

Multiple Thymoma with Myasthenia Gravis

Dong Hyun Seo, M.D.¹, Sukki Cho, M.D., Ph.D.^{1,2}

¹Department of Thoracic and Cardiovascular Surgery, Seoul National University Bundang Hospital,

²Department of Thoracic and Cardiovascular Surgery, Seoul National University College of Medicine

The actual incidence of multiple thymoma is unknown and rarely reported because it remains controversial whether the cases represent a disease of multicentric origin or a disease resulting from intrathymic metastasis. In this case, a patient underwent total thymectomy for multiple thymoma with myasthenia gravis via bilateral video-assisted thoracic surgery. A well-encapsulated multinodular cystic mass, measuring 57 mm×50 mm×22 mm in the right lobe of the thymus, and a well-encapsulated mass, measuring 32 mm×15 mm×14 mm in the left lobe, were found. Both tumors were type B2 thymoma. Few cases of multiple thymoma with myasthenia gravis have ever been reported in the literature. We report a case of synchronous multiple thymoma associated with myasthenia gravis.

Key words: 1. Thymoma
2. Thymectomy
3. Myasthenia gravis
4. Video-assisted thoracic surgery (VATS)
5. Thymus

Case report

A 47-year-old man, presenting with ptosis, visited the department of neurology at another hospital and was diagnosed with myasthenia gravis (MG) via a positive Jolly test and a positive Tensilon test. His level of anti-acetylcholine receptor antibodies was 2.0 nmol/L (normal <0.3 nmol/L). He was treated with 60 mg of pyridostigmine 3 times a day. A computed tomography (CT) scan of the chest showed 2 tumors present in the anterior mediastinum: a solid and cystic mass in the right anterior mediastinum and a solid nodular lesion in the left anterior mediastinum (Fig. 1). The anterior mediastinal masses were diagnosed as thymoma with MG. The patient was then transferred to department of thoracic and cardiovascular surgery of Seoul National University

Bundang Hospital for surgical treatment, having been diagnosed with ocular MG with multiple thymoma.

Under general anesthesia, a left-sided double-lumen endotracheal tube was positioned for selective lung ventilation. A thymectomy was performed via bilateral video-assisted thoracic surgery (VATS). First, the patient was placed in the left-semilateral decubitus position, and 3 ports were placed. The thoracoscopic port was placed along the anterior axillary line, at the fifth intercostal space. The second and third instrument ports, 5 mm each, were inserted under thoracoscopic vision, at the third intercostal space and distal clavicular line, and fifth intercostal space and distal clavicular line, respectively. The huge tumors were adherent to the pericardium, without invasion. Thymectomy was performed with a Harmonic scalpel (Ethicon Endo-Surgery Inc., Cincinnati,

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Corresponding author: Sukki Cho, Department of Thoracic and Cardiovascular Surgery, Seoul National University Bundang Hospital, 82 Gumi-ro 173beon-gil, Bundang-gu, Seongnam 13620, Korea
(Tel) 82-31-787-7132 (Fax) 82-31-787-4050 (E-mail) skcho@snuh.org

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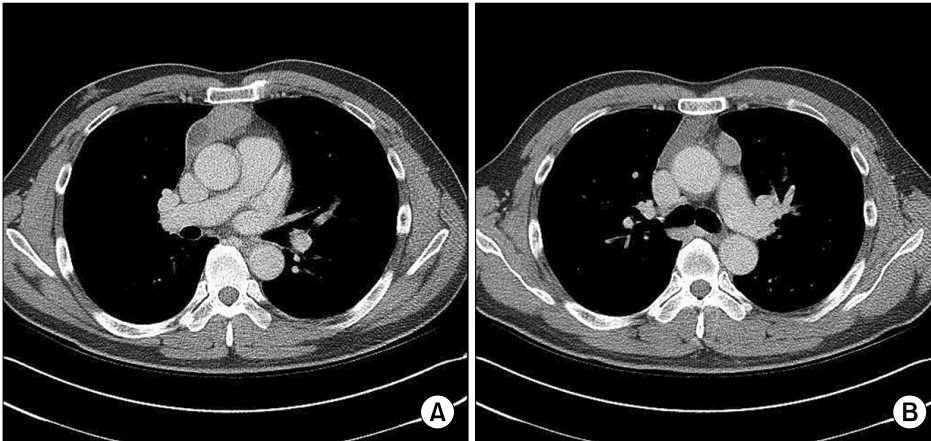


Fig. 1. Preoperative chest computed tomography. (A) A mass with mixed solid and cystic portions was found in the right anterior mediastinum. (B) A solid mass was found in the left mediastinum.

