Unusual Coronary Sinus Tumor in a Pregnant Woman
A Case Report With Literature Review

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SUMMARY
Primary coronary sinus tumors are extremely rare. Herein, we present a case of a pregnant woman with a primary myxoma in the coronary sinus (CS), which was diagnosed by echocardiography and computed tomography. We reviewed the literature and found two other primary CS tumors. We summarized the gender, ages, symptoms, diagnostic methods, associated anomalies, treatments, histologic findings, and outcomes of the 3 cases. Dyspnea was a common symptom of all 3 patients. Diagnostic methods included echocardiography, computed tomography, magnetic resonance imaging, and coronary angiography. Associated anomalies included coronary artery fistulas, coronary sinus orifice atresia with persistent left superior vena cava, intra-cardiac invasion, and pericardial effusion. The 3 histologic types of primary CS tumor were haemangioma, lymphoma, and myxoma. The 3 patients received proper treatment and had good therapeutic outcomes. (Int Heart J 2017; 58: 633-636)

Key words: Primary, Myxoma, Echocardiography, Computed tomography

Cardiac tumors are an uncommon imaging diagnosis, and are comprised of primary cardiac and metastatic tumors.1) Primary cardiac tumors are rare, with a reported prevalence of 0.001%–0.030% in an autopsy series, while metastatic tumors of the heart are reported to be 10–1000 times more common.2) Three-fourths of primary cardiac tumors are benign, and nearly one-half of benign heart tumors are cardiac myxomas (CMs). CMs can occur anywhere in the heart, but most commonly arise in the left atrium (60%–80%), followed by the right atrium (15%–28%), the right ventricle (8%), and the left ventricle (3%–4%).3,4) Herein, we present the case of a pregnant woman with a CM in the coronary sinus (CS).

CASE REPORT
A 32-year-old primigravida presented to the emergency room with sudden abdominal pain at 38-week gestation. She also had dyspnea without fever. On physical examination, her respiratory rate was 30/minute, pulse was 95/minute, blood pressure was 130/85 mmHg, and her temperature was 37.0°C. There were no murmurs on auscultation of the heart. The electrocardiogram showed a normal heart rhythm. Emergency echocardiographic ultrasound showed a dilated CS and pericardial effusion (PE) in the parasternal long axis view (Figure 1A). The dilated CS was filled with low echo. The 4 chambers of the heart did not have any abnormalities. This mass measured 3.97 × 3.01 cm in the apical 4-chamber view (Figure 1B). To rule out infective endocarditis, the white blood cell count, erythrocyte sedimentation rate, and C-reactive protein level were determined, which were all normal. To rule out a thrombus in the CS, parameters were determined, which were normal, including the D-dimer level (1.23 ug/mL). Therefore, there was a high likelihood that this mass was a tumor. Six hours later, she delivered a male infant with a birth weight of 3550 g by spontaneous vaginal delivery. To confirm the tumor type, contrast-enhanced chest computed tomography (CT) was performed 1 day later. Before contrast agent injection, it was revealed that a tumor with clear boundaries and a PE was present (Figure 2A). During the arterial phase, contrast-enhanced chest CT showed that the tumor had an overall attenuation lower than the myocardium (Figure 2B). During the delayed phase, the tumor demonstrated heterogeneous mild enhancement (Figure 2C). An abdominal CT was also performed; no abnormalities were detected. Considering the echocardiographic and CT imaging features, physicians surmised that this tumor was benign (probably a myxoma). Brain magnetic resonance imaging was normal without evidence of metastases (Figure 3). Surgery was performed 1 week later. A thoracotomy revealed a dilated CS in the atriocentral groove (Figure 4A). The dilated CS was incised with low echo. The tumor was attached to the anterior wall of the middle of the CS by a narrow pedicle. The tumor was exposed and removed. A photograph of the gross specimen showed a dark red tumor with a smooth surface (Figure 4B). Histologic sectioning con-
Figure 1. A: Echocardiography showing a dilated CS and pericardial effusion in the parasternal long axis view. B: The dilated CS was filled with low echo mass, which measured 3.97 × 3.01 cm in the apical four-chamber view. LA indicates left atrium; LV, left ventricle; PE, pericardial effusion; and RV, right ventricle; T, tumor.

Figure 2. A: Before contrast agent injection, contrast-enhanced chest computed tomography (CT) displayed a tumor with clear boundaries and a pericardial effusion. B: During the arterial phase, contrast-enhanced chest CT showed the tumor had an overall attenuation lower than the myocardium. C: During the delayed phase, the tumor demonstrated heterogeneous mild enhancement. T indicates tumor.

Figure 3. Brain magnetic resonance imaging was normal without evidence of metastasis.

Figure 4. A: A dilated CS with the tumor inside was revealed in the atrioventricular groove. B: Photograph of the gross specimen showing the dark red tumor with smooth surface. C: Histologic sectioning confirmed that the tumor was a myxoma. T indicates tumor.
have been reported.

Primary and metastatic types. Several metastatic CS tumors can determine the haemodynamic consequences of the tumor, morphologic appearance, location, and motion of tumors, and benign versus malignant tumors. Certain characteristics identified on CT imaging may help distinguish neoplastic versus non-neoplastic masses and provide additional diagnostic information and are useful for staging and treatment planning, particularly when surgical resection is being considered. Certain characteristics identified on CT imaging may help distinguish neoplastic versus non-neoplastic masses and benign versus malignant tumors. Sometimes, echocardiography may help echocardiography and CT.

