Non-Intubated Uniportal Subxiphoid VATS Thymectomy for Myasthenia Gravis: A Case Report with review of literature.

Shachee Bhattarai[1], Binay Thakur[1], Sagar Khatiwada[1], Manoj Tiwari[1], Mahesh Mani Adhikari[1], Sandeep Sapkota[1] Saujan Raut [1] Yogesh Regmi[2] Rashmi Thapa[2]

[1]Department of Thoracic Surgery, B.P. Koirala Memorial Cancer Hospital, Bharatpur, Nepal.
[2]Department of Anesthesiology, B.P. Koirala Memorial Cancer Hospital, Bharatpur, Nepal.

Abstract

Background: Myasthenia gravis (MG) is an autoimmune neuromuscular disorder characterized by fluctuating skeletal muscle weakness caused by antibodies against components of the neuromuscular junction, most commonly the acetylcholine receptor (AChR). Thymectomy is an established treatment for patients with MG, particularly those with AChR antibodies. Recently, uniportal subxiphoid has become the approach of choice offering improved postoperative recovery and reduced complications. Non-intubated thoracic surgery has also gained attention for minimizing anesthetic-related complications in MG patients.

Case: Twenty two year old female diagnosed with AChR-positive, muscle specific tyrosine kinase (MuSK) negative MG receiving pyridostigmine who underwent uniportal subxiphoid VATS (Video assisted thoracoscopic surgery) thymectomy using local anesthesia and laryngeal mask airway (LMA) without muscle relaxant at B.P. Koirala Memorial Cancer Hospital (BPKMCH), Nepal. Preoperative evaluation included neurological assessment, serological testing, and radiologic imaging. The procedure was successfully completed without conversion to open surgery or endotracheal intubation. Postoperatively, the patient experienced an uneventful recovery and at 6 months follow up she was symptom free with reduced dose of pyridostigmine.

Conclusion: Non-intubated uniportal subxiphoid VATS thymectomy is a safe and effective surgical option for MG patients. This technique may reduce postoperative complications and facilitate faster recovery compared with conventional intubated approaches. Our case highlights the potential benefits of minimally invasive thymectomy combined with non-intubated anesthesia in the management of myasthenia gravis.

Keywords: Myasthenia gravis, Uniportal, Subxiphoid, Thymectomy

Introduction

Myasthenia gravis is a chronic autoimmune disorder characterized by weakness and fatigability of skeletal muscles due to impaired neuromuscular transmission.[1] The pathophysiology involves antibodies directed against components of the postsynaptic membrane of the neuromuscular junction, most commonly the acetylcholine receptor (AChR), although other antibodies such as muscle-specific tyrosine kinase (MuSK) may also be involved. These antibodies disrupt neuromuscular transmission by reducing the number and function of acetylcholine receptors, resulting in muscle weakness and fatigue.[2]

The thymus plays a central role in the pathogenesis of MG, and thymic abnormalities such as thymic hyperplasia or thymoma are frequently observed in affected patients.[3] Consequently, thymectomy has become an important therapeutic option in the management of MG, particularly in patients with generalized disease and AChR antibodies. Clinical trials have demonstrated that thymectomy improves clinical outcomes and reduces the need for long-term immunosuppressive therapy compared with medical management alone.[4]

Traditionally, thymectomy was performed

Correspondence: Shachee Bhattarai, [1]Department of Surgical Oncology (Thoracic Unit). B.P. Koirala Memorial Cancer Hospital, Bharatpur, Nepal.

through a transsternal approach, which provided adequate surgical exposure but was associated with significant morbidity, postoperative pain and prolonged recovery.[5]2 patients (25% Over the past two decades, minimally invasive techniques such as subxiphoid, transcervical have been increasingly adopted, offering comparable oncologic outcomes with reduced postoperative pain, shorter hospital stay and improved cosmetic results.[6]

Among the minimally invasive approaches, uniportal subxiphoid thymectomy has emerged as a promising technique. This approach allows excellent visualization of the anterior mediastinum and both phrenic nerves, enabling complete resection of the thymus and surrounding fatty tissue while minimizing intercostal nerve injury.[7]

Another significant development in thoracic surgery is the use of non-intubated anesthesia techniques. Conventional thoracic surgery typically requires endotracheal intubation and mechanical ventilation with use of muscle relaxants, which may pose challenges in MG patients due to their unpredictable response to muscle relaxants and increased risk of postoperative respiratory failure. Non-intubated techniques with laryngeal mask airway (LMA) and local anesthesia avoids the use of neuromuscular blocking agents and reduces airway-related complications.[8]

In this report, we present a case of AChR-positive myasthenia gravis successfully treated with non-intubated uniportal subxiphoid VATS thymectomy at a tertiary cancer center in Nepal.

