

Anaesthetic management of a patient with Huntington's chorea undergoing robot-assisted nephron-sparing surgery

Sir,

Huntington's chorea (HC) is a rare, progressive, autosomal dominant neurodegenerative disease with prevalence of 5–7 in 1,00,000 people.^[1] We present a case of HC with bronchial asthma undergoing robot-assisted nephron-sparing surgery (NSS).

A 52-year-old, 50 kg female with history of abdominal pain for past 1 month was diagnosed to have multiple cystic lesions in the upper pole of the left kidney for which robot-assisted NSS was planned [Figure 1]. The patient was known case of HC for 5 years with choreiform movements, behavioural abnormalities and depression. She had a positive family history. Magnetic resonance imaging of the brain and genetic analysis confirmed diagnosis, and she was under neurology follow-up [Figure 2]. She was receiving tablet olanzapine 5 mg at bedtime and lorazepam

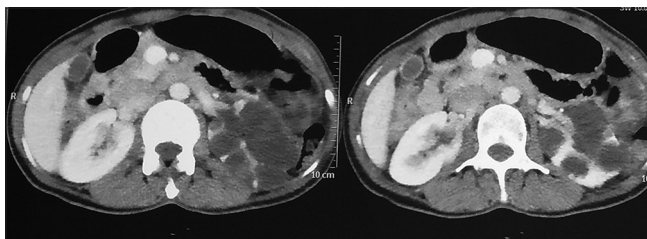


Figure 1: Contrast-enhanced computed tomography of the abdomen showing renal cyst

0.25 mg twice a day. Furthermore, she suffered from bronchial asthma for 2 years and was advised budesonide metered-dose inhaler for which she was non-compliant. Pre-anaesthetic evaluation revealed dysarthria and mild choreiform movements with no complaints of difficulty in swallowing. She refused nebulisation with bronchodilator in pre-operative period. She was advised to continue her medication, and tablet ranitidine 50 mg was given at night and morning of surgery.

After establishing intravenous access and attaching monitors (for electrocardiography, pulse oximetry, non-invasive blood pressure and train-of-four count [TOF]), she was administered injection fentanyl 50 µg and injection propofol 70 mg; trachea was intubated after giving injection rocuronium 50 mg intravenously. Intra-arterial cannula was placed in the right radial artery. Anaesthesia was maintained with mixture of oxygen, air and sevoflurane. The patient was turned to right lateral position; duration of surgery was 3 h and that of pneumoperitoneum was 2.5 h with about 300 ml blood loss. Intraoperatively, haemodynamics remained stable. Warm intravenous fluids and warming mattress were used to prevent

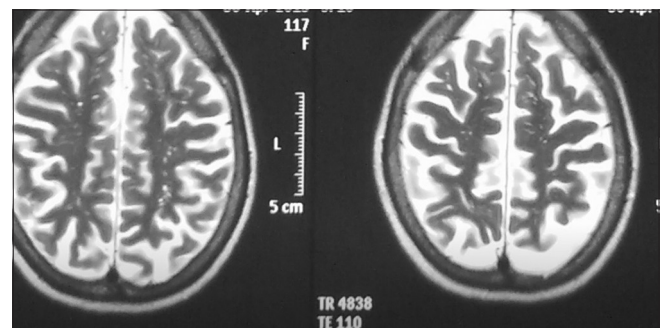


Figure 2: Magnetic resonance imaging of the brain showing gliosis

hypothermia. At the end of surgery, incision site was infiltrated with 15 ml of 0.2% ropivacaine, and intravenous ondansetron 8 mg was given as prophylaxis for post-operative nausea and vomiting. Trachea was extubated when the TOF count was 4 after administering neostigmine and glycopyrrolate. Post-operative analgesia was maintained with paracetamol 1 g IV 6th hourly and fentanyl boluses of 25 µg as rescue whenever visual analogue scale exceeded 3. There was no worsening of movements post-operatively and she was discharged after 10 days.

The overproduction of mutant protein, huntingtin is the cause of degeneration of neurons in caudate, putamen and cortex. Patients receive psychotropic medications (antipsychotics, antidepressants, benzodiazepines and antiepileptics) which can interact with anaesthetic agents.^[2] They may not be compliant with the treatment as also the case with our patient.

The fear of prolonged response to anaesthetic agents makes intraoperative management challenging. Prolonged apnoea and recovery from thiopentone, prolonged effect of benzodiazepines^[3,4] and prolonged apnoea with use of succinylcholine have been reported (with low levels of plasma cholinesterase).^[5] Contrary to above, in a large review of 11 patients, authors have reported normal response to thiopentone and propofol and did not observe prolonged amnesia with midazolam or abnormal response to succinylcholine.^[2] Safe use of total intravenous anaesthesia with remifentanyl and propofol is reported.^[6] Rocuronium use and reversal of neuromuscular blockade with sugammadex is reported, considering aggravation of choreiform movements with neostigmine and anticholinergics.^[7]

Robot-assisted surgery is advantageous due to lesser blood loss, decreased incidence of surgical complications, decreased pain scores and thereby shortened hospital stay. However, there are physiologic consequences due to prolonged pneumoperitoneum along with spatial restrictions and issues in proper patient positioning. Intraoperative management of patients undergoing robot-assisted procedure needs good communication between anaesthesiologist and surgeon.^[8]

Our case report emphasises extreme vigilance required in perioperative management of patients with HC undergoing robot-assisted surgery in view of disease-related neurological symptoms, lack of patient

cooperation, possible effects of certain anaesthetic agents prolonging the duration of anaesthesia and the concerns due to robot-assisted surgery.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Ankita Batra, Neeru Sahni, Uttam K Mete¹

Departments of Anaesthesia and Intensive Care and ¹Urology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Address for correspondence:

Dr. Neeru Sahni,
Department of Anaesthesia and Intensive Care, Postgraduate Institute of Medical Education and Research, Chandigarh - 160 012, India.
E-mail: neerunalin@yahoo.com

REFERENCES

1. Walker FO. Huntington's disease. *Lancet* 2007;369:218-28.
2. Kivela JE, Sprung J, Southorn PA, Watson JC, Weingarten TN. Anesthetic management of patients with Huntington disease. *Anesth Analg* 2010;110:515-23.
3. Davies DD. Abnormal response to anaesthesia in a case of Huntington's chorea. *Br J Anaesth* 1966;38:490-1.
4. Rodrigo MR. Huntington's chorea: Midazolam, a suitable induction agent? *Br J Anaesth* 1987;59:388-9.
5. Gualandi W, Bonfanti G. A case of prolonged apnea in Huntington's chorea. *Acta Anaesthesiol Scand* 1968;19 Suppl 6:235-8.
6. MacPherson P, Harper I, MacDonald I. Propofol and remifentanyl total intravenous anesthesia for a patient with Huntington disease. *J Clin Anesth* 2004;16:537-8.
7. Khan MH, Banerjee A. Anaesthesia and orphan disease: Sugammadex in a patient with Huntington's disease undergoing thyroid lobectomy. *Eur J Anaesthesiol* 2012;29:593-5.
8. Kakar PN, Das J, Roy PM, Pant V. Robotic invasion of operation theatre and associated anaesthetic issues: A review. *Indian J Anaesth* 2011;55:18-25.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/0019-5049.193702

How to cite this article: Batra A, Sahni N, Mete UK. Anaesthetic management of a patient with Huntington's chorea undergoing robot-assisted nephron-sparing surgery. *Indian J Anaesth* 2016;60:866-7.