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CLINICAL VIGNETTE

Anesthetic Management of a Patient with Lubag Syndrome

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Background

Lubag syndrome, also known as X-linked dystonia Parkinsonism, is a rare adult onset progressive neurodegenerative movement disorder that primarily involves the striatum in the brain.¹ It is categorized as a primary monogenic dystonia caused by a mutation in the TATA-binding protein associated factor (TAF-1) gene at the DYT3 locus of chromosome Xq13.1.² It is inherited in an X-linked recessive manner and has a high penetrance.

The condition is endemic in the Panay Island, in the Philippines, though there have been cases in people of maternal Filipino descent reported in other countries.³ The prevalence rate in the Philippines is 0.31 per 100,000, but increases to 5.74 per 100,000 on the island of Panay. Men are primarily affected with a gender ratio of 100:1.³ Patients are typically diagnosed after they become symptomatic during their third or fourth decade of life, and the disorder progresses until death usually in the fifth decade.³

Clinical presentation is variable with 94.3% of patients having progressive and severe focal dystonia, eventually leading to combined dystonia and Parkinson symptoms or Parkinsonism.³ Dystonic presentation includes blepharospasm, facial twitching or spasm, jaw opening or closing, involuntary tongue protrusion, torticollis, myoclonic and choreiform movements, dorsiflexion of the big toe, foot flexion, leg spasm, and gait dystonia. About 6% of patients present with symptoms of pure Parkinsonism, ie. bradykinesia, rigidity, and shuffling gait.³

Since there is no cure for Lubag syndrome, care is focused on symptomatic treatment. For dystonia, benzodiazepines, anticholinergics, and botox have been used, with limited supporting evidence.⁴ Tetrabenazine, zolpidem, and levodopa have shown more promise in improving symptoms.⁵ For patients refractory to medication, deep brain stimulation may provide improvement in voluntary motor control.⁶

Case Report

A 40-year-old Filipino male with Lubag dystonia (X-linked dystonia-parkinsonism) presented for direct laryngoscopy and biopsy of a vocal cord polyp, which was causing symptoms of dysphonia and mild dysphagia. The patient was diagnosed with Lubag dystonia approximately three years prior to presentation when he developed hemi-dystonia with parkinsonian features. Patient had a known family history of X-linked recessive

transmission of this syndrome and his birthplace was in Panay Island, Philippines.

His past medical history included well-controlled hypertension and prior tobacco use. The patient had no previous surgical or anesthesia history. He was on the following prescription medications: olmesartan, botox, sinemet, amlodipine, baclofen, clonazepam, trihexyphenidyl, and cannabis oil.

On physical exam, the patient had a height of 167 cm, weight of 49.6 kg, and BMI 17.8. His airway exam was notable for a Mallampati classification of III, decreased submental space of one fingerbreadth, but with adequate mouth opening and full neck range of motion. A bronchoscopic exam performed by the otolaryngologist revealed an omega shaped epiglottis, mobile vocal cords bilaterally with a large polypoid lesion along the right anterior commissure of the true vocal cords that did not appear to arise from the arytenoid cartilage. His neurologic exam was notable for rigidity in his upper extremities (right worse than left), frequent dystonic posturing of the neck with rotation, resting pill rolling tremor, and occasional dystonic posturing in the upper and lower extremities resulting in difficulties with gait.

On the day of his surgery, the patient took his outpatient oral medications Sinemet, amlodipine, and baclofen with a small sip of water, as instructed. He was taken to the operating room and placed on standard anesthesia monitors. After pre-oxygenation with 100% oxygen, general anesthesia was induced intravenously (IV) with midazolam 2mg, fentanyl 100 mcg, lidocaine 2% 50 mg, propofol 100mg, and rocuronium 50 mg. Patient was intubated via direct laryngoscopy with a MAC 4 blade using a size 5 laser endotracheal tube. Dexamethasone 10mg IV was given at the start of the case to reduce swelling from the vocal cord biopsy. Case duration was 46 minutes.

At the end of the case, the patient was given ondansetron 4mg IV to prevent post-operative nausea and vomiting. The neuromuscular blocking agent was reversed with glycopyrrolate 0.6mg IV and neostigmine 3mg IV after obtaining three twitches on the train-of-four test. After meeting all extubation criteria, patient was extubated uneventfully and taken to the post-anesthesia care unit for recovery. Patient was discharged home later the same day with outpatient follow-up in head and neck (ENT) surgery clinic.

Discussion

Patients with Lubag syndrome present distinct challenges to anesthesiologists given the variability of their neurologic manifestations. These patients should be approached similarly to patients with other dystonias and neurodegenerative disorders. Particular attention must be given to timing and administration of anti-psychotic medications and their potential interactions with anesthetics to prevent complications such as neuroleptic malignant syndrome.^{7,8} Dopamine agonists should be administered as close as possible to the time of surgery and restarted as soon as possible after surgery.⁷ Patients may have a temporary worsening in symptoms if these medications are withheld for extended period of time.⁸ Patients with sialorrhea can be treated with glycopyrrolate or Botox M injections of the parotid glands.⁸ For patients with dysphagia, aspiration precautions should be taken and suction should be readily available prior to induction of anesthesia.⁸

Typically, general anesthesia is safe for patients with severe dystonia. It is recommended to discontinue Monoamine oxidase (MAO)-B inhibitors for at least one week prior to surgery, to avoid halothane use with levodopa given the increased risk of cardiac sensitivity to catecholamines, and to use opioids with caution given the potential for worsening rigidity.⁸ Propofol generally has anti-parkinsonian effects, but can worsen dyskinesias and must be used with care in select patients.⁸ For patients who present with rigidity, frequent repositioning is recommended.⁸

Post-op nausea is best controlled with ondansetron, domperidone, or trimethobenzamide.⁸ Dopamine antagonists should be avoided. Opioids should be used with caution for controlling post-op pain, and meperidine should be given with caution in patients taking MAO inhibitors.

Our patient underwent direct laryngoscopy and biopsy of a vocal cord polyp under general anesthesia uneventfully. The patient did well under anesthesia with stable vital signs post-operatively and was discharged from the PACU on the same day.

Conclusion

We present the case of a middle age male with Lubag syndrome (X-linked dystonia parkinsonism) who underwent general anesthesia for biopsy of a vocal cord polyp. No complications were noted during the case or the post-operative period. We had discussed above the pre-operative, intra-operative, and post-operative anesthesia precautions for patients with Lubag syndrome undergoing surgery.

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