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CASE REPORT

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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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Abstract

Regional anesthesia can be a very safe option in patients with limb girdle muscular dystrophy undergoing lower abdominal surgeries as general anesthesia and volatile anesthetic agents are associated with increased risk of malignant hyperthermia and rhabdomyolysis.

KEYWORDS

general anesthesia, limb girdle muscular dystrophy, malignant hyperthermia, regional anesthesia

1 | INTRODUCTION

Limb-girdle muscular dystrophies (LGMDs) comprise a group of rare neuromuscular disorders, which primarily involve pelvic and shoulder girdle muscles[1] 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.

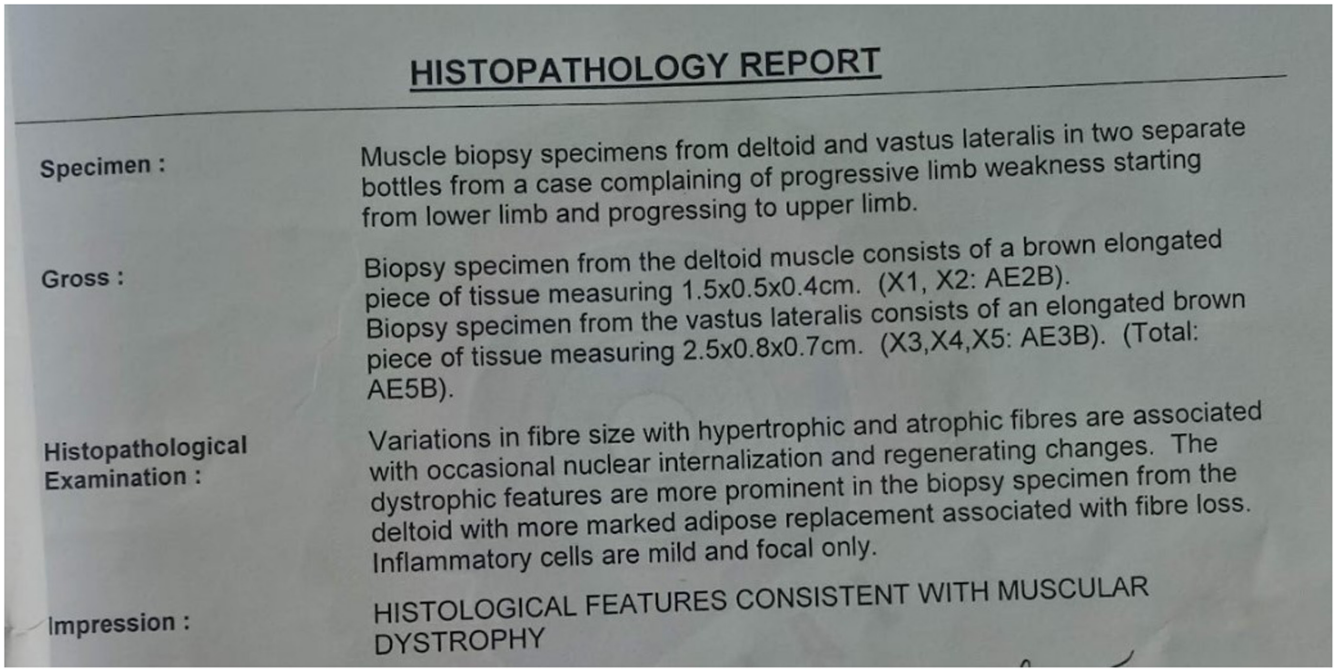
2 | CASE REPORT

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



HISTOPATHOLOGY REPORT

Specimen :	Muscle biopsy specimens from deltoid and vastus lateralis in two separate bottles from a case complaining of progressive limb weakness starting from lower limb and progressing to upper limb.
Gross :	Biopsy specimen from the deltoid muscle consists of a brown elongated piece of tissue measuring 1.5x0.5x0.4cm. (X1, X2: AE2B). Biopsy specimen from the vastus lateralis consists of an elongated brown piece of tissue measuring 2.5x0.8x0.7cm. (X3,X4,X5: AE3B). (Total: AE5B).
Histopathological Examination :	Variations in fibre size with hypertrophic and atrophic fibres are associated with occasional nuclear internalization and regenerating changes. The dystrophic features are more prominent in the biopsy specimen from the deltoid with more marked adipose replacement associated with fibre loss. Inflammatory cells are mild and focal only.
Impression :	HISTOLOGICAL FEATURES CONSISTENT WITH MUSCULAR DYSTROPHY

FIGURE 1 Muscle biopsy

dantrolene, we decided to proceed with regional anesthesia with combined spinal and epidural anesthesia (CSE).

