

Subxiphoid thoracoscopic thymectomy in a young female with seropositive Myasthenia Gravis: A case report

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ABSTRACT

Case: Myasthenia Gravis (MG) is an autoimmune disease that occurs at the neuromuscular junction and characterized by weakness of voluntary skeletal muscles which may be generalized or localized with an incidence of 4.1 to 30 cases per million person-years. MG is diagnosed clinically and by measuring the level of acetylcholine receptor antibodies, Muscle specific kinase antibodies and Low-density lipoprotein receptor related protein-4 antibodies. Patient is managed conservatively with cholinesterase inhibitor (pyridostigmine), prednisone, Mycophenolate mofetil and surgically by thymectomy. Here we present a case of a 27-year female who was known case of seropositive Myasthenia gravis for five years who underwent a novel technique of subxiphoid thymectomy.

Keywords: Myasthenia gravis, Thymectomy, Autoimmune disease

Introduction:

Myasthenia Gravis (MG) is an autoimmune disease that occurs at the neuromuscular junction and characterized by weakness of voluntary skeletal muscles which may be generalized or localized.¹The incidence of the disease is 4.1 to 30 cases per million person-years, and the prevalence rate ranges from 150 to 200 cases per million.² Recent epidemiological data indicates that 10%–15% of people with MG also have thymoma, and 20%–25% of thymomas are complicated with MG.³

The use of thymectomy with median sternotomy has gradually been supplanted by minimally invasive procedures that offer comparable efficacy, less trauma, and fewer complications. These procedures include subxiphoid approach, robotic-assisted (RATS), and standard video-assisted thoracoscopic surgery (VATS; unilateral or bilateral).⁴

Kido originally described the subxiphoid thymectomy in 1999, and since then, it has been effectively used to treat MG.⁴ We discuss a case of Myasthenia Gravis in a 27-year female who had undergone sub xiphoid

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thymectomy in B.P. Koirala Memorial cancer hospital (BPKMCH).

Case Presentation

A 27-year female presented to the outpatient clinic of Department of Thoracic surgery, BPKMCH with chief complaints of difficulty in swallowing and ocular muscle weakness for 5 years. She was diagnosed as seropositive (AChR >5 nmol/IL) and MuSK negative myasthenia gravis with Osserman severity index of Stage IIIb⁵ and hypothyroidism for 2 years. She had been taking acetylcholinesterase inhibitor (Pyridostigmine) 60 mg 4 times a day with Mycophenolate Mofetil 250 mg twice daily and Thyroxine 75mcg once daily for last 5 years. The symptoms were well controlled with the above medications. But due to the need for life long medications patient was seeking for an alternative option. Her Contrast enhanced computed tomography chest (CECT), revealed normal size (11mm) thymus gland, and density of Hounsfield (HU)+40 with smooth outline (Fig. 1). AChR was strongly positive (5.27nmol/L) while Anti-MuSK antibodies was negative (<0.18).

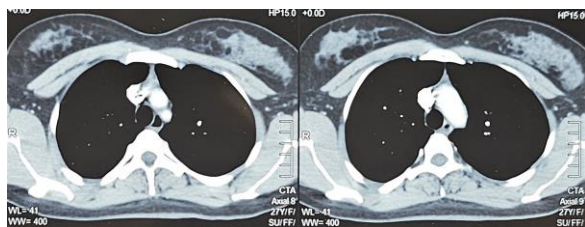


Fig:1 CECT Chest

We planned for surgical excision of thymus and patient underwent subxiphoid thymectomy.

Surgical Technique

After induction of general anesthesia patient was intubated with single lumen endotracheal tube in supine position with legs apart. The surgeon stood between the patient's legs while the camera surgeon stood on the right side of the patient (Fig. 2).

