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Secondary Cardiac Tumor in Children

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Abstract. We describe our clinical experience of eight cases of secondary cardiac tumor. The pathology of the tumors were lymphoma (three), Wilms' tumor (two), malignant teratoma (one), neuroblastoma (one), and pleuropulmonary blastoma (one). Metastatic sites were the right atrium in Wilms' tumor and neuroblastoma, the left atrium in pleuropulmonary blastoma and malignant teratoma, and multiple sites in lymphoma. Primary masses in the mediastinum extended directly to the heart (three lymphoma, malignant teratoma, pleuropulmonary blastoma). Wilms' tumor and neuroblastoma showed cardiac metastases through the inferior vena cava. Many cases revealed vague abnormal cardiovascular findings (symptoms in six; physical signs in five). In five cases surgery was performed to relieve the possible obstruction to flow and to identify the pathology (lymphoma in three, Wilms' tumor in one, and malignant teratoma in one). Chemotherapy prior to operation resulted in the disappearance of the intracardiac masses in each case of Wilms' tumor and pleuropulmonary blastoma. All three patients with lymphoma died immediately after operation. Four died of multiple metastases or *Pneumocystis* pneumonia several months after operation. This study indicates that suspicion of a secondary cardiac tumor is crucial to early diagnosis. Because of the poor postoperative outcome, surgery for secondary cardiac tumors should be done cautiously only in cases with definite hemodynamic decompensation.

Key words: Secondary cardiac tumor — Children

The incidence of cardiac metastases is rising, probably due to the increasing incidence of cancer and the fact that patients are living longer with their disease. The improvement in diagnostic methods such as echocardiography and magnetic resonance imaging (MRI) makes it possible to detect early cardiac metastases with or without clinical symptoms.

The incidence of secondary cardiac tumors ranges up to 20% in adults [1, 3]. However, information on metastatic involvement of the cardiovascular system in childhood is scanty. The pattern of secondary cardiac tumors in children seems different from that in adults because of the rarity of carcinomas and malignant melanoma in children and of neuroblastoma in adults [2, 4].

The purpose of this study was to describe the clinical features and therapeutic experience of eight cases of cardiac metastases. These were either suspected as a result of their cardiovascular manifestations (six cases) or found incidentally by radiological studies such as computed tomography (two cases).

Patients and Methods

On retrospective examination of the medical records at Seoul National University Children's Hospital between January 1986 and October 1997, eight cases of secondary cardiac tumor were found. Patients consisted of five males and three females, ranging in age from 2 to 11 years.

Results

Tables 1 and 2 give the clinical data and outcome for each of the eight children.

Presenting Features

Six patients presented with cardiovascular symptoms such as dyspnea, tachypnea and chest pain. The other

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Table 1. Clinical and radiological findings of secondary cardiac tumors

Case no.	Age at diagnosis	Sex	Cardiovascular symptoms	Radiological findings
1	2 years	M	Asymptomatic	MRI: right kidney mass and IVC thrombosis-like lesion extending to right atrium
2	3 years	M	Asymptomatic	CT: right kidney mass and IVC thrombosis-like lesion extending to right atrium
3	2 years 10 months	F	Dyspnea	CT: right thoracic and mediastinal mass
4	4 years 8 months	F	Right chest pain and cough	CT: huge right thoracic mass
5	4 years 11 months	M	Dyspnea	CT: right adrenal mass extending via the IVC to the right atrium
6	3 years 2 months	F	Chest pain and tachypnea, respiratory distress	Cardiomegaly
7	11 years	M	Dyspnea	MRI: thickening of right ventricle, RVOT, interventricular septum
8	3 years	M	Dyspnea on exertion and cough	Mediastinal widening, cardiomegaly

M, male; F, female; MRI, magnetic resonance imaging; IVC, inferior vena cava; CT, computed tomography; RVOT, right ventricular outflow tract.