Operating room was prepared ensuring breathing circuits were replaced with a new one and the vaporizers 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 interspace 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, intraoperative 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 postoperative 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 neurological deficits at the time of discharge and on follow-up at 1 week were noted.

3 | DISCUSSION

Limb-girdle muscular dystrophie comprises a group of genetic disorders[3–5] primarily caused by mutation leading to abnormal protein synthesis localized to various parts of the muscle fiber.[2] The classic pathologic change seen is degeneration and necrosis of muscle fibers, which are replaced by connective tissue and fat.[6] 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 hyperkalemia and rhabdomyolysis.[7–9] Even though propofol, etomidate, and opioids have not been known to trigger malignant hyperthermia,[10] some myopathic patients may be more sensitive.[11] Therefore, we chose regional anesthesia with CSE technique for surgery in this patient.

As we went through PubMed searching for articles related to anesthetic management in patients with muscular dystrophy, we reviewed six case reports, which involved 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 adenoidectomy, and the last one had undergone replacement 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, neuromuscular blockade monitoring was routinely recommended. Suggamadex was used for reversing the muscle relaxant. Our anesthetic strategy was basically planned considering the techniques used in these reports. We also considered the fact that use of TIVA cannot completely avoid the risk of malignant hyperthermia.[12]

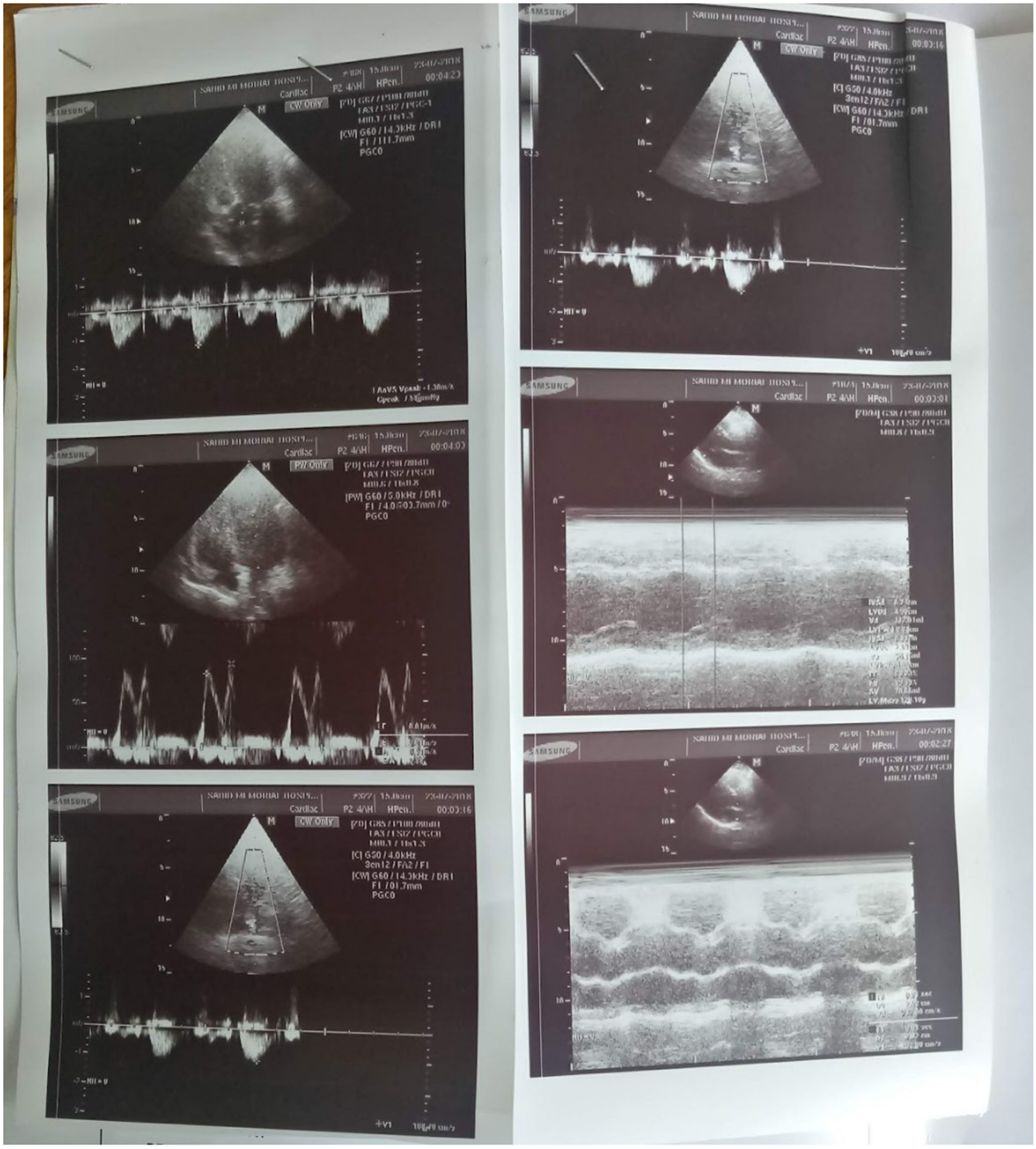
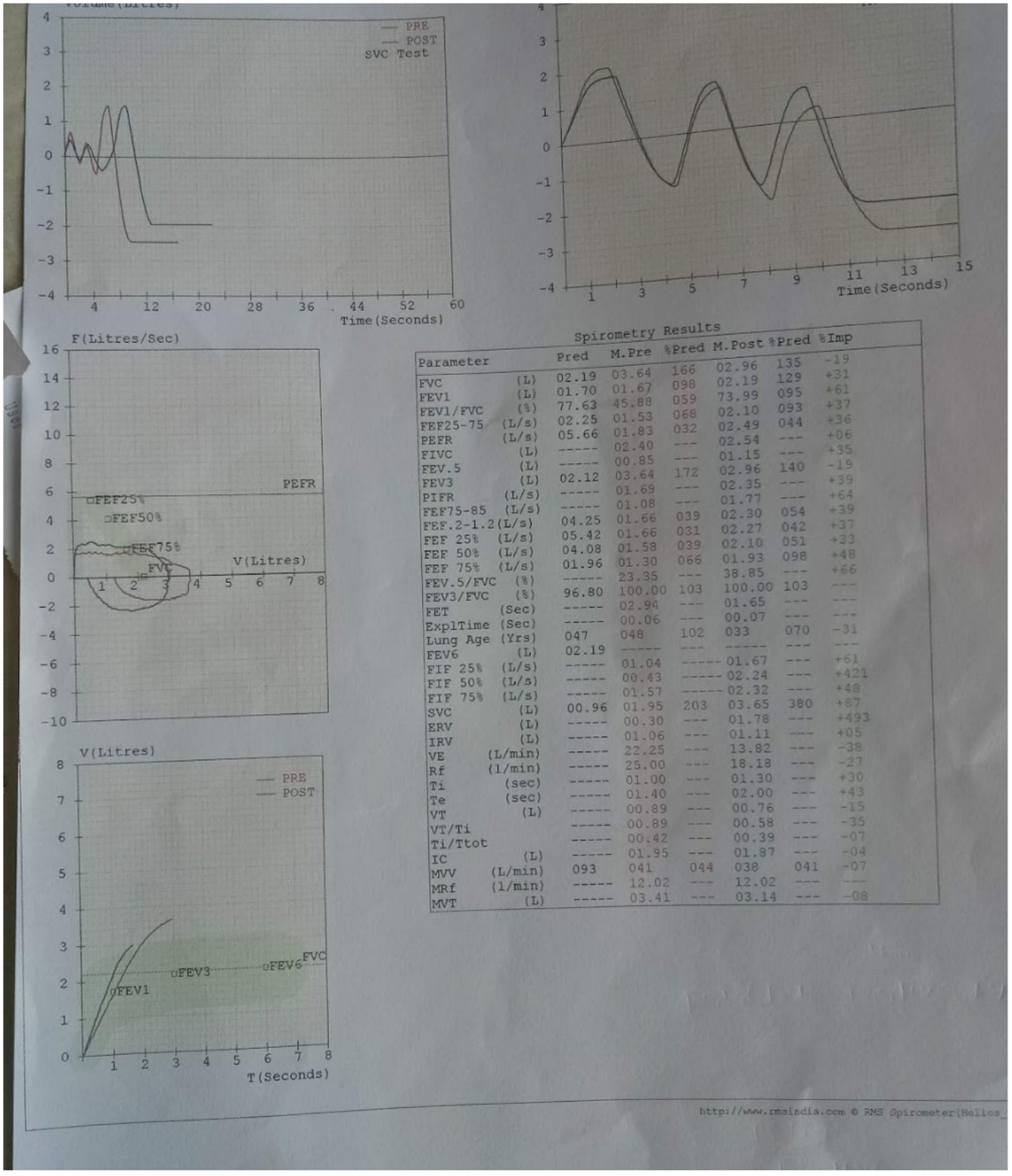


