

## CASE REPORT

# Radio Frequency Ablation of Globus Pallidus for Meige's Syndrome: An Anaesthetic Challenge

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**ABSTRACT**

**Background:** Meige syndrome is a rare neurological condition characterized by oromandibular dystonia and blepharospasm. Treatment modalities includes medications like anticholinergics, dopamine antagonist, GABA agonist, physical or occupational therapy, baclofen injections, and surgical interventions like ablation or deep brain stimulation of Globus pallidus internus nuclei [GPI].

**Case Details:** We present a case of 31 year old male patient with abnormal involuntary movement of head, neck, left shoulder and arm since three years, diagnosed as "Meige syndrome". Patient was planned for right GPI radiofrequency ablation [RFA]. Thorough preoperative assessment was done. Significant findings were severe head and neck dystonia, and anticipated difficult airway. Surgery was planned under monitored anesthesia care with titrated sedation to allow for intra-operative neurologic testing. On the day of surgery, patient had severe cervical dystonia, not subsiding even after sedation with dexmedetomidine, requiring change of anaesthesia plan to general anaesthesia with secured airway as an airway intervention would have been difficult after application of the head frame. Standard balanced anaesthesia with nasal endotracheal tube was administered. BIS was used to monitor depth of anaesthesia. Surgeon required intraoperative testing during RFA of right GPI. About 20 minutes prior to testing, anaesthetic agents were stopped and BIS around 90 was documented. Patient cooperated well for neurological testing during localization and RFA of right GPI. After RFA, general anesthesia was continued. Procedure was uneventful, patient was extubated and shifted to ward after observation in recovery room and discharged on post-operative day three.

**Discussion and Clinical relevance:** RFA of GPI is a modality of treatment for patient with Meige syndrome. Anaesthesia management may be challenging due to exacerbation of the abnormal involuntary movements during surgery. Anesthesia

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may be in form of monitored anesthesia care with sedation or general anesthesia with intra-operative testing during lesioning. Anesthesia drugs need to be titrated to facilitate awake testing during radio-frequency ablation procedure.

**Conclusion:** Exacerbation of dystonia may necessitate change of anesthesia plan to general anesthesia with awake testing during lesioning. Patient must be counselled in preoperative period to cooperate for testing. Team work, planning and patient counselling for cooperation during procedure, aids in safe conduct of procedure.

#### KEYWORDS

• Meige syndrome • Cervical dystonia • Radiofrequency ablation of globus pallidus • General anaesthesia

## INTRODUCTION

Dystonia is a movement disorder characterized by abnormal, sustained muscle contractions leading to abnormal posture and /or repetitive movements.<sup>1</sup> Meige syndrome is a rare neurological disorder, characterized by involuntary, forceful contractions of the muscles of the face. Patient presents with symptoms of oro-facial dystonias like difficulty in opening mouth, deviation of the jaw, grinding of teeth and blepharospasm<sup>2,3</sup> Exact etiology for Meige syndrome is not known, but it is related to abnormalities in the basal ganglion.<sup>3</sup> There is no complete cure for Meige syndrome. Treatment goals are to reduce symptoms and improve the functional quality of life. Medical Management includes anticholinergics, dopamine antagonist, GABA agonist and baclofen injections.<sup>4</sup> Surgical modalities for the treatment include ablation or deep brain stimulation (DBS) of the Globus pallidus internus nuclei (GPI). We present a case of a patient with Meige syndrome posted for radiofrequency ablation (RFA) of right GPI.

## CASE DETAILS

A 31 year old male patient, weighing 64 kg presented with abnormal involuntary movement of head, neck, left shoulder, arm and left hemi-dystonia since three years. He also had slurring of speech. He was on Tetrabenazine 25 mg and Clonazepam 0.5 mg per day. He was diagnosed case of "Meige syndrome" and was advised deep brain stimulation or radiofrequency ablation (RFA) of right globus pallidus internus (GPI) nuclei. He was scheduled for RFA of right GPI nuclei. A thorough preoperative anaesthetic evaluation was conducted. Airway assessment was suggestive of "anticipated difficult airway"

in view of Mallampati Grade 3 and restricted neck extension. All other investigations were within normal limits. He was explained about need for intra-operative testing during surgery. He was counselled for surgery under local anaesthesia to facilitate intra-operative testing to rule out adverse effects during ablation. Appropriate consents were obtained and standard fasting guidelines were followed. Operation theatre (OT) was prepared taking into consideration patient's condition and plan for testing during RFA of the right GPI. Anaesthesia plan was monitored anesthesia care with local anaesthetic infiltration at site pins of the Leksell stereotactic head frame and burr hole.

Standard American society of Anesthesiologist (ASA) monitoring was instituted. A large bore intra-venous access (IV) was secured. Patient had severe exacerbation of involuntary movements of head and neck, making application of the head frame difficult. Sedation with injection Dexmedetomidine was instituted, but there was no decrease in involuntary movements. Further sedation may have required airway intervention, which would have been difficult with head frame applied and anticipated difficult airway. Anesthesia plan was changed to general anaesthesia with endotracheal tube (ETT) with intra-operative waking up the patient to aid intra-operative neurologic testing. Patient was counselled about the waking up during surgery to allow for the same. General anaesthesia was induced with fentanyl 1 mcg/kg, Propofol 2.5 mg/kg and Cisatracurium 0.2mg/kg. Airway was secured with seven size cuffed nasal ETT. Anaesthesia was maintained with sevoflurane with air plus oxygen, along with dexmedetomidine 0.5 mcg/kg/hr, titrated infusion of propofol and