In our case, the CS mass was unlikely to be a vegetation because such patients usually have a history of CS endothelial injury caused by right heart procedures or cardiac surgery. Spontaneous CS thrombosis is extremely rare, and is usually associated with atrial fibrillation. None of the above-mentioned findings were present in our patient. After considering that this CS mass was a tumor, we excluded the tumor as metastatic by chest and abdominal CT. Next, we determined the tumor type (benign or malignant). In the absence of invasive echocardiographic imaging and based on the good physical condition of this patient, we concluded that the tumor was benign. Contrast-enhanced chest CT helped identify the types of benign primary cardiac tumors.

The 3 histologic types of the primary CS tumors were haemangioma, lymphoma, and myxoma.

Treatment of primary CS tumors is like that for other cardiac tumors: in most patients with benign tumors, or in those patients with resectable primary malignant disease and no evidence of metastasis, surgery is generally indicated to improve prognosis. Patients with a primary cardiac lymphoma are usually treated with chemotherapy and radiotherapy rather than surgery. All 3 patients had good outcomes: two recovered well after surgery; and the other patient with dyspnea was markedly improved after chemotherapy.

In conclusion, primary CS tumors can be detected by multiple imaging methods. Proper treatment may achieve a good therapeutic outcome.

**REFERENCES**


**Table. Summary of Literature Involving the Primary CS Tumors**

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<td>Our case</td>
<td>32, Female</td>
<td>Dyspnea</td>
<td>TTE, CT</td>
<td>PE</td>
<td>Surgery</td>
<td>Myxoma</td>
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CAF indicates coronary artery fistula; CS, coronary sinus; CSOA, coronary sinus orifice atresia; CT, computed tomography; MRI, magnetic resonance imaging; PE, pericardial effusion; PLSVC, persistent left superior vena cava; TEE, transesophageal echocardiography; and TTE, transthoracic echocardiography.

**DISCUSSION**

A cardiac mass in the CS is extremely rare, and includes thrombi, vegetations, and tumors. Tumors are divided into primary and metastatic types. Several metastatic CS tumors have been reported. To the best of our knowledge, only 3 primary CS tumors have been reported, including the case herein. Our case was the only one involving a myxoma. The gender, ages, symptoms, diagnostic methods, associated anomalies, treatments, histologic findings, and outcomes of the 3 cases are shown in the Table. The most common symptom of the patients was dyspnea. Dyspnea may be caused by obstruction of the tumor. Normal CS pressure is approximately 0 mmHg; when CS obstruction occurs (CS stenosis or CS orifice atresia), the CS pressure may increase to 10–25 mmHg. The increased CS pressure could cause left ventricle diastolic dysfunction and an elevated filling pressure, thus indirectly leading to dyspnea.

The diagnostic methods included echocardiography (transesophageal echocardiography and transthoracic echocardiography), CT, magnetic resonance imaging (MRI), and coronary angiography. Echocardiography is an ideal initial imaging modality because it is simple, non-invasive, widely available, and has a low cost. Transesophageal echocardiography is the diagnostic test of choice for benign cardiac tumors, and has a 93.3% diagnostic accuracy. Echocardiography delineates the morphologic appearance, location, and motion of tumors, and can determine the haemodynamic consequences of the tumor, if any. The echocardiographic appearance of cardiac tumors may accurately predict tumor type (benign or malignant). CT and MRI are complimentary techniques that provide additional diagnostic information and are useful for staging and treatment planning, particularly when surgical resection is being considered. Certain characteristics identified on CT imaging may help distinguish neoplastic versus non-neoplastic masses and benign versus malignant tumors. Sometimes, electrocardiography may help echocardiography and CT. In our case, the CS mass was unlikely to be a vegetation because such patients usually have fevers, congenital heart disease, or a history of intravenous drug use. A CS mass is unlikely to be a thrombus because such patients usually have a history of CS endothelial injury caused by right heart procedures or cardiac surgery. Spontaneous CS thrombosis is extremely rare, and is usually associated with atrial fibrillation. None of the above-mentioned findings were present in our patient. After considering that this CS mass was a tumor, we excluded the tumor as metastatic by chest and abdominal CT. Next, we determined the tumor type (benign or malignant). In the absence of invasive echocardiographic imaging and based on the good physical condition of this patient, we concluded that the tumor was benign. Contrast-enhanced chest CT helped identify the types of benign primary cardiac tumors.

Associated anomalies included coronary artery fistula, CS orifice atresia with persistent left superior vena cava, intracardiac invasion, and PE. There are no special anomalies associated with primary CS tumors, according to the literature.

The 3 histologic types of the primary CS tumors were haemangioma, lymphoma, and myxoma.

Treatment of primary CS tumors is like that for other cardiac tumors: in most patients with benign tumors, or in those patients with resectable primary malignant disease and no evidence of metastasis, surgery is generally indicated to improve prognosis. Patients with a primary cardiac lymphoma are usually treated with chemotherapy and radiotherapy rather than surgery. All 3 patients had good outcomes: two recovered well after surgery; and the other patient with dyspnea was markedly improved after chemotherapy.

In conclusion, primary CS tumors can be detected by multiple imaging methods. Proper treatment may achieve a good therapeutic outcome.
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