Case Presentation

A 22-year-old female presented with progressive muscle weakness and fatigue involving the ocular and proximal limb muscles. Her symptoms included intermittent ptosis, generalized fatigue, difficulty in speaking and swallowing, and difficulty performing prolonged physical activities. The symptoms worsened with exertion and improved with rest.

With suspicion for myasthenia gravis, laboratory testing was done which revealed positive AChR antibodies, while MuSK antibodies were negative. Based on clinical findings and serologic testing, a diagnosis of AChR-positive MG with Osserman stage III was established.

The patient was initially managed medically with pyridostigmine 60 mg orally four times daily (QID), which resulted in partial symptomatic improvement. Imaging with contrast enhanced computed tomography (CECT) of the chest demonstrated no evidence of invasive thymoma. (Figure 1)

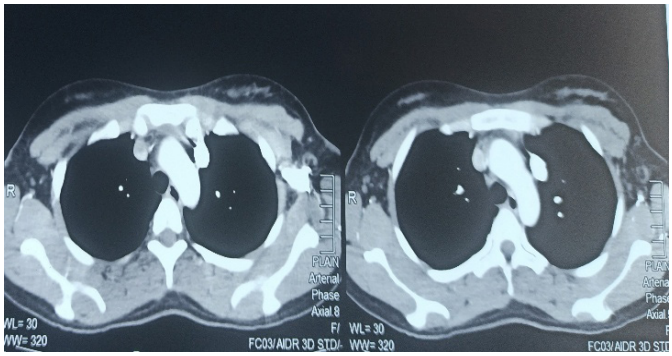

Figure 1 : CECT Chest showing normal thymus without thymoma.

Due to inconvenience and financial constraints for life long therapy of pyridostigmine, patient was referred to our center for an alternative treatment option. Following multidisciplinary evaluation involving neurologists, thoracic surgeons, and anesthesiologists, surgical thymectomy was recommended as part of definitive management. Then patient was scheduled for minimally invasive VATS thymectomy using uniportal subxiphoid approach.

Surgical and Anesthetic Technique

Anesthesia was administered, induction was done with midazolam, fentanyl and propofol then LMA was introduced for ventilation. Maintenance of anesthesia done with infusion for propofol and dexmedetomidine with intermittent ketamine and fentanyl. Continuous monitoring included electrocardiography, pulse oximetry, end-tidal carbon dioxide and arterial blood pressure.

After induction of anesthesia, the patient was placed in the supine position with their legs abducted markings for incision followed by painting and draping was done. The surgeon stood between the patient's legs and the assistant on the patient's right side to operate the camera.

A small vertical incision about 3 centimeter (cm) (Figure 2) was made which was, 1 cm below the lower edge of the xiphoid process after infiltration of local anesthesia with mixture of lidocaine and bupivacaine. Blunt finger dissection created a

retrosternal tunnel between the incision and the thoracic cavity and a wound protector was placed to further optimize the view.

During the operation, a 10-mm 30 degree thoracoscope was placed via uniport and several thoracoscopic instruments could be simultaneously fitted into the uniport. Slight elevation of the sternum using retractor with traction frame was done to improve exposure of the anterior mediastinum. (Figure 3)

Figure 2 : Skin marking for subxiphoid uniportal approach

The right and left mediastinal pleura were cut near the sternal surface up to the level of the right and left internal thoracic veins, right side first and then the left side, as the right mediastinal pleura is easier to open. During spontaneous ventilation, a gradual and natural collapse of the lung occurred as a result of natural pneumothorax. This allowed maximal visualization after creating the incisions and opening the pleura.

The lower edge of the thymus and the connecting pericardial fat were easily visualized and a thymectomy was started caudally and proceeded gradually in cranial direction. The pericardial fat as well as the right epiphrenic fat pad was dissected from the pericardium and diaphragm with the right phrenic nerve being the dorsal boundary of dissection. The dissection of the pericardial fat is preceded upwards under thoracoscopic control in an en-bloc fashion, avoiding any attempt to dissect the thymus tissue. As the left mediastinal pleura was opened at a relatively early stage of the procedure, the dissected specimen could be moved out of vision (i.e. from the right into the left chest cavity) in order to facilitate the maneuver substantially.

Then cranially, the thymic veins and arteries were cauterized and divided after proper identification of left innominate vein and ascending aorta. Besides carefully dissecting the prepericardial fat pad, skeletonization of superior vena cava, both innominate veins and the aorta is an objective to remove thymic horns.

Figure 3 : Uniportal subxiphoid VATS thymectomy setup

With the thyro-thymic ligaments as the sole attachments from the cranial side, the upper poles were divided close to the thyroid gland. Dissection of the specimen along the left phrenic nerve was performed similarly as on the right side. At the end of surgery, a clean post-operative field devoid of any fatty tissue from right to left phrenic nerve laterally, both hemidiaphragms caudally, pericardium, innominate veins, aorta posteriorly, sternum anteriorly and suprasternal notch superiorly was achieved.

