

Surgical Treatment of an Invasive Thymoma with Intracaval and Intracardiac Extension

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A 53-year-old male with an invasive thymoma extending to the superior vena and right atrium, presenting as superior vena cava syndrome is herein reported. However invasive thymoma with this growth pattern is extremely rare. In this case, the tumor was successfully resected via median sternotomy with cardiopulmonary bypass. After 17 months of follow-up, the patient was still free from any signs and symptoms indicative of superior vena cava syndrome, but recurrent tumor in the right pleura was observed on the follow-up chest computed tomography.

Key Words: Invasive thymoma • Right atrium • Superior vena cava • Surgery

INTRODUCTION

Thymoma is one the most common primary tumors in the mediastinum, accounting for 45% of anterior mediastinal masses in adults.¹ Thymomas are classified as invasive or noninvasive, depending on whether or not there is capsular invasion and/or anatomical extension.^{1,2} Invasive thymomas, comprising approximately 30% of thymomas, commonly infiltrate mediastinal organs such as the pleura, lungs, and pericardium, but rarely invade the superior vena cava.² Invasive thymoma with intravascular invasion of the superior vena cava (SVC) extending to the right atrium (RA) is extremely rare; to our knowledge, only 29 cases have been reported in the literature aside from this report.³ We report here another case of invasive thymoma with this growth pat-

tern, where the tumor was completely resected with the aid of cardiopulmonary bypass technique.

CASE REPORT

In April 2014, a 53-year-old male presented to our hospital with facial and neck swelling and dyspnea of one month's duration. The patient had also loss 4 kg within the 3 months prior to this admission. Seven years earlier (April, 2007), an anterior mediastinal mass measuring 8.9 cm at the longest dimension was incidentally discovered on computed tomography (CT), when the patient was hospitalized for surgery of the cervical spine. At that time, a thymoma was impressed but there was no histopathological diagnosis. Subsequently, he lost to follow-up due to no obvious symptom, and he thereafter self-administered herbal drugs. In November 2009, anterior chest pain was noted and follow-up CT of the chest showed the anterior mediastinal tumor was 4.53 cm at its longest dimension. In May 2010, follow-up chest CT showed the mass was 3.95 cm at the longest dimension. Physical examination showed facial swelling and distended neck veins. However, no abnormality was detected in laboratory examination. Chest CT revealed an anterior mediastinal mass (measuring 8 × 9 cm) invading the brachiocephalic veins and the SVC and ex-

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tending to the RA (Figure 1). Echocardiography confirmed a 3.3×2.7 cm mass in the RA. The diagnosis of an invasive thymoma invading the brachiocephalic veins and SVC and extending into the RA was made. Due to its extensive nature, a decision was made to surgically remove the tumor, without any preoperative chemotherapy or radiation therapy to avoid preoperative neoplastic embolization and to resolve the SVC syndrome.

Tumor resection was performed via median sternotomy under general anesthesia. A firm mass was found in the right anterior mediastinum invading the right brachiocephalic vein and SVC. The tumor encased the right phrenic nerve and infiltrated the right middle lobe of the lung. Another 2×2 cm tumor lesion was located 2 cm inferior to the largest lesion. The two anterior mediastinal tumors were dissected from the surrounding tissue en bloc until direct invasion to the pericardium, the SVC, and the right atrium were encountered. The pericardial cradle was created for adequate exposure, and a generous patch of autologous pericardium was harvested for later vascular reconstruction. Because the lumen of SVC and brachiocephalic vein were occluded by the tumor, the cardiopulmonary bypass was established by cannulating the left brachiocephalic vein, the inferior vena cava, and the ascending aorta. The right brachiocephalic vein was transected before tumor thrombi. The stump of the brachiocephalic vein and lateral wall of the SVC that was directly in contact with tumor was resected. The RA was opened and a tumor thrombus (measuring $7 \times 7 \times 4$ cm) was seen in the RA, without invading any cardiac structures (Figure 2A). The intracardiac portion of the mass was completely removed. The SVC and the right atrial wall defect were repaired with bovine pericardial patch. A pericardial tube

was made from the autologous pericardium and was interposed between the left brachiocephalic vein and the right atrium (Figure 2B). The right phrenic nerve was sacrificed. Thereafter, the patient was uneventfully weaned off cardiopulmonary bypass.

