at L3-L4 interspace. The level of block was noted
at T4 at 5 min. The surgery was started. Level of block was
assessed hourly and at 1 h 30 min, she complained of pain
at surgical site. Epidural was activated with 10 ml 0.25%
plain bupivacaine and 4 ml 1% lignocaine. After this, in-
traoperative period was uneventful, and she was shifted to
postoperative ward. About 0.0625% plain bupivacaine via
epidural at 6 ml/h was used for postoperative analgesia,
and vitals were monitored every 30 min. On the first post-
operative day, she looked comfortable with visual analog
scale (VAS) pain score 2 at rest and 4 on movement. She
was discharged on fifth postoperative day. No new neuro-
logical deficits at the time of discharge and on follow-up
at 1 week were noted.

3 | DISCUSSION

Limb-girdle muscular dystrophy comprises a group of ge-
etic disorders primarily caused by mutation leading to
abnormal protein synthesis localized to various parts of
the muscle fiber. The classic pathologic change seen is
degeneration and necrosis of muscle fibers, which are re-
placed by connective tissue and fat. However, the biopsy
findings may not be specific, thereby requiring genetic
testing for definitive diagnosis.

Use of volatile anesthetics and muscle relaxants such
as succinylcholine may lead to life-threatening hyper-
kalemia and rhabdomyolysis. Even though propofol,
etomidate, and opioids have not been known to trigger
malignant hyperthermia, some myopathic patients may
be more sensitive. Therefore, we chose regional anesthe-
sia with CSE technique for surgery in this patient.

As we went through PubMed searching for articles
related to anesthetic management in patients with mus-
cular dystrophy, we reviewed six case reports, which in-
volved surgeries of lower limbs and abdomen. In those
case reports, two patients had undergone laparoscopic
cholecystectomy, one had undergone appendectomy,
two had undergone caesarean section, one had an ad-
enoidectomy, and the last one had undergone replace-
ment of ascending aorta. Four surgeries were performed
under TIVA, and the remaining were performed under
neuraxial anesthesia. Cases performed under TIVA
used propofol and remifentanil as anesthetic agent
and if muscle relaxants had to be used, neuromuscu-
lar blockade monitoring was routinely recommended.
Suggamadex was used for reversing the muscle relaxant.
Our anesthetic strategy was basically planned consider-
ing the techniques used in these reports. We also consid-
ered the fact that use of TIVA cannot completely avoid
the risk of malignant hyperthermia.
FIGURE 2  Echocardiography
FIGURE 3  Pulmonary function test
CONCLUSION

For a safe anesthesia, carefully conducted CSE can be a safe option in patients with LGMDs but a very careful planning, workup, and multidisciplinary teamwork is of utmost importance.

AUTHOR CONTRIBUTIONS

Sagar Devkota conceptualized the study. Sangeeta Shrestha and Tara Gurung were in charge of the case, and they reviewed and edited the manuscript. Sagar Devkota wrote the original manuscript and reviewed and edited the manuscript. Saurav Shrestha corrected and edited the original manuscript.

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CONFLICT OF INTEREST

None.

DATA AVAILABILITY STATEMENT

None.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal’s patient consent policy.

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