Finally, the specimen was placed in an endobag and removed through the subxiphoid incision (Figure 4). A single chest tube was inserted through the subxiphoid incision. The subxiphoid incision was closed in standard manner.

Throughout the procedure, the patient maintained on LMA with stable oxygen saturation and hemodynamic parameters. No neuromuscular blocking agents or intubation were required. The surgery was completed successfully with operative time of 165 minutes with negligible intraoperative blood loss, without intraoperative complications or the need for conversion to open surgery.

Postoperative course

The patient was transferred to the recovery unit

in stable condition. Postoperative monitoring focused on respiratory status due to the risk of myasthenic crisis following thymectomy. The patient experienced an uneventful recovery without respiratory distress, prolonged ventilation, or neurological deterioration. She was mobilized early, chest tube removed on 2nd postoperative day and discharged on 7th postoperative day.

Following surgery, the patient showed improvement in symptoms, and the dose of pyridostigmine was reduced from 60 mg four times daily to twice daily. In following days her histopathological report revealed thymic hyperplasia. In 6 months follow up she had remarkable improvement, she was then advised to discontinue the drug completely. Now she is symptom free and in regular follow-up.

Discussion

Myasthenia gravis is strongly associated with thymic pathology, including thymic hyperplasia and thymoma. The thymus is believed to play a central role in the autoimmune process by promoting the production of autoantibodies against neuromuscular junction components.[3] Consequently, thymectomy is widely accepted as a therapeutic intervention in MG.[4]

Figure 4 : Total Thymectomy Specimen (Thymus gland along with mediastinal fat)

Several studies have demonstrated that thymectomy can lead to significant improvement in MG symptoms and reduce the need for long-term immunosuppressive therapy.[9] Randomized clinical trials have shown improved clinical outcomes, including lower disease severity scores and decreased corticosteroid requirements in patients undergoing thymectomy compared with medical treatment alone.[10]

Historically, thymectomy was performed through median sternotomy, which provided excellent surgical exposure but was associated with significant morbidity.[5] Minimally invasive techniques such as VATS have revolutionized the surgical management of thymic diseases but their main disadvantage is inadequate visualization of contralateral side of phrenic nerve as well as tissue located in another side of mediastinum which may lead to incomplete excision and hence recurrence. Hence, quite often either bilateral VATS is used or an extra contralateral telescopic port is inserted from contralateral side.[7]

Among minimally invasive techniques, uniportal subxiphoid approach offers several advantages. It provides a central view of the mediastinum, allowing simultaneous visualization of both pleural cavities and phrenic nerves, hence, least probability of leaving behind any thymic tissue.[11] Additionally, this approach avoids intercostal incisions, thereby reducing postoperative pain and intercostal nerve injury.[12]

The anesthetic management of patients with MG undergoing thymectomy is challenging because of their increased sensitivity to neuromuscular blocking agents and the risk of postoperative respiratory failure. Conventional thoracic surgery often requires tracheal intubation and one-lung ventilation, which can increase postoperative complications.[1]

Non-intubated thoracic surgery has emerged as an alternative approach. This technique eliminates the need for neuromuscular blocking agents and reduces airway trauma associated with intubation. Studies have shown that non-intubated thymectomy may reduce postoperative myasthenic crisis and shorten hospital stay.[8] Furthermore, avoidance of intubation may decrease complications such as postoperative dysphagia, sore throat and tracheal injury.

Another important aspect of thymectomy in MG is the long-term reduction in medication requirements. Previous studies have demonstrated significant reductions in pyridostigmine and immunosuppressive drug doses following successful

thymectomy, with some reports showing more than 70% reduction within two years.[9] In our case, early postoperative improvement was observed with a reduction in pyridostigmine dosage with complete withdrawal of drug in 6 months period. Although long-term follow-up is required to evaluate sustained remission, this early response is encouraging.

To our knowledge we performed the first nonintubated uniportal subxiphoid VATS thymectomy in Nepal, which was successfully done without intraoperative complications or conversion to open surgery. This case also highlights the feasibility of performing advanced minimally invasive thoracic surgery in resource limited settings such as Nepal. However, careful patient selection is essential for non-intubated thoracic surgery. Patients with severe obesity, significant pulmonary disease or anticipated difficult airway may not be suitable candidates. Additionally, close collaboration between surgeons, anesthesiologists, and neurologists is critical for optimal perioperative management.

Conclusion

Non-intubated uniportal subxiphoid VATS thymectomy is a safe and effective minimally invasive technique for the surgical management of myasthenia gravis. This approach offers several advantages, including reduced surgical trauma, avoidance of neuromuscular blocking agents and potentially faster postoperative recovery.

Our case demonstrates that this advanced surgical technique can be successfully performed in a tertiary care center in Nepal, highlighting its feasibility and potential benefits for selected MG patients. Further studies with larger patient cohorts and long-term follow-up are required to validate the advantages of this approach.

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