Case Report

Primary cardiac fibroma in an infant: Computed tomography and magnetic resonance imaging findings

Hsien-Tzu Liu a, Chui-Mei Tiu a,b,c,*, Zen-Chung Weng d, Yi-Hong Chou a,b, Huai-Cheng Hsueh c, Ming-Hsun Lee c, Tse-Kai Tseng c, Cheng-Yen Chang a,b

a Department of Radiology, Taipei Veterans General Hospital, Taipei, Taiwan, ROC
b Department of Radiology, National Yang-Ming University, Taipei, Taiwan, ROC
c Department of Radiology, Lotung Poh-Ai Hospital, Lotung, Ilan, Taiwan, ROC
d Division of Cardiovascular Surgery, Department of Surgery, Taipei Veterans General Hospital, Taipei, Taiwan, ROC

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Abstract

Cardiac fibromas (CFs) are benign primary tumors that typically occur during childhood and may be asymptomatic. However, due to the proximity of CFs to the cardiac structure, significant morbidity and mortality may also be anticipated. CFs do not show spontaneous regression and surgical resection generally remains the treatment of choice for these tumors in children. Thus, it is important to take aggressive steps to obtain accurate pretreatment image diagnosis. A full-term male infant was presented to our facility suffering from shortness of breath, after an episode of upper respiratory tract infection at age 1.5 months. Subsequent chest X-ray revealed widening of the mediastinum and trachea deviation. Cardiogenic pathology was suspected. Computed tomography and magnetic resonance imaging were performed, and we confirmed a diagnosis of benign CF. Thoracotomy biopsy of the tumor confirmed the pathological diagnosis.

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1. Introduction

Primary cardiac tumors are rarely encountered in pediatric practice. When faced with a child suspected of cardiac neogrowth, noninvasive imaging should be undertaken as the first step of assessment. We report a case of primary cardiac tumor, its presentation, image evaluation, and final outcome, along with a review of the literature, focusing on computed tomography (CT) and magnetic resonance imaging (MRI) findings.

2. Case report

A 1.5-month old male infant presented with irritable crying and shortness of breath after an episode of upper respiratory tract infection. Chest X-ray provided evidence of a space-occupying lesion in the mediastinum (Fig. 1). Further echocardiographic examination showed a tumor mass in the left ventricle, with compression of the right ventricle and aortic outflow tract obstruction. Pericardial effusion was also found. Further imaging with CT and MRI was performed.

Low-dose CT revealed cardiomegaly with a homogeneous solid tumor measuring up to 4 cm in diameter and occupying the left ventricular cavity. Tumor infiltration of the interventricular septum was also demonstrated. Furthermore, there was compression of the right ventricle and left main bronchus, resulting in near total collapse of the left upper lung (Fig. 2).
After contrast injection, enhancement of the tumor was displayed, and a large feeding vessel was discovered at the early arterial phase (Figs. 3A and 3B). Contrast-enhanced MRI revealed similar findings of a hypervascular infiltrative tumor occupying the left ventricle. Signal intensity of the tumor mass could be readily differentiated from the surrounding myocardium, except for intramural involvement at the interventricular septum. Post-contrast T1-weighed imaging showed strong enhancement, confirming its hypervascular nature (Fig. 4). Later, tumor biopsy via thoracotomy was done, and pathology confirmed the diagnosis of cardiac fibroma (CF).

Surgery was not indicated at that time due to the lack of immediate risk of cardiac decompensation. However, due to the child’s persistent wheezing and dyspnea, bronchoscope-assisted left main bronchus stenting was done as palliative treatment for tumor-compression-related laryngomalacia.

During the patient’s entire course of hospitalization, electrocardiographic monitoring showed multiple ventricular...

Fig. 1. Chest X-ray on presentation depicts significant cardiomegaly in correlation with age, and rightward deviation of the trachea, suggesting a mediastinal space-occupying lesion.

Fig. 2. Post-contrast, arterial phase, axial computed tomography. A four-chamber view demonstrates cardiomegaly with a space-occupying lesion in the left ventricle. There is a significant mass effect on surrounding structures with heart axis rotation, interventricular septum deviation, and left ventricle outflow tract obstruction. No foci of calcification are identified.

