Case Series

Radical thymectomy in Myasthenia Gravis through partial sternotomy approach: A report on three patients

Venkatesa Kumar Anakaputhur Rajan[1*], Anton Prem Thilak[1], Chandrasekar Purushothaman[2] Arun Kumar Govindarajan[2], Siva kumaran Subbarayan[2], Harikrishnan Subramanian[2], Raghuram Arani Raghavendrarao[1]

[1]*Department of Cardiothoracic Surgery, Kauvery Hospital, Alwarpet, Chennai*

[2]*Department of Cardiac Anaesthesia, Kauvery Hospital, Alwarpet, Chennai*

*Corresponding Author: **M:** +919566823267; **Email**: dr.venkatesakumar@gmail.com

Abstract

Myasthenia Gravis is one of the most common neuromuscular junction disorders caused by autoantibodies against acetylcholine receptors on the postsynaptic membrane. This leads to progressive weakness of muscles. Thymic hyperplasia is associated with myasthenia in 7 out of 10 patients, while thymoma is seen in 1 out of 10 patients. Several studies have published the advantage of performing a radical thymectomy in patients with thymic hyperplasia or thymoma to induce remission in patients with myasthenia gravis. The transsternal approach is the standard approach for performing radical thymectomy. Recently, minimally invasive approaches have been increasingly used to reduce the morbidity of performing a complete sternotomy. Robotic-assisted, video-assisted, and partial sternotomy approaches are the available options. In our study, we are explaining the technique and advantages of performing radical thymectomy through a partial sternotomy approach.

Keywords: Radical thymectomy; Thymoma; Thymic hyperplasia; Myasthenia gravis; Partial sternotomy

Background

Myasthenia Gravis (MG) is one of the common neuromuscular junction disorders caused by autoantibodies against acetylcholine receptors on the postsynaptic membrane, leading to progressive skeletal muscle weakness.

Thymic hyperplasia is associated with myasthenia in 7 out of 10 patients, while thymoma is seen in 1 out of 10 patients[1]. Thymectomy is a well-known procedure done as a part of surgical management in MG. Herein, we present a case of myasthenia gravis with thymic hyperplasia managed with radical thymectomy through a partial sternotomy approach.

Case Presentation

A 57-year-old gentleman (patient 1), without known comorbidities, developed slurring of speech and difficulty swallowing, which were insidious in onset and progressive. Later, he started to experience difficulty breathing at rest, for which he was admitted to the emergency department elsewhere and was diagnosed with myasthenia gravis. He was started on oral pyridostigmine and prednisolone tablets. He recovered from the symptoms and came to our institution for further management. His CT chest showed thymic hyperplasia without any evidence of thymoma. He was referred to us for a thymectomy. Considering the delay in wound healing in patients with high doses of steroids, surgery was postponed until the steroid was reduced to a considerably low dose. Routine preoperative workup was done, and he was planned for radical thymectomy. He underwent radical thymectomy through an upper partial sternotomy approach.

Surgical Technique

Skin was incised to about 7 cm (Fig. 1). After dissecting the suprasternal notch, a sternotomy was performed with an oscillating saw until the 4th intercostal space. A transverse sternotomy was performed in the 4th intercostal space in an inverted T shape. Even though exposure of the thymus was adequate with a partial sternotomy until the 3rd ICS, we extended it until the 4th ICS for better exposure of the inferior horns of the thymus, extending till the diaphragm. A radical thymectomy was performed by removing all fibrofatty tissue from the right to the left phrenic and between the innominate vein and diaphragm, including the cervical horns. The thymectomy specimen is shown in Fig. 2.

Fig. (1). Partial sternotomy scar taken after discharge during follow up. Skin incision measured as 7 cm (Fig. 1B).

Fig. (2). Radical thymectomy specimen-bilobed thymus with superior/cervical (marked with yellow arrow) and inferior horn (marked with red arrow).

Hemostasis was performed, and the sternum was closed with five wires. In this technique, the sternum is divided into three portions, and the stability of the sternum is achieved by crisscross wiring, as shown in Fig. 3. After the surgery, he was extubated within one hour after shifting to the ICU. After removing bilateral pleural drains, he was shifted to the ward on the first postoperative day. He got discharged on postoperative day 3. Histopathology reports confirmed the presence of thymic hyperplasia with no evidence of thymoma.

