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AN INTERESTING CASE OF THYMOMA ASSOCIATED MYASTHENIA GRAVIS TREATED WITH BILATERAL VATS RADICAL THYMECTOMY

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Myasthenia gravis, thymoma, VATS, radical thymectomy.

Abstract

A 59 years lady presented with complaints of drooping of eyelids, double vision and fatigability in hands. Neurological evaluation confirmed myasthenia gravis. She was found positive for anti-acetylcholine receptor antibody (anti-AChRAb). Imaging study detected a thymic tumour. CT guided FNAC from the lesion in the anterior mediastinum confirmed it to be a thymoma. She was managed by bilateral VATS (Video Assisted Thoracoscopic Surgery) radical thymectomy & anti-cholinesterase and steroid medications. She is currently 20 months following thymectomy. She is clinically asymptomatic with pyridostigmine 30 mg a day qualifying for MGFA (Myasthenia Gravis Foundation of America) post-intervention status minimal manifestation (MM-2). Hence, an early and radical treatment by minimally invasive technique may have good outcome in patients with thymomatous myasthenia gravis.

Introduction

Myasthenia Gravis (MG) is an autoimmune disorder resulting in easy fatigability of voluntary muscles due to the production of anti-acetylcholine receptor antibody (anti-AChRAb) and reduction of available nicotinic acetylcholine receptors (AChR) at neuromuscular junction. The muscle weakness can occur in the extraocular, bulbar, neck, limb girdles, distal limbs and trunk muscles [1]. Of all patients with MG, about 10-15% have thymoma. The diagnosis is made based on history, challenge test (neostigmine or edrophonium test), repetitive nerve stimulation tests and autoimmune antibodies (anti-AChR or anti-Musk antibodies).

Thymoma is graded according to the World Health Organization (WHO) histological classification and is classified by the Masaoka staging system [2,3]. The severity of MG is evaluated according to MGFA (Myasthenia Gravis Foundation of America) clinical classification system [4].

MG is sometimes identified as having ocular and generalized form, although one is not exclusive of the other and the ocular form is considered an initial, milder form of illness that progresses to the more severe form in most but not all patients [5].

Case Report

A 59 years lady who is a known hypertensive presented to the clinic with complaints of drooping of eyelids, double vision and fatigability in hands for three months. She was evaluated by neurologist and diagnosed clinically as myasthenia gravis, MGFA class IIa. Repetitive nerve stimulation study revealed decrement in response of bilateral

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orbicularis oculi and trapezius muscles. She was found positive for anti-AChR antibodies. A contrast enhanced computed tomography (CECT) of the chest showed a well-defined lesion of size 34 x 28 mm in the anterior mediastinum at the level of aortic root with mild heterogeneous contrast enhancement [Figure 1a]. CT guided FNAC from the lesion confirmed it to be a thymoma. The patient was optimized for surgery with medications to control myasthenic symptoms. Her symptoms were controlled with pyridostigmine 60 mg three times and prednisolone 15 mg a day.

VATS radical thymectomy was performed using bilateral thoracoscopic approach under elective single lung ventilation using a left sided double lumen endotracheal tube. Patient position on the operation table was supine with head end elevated to thirty degree (low fowler's position). Mechanical compression device was applied for DVT (Deep Venous Thrombosis) prophylaxis in both lower limbs. Upper limbs were supported to avoid injury to the brachial plexus. One 10 mm port was used for introduction of thoracoscope at anterior axillary line at sixth intercostal space. Two additional 5 mm ports were used for introduction of thoracoscopic instruments at 3rd intercostal space along anterior axillary line and 6th intercostal space along midclavicular line. The port position was similar on both sides of the chest [Figure 2]. Insufflation of CO_2 into pleural cavity was employed to improve visualization of the operating field. The major part of the operative procedure to excise the thymus gland was done from left side of the chest. Dissection of all the perithymic fatty tissue with the thymus gland was carried out from neck to diaphragm and from one phrenic nerve to the other, preserving both phrenic nerves. Thymoma of size 3 x 4 cm was seen arising from the right lobe of thymus gland [Figure 1b]. The excised thymus gland specimen was retrieved in an endobag and delivered by enlarging a right sided 5 mm port. A single 24 Fr chest tube was kept in right pleural cavity which was removed on day 1 following the surgery. Oral diet was resumed on the same day of surgery and she was ambulant on postoperative day 1. She was discharged from the hospital on second postoperative day with the advice to continue medications advised by the neurologist. The histopathology report showed WHO type AB thymoma with no capsular or vascular invasion. The thymoma was classified as Masaoka stage I.
She is currently 20 months following thymectomy and doing well on regular follow-up visits to our clinic. Steroid was stopped by gradually tapering the dose. She is clinically asymptomatic with pyridostigmine 30 mg a day qualifying for MGFA post intervention status minimal manifestations (MM-2).

