CARDIAC TAMPOONADE CAUSED BY INTRAPERICARDIAL YOLK SAC TUMOR IN A BOY

Tien-Chi Liang, Meng-Yuo Lu, Shyn-je Chien, Frank L. Lu, and Kai-Hsin Lin

Abstract: Yolk sac tumor rarely involves the pericardium. A 17-year-old boy had a mediastinal mass leading to total obstruction of the superior vena cava with cardiac tamponade and pleural effusion. Further imaging studies and biopsy showed that the origin was a yolk sac tumor. After surgery and adjuvant chemotherapy, no recurrence was noted during a follow-up of 20 months. This report describes the magnetic resonance imaging characteristics of pericardial yolk sac tumor.

Key words: yolk sac tumor intrapericardial tumor cardiac tamponade superior vena cava syndrome

Endodermal sinus tumor partially or completely recapitulates the structure of the yolk sac, so it is also named yolk sac tumor. It usually develops in the ovary or testis. Less common sites are the sacrococcygeal area, anterior mediastinum, pineal region, and vagina in infants. Rare cases of primary yolk sac tumor have also been reported in the stomach, liver, vulva, retroperitoneum, and prostate and on the face. Intracardial or intrapericardial yolk sac tumors have occasionally been reported [1–6]. Three of the six reported cases were intrapericardial yolk sac tumors, all of which occurred in young girls, 14 months old [1], 18 months old [2], and 3 years old [3], respectively. We report the case of a boy with intrapericardial yolk sac tumor with the unusual presentation of cardiac tamponade combined with total occlusion of the superior vena cava (SVC).

Case Report

A 17-year-old boy complained of low-grade fever, cough, exertional dyspnea, and substernal pain for 1 week. Cardiomegaly and widened mediastinum were noted on both chest roentgenogram and lateral films at admission. He presented with mild plethora, distant heart sound, engorged jugular vein, and paradoxical pulse. Swelling and edematous changes of the face and upper extremities were also noted. Electrocardiography revealed low voltage in all leads and echocardiography showed massive pericardial effusion with a mass lesion between the epicardium and myocardium. About 1,020 mL of bloody, non-coagulable pericardial fluid was drained by pericardiocentesis. Computerized tomography (CT) revealed a huge mediastinal tumor invading the right atrioventricular (AV) groove and partially enveloping the right atrium (RA), aortic root, and pulmonary artery (PA). Pleural effusion with atelectasis of lower lungs was also noted on both sides. Total occlusion of the lower SVC with prominent collaterals on the chest wall was recognized (Fig. 1).

Magnetic resonance (MR) imaging showed that the tumor encased the innominate vein and the distal part of right SVC-RA junction (Fig. 2). Part of the venous blood drained through the azygos vein down to the infradiaphragmatic inferior vena cava (IVC), and the rest of it through prominent collateral vessels on the chest wall (Fig. 3). Elevated alpha-fetoprotein (α-FP) of 9,580 ng/mL and β-subunit of human chorionic gonadotropin of 446.1 mIU/mL suggested a germ cell tumor.

Surgical resection and excisional biopsy were arranged. Intact thymus was noted and a 20 x 15 cm intrapericardial mass which encased the RA, aorta, PA, SVC and innominate vein was found during surgery. The tumor was tightly attached to the base of the heart, so only debulking surgery could be performed. Microscopically, the partially necrotic tumor was composed of flat or cuboidal cells arranged in reticular or microcystic areas. Pseudopapillary processes
with central vessels (Schiller-Duval bodies) were also seen. α-FP was demonstrated by immunohistochemistry study (Fig. 4).

α-FP abruptly dropped to 3,140 ng/mL 2 days after surgery. Plethora and edema of the face and upper extremities disappeared gradually. The patient received adjuvant chemotherapy including bleomycin, etoposide, and cisplatin. After chemotherapy for 2 months, his α-FP returned to normal (< 20 ng/mL). No recurrence was found during regular follow-up at our outpatient department for more than 20 months.

**Discussion**

Sarcoma is the most common primary malignant tumor of the heart, accounting for about 95% of cases. Intrapericardial or intracardial yolk sac tumors are extremely rare with only six cases previously reported (Table) [1–6].

Yolk sac tumor is a germ cell tumor subtype that presents as a highly malignant neoplasm showing overgrowth of the yolk sac endoderm associated with extraembryonic mesoblast. Approximately 10 to 15% of yolk sac tumors occur in an extragonadal site, the most frequent of which is the anterosuperior mediastinum [7].