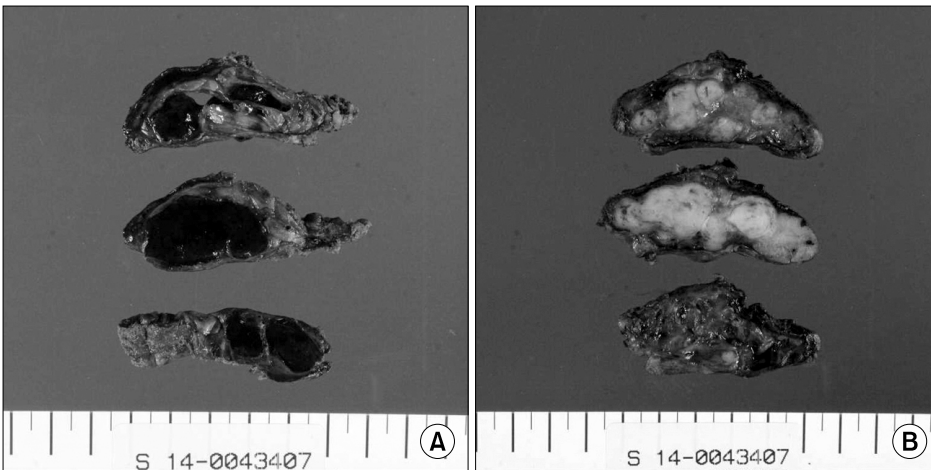


Fig. 2. Pathologic findings. (A) A well-encapsulated multinodular cystic mass, whitish-grey in color, measuring 57 mm×50 mm×22 mm, was found in the right lobe of the thymus. (B) A well-encapsulated mass, measuring 32 mm×15 mm×14 mm, was found in the left lobe of the thymus.

OH, USA). From the diaphragm to the right upper pole, all thymic tissues, including the mediastinal and pericardial fat, were resected. Then, 3 ports were positioned on the left side similarly to those on the right. After completion of the left-side dissection, the entirely resected thymic tumors were placed into an endoscopic retrieval bag for removal. In the right lobe of the thymus, a well-encapsulated multinodular cystic mass, whitish-gray in color, measuring 57 mm×50 mm×22 mm was found (Fig. 2A, right side). In the left lobe of the thymus, a well-encapsulated mass, measuring 32 mm×15 mm×14 mm was found (Fig. 2B, left side). A pathological diagnosis confirmed type B2 thymoma for both resected tumors. The patient was discharged without any postoperative complications. At 9 months after operation, the symptoms of his ocular MG were relieved and no symptoms related to MG were observed. Although no fol-

low-up examination of the level of anti-acetylcholine receptor antibodies was performed, the dosage of pyridostigmine was reduced to 60 mg once a day.

Discussion

Neoplastic thymic epithelial cells consist thymoma, and it contains diverse lymphocytes. For such reasons, thymoma is known to have various characteristics [1]. However, there are rare reports of actual incidence of multiple thymomas because it is still undecided if the cases represent a disease of multi-centric origin or intra-thymic metastasis [2].

Thymus or a thymoma is a factor in the pathogenesis of myasthenia gravis, and thymoma occurs in approximately 10% of MG patients in general [3]. Multiple thymoma is rare, with a reported incidence rate of 1.1%–2.2% [4,5]; furthermore, MG combined

with multiple thymoma is very rare. It remains controversial as to whether cases of multiple thymoma derive from a multicentric origin or result from intrathymic metastasis. Bernatz et al. [4] reported 3 cases of multiple thymoma; it was difficult to determine whether these cases were multicentric thymoma because they presented no findings regarding their histologic characteristics. Nomori et al. [6] reported 1 case of multiple thymoma with MG, and they suggested the possibility of intrathymic metastasis rather than multicentric thymoma by histological, morphometric, and immunohistochemical findings. Several reports suggest that histological, morphometric, and immunohistochemical analyses are useful in characterizing thymomas. In addition, Inoue et al. [7] used micro-dissection or culture of thymoma-epithelial cells to prove the difference of LOH and FISH among histological types of thymoma. However, due to the diversity of lymphocytes in the thymoma-epithelial cells, LOH and FISH have limitations in their examinations of thymoma [2]. Therefore, those examination tools are rarely used to distinguish multiple thymoma in the routine protocol of many centers. The followings characterize multicentric development: (1) there are usually less than three thymomas, (2) the sizes of thymomas are similar, and (3) thymomas are often Masaoka stage I, thus, unable to spread into the thymic tissue [2]. Therefore, our patient was considered to have multicentric thymoma, as he exhibited all 3 aforementioned characteristics.

Nomori et al. [6] recommends an extended thymectomy—a complete thymectomy, which surgically removes all fatty tissue from the right phrenic nerve to the left phrenic nerve and from the diaphragm up to the thyrothymic ligaments. The goal of the procedure is to prevent postoperative tumor recurrence and myasthenic crisis. In our case, an extended thymectomy was performed for MG and to prevent the possibility of intrathymic metastasis and higher frequency of recurrence. Regarding the opti-

mal treatment for multiple thymoma, with or without MG, an extended thymectomy should be performed, because it is difficult to clearly distinguish multicentric thymoma from intrathymic metastasis. Due to the presence of MG and 2 thymomas on chest CT in our patient, we planned to resect the fat tissue around the thymus as much as possible.

In conclusion, we reported a rare case of multiple thymoma accompanied by MG, whereby an extended thymectomy was performed via bilateral VATS. According to the pathologic report, the resected masses were confirmed as multicentric thymoma, rather than intrathymic metastasis.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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