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CASE REPORT

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Superior vena cava obstruction following pericardial effusion absorption in the presence of a pericardial teratoma: a case report

Kai Guo[1] and Hui Wang[1*]

Abstract

Intrapericardial teratoma is a rare tumor that usually presents in neonates or during infancy because of the associated high degree of pericardial effusion, cardiac compression and severe respiratory distress. In this paper, we report a rare case of intrapericardial teratoma that was incidentally discovered in an infant with superior vena cava obstruction following pericardial effusion absorption. Echocardiography and thoracic computed tomography angiography revealed that the intrapericardial mass obviously suppressed the superior vena cava. The tumor was resected surgically. Histopathological examination confirmed the diagnosis of mature teratoma.

Keywords Intrapericardial teratoma, Superior vena cava obstruction, Thoracic computed tomography angiography

Introduction

Intrapericardial teratoma is a rare, congenital condition. These are usually pedunculated, primary cardiac tumors [1]. They usually present in neonates or during infancy because of the associated large pericardial effusion, cardiac compression and severe respiratory distress but rarely complicated by superior vena cava obstruction [2–4]. We report a rare case of intrapericardial teratoma that was incidentally discovered in an infant with superior vena cava obstruction following pericardial effusion reduction.

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Case presentation

4-month-old boy presented with 3 days of dry cough. The patient underwent chest radiography, which revealed cardiomegaly with a cardiothoracic ratio of 0.79 (Fig. 1). To determine the cause of the cardiomegaly, he underwent echocardiography, which confirmed the presence of massive pericardial effusion. Physical examination revealed no pedal edema, puffiness of the face or indrawing of the chest cavity with inspiration. The liver and spleen were not palpable. Instead of pericardiocentesis, diuretic therapy was used to drain the pericardial effusion. After 2 days of diuretic therapy, the patient's facial puffiness was surprising. To determine the reason, the patient underwent echocardiography, which unexpectedly revealed a well-defined, multilocular, intrapericardial cystic mass with little pericardial effusion compared with the former. The mass containing solid and lipid densities compressed the superior vena cava. (Fig. 2). Thoracic computed tomography angiography revealed that the mass originated from the anterolateral portion