Only three cases of intrapericardial yolk sac tumors have been previously reported. The first case was reported in 1987 by Nelson and Stenzel in a 14-month-old girl who presented with systolic murmur, cardiomegaly, and heart failure with pericardial effusion [1]. The other two cases, also both in young girls, were reported by Bath et al [2] and Sicari et al [3] with the manifestations of respiratory distress and myocarditis, respectively. The findings of chest roentgenography and echocardiography were described, but only one of these case reports included CT findings.
Cardiac Tamponade in Intrapericardial Yolk Sac Tumor

Fig. 3. Contrast-enhanced magnetic resonance angiography in the right anterior oblique view shows absence of the right brachiocephalic vein and the superior vena cava. The drainage from the right neck and upper extremity goes through either the prominent collaterals in the chest wall (white arrows) or passes to the left side via the jugular plexus. The left innominate vein is patent but blood drains down through the azygos vein (black arrows) first. Anastomosis between the azygos plexus with the inferior vena cava reconstitutes the venous return to the right atrium.

Fig. 4. Schiller-Duval bodies and the reticular pattern of tumor cells are typical of a yolk sac tumor, as seen in this field. (Haematoxylin and eosin, original magnification x 33)

[2]. This case is the report of an intrapericardial yolk sac tumor in a boy. He had an unusual presentation of cardiac tamponade combined with SVC total occlusion that was demonstrated by detailed MR imaging.

Chest MR imaging was used to determine the nature of the lesion and its relationship between adjacent structures. MR imaging provides better resolution for tissue characterization than chest roentgenography, two-dimensional echocardiography, and CT. MR angiography delineated vascular structures and abnormalities in this patient. Abdominal CT and bone scan excluded an extrapericardial origin or extension of the tumor and showed it to be a primary intrapericardial yolk sac tumor.

Table. Characteristics of previously reported cases of intrapericardial and intracardial yolk sac tumor and the present case

<table>
<thead>
<tr>
<th>Ref</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Initial symptoms</th>
<th>Detailed image</th>
<th>Treatment</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>[1]</td>
<td>14 mo</td>
<td>F</td>
<td>Intrapericardial</td>
<td>Irritability, pallor, abdominal distension</td>
<td>Echo</td>
<td>Surgery, post-op C/ T</td>
<td>Recurrent 2 mo later; died 24 mo later</td>
</tr>
<tr>
<td>[2]</td>
<td>18 mo</td>
<td>F</td>
<td>Intrapericardial</td>
<td>Lethargy, anorexia, fever, tachypnea</td>
<td>Echo</td>
<td>Surgery, post-op C/ T</td>
<td>Disease-free survival (&gt; 1 yr)</td>
</tr>
<tr>
<td>[3]</td>
<td>3 yr</td>
<td>F</td>
<td>Intrapericardial</td>
<td>Abdominal discomfort, lethargy, respiratory distress</td>
<td>Echo</td>
<td>Nil</td>
<td>Died at diagnosis</td>
</tr>
<tr>
<td>[4]</td>
<td>15 mo</td>
<td>F</td>
<td>Intracardial</td>
<td>Periorbital swelling</td>
<td>Echo, CT</td>
<td>Surgery, post-op C/ T</td>
<td>Disease-free survival (several mo)</td>
</tr>
<tr>
<td>[5]</td>
<td>2 yr</td>
<td>F</td>
<td>Intracardial</td>
<td>Pale, mild facial puffiness, low volume pulse</td>
<td>Echo</td>
<td>Surgery, post-op C/ T</td>
<td>Died 4 mo later</td>
</tr>
<tr>
<td>[6]</td>
<td>2.5 yr</td>
<td>F</td>
<td>Intracardial</td>
<td>Fever</td>
<td>Echo</td>
<td>Surgery, post-op C/ T</td>
<td>Disease-free survival (&gt; 14 mo)</td>
</tr>
<tr>
<td>This case</td>
<td>17 yr</td>
<td>M</td>
<td>Intrapericardial</td>
<td>Fever, exertional dyspnea</td>
<td>Echo, CT, MRI</td>
<td>Surgery, post-op C/ T</td>
<td>Disease-free survival (&gt; 20 mo)</td>
</tr>
</tbody>
</table>

Ref = reference; F = female; Echo = echocardiography; CT = computerized tomography; C/ T = chemotherapy; M = male; MRI = magnetic resonance imaging.
Total occlusion of SVC has rarely been reported as the initial presentation in intrapericardial tumors. Janin et al found only one cardiac tumor in a review of 175 cases of SVC syndrome [8]. Our patient did not undergo central venous catheterization of the jugular vein at admission due to the risk of injury.

Previous reports indicate a poor prognosis for patients with intrapericardial yolk sac tumor. The first reported case died 1 year after diagnosis [1]. The second reported case died at the time of diagnosis [3]. Our patient was well at 20 months after diagnosis. Including this case, the reported mortality rate for patients with intrapericardial and intracardial tumor is 42.86% (3/7 cases) (Table).

This is the first case report of intrapericardial yolk sac tumor in a male, and it occurred in an older patient than in previous reports. The unusual presentation with cardiac tamponade and pleural effusion due to SVC total occlusion by an intrapericardial yolk sac tumor can easily be missed and should be included in the differential diagnosis. More detailed imaging studies such as MR imaging can help in the evaluation of tumor involvement, planning for operation, and follow-up.

References