Fig. 3. (A) Post-contrast, arterial phase, axial computed tomography (CT) depicts a large tumor feeding vessel (arrow) branching from the left coronary artery and penetrating into the left ventricle tumor mass, indicating its hypervascular nature. (B) Post-contrast, arterial phase, coronal reconstructed CT, four-chamber view. The tumor feeding vessel is seen en face, entering the cardiac tumor at its superior aspect.

Fig. 4. T1-weighed, post-contrast, fat saturation, axial magnetic resonance imaging shows strong heterogeneous enhancement of the cardiac tumor, and ill-defined differentiation between the tumor and adjacent ventricle wall.
premature complex and repetitive episodes of accelerated ventricular rhythm, despite the use of antiarrhythmic agents. This can be explained as the effect of tumor extension into the conduction system, which was compatible with the imaging finding of interventricular septum involvement.

Before surgical intervention could be achieved, fatal irreversible ventricular tachycardia developed at the age of 4 months.

3. Discussion

Primary cardiac tumors are rare in pediatric practice with a prevalence of 0.0017–0.28% in autopsy series. Among these, benign tumors make up the majority, whereas ~10% are malignant. The most commonly found benign pediatric cardiac tumor is rhabdomyoma, which comprises >60% of the total number of benign primary tumors. CF and cardiac teratoma are the second and third most frequently occurring tumors, respectively.1–4 A third of these patients are diagnosed at <1 year of age.3 The mean age of presentation is 13 years.5

Fibromas are described as solitary, fibrous tumors arising from the connective tissue fibroblasts of the left ventricular free wall or the interventricular septum. They are dormant and have not been documented to show spontaneous regression over time. The average size of these tumors ranges from 1 cm to 10 cm.3,4 Presentation of primary heart tumors is mainly associated with the classic cardiac symptoms resulting from blood flow obstruction, cardiac strain, and other constitutional symptoms. CF, due to its predilection in the ventricular free wall and septum, has the additional involvement of the cardiac conduction system and may result in ventricular arrhythmia, and ultimately, sudden death.6

Tumor biopsy with histological assessment is the gold standard for tumor diagnosis. With the increasingly accelerated development of imaging modalities, noninvasive imaging studies including echocardiography, CT, and MRI have become widely used as first-line assessment tools of cardiac tumors, whereas cardiac catheterization is rarely indicated.1

CF appears as a well-demarcated, homogeneous mass with iso-to hyperintensity in comparison with adjacent myocardium on T1-weighted images, and hyperintense on T2-weighted images of MRI.3,5 Delayed enhancement on post-contrast T1-weighted imaging has been described.5 On CT imaging, the tumor appears as a solitary tumor with a ventricular mural origin. It is mainly hypervascular with homogeneous or heterogeneous enhancement after contrast administration. Large feeding vessels of the tumor are occasionally identified. CT is optimal for investigating foci of calcification, reflecting areas of poor blood supply seen in 25% of the cases.3–5

Surgical resection remains the treatment of choice despite its high mortality rate and relatively poor late outcome in patients with large masses. However, some benefit arising from subtotal resection, staged surgery, or palliative shunting has been reported.3

In conclusion, CF should be suspected in neonates, infants, and children with symptoms of cardiac decompensation or ventricular arrhythmia. As in our case, when we suspected a mediastinal mass lesion as seen first on the chest X-ray, we performed a further noninvasive study utilizing both CT and MRI. With CT, reconstructed images provide excellent anatomic demonstration, whereas MRI clarifies tissue characteristics and information about tumor vascularity by contrast-enhanced studies; this accumulated diagnostic information can better enable practitioners to make a timely finding of CF. However, upon CT examination for younger patients, radiation burden is a particularly important issue and a low-dose protocol should be undertaken whenever possible without compromising image quality.

A spectrum of possibilities can be proposed in the setting of a primary cardiac tumor. The differential diagnosis of CF is based largely on patient age, with imaging providing additional information, including a solitary mass with typical intramural origin, predominantly occupying the ventricle, and heterogeneous contrast enhancement suggesting the hypervascular nature of this tumor. Additionally, calcified central areas of impaired perfusion and tumor feeding vessels may or may not be seen. Given the above information, CF becomes a favored prospective diagnosis.

References