Table 2. Clinical diagnosis and outcome of secondary cardiac tumors

Case no.	Echocardiography	Initial treatment	Outcome	Pathology
1	Mass in RA	Chemotherapy: response (+)	Death: multiple metastases after 18 months	Wilms' tumor
2	Mass in RA nearly obstructing the inflow of RV	Surgical removal	Alive: follow-up for 7 months	Wilms' tumor
3	SVC obstruction; masses in LA, right pulmonary artery and vein	Chemotherapy: response (+)	Death: PCP after 11 weeks	Pleuropulmonary blastoma
4	Mass in LA obstructing the inflow of LV	Surgical removal	Death: PCP after 3 months	Malignant teratoma
5	Mass in RA	Chemotherapy: no response	Death: multiple metastases after 6 months	Neuroblastoma
6	Multiple tumors in RA, interatrial septum, RVOT, and posterior septal wall of LV; pericardial effusion	Surgical removal	Death: postoperative deterioration 2 days later	Lymphoma, Burkitt type
7	Mass in RVOT; thickening of free wall and septum of RV; pericardial effusion	Surgical removal	Death: immediately after operation	Lymphoma
8	Narrowing of right pulmonary artery and compression of LA and LV	Exploratory operation	Death: postoperative deterioration 1 week later	Lymphoma, Burkitt type

RV, right ventricle; SVC, superior vena cava; RA, right atrium; RVOT, right ventricular outflow tract; LA, left atrium; LV, left ventricle; PCP, *Pneumocystis carinii* pneumonia.

two patients (cases 1 and 2) did not show any cardiovascular symptoms. Physical examination revealed a low-grade systolic murmur in four patients (cases 2, 6, 7, and 8) and a low-pitched sound after the S2 sound that was suggestive of tumor plop (case 4).

Location and Extension of Tumors

Primary sites of tumors in our patients were the right adrenal gland in one case (case 5), the right kidney in two cases (cases 1 and 2) and the mediastinum in five cases (cases 3, 4, 6, 7, and 8).

Intracardiac masses were diagnosed by echocardiography in seven cases. The patient with lymphoma (case 8) showed no definite intracardiac mass; instead a compressed left ventricle, left atrium and right pulmonary artery were seen on echocardiography. The extension and metastasis of the tumor were evaluated by computed tomography (CT) in four cases and magnetic resonance imaging (MRI) in two cases.

In the two cases of Wilms' tumor, a renal mass

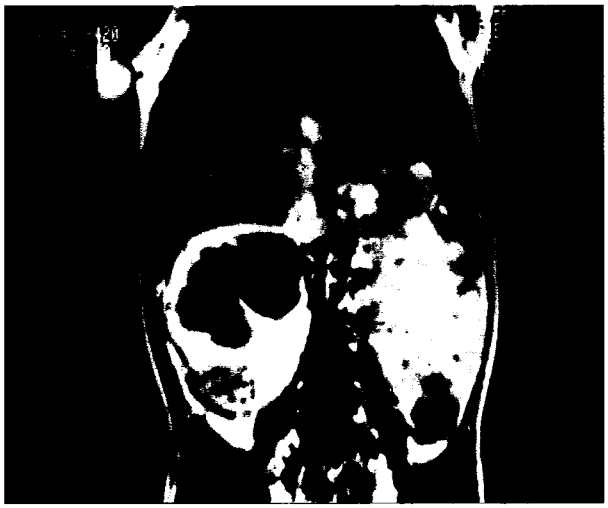

Fig. 1. Case 1. In this case of Wilms' tumor, MRI (coronal image) shows a huge right renal tumor with cystic portions. The tumor extends to the inferior vena cava and right atrium.

Fig. 2. Case 2. Apical four-chamber view echocardiogram in a child with Wilms' tumor demonstrating a large echo-dense mass in the right atrium.