Pathologic examination revealed a type B2 thymoma (World Health Organization classification), which had directly invaded the right brachiocephalic vein, from where the tumor thrombus extended into the SVC and the RA. Postoperatively, the patient was referred to the oncology department for adjunct radiotherapy and chemotherapy. Radiation therapy was delivered to the mediastinum in a dose of 54 Gy/30 fractions within 36 days between August 5 and September 25, 2014. He was initially treated with chemotherapy consisting of cisplatin (40 mg/m^2) for a total of six cycles in the same period as radiation therapy. In addition, adjuvant chemotherapy consisting of combined etoposide (100 mg/m^2) and cisplatin (40 mg/m^2) for a total of four to six cycles between October 22, 2014 and September 2, 2015. After 17 months of follow-up, the patient was free from any signs and symptoms indicative of superior vena cava syndrome, but recurrent tumor in the right pleura was observed on the follow-up chest CT. The recurrent pleural tumor (measuring $8 \times 7 \times 3$ cm) was completely resected by endoscopic technique by a general surgeon. The autologous pericardial conduit was still patent (Figure 2C).

DISCUSSION

Invasive thymoma with intravascular invasion of the SVC extending into the RA is extremely rare. Most pa-



Figure 1. Preoperative chest computed tomography (CT) scans showing a mediastinal tumor invading the superior vena cava and filling the right atrium.

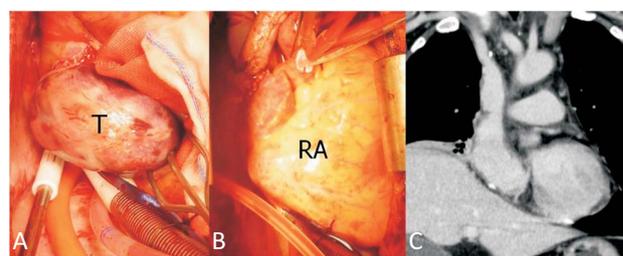


Figure 2. (A) A large tumor (T) measuring $7 \times 7 \times 4$ cm in the right atrium. (B) An autologous pericardial conduit was interposed between the left brachiocephalic vein and the right atrium (RA). (C) Follow-up chest CT scans at 17 months after the operation showing patency of the vascular reconstruction (pericardial tube).

tients with invasive thymoma will present as a localized tumor, in which surgical resection is considered the primary therapy. In cases of advanced invasive thymoma, various therapies, including surgery, radiotherapy, and chemotherapy are being used.⁴⁻⁶ The goal of surgery in such cases is to restore blood flow from the brachiocephalic veins to the RA and to rapidly resolve SVC syndrome. One retrospective study analyzed the survival data in 165 patients with malignant thymoma.⁷ In that study, patients with invasive thymoma survived a shorter period than patients with noninvasive tumors (67% vs. 85% at five years). When the tumor could be radically resected, there was no difference in survival rates between these two groups. Complete resection of tumor is thus a critical component in improving the outcome of these patients. In our case, resection of tumor was considered mandatory due to clinical symptoms and the risk of neoplastic embolization.

In most reported cases of surgical resection of invasive thymoma with intracardiac extension, cardiopulmonary bypass is required as was in our case. Cases in which tumor resection was performed without cardiopulmonary bypass included partial resection of the lesion,⁸ a tumor that only protruded into the atrium without adhesion,⁹ a tumor that was reduced by preoperative radiotherapy,¹⁰ and performance of a transient cava-pulmonary shunt.^{2,4} More than half of the cases (19 cases) were alive after follow-up of at least 6 months.³ Therefore, similar to our case, a favorable prognosis is likely to be obtained after successful resection in combination with chemoradiotherapy.

After resection of the tumor, vascular wall reconstruction is typically necessary. Reconstructions of the SVC using both biologic and synthetic materials, such as autologous or bovine pericardium, the saphenous vein, a spiral vein graft, and expanded polytetrafluoroethylene graft replacement or patch angioplasty have been reported. In our case, the tumor was resected en bloc

with no more tumor thrombus in the SVC, brachiocephalic vein, and RA, and the vessels and RA defect were reconstructed with autologous pericardial patch.

In conclusion, we herein have reported a rare case with invasive thymoma extending to the SVC and the RA. Radical tumor resection and reconstruction of the SVC via cardiopulmonary bypass can improve early survival. The patients, however, must be carefully followed for any evidence of long-term recurrence.

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