Hemangioendothelioma of the right atrial appendage associated with pericardial effusion in an infant

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Background: **Cardiac hemangioma is a rare cause for pericardial effusion. We present a case of hemangioendothelioma of the right atrial appendage associated with pericardial effusion in an infant.**

Methods: **The patient was a 49-day-old infant transferred to our department because of the presence of pericardial effusion. Two-dimensional transthoracic echocardiography revealed moderate pericardial effusion and a 24 mm × 16 mm hypoechoic mass located on the right side of the right ventricular outflow tract and in front of the aorta.**

Results: **The infant underwent an exploratory median sternotomy. In the pericardial cavity, 120 mL transudate was observed. A 15 mm × 15 mm encapsulated mass of soft tissue was located in the pericardial cavity and involved the right atrial appendage. The tumor with right atrial appendage was completely removed. Pathological examination revealed wide sessile implant basis of the tumor into the the myocardium of the right atrial appendage, with no affection to the endocardium. Hemangioendothelioma was confirmed histopathologically. Echocardiographic examination 2 years after operation revealed that the infant was free from tumor recurrence.**

Conclusions: **Pericardial effusion may be caused by hemangioma of the right atrial appendage. The diagnosis of cardiac hemangioma is based on imaging examination and histopathological studies.**

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Key words: **cardiac hemangioma; infant; pericardial effusion; right atrial appendage**

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Introduction

Cardiac hemangioma is a rare cardiac tumor. [1,2] Primary cardiac hemangioendothelioma in infancy is extremely rare. Here we present a case of pericardial effusion caused by hemangioendothelioma in the right atrial appendage in an infant.

Case report

A 49-day-old female infant was transferred to our department for the presence of pericardial effusion. The infant was born at 38 weeks, weighed 2 kg, had no history of shock, dyspnea or arrhythmia, and no family history of congenital heart disease.

On admission, she was afebrile, with a heart rate of 148 beats/min and a respiratory rate of 48 breaths/min with respiratory distress. Her blood pressure was 94/57 mmHg. All peripheral pulses were well felt. Pallor, icterus, cyanosis, and edema were absent. Systemic examination revealed normal heart sounds with soft murmur in the apex. Examination of the respiratory system showed nothing abnormal. There was no hepatosplenomegaly or hemangioma cutis. The infant had mild hypertonia and brisk knee reflexes.

Laboratory examination revealed normal renal and liver functions. Blood routine and blood gas examination were also normal. Chest radiography demonstrated cardiomegaly, but electrocardiogram was normal. A chest CT scan revealed mild pericardial effusion. No pulmonary pathology or abnormal cardiac mass was found by CT. Two-dimensional transthoracic echocardiography revealed moderate pericardial effusion and a 24 mm × 16 mm hypoechoic mass located at the right side of the right ventricular outflow tract and in front of the aorta (Fig. 1). Color Doppler flow imaging showed abnormal arterial

flow patters within and around the mass. No other coronary abnormalities were found. Initially, the mass was considered as a myxoma. Two days after hospitalization, the infant underwent an open-heart surgery without cardiopulmonary bypass. About 120 mL transudate was seen in the pericardial cavity and drained. A 15 mm × 15 mm encapsulated mass of soft tissue was located in the pericardial cavity and involved the right atrial appendage. The tumor with right atrial appendage was completely removed. Pathological examination of the resected specimens revealed wide sessile implant basis of the tumor into the myocardium of the right atrial appendage, with no affection to the endocardium. The cut surface of the mass was reddish and showed abundant blood vessels in the central area. Microscopically, the tumor showed a lot of cavities surrounded by multiple, dilated, thin-walled vessels, and endothelial cells proliferated and swelled, projecting into the cavities. Hyperplasia of papillary endothelial cells was seen in part of the caverns.

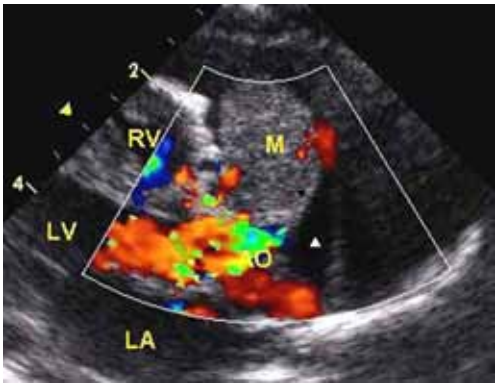


Fig. 1. Echocardiography revealed the heart was surrounded by a moderate amount of pericardial effusion (▲), and a medium echo mass (↑) with a diameter of 24 mm × 16 mm on right of the right ventricular outflow tract and in front of the aorta.