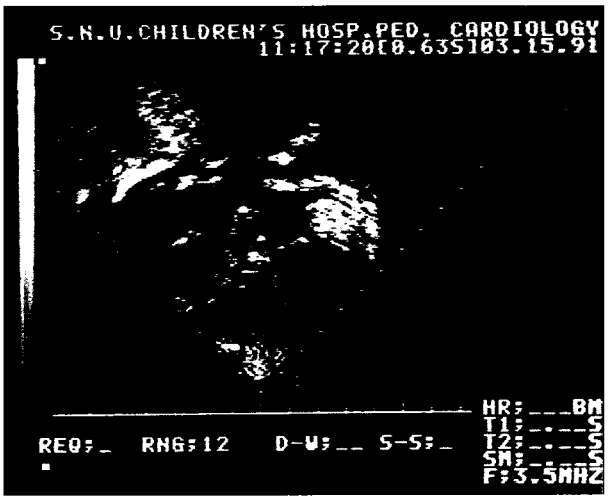

Fig. 3. Apical four-chamber view echocardiogram in a child with lymphoma demonstrating a large echogenic mass in the right atrium with possible obstruction to the flow into the right ventricle and another echogenic mass in the interatrial septum and posterior septal wall of the left ventricle.

extended through the inferior vena cava to the right atrium (cases 1, 2) (Figs. 1 and 2). In one case of neuroblastoma, a right adrenal mass extended through the inferior vena cava to the right atrium (case 5). In another two cases, an intracardiac mass was located in the left atrium obstructing the inflow of the left ventricle (cases 3 and 4). In the two cases of lymphoma, multiple cardiac site involvement resulted in an obstruction of blood flow into and out of both ventricles (Fig. 3).

Pathology and Outcome

Pathological studies revealed malignant teratoma (one), neuroblastoma (one), pleuropulmonary blastoma (one), Wilms' tumor (two), and non-Hodgkin's lymphoma (three). Initially, three patients (cases 1, 3, and 5) were treated with chemotherapy and the other five (cases 2, 4, 6, 7, and 8) with operative removal of the cardiac tumor. In each case of Wilms' tumor (case 1) and pleuropulmonary blastoma (case 3), chemotherapy resulted in the disappearance of the intracardiac mass. In the case of neuroblastoma, chemotherapy left a residual mass in the right atrium that was detected later when the operation to remove residual adrenal mass and inferior vena caval extension was performed (case 5). The initial operations were performed to relieve obstruction (cases 2, 4, 6, and 7) or to identify tumor pathology (case 8). Except in the patient with Burkitt's lymphoma (case 8), as much as possible of the cardiac mass was removed at operation. In case 8 the mass was left intact because of an undiagnosed infiltration into the left atrium. Chemotherapy after the operation resulted in a significant reduction in the size of the mass.

Only one patient with Wilms' tumor is still alive (case 2). Two patients died of *Pneumocystis carinii* pneumonia about 3 months postoperatively (cases 3 and 4). Two patients died of multiple metastases 6 and 18 months after detection (cases 1 and 5). All three patients with lymphoma died soon after open-heart surgery.

Discussion

With improvement in therapeutic procedures and the resultant prolongation of survival, the incidence of cardiac

metastases is likely to increase [3]. However, early diagnosis of cardiac metastases is very difficult in most cases, due to the insidious nature of cardiac lesions [7]. Unexplained rapid cardiac enlargement, symptoms of cardiac dysfunction, congestive heart failure and cardiac arrhythmia in cancer patients with no previous history of heart disease should alert clinicians to the possibility of cardiac metastases [3, 5, 8].

In our six patients with cardiopulmonary symptoms the dyspnea, tachypnea or chest pain were thought to result from their deteriorating general condition. The cardiopulmonary symptoms were more profound in the patients with mediastinal primary tumors. In addition, four patients presented with systolic murmur and one with a tumor plop-like sound, all of these findings being regarded as insignificant. In two asymptomatic cases, a cardiac lesion was suspected on CT of the abdomen and was confirmed by echocardiography. The difficulty of early detection of cardiac metastases is shown in our experience.

