ANESTHETIC MANAGEMENT OF A PATIENT WITH DEEP BRAIN STIMULATION IMPLANT FOR GENERALIZED DYSTONIA: A CASE REPORT

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Abstract

Background: Generalized dystonia is a hereditary neurological syndrome with early onset, characterized by repetitive movements and muscle contractions leading to abnormal posture. A growing number of patients with dystonia resort to Deep Brain Stimulation (DBS) implantation as a form of therapy that will improve their quality of life as well as their motor dysfunction.

Case: We present the case of a 45 year old female with generalized dystonia and DBS scheduled for debulking of ovarian cancer. Anesthetic management of patients with dystonia and specific considerations regarding brain implantable devices are being reported.

Conclusion: Anesthetists should begin to understand both the function and the possible perioperative adverse effects of DBS devices as more and more patients will either proceed to their implantation or will present for elective procedures while having them already implanted.

Keywords: General anesthesia; Anesthetic Management; DBS; Dystonia; Electrocautery.

Introduction

Dystonia is a neurological syndrome characterized by involuntary, prolonged muscle contractions that produce, either alone or in combination, twisting movements, repetitive movements, or abnormal postures.[1] The contractions may be sustained or intermittent, movements may be patterned or in some cases resembling tremor and tend to be worsened during voluntary action. Classification may vary depending on whether it is based on etiology, clinical features, time of onset, concomitant degenerative movement disorders and body parts affected. Our patient suffered from generalised dystonia, a subgroup with early childhood onset. It is the most common form of hereditary dystonia, inherited with an autosomal dominant way, with the responsible abnormal gene called DYT1 mapped to chromosome 9q34. While treatment was previously restricted to oral medications with often intolerable side effects, the widespread application of deep brain stimulation (DBS) the past decade has led to great improvement in the quality of life of these patients.[2] Approval and written informed consent was received from the patient.

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Case Report

We present the case of a 45-year-old female who was scheduled for debulking of ovarian cancer. She had been treated for generalised dystonia from childhood with symptoms including twisting and repetitive movement of her upper and lower limbs. As her clinical presentation was poorly controlled by pharmacological regimens, DBS implantation surgery was performed in 2009 with no perioperative complications. The DBS device was implanted on the left side of her thoracic wall stimulating the globus pallitus interna (GPi DBS). Since then it had been regularly checked every year for normal function and battery. Her treatment also included 5mg of trihexyphenidyl for a better control of her voluntary muscle movement.

A careful preoperative evaluation was performed in order to assess all issues related to her dystonia. Our clinical examination showed a Mallampati score of II, with a slightly reduced thyromental distance (5 cm) and a reduced mouth opening (two of patient's fingers). Neck extension was adequate.

After communication with the technician responsible for her implanted GPi DBS device, it was decided that his presence was needed in the operation theatre in case the device needed to be switched off.

Upon arrival at the operation room standard monitoring, venous access and catheterization of the right radial artery for invasive blood pressure measurement was installed at the patient. Premedication of 1mg of midazolam was given intravenously.

Anesthesia induction was achieved with 230mg of propofol, 100mcg of fentanyl and 50mg of rocuronium. Bag mask ventilation was performed for 90 seconds without difficulty and a 7mm cuffed tracheal tube was inserted during videolaryngoscopy with CMAC. For maintenance, general inhalational anesthesia was preferred with the use of sevoflurane 1% and an oxygen-N_20 mixture delivered at 1:1 ratio aiming for a MAC of 1. Intraoperatively incremental doses of 50mcg fentanyl were given in order to maintain adequate levels of analgesia as well as 1g of paracetamol, 75mg of diclofenac and 8mg of morphine as part of the multimodal analgesia plan.

An effort was made by the surgical team to work with bipolar cautery, however there was dissatisfaction with the results and a decision to continue with monopolar cautery was made. The technician in charge switched off the neuro-stimulator before the change of the cautery and after induction so as to avoid any exacerbation of dystonia. While changing the cautery from bipolar to monopolar it was made sure that the current path between the pad and active electrode of the electrocautery/surgery system was as far away as possible from the DBS conductive path.

The procedure was uneventful with the patient maintaining stable blood pressure and heart rate with normal Spo_2 and $EtCO_2$ measurements.

