Thymic Carcinoma Presenting as an Intraluminal Growth into the Great Vessels and the Cardiac Cavity

Koji Yamazaki, MD,1 Sadanori Takeo, MD,1 Morishige Takeshita, MD,2 and Keizo Sugimachi, MD3

A rare case of thymic carcinoma with an intraluminal growth to the left brachiocephalic vein, the superior vena cava and the right atrium is described. Venogram and cardiac echogram were useful for evaluating the vascular permeation of the tumor. The patient underwent successful en bloc excision of the tumor under conditions of cardiopulmonary bypass. (Ann Thorac Cardiovasc Surg 2002; 8: 163–6)

Key words: thymic carcinoma, intraluminal growth, superior vena cava syndrome

Introduction

Thymic tumors commonly cause superior vena cava (SVC) syndrome by extrinsic compression of the SVC, but less commonly by invasion. Intraluminal permeation to the great vessels is the most uncommon form of causing SVC syndrome. Only eight reports of cases presenting such rare conditions have been published.

Case Report

A 72-year-old Japanese woman was referred to our hospital for advanced dyspnea and edema of face and upper extremities. A chest X-ray, computed tomography (CT) and magnetic resonance imaging (MRI) of the chest revealed an anterior mediastinal mass occupying the left brachiocephalic vein (LBCV) and SVC and extending to the right atrium (RA). A venogram from the right jugular vein revealed a filling defect of the SVC (Fig. 1). A venogram from the left brachial vein showed a total defect of the LBCV. The venous flow to the RA was via the hemi-azygos vein and some collaterals (Fig. 2). With a venogram from the inferior vena cava, the mass extended to the RA. Transesophageal cardiac echogram also revealed the SVC to be completely occupied with a low echoic tumor and the tumor was protruded and flapping into the right atrial cavity. The head of the tumor was 2.0×1.8 cm in the RA (Fig. 3). The preoperative histological diagnosis of the mass proved to be thymoma based on a percutaneous needle biopsy. There was no sign of any metastases to any other organ and no typical symptoms suggesting myasthenia gravis. The preoperative diagnosis was stage III thymoma with Masaoka’s classification.1)

The patient underwent surgery with no preoperative treatments. The median sternotomy revealed that the SVC and RA had been invaded anteriorly by an anterior mediastinal tumor. Under cardiopulmonary bypass, the RA was opened to reveal that the tumor had invaded the superior area of the right atrial endocardium and protruded into the atrial cavity, but without invasion. The right brachiocephalic vein (RBCV) was intact and the tumor was resected with the entire length of the SVC and the antero-lateral side of the RA. Vascular reconstruction was done between the RBCV and RA with a ringed polytetrafluoroethylene graft. A pace maker was not required.

The resected specimen was 19.0×6.5×3.2 cm (Fig. 4). The tumor occupied the LBCV and SVC with a prominent permeation into the intima and media of these vessels. It also invaded the right atrial wall but stumps of the resected specimen were free of tumor cells. The final pathological diagnosis was well differentiated thymic
epithelial cell carcinoma (Fig. 5).

The postoperative course was uneventful and she is alive without relapse 10 months after the surgery.

Comment

Thymic tumors commonly invade the capsule and the adjacent organs such as the pleura, pericardium, lung or great vessels. SVC obstruction is commonly caused by extrinsic compression of the tumor, but less commonly by invasion. To our knowledge, only eight cases including ours have been reported in the English literature with thymic tumors presenting with intraluminal growth to large vessels.

To investigate the area of invasion, chest X-ray, CT scan, MRI, cardiac echogram and venous angiogram were
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Carried out. The venous angiogram was the most helpful in assessing the invasion of vessels. In cases of an intraluminal occlusion in the SVC, the venous angiograms from 3 directions, bilateral cephalic veins and the inferior vena cava, are necessary to determine the pointed head of the tumor. The cardiac echogram was also helpful for confirming the diagnosis.

It has not been well documented, in previously reported cases, as to where the thymic tumors entered the vessels. In our patient, the anterior wall of the SVC and RA were most severely affected by the tumor. However, there were no pathological findings of tumor cells penetrating the atrial cavity which was solid and encapsulated with no involvement of adjacent tissue.

Table 1. Selected treatments and outcomes for thymic tumors with an intraluminal growth

<table>
<thead>
<tr>
<th>Author</th>
<th>Involved vessels</th>
<th>Resection</th>
<th>Adjuvant therapy</th>
<th>Survival (months)</th>
</tr>
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<tbody>
<tr>
<td>Airan²</td>
<td>SVC, RA, RV</td>
<td>Incomplete†</td>
<td>RT</td>
<td></td>
</tr>
<tr>
<td>Yokoi⁷</td>
<td>LBCV, SVC, RA</td>
<td>Complete†</td>
<td>CT, RT</td>
<td>24</td>
</tr>
<tr>
<td>Missault⁶</td>
<td>SVC, RA</td>
<td>Incomplete</td>
<td>RT</td>
<td>24</td>
</tr>
<tr>
<td>Okereke³</td>
<td>LBCV, SVC, RA, IVC</td>
<td>Complete†</td>
<td>None</td>
<td>12</td>
</tr>
<tr>
<td>Filippone⁸</td>
<td>LBCV, SVC, RA</td>
<td>Complete†</td>
<td>CT</td>
<td>24</td>
</tr>
<tr>
<td>Gleeson⁴</td>
<td>LBCV, SVC, RA</td>
<td>Complete</td>
<td>None</td>
<td>12</td>
</tr>
<tr>
<td>Minato³</td>
<td>RBCV, LBCV, SVC, RA</td>
<td>Complete†</td>
<td>CT</td>
<td>29</td>
</tr>
<tr>
<td>Current case</td>
<td>LBCV, SVC, RA</td>
<td>Complete†</td>
<td>None</td>
<td>10</td>
</tr>
</tbody>
</table>

SVC: superior vena cava, RA: right atrium, RV: right ventricle, IVC: inferior vena cava, LBCV: left brachiocephalic vein, RBCV: right brachiocephalic vein. CT: chemotherapy, RT: radiotherapy
†: Surgery was done under conditions of cardiopulmonary bypass.
through the SVC or the right atrial wall. In addition, it is questionable that the tumor would grow against an adverse current to the LBCV. On the other hand, histological examination revealed that the tumor cells severely permeated into the thymic veins. Thus we suggest that the thymic veins that drain the LBCV are possible entrance routes of the tumor. The tumor cells might have entered through them and grown downstream to the RA with the blood flow.

For an advanced stage of malignant thymic tumor, complete resection, including normal thymic tissue and invaded tissue can lead to a good outcome. We completely resected the malignant thymic tumor together with the LBCV, SVC and the upper part of RA, under the conditions of cardiopulmonary bypass. Removal of the SVC occlusion led to a dramatic improvement in the edema of the face and upper extremities. Selected therapies and their outcomes of eight reported cases including ours are given in Table 1. Patients who underwent a complete resection had a better prognosis. Radiotherapy and chemotherapy also can achieve high survival rates. However, on the basis of the small number of patients reported, we cannot assess the influence of adjuvant therapy for thymic tumors which grow intraluminally into great vessels, hence radical excision is, apparently the most important procedure to achieve a good prognosis. In such cases, cardiopulmonary bypass facilitates the radical excision.

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References