FIGURE 2 Echocardiography



Parameter		Pred	M.Pre	%Pred	M.Post	%Pred	%Imp
FVC	(L)	02.19	03.64	166	02.96	135	-19
FEV1	(L)	01.70	01.67	098	02.19	129	+31
FEV1/FVC	(%)	77.63	45.88	059	73.99	095	+61
FEF25-75	(L/s)	02.25	01.53	068	02.10	093	+37
PEFR	(L/s)	05.66	01.83	032	02.49	044	+36
FIVC	(L)	-----	02.40	---	02.54	---	+06
FEV.5	(L)	-----	00.85	---	01.15	---	+35
FEV3	(L)	02.12	03.64	172	02.96	140	-19
PIFR	(L/s)	-----	01.69	---	02.35	---	+39
FEF75-85	(L/s)	-----	01.08	---	01.77	---	+64
FEF.2-1.2	(L/s)	04.25	01.66	039	02.30	054	+39
FEF 25%	(L/s)	05.42	01.66	031	02.27	042	+37
FEF 50%	(L/s)	04.08	01.58	039	02.10	051	+33
FEF 75%	(L/s)	01.96	01.30	066	01.93	098	+48
FEV.5/FVC	(%)	-----	23.35	---	38.85	---	+66
FEV3/FVC	(%)	96.80	100.00	103	100.00	103	---
FET	(Sec)	-----	02.94	---	01.65	---	---
ExplTime	(Sec)	-----	00.06	---	00.07	---	---
Lung Age	(Yrs)	047	048	102	033	070	-31
FEV6	(L)	02.19	-----	---	-----	---	---
FIF 25%	(L/s)	-----	01.04	-----	01.67	---	+61
FIF 50%	(L/s)	-----	00.43	-----	02.24	---	+421
FIF 75%	(L/s)	-----	01.57	-----	02.32	---	+48
SVC	(L)	00.96	01.95	203	03.65	380	+87
ERV	(L)	-----	00.30	---	01.78	---	+493
IRV	(L)	-----	01.06	---	01.11	---	+05
VE	(L/min)	-----	22.25	---	13.82	---	-38
Rf	(1/min)	-----	25.00	---	18.18	---	-27
Ti	(sec)	-----	01.00	---	01.30	---	+30
Te	(sec)	-----	01.40	---	02.00	---	+43
VT	(L)	-----	00.89	---	00.76	---	-15
VT/Ti		-----	00.89	---	00.58	---	-35
Ti/Ttot		-----	00.42	---	00.39	---	-07
IC	(L)	-----	01.95	---	01.87	---	-04
MVV	(L/min)	093	041	044	038	041	-07
MRf	(1/min)	-----	12.02	---	12.02	---	---
MVT	(L)	-----	03.41	---	03.14	---	-08

FIGURE 3 Pulmonary function test

4 | 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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No financial burden to the patient.

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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