When four responses to TOF were seen, the DBS device was switched on again and the neuromuscular block was antagonized with $2mg.kg^{-1}$ suggamadex. No signs of dystonia were observed during the emergence procedure and the patient was extubated successfully. She remained under observation in the post anesthesia care unit and then was transferred to the ward where instructions for post-operative analgesia were given.

Discussion

The term dystonia corresponds to a broad spectrum of movement disorders in which sustained involuntary muscle contractions lead to abnormal postures or repetitive movements.[3] Our patient suffered from a specific type of primary dystonia called generalised dystonia or DYT1 related dystonia. Standard therapy treatment includes botulinum toxin types A and B, anticholinergics, benzodiazepines, baclofen, dopamine blocking and depleting agents, sensory motor behavioral intervention therapy and surgery.[1,4] Surgical interventions vary from stereotaxic pallidotomy and thalamotomy to neuro-stimulator implantation. As described for refractory dystonia,[5] in our case bilateral globus pallidus internus deep brain stimulator (GPi DBS) was implanted, improving the patient's motor control.

Preoperative evaluation of patients with dystonia should focus on the subtype from which the patient suffers, the degree and the severity of one's dystonic movements, and the presence of comorbidities.[3] A neurological evaluation of the patient's baseline neurocognitive function is necessary. Our patient was

under close surveillance, so no contraindication was presented towards her proceeding with the surgery.

Thorough airway examination and early preparedness for potential difficult bag mask ventilation and intubation is of crucial importance. Symptoms involving the oropharynx and the cervical part of the spine along with reduced neck and jaw mobility and mouth opening may hinder direct laryngoscopy.[3] A videolaryngoscope was preferred as we wanted to minimize the likelihood of a failed intubation technique and achieve a better view of the tube insertion through the vocal cords, because patients with dystonia are under greater danger of aspiration and laryngospasm.[5]

As in patients with Parkinson's disease, a possibility of restrictive lung disease and low coughing capacity indicates the need of a pulmonary evaluation, which in our case was within normal values.

No type of anesthesia appears to be contraindicated in patients with dystonia.[4,6] Spasms are abolished both by the use of neuromuscular blockade and the inhalation of N_2O in concentrations above 50%. We used propofol for induction and inhalational anesthesia with sevoflurane and an oxygen- N_2O mixture for maintenance of anesthesia. Monitoring neuromuscular blockade with peripheral nerve stimulators seems to be acceptable as long as it is ensured that the path of the electrical stimulation does not pass through the DBS system.[6,7] The same rule is applied when a peripheral nerve stimulator is used as assistance to a peripheral nerve block.[7]

In terms of the DBS system itself, it consists of several components like the intracranial electrodes and the pulse generator, implanted into the chest in the infraclavicular area.[7] Chest X-ray for identification of the nature of the device and its wires is deemed necessary preoperativel.[7] With a battery spanning from two to five years,[8] a DBS technician has to conduct a detailed control of the device's settings. In our case the technician was present throughout the whole procedure and was responsible for the turn-off after induction and the turn-on before emergence. This timing aims to avoid the recurrence of episodes that will hinder the anesthetist's work and provoke discomfort to the patient. During the procedure, artifacts may be seen in the ECG recordings, requiring DBS system deactivation for a proper interpretation.[4,7]

During surgery, the use of bipolar cautery seems to be the norm, at the minimum power settings with short intermittent irregular bursts[7-9] because the electrical current is confined between the two electrodes of the electrosurgical unit. In contrast during monopolar mode the current generated through the electrode enters the patient's body, reaching the grounding pad and completing the circuit. The danger with cautery equally involves damage to the implantable pulse generator (IPG) causing altered stimulation or even deactivation of the device and overheating of the DBS electrode tip inside the brain[6,7] leading to severe brain injuries in some cases.[10] In our case, the procedure was impossible to be performed with the use of bipolar cautery so a switch to monopolar was made, after DBS deactivation, taking into account all the safety measures mentioned in the literature, like placing the grounding pad so that current path between cautery electrode and pad doesn't pass through DBS system and using a low voltage mode in the lowest possible power setting.[6,7] We faced no difficulties with reactivating the DBS system, the patient's dystonic movements were well controlled postoperatively and no injury or worsening of our patient's condition was observed.

We suggest a close collaboration between surgeons, anesthetists and neurologists when dealing with such cases. Technicians should be informed in time for evaluating implantable devices. Anesthetists should be able to handle such devices intra and postoperatively as they become widely more popular among our patients.

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Conflicts of interest: All authors declare that there are no conflicts of interest.

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