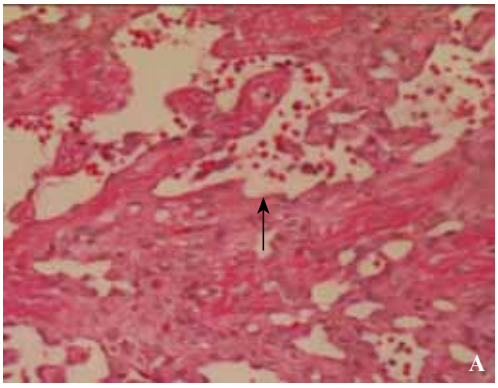
Immunohistochemically, tumor-forming capillary endothelial cells were positive for Vimentin and CD34 staining. An infantile hemangioendothelioma was confirmed histopathologically (Fig. 2).

The infant was uneventfully discharged 2 weeks after the operation. A repeated echocardiography showed no pericardial effusion or mass. She has been followed up for 2 years without any evidence of echocardiographic recurrence.

Discussion

Cardiac hemangioma is a rare tumor of the heart that can affect patients of different ages, but it is often benign in children. The tumor may involve the endocardium, myocardium or epicardium. Pathologically, cardiac hemangiomas are classified into cavernous, capillary and arteriovenous types. The incidence of primary cardiac tumor is estimated to be between 0.001% and 0.03% at autopsy. Among benign primary tumors of the heart, hemangiomas account for 5% to 10%.[2] Symptoms of cardiac hemangioma are dependent primarily on the anatomic location, size, and extension of the tumor. In most cases, cardiac hemangiomas are asymptomatic. In symptomatic patients, the tumor can cause arrhythmia, congestive heart failure, coronary insufficiency, pericardial effusion,[2,3] and sudden death.[4] The presence of multiple hemangiomas in the skin should be suspicious of cardiac hemangioma in neonates.[5] Cardiac hemangioma is usually diagnosed by noninvasive imaging examinations such as echocardiography, CT, and MRI.[6]

Non-hydropic pericardial effusion in newborns or infants is uncommon.[1] It may present with compressive symptoms, but is usually asymptomatic. In the presence of an enlarged cardiac silhouette on chest X-ray films, the possibility of a pericardial effusion should be



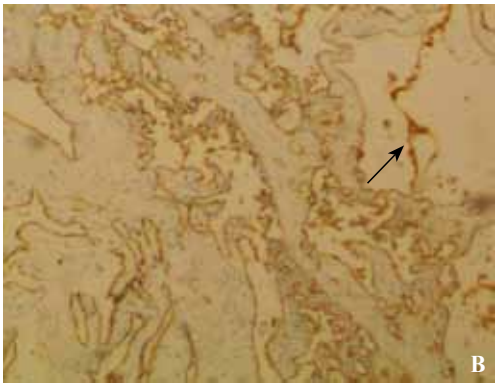


Fig. 2. A: Histopathologically, the tumor showed a lot of caverns with multiple, dilated, thin-walled vessels (arrow), and hyperplasia of papillary endothelial cells was noted in part of the caverns (HE, original magnification × 400). **B:** Immunohistochemically, tumor-forming capillary endothelial cells showed a strong positive CD34 staining (arrow).

considered. Transthoracic echocardiography and other image examinations are necessary. Pericardial effusion is not a common symptom of cardiac hemangioma. In rare cases, cardiac hemangioma may cause cardiac tamponade, which needs emergent surgery.[7]

Cardiac hemangioma of the right atrial appendage associated with pericardial effusion is extremely rare in infancy. To our knowledge, our patient might be the first infant who had the hemangioma arising from the right appendage and growing into the pericardial cavity with resultant pericardial effusion. Diagnosis is made by echocardiography and the tumor was successfully removed. However, as shown in our patient, extraatrial growth of the tumor makes the cardiac tumor difficult to be distinguished from a paracardiac mass.

Cardiac hemangioma has an unpredictable outcome, and not all of such patients have a favorable prognosis. Successful resection can be made only when hemangioma is well circumscribed and small.[8] For patients undergoing surgical excision, follow-up with regular echocardiography is recommended for the detection of tumor recurrence.[9]

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