Surgery was performed at the time of diagnosis in five of our cases to relieve the possible obstruction of blood flow or to identify tumor pathology. Although operations were performed to prevent the possible hemodynamic deterioration and to excise the tumor mass, the outcome was very poor. Only one patient with Wilms' tumor is still alive. The other patients died early after operation (immediate perioperative days in three cases of lymphoma; *Pneumocystis* pneumonia infection 3 months postoperatively in each case of pleuropulmonary blastoma and malignant teratoma). The surgical intervention did not improve the prognosis of the underlying tumors, which implies that a different approach rather than early surgery should be considered even in cases that would result in possible hemodynamic deterioration. One possible solution is that if the hemodynamics is tolerable, a rapid pathological diagnosis should be done primarily by means of less traumatic methods such as endocardial biopsy or thoracotomy to acquire tissue. If the tumor is determined to be sensitive to chemotherapy and/or radiotherapy, these treatment modalities could be chosen prior to major operation. If the hemodynamics is definitely compromised or the tumor is resistant to chemotherapy and/or radiotherapy, an operation should be performed rapidly to relieve the obstructive flow and to reduce the mass. Our experiences of initial chemotherapy in three cases without seriously obstructive lesions support the previous proposal: disappearance of intracardiac masses was confirmed in two cases of Wilms' tumors and a marked reduction of the tumor mass in the case of neuroblastoma. The perioperative mortality in lymphoma cases is thought to be triggered by additional operative cardiac injury.

Since the detection of cardiac tumors is difficult due to the generally vague cardiovascular symptoms or signs, early suspicion is very important. A thorough cardiovascular evaluation by meticulous echocardiographic examination should be done in every suspicious case. Antemortem diagnosis of cardiac metastases retains a great practical significance since it is immediately connected with the problem of operability as well as with the choice of the most rational method of surgical intervention or conservative methods of treatment [6].

We conclude from these experience that one should be alert to the possibility of cardiac metastases and try to make a definite pathological diagnosis through aggressive diagnostic approaches. Once cardiac metastasis occurs, the prognosis is grave. The protocol of early operation to remove cardiac tumor or to relieve possible obstruction in all cases should be redefined. Only in cases with definitely decompensated hemodynamics should early operation be considered prior to other therapeutic modalities. In tumors resistant to chemotherapy and/or radiotherapy, operative removal of the cardiac tumor could be performed in selected cases.

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References

1. Burnett RC, Shimkin MB (1954) Secondary neoplasms of the heart. *Arch Intern Med 93*:205–218
2. Chan HSL, Sonley MJ, Moes CAF, et al. (1985) Primary and secondary tumors of childhood involving the heart, pericardium, and great vessels: a report of 75 cases and review of the literature. *Cancer 56*:825–836
3. Mukai K, Shinkai T, Tominaga K, Shimosato Y (1988) The incidence of secondary tumors of the heart and pericardium: a 10-year study. *Jpn J Clin Oncol 18*:195–201
4. Nadas AS, Ellison RC (1968) Cardiac tumors in infancy. *Am J Cardiol 21*:363–366
5. Nakayama R, Yoneyama T, Takatani O, Kimura K (1966) A study of metastatic tumors to the heart, pericardium and great vessels. I. Incidence of metastases to the heart, pericardium and great vessels. *Jpn Heart J 7*:227–234
6. Skhvatsabaja LV (1986) Secondary malignant lesions of the heart and pericardium in neoplastic disease. *Oncology 43*:103–106
7. Smith C (1986) Tumors of the heart. *Arch Pathol Lab Med 110*: 371–374
8. Vaughn ED, Crosby IK, Tegtmeyer CJ (1977) Nephroblastoma with right atrial extension: preoperative diagnosis and management. *J Urol 117*:530–533