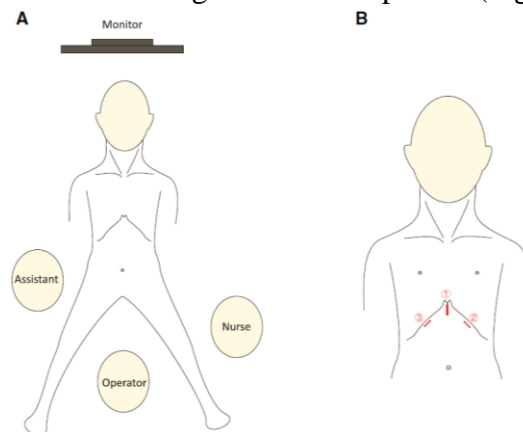


Figure 2. 1: 10mm sub xiphoid incision for camera and CO₂; 2,3: two 5mm working port for graspers and energy device

A midline retrosternal tunnel was created by blunt finger dissection, and the pleura mediastinal was pushed to both sides at the costal arch. Two 5mm working port were created at the intersection point of bilateral midclavicular line and subcostal margin. A 10-mm 30-degree angled thoracoscope and capnotherax of 6mm Hg pressure with 5-6L/min CO₂ were used during the operation. The right pleural cavity was opened, the substernal space was examined, and the fat pad in the pericardiophrenic sinus was separated from the pericardium. In the similar manner, left pleura is opened and left pleural cavity is entered. Thymic tissue along with the fat pad is mobilized from the

level of both diaphragms to the level of thyrothymic ligament. The lateral margins of the dissection were the phrenic nerves in both sides. During the course of dissection, the thymic tissue was carefully lifted away from the SVC, ascending aorta and bilateral brachiocephalic veins. Thymic veins draining into left brachiocephalic vein were ligated and divided. Thymus (Fig. 3) was placed in a plastic bag and removed from the mediastinum via the sub xiphoid port. A right sided underwater seal chest drain was inserted.



Figure 3. En-bloc specimen of Thymus gland. A, B, C, D; right superior, left superior, left inferior, right inferior horn

Postoperative care and evaluation

Patient was managed in post operative ward with intravenous antibiotics and analgesic, shifted to ward on 1st post operative day.

Chest drain was removed on 3rd post operative day and patient was discharged on 5th day of surgery in hemodynamically stable condition. The same medications used preoperatively were postoperatively restarted to control MG. During discharge the dose of Pyridostigmine was reduced to 60mg twice a day.

Follow up

Patient visited to our outpatient unit 1 month after surgery without any complaints. Final Histology showed normal thymic gland. The doses of the medications have been further reduced and we are planning to stop the medications completely in subsequent 2-3 months duration.

Discussion

Here, we describe thymectomy using minimally invasive sub xiphoid approach. In addition to the added benefit of angles and flexibility, camera hopping allows us to visualize phrenic nerves with exceptional clarity. To the best of our knowledge, this is the first reported case of subxiphoid thymectomy from Nepal. China, Japan, and Poland have produced the majority of the previous literature on minimally invasive subxiphoid thymectomy.⁶ The gold standard for treating anterior mediastinal tumors and myasthenia has been median sternotomy, which was initially described by Blalock et al. in 1939.⁷ With the advancement of video-assisted surgery and endoscopic instrument VATS thymectomy can be performed by lateral intercoastal approach but surgeons have challenges while attempting a full dissection of the mediastinal adipose tissue and locating the contralateral phrenic nerve. Francesco Paolo Caronia has described the

uniportal bilateral VATS thymectomy but it was associated with significant post operative pain and intercostal nerve damage.⁸ In 2002, Hsu et al. described their first experience of sub xiphoid thymectomy for MG, since then it has gained its popularity due to excellent visualization of both phrenic nerves, less post operative pain and early post operative recovery.⁹ There has been two studies done in Nepal regarding VATS thymectomy for MG. In a study done by Thapa et.al in 2014, 12 patients underwent VATS thymectomy while 6 patients underwent thymectomy via sternotomy for MG, they found that overall hospital stay was shorter in VATS group as compared to sternotomy group (7.8±4.2 vs 4.6±1.8).¹⁰ Similarly in 2021, another study from Manmohan cardiothoracic vascular and transplant center by Khadka et.al reported that 25 patients who underwent VATS thymectomy has lesser hospital stay as compared to thymectomy via sternotomy (5.3±2.9 vs 7.0±3.8).¹¹

Conclusion

To summarize, to the best of our knowledge this is the first case reported in Nepal. Sub xiphoid thymectomy is a cutting-edge, minimally invasive technique that reduces postoperative discomfort, prevents intercostal neuropathy after surgery and removes precisely entire thymic tissue.

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