ANAE ThESIA AND HUNTINGTON’S CHOREA

Dr. Anurag Yadava¹ Dr. Pradip K. Battacharya²
Dr. Rajnish K. Jain³ Dr. Ramesh C. Agarwal⁴

SUMMARY

Huntington’s chorea is a very rare degenerative disease of the nervous system. Reported anaesthesia experience with Huntington’s chorea is very limited. Patients with Huntington’s chorea are at higher risk of intraoperative complications. Successful anaesthetic management of a case of Huntington’s chorea posted for a urological procedure is presented in this report.

Keywords : Huntington’s disease, Ataxia, Dementia.

Huntington’s chorea is a rare hereditary disorder of the nervous system. It was first described by the American physician George Huntington in 1872, who used data from families living in Huntington county, New York. We report the successful anaesthetic management of a patient with Huntington’s chorea posted for surgical management of ureteric calculus.

Case report

A 42 year old male with left ureteric calculus was admitted. He was a known case of Huntington’s chorea for the last four years and was not taking any significant medication. After proper neurology consultation, the patient was put on tab. tetrabenazine 25 mg daily. The patient was cachectic, had progressive mental deterioration, ataxia and gross abnormal chorea movements. His physical examination revealed a pulse rate of 86/minute and blood pressure of 130/86 mmhg. His other investigations like plasma glucose, urea, electrolytes, blood count, CXR and ECG were within normal limits. Intravenous pyelography revealed complete duplication of ureter and the stone was impacted at vesicoureteric junction of lower moiety ureter. The risk of anaesthesia was explained and informed consent was obtained.

Tab. Tetrabenazine was continued through the day of surgery. Tab. ranitidine 150 mg and tab. ondansetron 8 mg were given night before surgery. Patient was shifted to OT and intravenous line was secured. Patient’s vital signs were monitored with NIBP, ECG and Pulse oximeter. The patient was preoxygenated with 100% O₂ for 3 minutes a rapid sequence induction using inj. propofol 60 mg and inj. rocuronium 40 mg I.V. with cricoid pressure applied was done and intubation was done with a 8.5 mm cuffed endotracheal tube. The cricoid pressure was released only after inflating the ET cuff.

Anaesthesia was maintained with inj. fentanyl 100 mg and isoflurane 0.8 – 1.0 %. To determine the degree of muscle relaxation, we used train of four (TOF) monitoring. After forty minutes, all four twitches of the TOF returned to control level and first increment of inj. rocuronium 10 mg was given. After 25 minutes, a second increment of inj. rocuronium was given as four twitches were detectable again and the TOF ratio was > 0.75.

He underwent lower moiety ureteric meatotomy with URS Tripsy and DJ Stenting.

All the vital signs were maintained intraoperatively. At the end of the surgery which lasted 100 minutes, the TOF recovered to control values and isoflurane was discontinued. The patient was extubated uneventfully with reversal using inj. neostigmine 2.5 mg and glycopyrrolate 0.4 mg I.V.

Immediate postoperative period was uneventful. A week later the stent was removed under local anaesthesia and the patient was discharged on fifteenth postoperative day.

Discussion

Huntington’s disease is a premature neurodegenerative disease characterized by marked atrophy of the basal ganglia, particularly the caudate nucleus. It is a genetic disease transmitted in an autosomal dominant pattern.¹ The Huntington’s disease gene has been identified and is expressed when the patient is in adulthood (35-40 year of age). Early onset of symptoms is associated with more rapid and severe progression of the disease.
Disordered movement and dementia are the clinical hallmarks of the disease. Movement abnormalities can involve the extremities, trunk, face, eyes, mouth, oropharynx and respiratory muscles, thereby leading to an increased risk of aspiration pneumonitis and consequently antiaspiration maneuvers must be used. The disease progresses for several years and accompanying mental depression makes suicide a frequent occurrence.2

Experience with the management of anaesthesia in Huntington’s chorea is too limited to propose specific drugs or techniques. Reported anaesthesia experience in patients with Huntington’s disease is largely anecdotal and consists of a small number of case reports and letters.3-11 Although there are no specific contraindications to using intravenous or inhaled anaesthetics, delayed awakening and generalized tonic spasms have been reported after administration of thiopental.7 In contrast, a normal response with rapid recovery has been reported with propofol in patients with Huntington’s disease.12-14

It has been suggested that these patients may be sensitive to the effects of non-depolarizing muscle relaxants although the reported response to atracurium has been normal.13,15 Interestingly, the incidence of decreased pseudocholinesterase activity is substantially higher in Huntington’s disease patients than in normal population.6 This may explain the single report of prolonged response to succinylcholine. The use of sevoflurane and mivacurium for general anaesthesia in patients suffering from Huntington’s disease seems to be effective and safe.16

We used glycopyrrolate in this case perioperatively since from Huntington’s disease seems to be effective and safe.16 We used glycopyrrolate in this case perioperatively since mivacurium for general anaesthesia in patients suffering from Huntington’s disease than in normal population.5 Prolonged apnea following suxamethonium can be averted by using rocuronium when rapid sequence induction becomes necessary.17 For lower abdominal and perineal surgery, spinal anaesthesia is a tangible alternative although achieving proper position and avoidance of trauma remains difficult in these patients.11

Factors of considerable concern to the anaesthesiologist who treats patients with Huntington’s disease may include how to treat these frail elderly people incapable of cooperation, how to treat patients suffering from malnourishment and how to treat patients with an increased risk for aspiration or exaggerated response to sodium thiopental and succinylcholine. Other concerns regarding the anaesthetic management are determined by the level of physiologic debilitation in the individual patient.

References