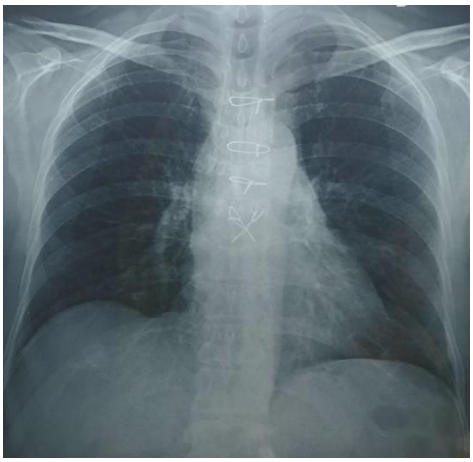

Fig (3). Chest X ray PA view showing 5 wires with last cross wiring technique.

We have operated on three patients so far through a partial sternotomy approach. The details and clinical outcomes have been mentioned in Table 1.

Table 1. Clinical details of patients who underwent partial sternotomy radical thymectomy in our institution.

Patient No.	Age/Sex	Diagnosis/Subtype	Surgery	Postop ventilation time (h)	ICU stay (d)	Hospital stay (d)
1	57/M	Generalised MG	Radical thymectomy	1	1	3
2	55/M	Ocular MG + AChR +ve	Radical thymectomy + Thymoma excision	2	1	4
3	47/M	Ocular MG + AChR +ve	Radical thymectomy + Thymoma excision	0 (On table extubation)	1	5

Discussion

Myasthenia gravis is an autoimmune disorder affecting the neuromuscular junction that leads to progressive weakness of skeletal muscles. It usually affects the muscles of the eye, throat, and extremities. Involvement of respiratory muscles can lead to distress and might require emergency intervention [2].

Classification of Myasthenia Gravis

Depending on the clinical features and the type of antibodies involved, MG can be classified into various subgroups. Each group responds differently to treatment and hence carries a prognostic value [3]

(1) Early-onset MG: age at onset less than 50 years with thymic hyperplasia
(2) Late-onset MG: age at onset greater than 50 years with thymic atrophy
(3) Thymoma-associated MG
(4) MG with anti-MuSK antibodies
(5) Ocular MG: Symptoms only from periocular muscles
(6) MG with no detectable AChR or MuSK antibodies

The anti-AChR Ab test is very specific, and it confirms the diagnosis in patients with classical clinical findings. It is present in four-fifths of patients with generalized MG and only in half of the patients with pure ocular MG. The rest of the patients, about 5 to 10%, would demonstrate anti-MuSK antibodies [2]. In our patient, the symptoms were insidious in onset and progressive, with the involvement of respiratory muscles causing distress. This belongs to the generalized MG subtype with AChR antibody positivity. Thymectomy is known to be beneficial in these groups of patients.

Treatment Options

Myasthenia gravis can be medically managed using anticholinesterase inhibitors (pyridostigmine), corticosteroids (prednisolone), other immunosuppressants (azathioprine, mycophenolate mofetil, cyclosporine, methotrexate, cyclophosphamide, rituximab), and rapid-acting immunotherapies like plasma exchange, intravenous immunoglobulins, and eculizumab [3,4]. The only surgical option available in these cases is radical thymectomy with or without thymoma excision.

Several studies have proven the definitive advantage of performing radical thymectomy with or without thymoma excision in patients with myasthenia gravis. All the subtypes of MG have shown clinical improvement

and decreased incidence of recurrence after thymectomy, except for the non-thymomatous MG type associated with anti-musk ab positive status. [1]

Thymectomy for MG can be performed via the following surgical approaches:

(1) Transsternal approach: this is the standard approach for radical thymectomy, which involves a complete sternotomy. The advantages include excellent exposure, mainly indicated if there is a large tumour, and in cases of tumour invasion to major vascular structures, such as the pericardium or heart. But the disadvantages include a big scar, pain, and slow recovery after sternotomy and wound healing problems, since all these patients are managed with high doses of steroids,

(2) Transcervical approaches and combined approaches are not used these days.