bolus dose of cisatracurium 0.015 mg/kg. Depth of anesthesia was monitored using BIS intra-operatively. Later head frame was attached, followed by CT scan and then patient was taken in OT for right GPI ablation. Surgical draping was done to allow for awaking up of the patient and conduct neurologic testing. (Figure 1) Around 20 minutes prior to testing required, dexmedetomidine, propofol and sevoflurane was stopped. When BIS was around 90, patient was asked to follow commands. (Figure 2) Macrostimulation was done to test for side-effects. After appropriate testing, radiofrequency ablation of right GPI nuclei was done with 70 degree temperature for 40 seconds. Then general anesthesia was continued with fentanyl bolus, sevofurane in air+oxygen and propofol infusion. Intra-operative course was uneventful. After the surgery patient was extubated and observed in post-anesthesia care unit for an hour, and then shifted to ward. Dystonias were reduced post-operatively. He had uneventful course and was discharged from hospital on post-operative day three.

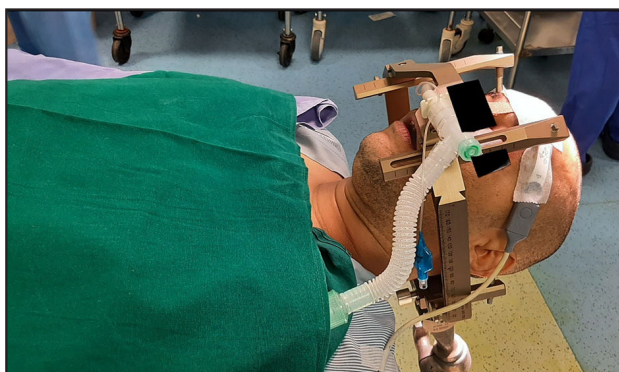


Figure 1:

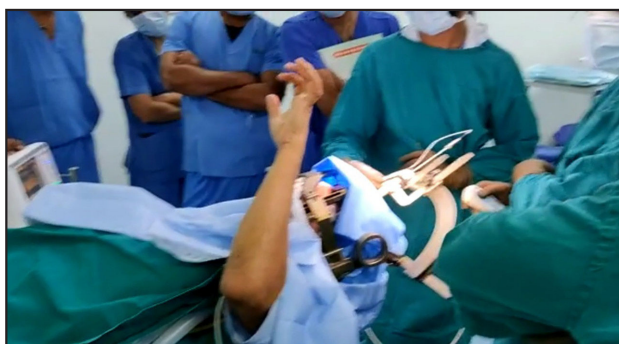


Figure 2:

### Discussion and literature review

Stereotactic functional neurosurgery either in form of lesioning or DBS of GPI is considered for treatment of Meige syndrome in patients,

who are refractory to the medical management or have side-effects related to it.<sup>1</sup> Though DBS is preferred modality, lesioning may be considered in some group of patients who have financial constraints, cannot be followed-up or at high risk of infection.<sup>5</sup> Our patient was planned for RFA of the right GPI.

Anaesthesia goals include adequate operating condition, comfortable and co-operative patient to facilitate intra-operative neurologic testing to localize target and prevent adverse effects, a safe airway and hemodynamic stability.<sup>6</sup> Macrostimulation testing to see clinical benefits and adverse effects requires an awake, co-operative patient.<sup>6</sup> Monitored anaesthesia care with local infiltration or conscious sedation may be used. General anesthesia may be required in some group of patients like those with psychiatric dysfunction, severe dystonia, anxiety or pediatric patients.<sup>6</sup> Our patient had severe cervical dystonia on the day of surgery. Stress may cause exacerbation of dystonias in some patient. Jahngir et al suggest, most patients will have sensory tricks in form of sensory stimuli, like sleeping, relaxing, talking, walking, yawning or drinking beverages, that can alleviate their dystonias.<sup>7</sup> Unfortunately our patient had no specific sensory trick to alleviate his symptoms. Dexmedetomidine is considered an effective agent for sedation in these cases.<sup>8</sup> There was no decrease in dystonia even after sedation with dexmedetomidine in our case. Administration of further sedation may compromise the airway, necessitating need to secure the airway. Airway management may be a challenging after application of stereotactic frame due to limited access to patient head.<sup>9</sup> Hence, we decided to proceed with general anesthesia with ETT for the surgery. But general anaesthesia may interfere with clinical testing and recognition of adverse effects due to stimulation of adjacent structures.<sup>6</sup> Under GA location of electrodes may be confirmed with Magnetic resonance imaging MRI without stimulation.<sup>6,10</sup> Visual evoked potentials may be used in patients undergoing DBS under GA.<sup>6</sup> As ablation was planned, our surgeon wanted to confirm accurate positioning with macrostimulation and testing to ensure no adverse effects. Asleep-awake-asleep technique may be used to aid macrostimulation testing in such cases.<sup>6</sup> We titrated the anaesthesia to allow for intra-operative waking up of the patient to co-

operate for testing during macrostimulation and RFA.

## CONCLUSION

Surgical modalities of treatment for Meige syndrome may be DBS or RFA of GPI. Exacerbation of the abnormal involuntary movements in peri-operative period makes anaesthesia management challenging in these patients. Anesthesia may be in form of monitored anesthesia care with sedation or general anesthesia with intra-operative testing to avoid adverse effects related to pallidotomy. Patient preparation and counselling to facilitate intra-operative testing is very important. Team work, planning and patient counselling for cooperation during procedure, aids in safe conduct of procedure.

**Conflicting Interest:** Nil

**Source(s) of support:** Nil

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