

Thoracoscopic Extended Thymothymectomy for Myasthenia Gravis with Aplastic Anemia

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It is very rare for both myasthenia gravis and aplastic anemia to be complicated with a thymoma. A 74-year-old female was diagnosed to have aplastic anemia with pancytopenia and systemic myasthenia gravis with severe restrictive respiratory dysfunction. Chest CT showed a 5-cm diameter thymoma. After platelets and packed red blood cells were transfused before surgery, an extended thymothymectomy was performed with a bilateral thoracoscopic approach. The thymoma was noninvasive, stage I, and was classified as B1 according to the World Health Organization classification. After the operation, the patient was managed on artificial ventilation with no complications. The myasthenia gravis remitted with the concomitant administration of steroids and immunosuppressants, but the aplastic anemia was not ameliorated. A thoracoscopic thymothymectomy for such a high-risk case of infection and respiratory distress is appropriate surgical procedure, but the complications associated with aplastic anemia are intractable. (Ann Thorac Cardiovasc Surg 2009; 15: 328–331)

Key words: thoracoscopic thymothymectomy, thymoma, myasthenia gravis, aplastic anemia

Introduction

Thymoma is often complicated by autoimmune diseases, but the complication of both myasthenia gravis and aplastic anemia is very rare. In this study, an extended thymothymectomy with a bilateral thoracoscopic approach was performed, and postoperative management was good without the supervention of infection. Thoracoscopic surgery is particularly appropriate for high-risk cases of the thymoma with severe complications, but it is believed that aplastic anemia is intractable.

Case Report

The case was a 74-year-old female. An abnormality was

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identified in a respiratory function test during a detailed examination of aplastic anemia, which was diagnosed to be myasthenia gravis (Myasthenia Gravis Foundation of America [MGFA] classification IVb). The patient was also complicated by diabetes. Chest CT (Fig. 1) showed a well-defined thymoma that was 5 cm in diameter at the right side in the anterior mediastinum. In a blood test, pancytopenia was detected with 1,100/ μ l of white blood cells, 197×10^4 / μ l of red blood cells, 6.3 mg/dl of hemoglobin (Hgb), and 3.6×10^4 / μ l of platelets, and the antiacetylcholine receptor-binding antibodies showed a high value of 149 nmol/L (normally 0.2 nmol/L or less). In the respiratory function test, a severe restrictive disorder was detected with 1,090 ml of vital capacity (%VC: 52.7%) and 800 ml of forced expiratory volume per second (FEV_{1.0}%; 89.9%). In regard to surgery, it was decided that a thymothymectomy with a thoracoscopic approach would be suitable because of the need to avoid the onset of an infection. After platelets and packed red blood cells were transfused immediately before the operation, the anterior chest wall was subcutaneously suspended with Kirschner wire in a supine position to allow thoracoscopic surgery with a bilateral four-port transperitoneal

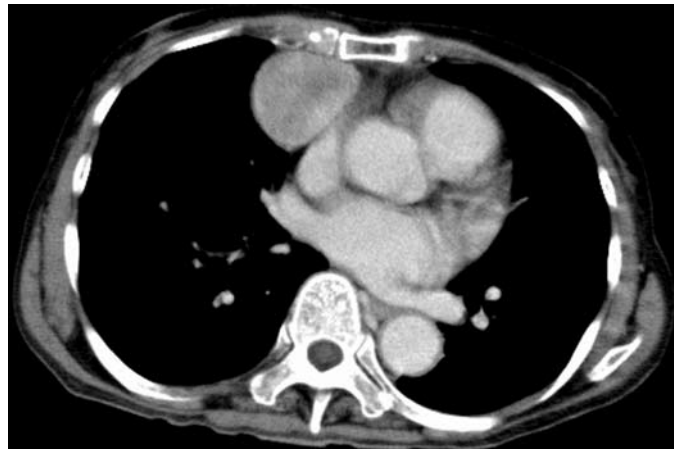


Fig. 1. Chest-enhanced CT revealed a 5-cm tumor in the right anterior mediastinum, and its inside showed nonuniform contrast findings.

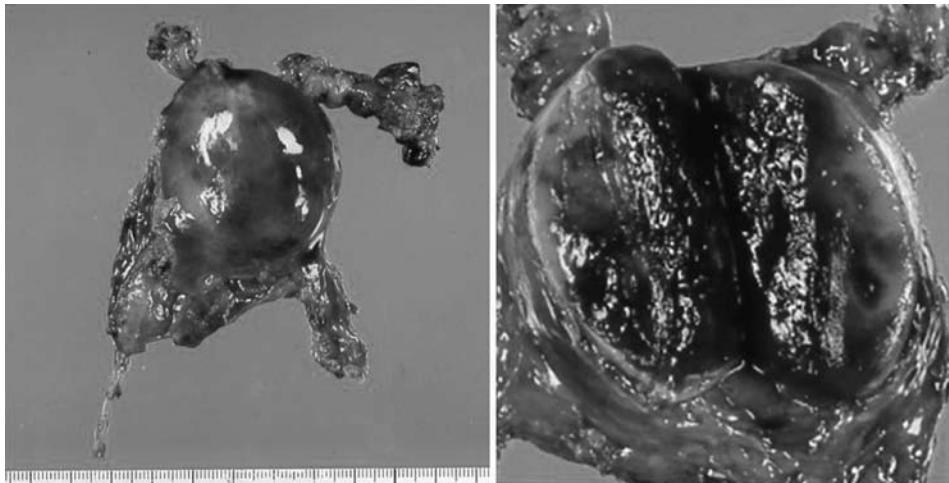


Fig. 2. A resected specimen showed that the thymoma measured 55 × 50 mm and that it was a non-invasive stage I, lymphocytic predominant type, and B1 according to the WHO classification.