(3) Partial sternotomy approach: Fernandes et al. [5] studied the outcomes of performing thymectomy through a partial sternotomy approach for MG. Partial sternotomy was performed until the 4th ICS, and transverse extension (inverted T) was not performed in this series. There were no complications related to surgery like phrenic nerve palsy, major blood loss, prolonged ventilation, or pleural effusion requiring ICD drainage for a long duration in this study. In our patients, an inverted T-shaped partial sternotomy was performed that gave better exposure to the structures in the mediastinum. But meticulous sternal wiring techniques are required to provide adequate stabilization of the sternum.

(4) Minimally invasive thoracoscopic approach: video-assisted thoracoscopic surgery (VATS) or robotic-assisted thoracoscopic surgery (RATS) Both approaches involve the use of a thoracoscope and have gained a lot of attention in the last decade due to less pain, faster postoperative recovery, early mobilization, and less blood loss. But disadvantages include intercostal neuralgia, incomplete

resection, injury to structures causing conversion to sternotomy, and phrenic nerve injury.

Rowse et al. conducted a study comparing the VATS versus RATS approach for thymectomy [6]. The mean operative time and blood loss were significantly less in the VATS group. But there was no difference in hospital stay or ICU stay between the groups. The mean hospital stay was 1.5 days (range 1-4) in the VATS group, while the RATS group had a mean stay of 2 days (range 1–5 days). They have also reported a few postoperative complications, like phrenic nerve palsy, pericarditis, atrial fibrillation, and pleural effusion, in a few patients.

The hospital stay is definitely shorter compared to patients undergoing partial sternotomies. The incidence of complications related to surgery is negligible with a partial sternotomy approach [5]. VATS and RATS approaches require expensive instruments and a steep learning curve, while partial sternotomy provides adequate exposure, is cost-effective, and is easy to perform with a lower incidence of surgery-related complications.

In our institution, patients diagnosed with myasthenia gravis are treated medically. Once the symptoms are well controlled and the steroid dose is reduced considerably, surgery is offered. If patients have symptoms of myasthenia, and if surgery is performed at that stage, they are at high risk for prolonged ventilation and can develop myasthenic crisis. So, medical control of symptoms will provide good surgical outcomes. Radical thymectomy with or without thymoma excision through a partial sternotomy approach is offered to these patients. We believe that this approach is a cost-effective alternative to VATS or RATS in terms of fewer procedure-related complications and a complete excision of the thymus.

Conclusion

Myasthenia gravis is a serious neurological condition that needs optimal treatment to prevent dreadful complications. Surgical management provides clinical improvement and reduces the incidence of remission. Minimally invasive approaches have proven to be superior in terms of less pain and faster recovery. The partial

sternotomy approach is a cost-effective alternative among minimally invasive approaches. A prospective study comparing the clinical outcomes with follow-up of the remission-free period between the partial sternotomy and VATS/RATS approaches would shed light on this matter.

Authors Contributions

Venkatesa kumar Anakaputhur Rajan and Arani Raghavendrarao Raghuram wrote the first draft of the article. All other co-authors helped in revising and have approved the final draft of the case study.

Conflict of interest

None declared

References

[1] Aljaafari D, et al. Thymectomy in Myasthenia Gravis: A narrative review. Saudi J Med Med Sci. 2022;10(2):97–104.

[2] Beloor Suresh A, Asuncion RMD. Myasthenia Gravis. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 [cited 2023 Aug 17]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK559331/

[3] Gilhus NE, et al. Myasthenia gravis: a review of available treatment approaches. Autoimmune Dis. 2011;2011:847393.

[4] Farmakidis C, et al. Treatment of Myasthenia Gravis. Neurol Clin. 2018;36(2):311–37.

[5] Pêgo-Fernandes PM, et al. Thymectomy by partial sternotomy for the treatment of myasthenia gravis. Ann Thorac Surg. 2002;74(1):204–8.

[6] Rowse PG, et al. Minimally invasive thymectomy: the Mayo Clinic experience. Ann Cardiothorac Surg. 2015;4(6):519–26.