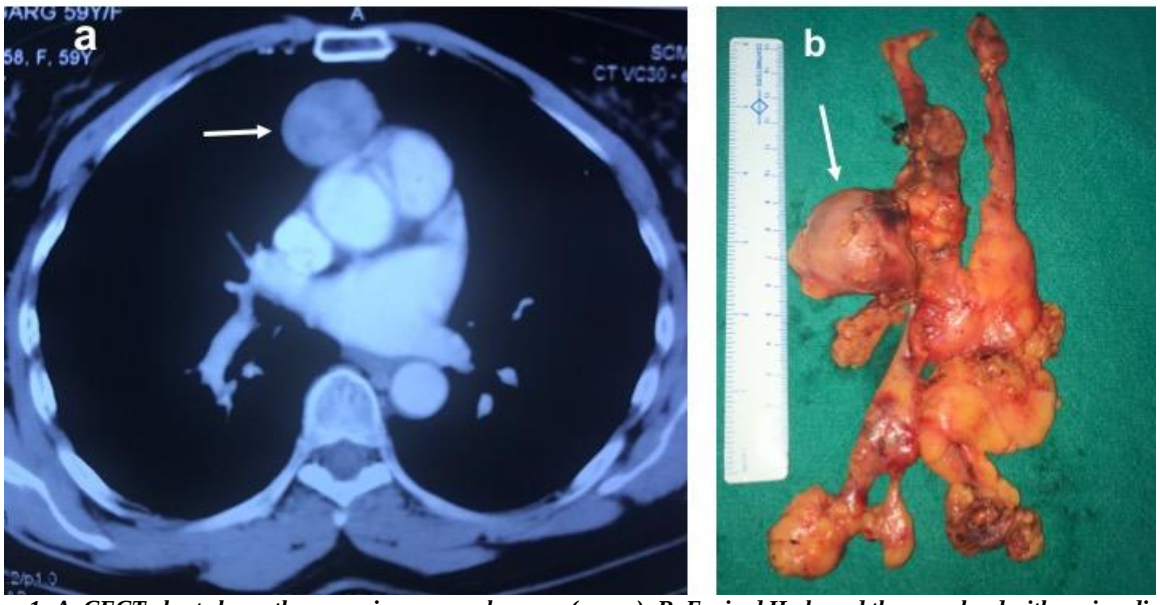

Figure 1: A. CECT chest shows thymoma in prevascular area (arrow). B. Excised H-shaped thymus gland with pericardial fat pad with thymoma (arrow) arising from the right lobe of

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thymus gland.

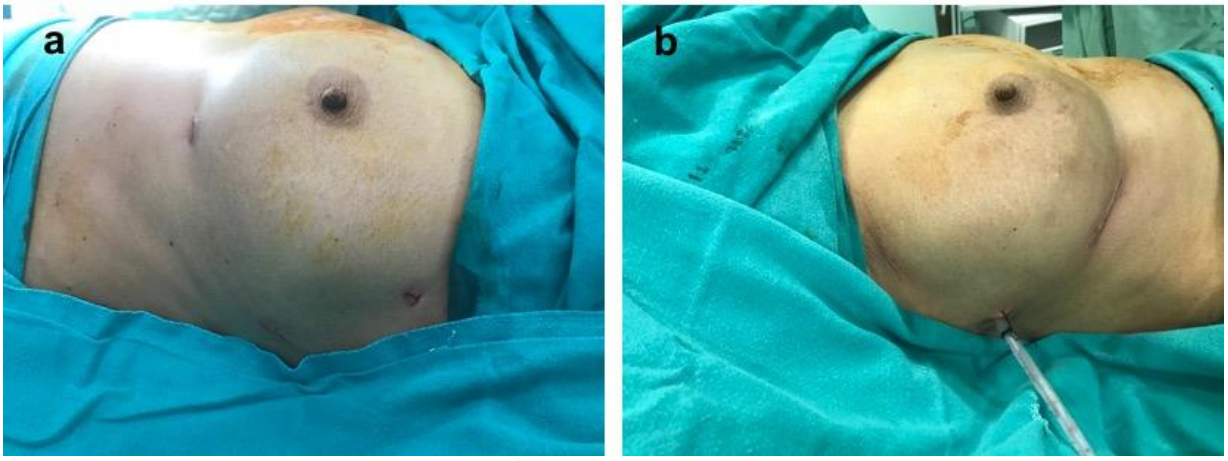

Figure 2: Port sites on left side (Figure 2a) and right side (Figure 2b).

Discussion

Thymoma-associated MG (T-MG) is identified as a tumor originating from thymic epithelial cells; most being cortical subtype (World Health Organization Type B) [2]. Because cortical thymoma usually has some morphological similarities with the thymic cortex, they share the ability to propagate the maturation of immature naive CD4 T cells and spread mature naive T cells into the periphery. The pathogenesis of T-MG suggests that epithelial neoplastic cells encircled by maturing T cells can express epitopes cross-reactive with skeletal muscle proteins, such as acetylcholine receptors [6].

MG was previously considered an adverse prognostic factor in case of thymoma because of increasing risk for surgery and a higher perioperative mortality [7]. On the contrary, in recent series, myasthenic symptoms are a favorable prognostic factor in case of thymoma [1,3,8]. The change is a consequence of several factors: the improvements of the medical therapy of MG, an earlier diagnosis with less advanced stage of thymoma and the hypothetical beneficial effect of steroids, widely used for the treatment of both diseases [9].

Neurological outcome may reflect a favorable oncological outcome and both depend on the radicality of thymectomy. Patients with the association of MG and thymoma who achieve a complete remission after the thymectomy may be considered at good prognosis, while patients who maintain some symptoms or require some medications should undergo a stricter radiological follow-up [10].

Our technique of doing radical thymectomy is always a bilateral thoracoscopic approach. The reason for adopting this technique is optimal visualization of both phrenic nerves and pericardial fatty tissue. We believe that, visualization of nerves and fatty tissue in bilateral thoracoscopy provides good safety margin for the operative procedure.

Conclusion

There is a known and established relationship between thymoma and myasthenia gravis. Although thymomatous myasthenia gravis tends to have a difficult clinical course and poor prognosis, early and aggressive treatment by a multidisciplinary (thoracic surgeon and neurologist) team may improve the outcome of these patients.

Conflict of Interest: The authors declare that they have no conflict of interest.

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