approach. Starting from the right side, we sufficiently excised mediastinal fat while maintaining a healthy margin from the thymoma. An excision was made from the left side in the same fashion so that the fat around the left brachiocephalic vein would not be left to complete an extended thymothymectomy with one piece from the right side (Fig. 2). The thymoma measured 55 × 50 mm with no infiltration into surrounding tissue, which was at stage I and stage B1 according to the World Health Organization (WHO) classification. The duration of the operation was 218 minutes, and the amount of bleeding was 50 grams. After the operation, management with artificial ventilation and blood transfusion was performed, and no infection was observed during the course of treatment (Fig. 3). The concomitant therapy of an anti-

acetylcholine esterase drug and a steroid was administered, and 1 year later the myasthenia gravis had improved significantly with 1,680 ml of vital capacity (%VC: 82.4%) and 2.2 nmol/L of antiacetylcholine receptor antibodies. However, in regard to the aplastic anemia, the pancytopenia has not yet ameliorated with 400/ μ l of white blood cells, 215×10^4 / μ l of red blood cells, 6.8 mg/dl of Hgb, and 0.7×10^4 / μ l of platelets.

Discussion

It is well known that thymomas can be complicated with such autoimmune diseases as myasthenia gravis and pure red cell aplasia,¹ but a complication of aplastic anemia is rare.² It is even rarer for thymomas to be complicated

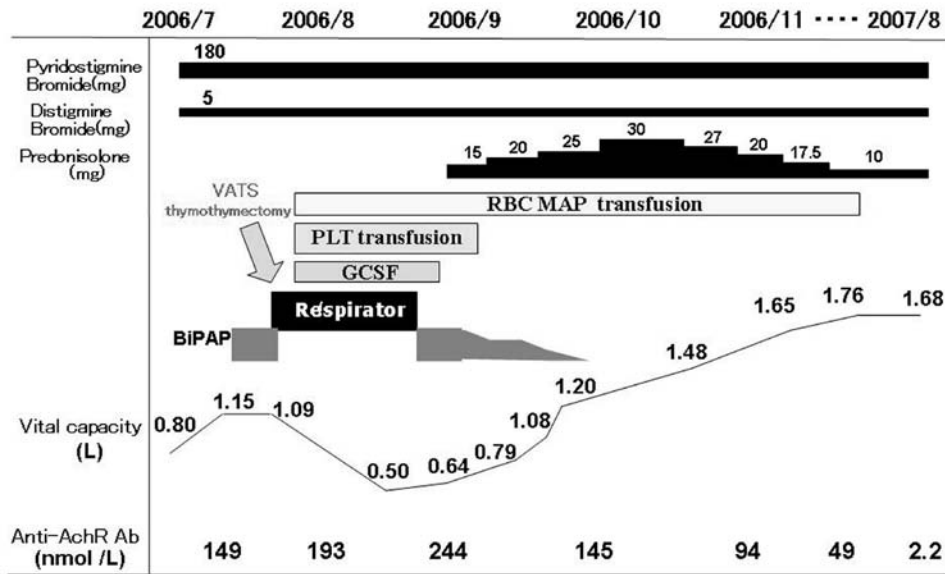


Fig. 3. After the operation, management with artificial ventilation, red blood cell transfusion, platelet transfusion, and the administration of a GCSF preparation was performed, the clinical course was good with no sign of infection. Concomitant therapy with an immunoglobulin preparation, steroids, and immunosuppressants was administered, and the serum antiacetylcholine antibodies decreased while vital capacity also improved.

VATS, video-assisted thoracic surgery; RBC, red blood cell; MAP, mannitol-adenine-phosphate; PLT, practice of platelet; GCSF, granulocyte-colony stimulating factor; BiPAP, biphasic positive airway pressure.

with both myasthenia gravis and aplastic anemia, which has been reported in only a few cases in medical literature after the removal of thymomas.³ It has been reported that the usefulness of a thoracoscopic extended thymothymectomy for myasthenia gravis is equivalent to that of a midsternal approach, which has recently become more widely used.⁴ For cases complicated with thymoma, Cheng et al.⁵ reported this method to be more useful than a thoracotomy in stages I or II thymoma. Our case was a thymoma that measured about 5 cm in diameter, but it was diagnosed to be noninvasive; thus thoracoscopic surgery was performed. Using a bilateral approach, we sufficiently excised fat around the left brachiocephalic vein, and we excised the thymoma in one piece with the mediastinal parietal pleura. Considering the increased susceptibility to infection resulting from pancytopenia and the decreased respiratory function, we believed that performing the operation without a midsternal incision made postoperative management easier. Until now it has been said that the effect of an extended thymothymectomy on pure red cell aplasia is inferior to that on myasthenia gravis,^{6,7} and we suppose that similar results will be

induced in regard to aplastic anemia, which is a blood disorder. As for its reasons, Ritchie et al.³ showed a case in which aplastic anemia developed as a late-stage complication after a thymoma had been cured, and in their report this was caused mainly by the impact of the long-term surviving autoreactive T-cell population produced by the thymoma. In our case, the thymoma shows no recurrence by 1 year after the operation, and the myasthenia gravis has also improved significantly, but the pancytopenia has not yet improved. For aplastic anemia, allogeneic stem cell transplantation,⁸ and drug therapies,⁹ such as anti-lymphocyte globulin, cyclosporine A, and granulocyte-colony-stimulating factors, have all been reported to be effective; however, further study is required. Although the thoracoscopic approach for thymomas with myasthenia gravis, which shows a high risk of infection and is also associated with an advanced respiratory dysfunction, helps to make perioperative management easier, this is considered to be a valuable case indicating that the complication of aplastic anemia may be intractable even after a thymoma has been removed.

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