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Case Report

Anesthetic management of a myotonic dystrophy patient with paraganglionoma ☆

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Abstract Myotonic dystrophy (DM), though rare, can significantly complicate anesthesia due to muscular and extra-muscular involvement. When this condition is compounded by a pheochromocytoma, anesthetizing such patients becomes extra challenging. We present a case report of a 61-year-old lady with congenital DM, with the whole gamut of associated features, was diagnosed with a noradrenaline secreting paraganglionoma following investigation of refractory hypertension. We anesthetized her for an open resection of the lesion. The conduct of anesthesia and recovery of this patient is described. Our experience suggests that anesthetizing these patients though challenging can be safely managed with relaxant general anesthesia and epidural analgesia with meticulous care pre, intra and post-surgical intervention.

1. Introduction

Much attention is required to manage myotonic dystrophy (DM) for anesthesia and surgery. DM is associated with prolonged duration of action of drugs used during anesthesia and may also cause dangerous interactions such as severe arrhythmia, significantly limiting the choice of anesthetic and adjuvant drugs. At the same time, management of blood pressure (BP) and heart rate (HR) during pheochromocytoma resection requires meticulous care as severe hypertension and tachycardia can result in catastrophic complications [1]. Although there are quite a few publications on the anesthetic management of individual conditions, we could only find one case report in the Japanese literature where the anesthetic management for the combination of DM and pheochromocytoma was described [2]. Differences in technique are summarized in Table 1. The conduct of anesthesia and recovery of this patient is described.

2. Case history

A 61-year-old lady was diagnosed with pheochromocytoma when investigated for uncontrolled hypertension over a

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Table 1 Differences between our technique when compared with that previously reported.

	Ando et al. [2] report	Our case report
Case	54-yr.-old male DM diagnosed at age of 52	61-yr.-old female DM diagnosed at age of 40
Type of lesion	**Pheochromocytoma** - 5 cm cystic right adrenal mass Noradrenaline secreting	**Paraganglionoma** - 4 cm mass located between IVC and aorta Noradrenaline secreting
Premedication	GTN patch	none
Venous Access	Swan Ganz catheter in left internal jugular vein	4-lumen central line in right internal jugular vein
Epidural	Yes (0.2% ropivacaine)	Yes (0.25% bupavicaine + fentanyl)
Anesthesia		
Type	General anesthesia	General anesthesia
Induction	Propofol (2 mg/kg)	Fentanyl, propofol (2 mg/kg), vecuronium
Neuromuscular blocker	Not used	Vecuronium
Maintenance of anesthesia	Nitrous oxide + TIVA with propofol at 3 mg/kg/hr	Sevoflurane in air-O_2 mixture (FiO_2 0.8) + intermittent vecuronium (2 mg × 2)
Use of opiates	Not used	Fentanyl used both systemically and epidurally
Surgical Aspects		
Type of surgery	Laparoscopic	Laparotomy
Adverse events during surgery	Metabolic acidosis thought to be due to cyanide toxicity	None
Surgical duration	9 hours and 20 minutes	~3 hours
Estimated blood loss	Approximately 800 mL	Approximately 250 mL
Blood pressure control		
During dissection of tumor	Doxasosin mesylate 2 mg Landiolol hydrochloride (5 mg/hr) initially, then GTN and nicardipine	Phentolamine 4 mg in total
Following dissection of tumor	Catecholamines not required; no comment about fluid challenges	Fluids (1 L crystalloid and 500 mL colloid) and metaraminol 0.5 mg and ephedrine 6 mg
Magnesium sulfate	Not used	Not used
Miscellaneous		
Time to extubation	14 days	2 days
Time to discharge	Not mentioned	11 days

DM – myotonic dystrophy; GTN – glyceryl trinitrate; FiO_2 – fractional inspired O_2.

12-month period. This was in the background of congenital myotonic dystrophy (DM) with significant muscular and extra-muscular involvement manifested by early cataracts, cardiac conduction abnormalities requiring permanent pacemaker, fatty liver, previous miscarriages, cognitive problems with executive dysfunction, frontal baldness with bossed forehead, high-arched palate with a receding jaw and endocrine dysfunction in the form of insulin resistance and now a newly diagnosed pheochromocytoma.

Investigations revealed elevated urinary metanephrines and catecholamines with plasma metanephrines >10 times the upper limit of normal (Table 2). High plasma noradrenaline levels with normal adrenaline levels were also observed. A CT scan (Fig. 1a) of her abdomen localized a 4 cm soft tissue lesion lying between the inferior vena cava and the aorta just above the right renal artery and a MIBG (iodine-123-meta-iodobenzylguanidine) scan (Fig. 1b) revealed a hyper-functioning area in the same area confirming a paraganglionoma. Her hypertension was managed initially with α-blockers (phenoxybenzamine 20 mg twice daily). A non-selective β-blocker (propranolol 20 mg thrice daily) was added once the α-adrenergic blockade was established. Salt tablets were subsequently introduced to expand her volume status to treat her symptomatic postural hypotension.

Pre-anesthetic assessment a month prior to the surgery revealed history of repeated falls due to her generalized weakness compounded by the postural hypotension from the α-blocker. Her metabolic equivalent for task score (METS; a simple and practical concept that represents the energy cost of physical activities as a multiple of the resting metabolic rate [3]) was 2 as she was barely able to carry out activities of daily living, due to weakness. She had never smoked and no recent alcohol consumption. On examination, her pulse was 60 bpm with a supine BP 95/64 mmHg and SpO_2 of 95% on