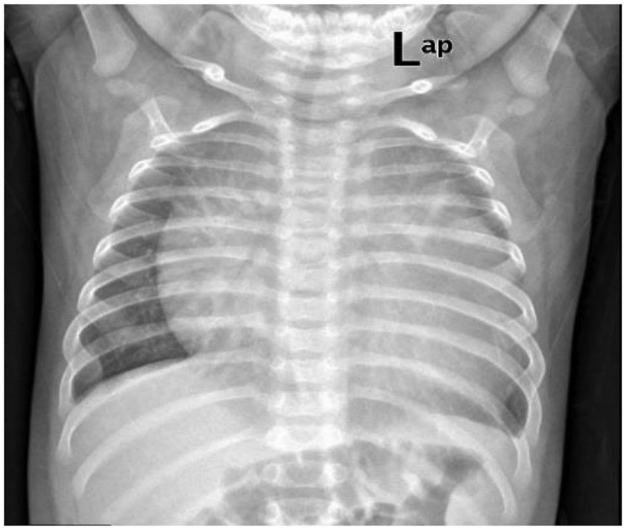

Fig. 1 Chest X-ray image showing cardiomegaly

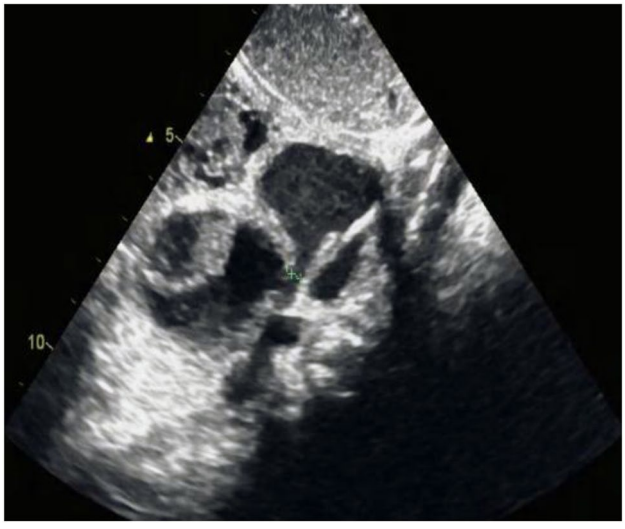

Fig. 2 Two-dimensional colour Doppler echocardiogram showing a tumour that compresses the superior vena cava

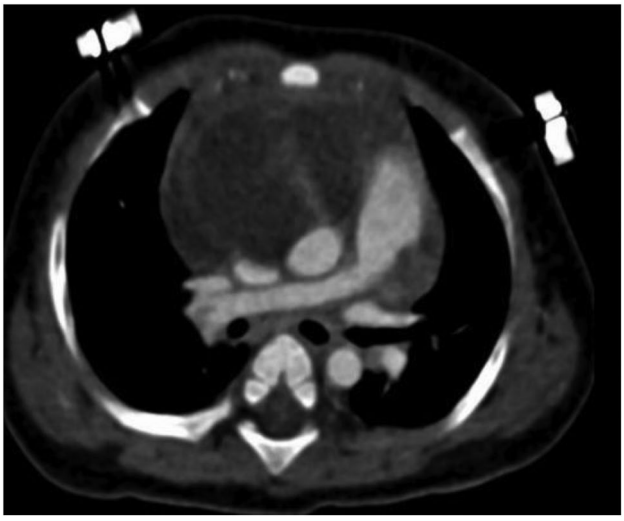

Fig. 3 Thoracic computed tomography angiography image showing the tumor that displaces the superior vena cava, ascending aorta, and pulmonary trunk

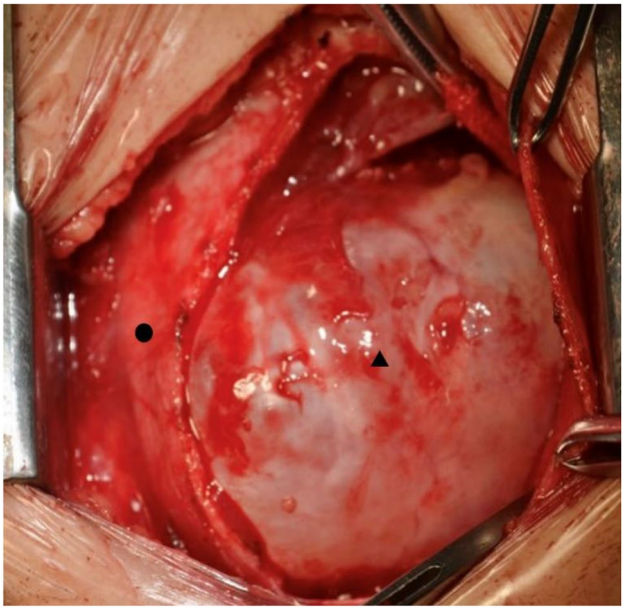

Fig. 4 Intraoperative aspect of the tumor covering the heart surface.(●: pericardium, ▲: pericardial teratoma)

of the ascending aorta. To alleviate the patient's condition and relieve the superior vena cava compression, we have decided to perform surgical intervention. Median sternotomy and pericardiotomy revealed a large encapsulated mass, 6.0 × 5.5 × 4.0 cm in size, occupying a major portion of the pericardial space. The mass was primarily connected to the aorta, obviously compressing the superior vena cava and displacing the right atrium posteriorly (Fig. 3). A portion of the adventitia of the aorta was resected along with the mass. The excision was macroscopically completed with no damage to other structures without cardiopulmonary bypass (Fig. 4). Histopathological examination revealed a mature teratoma. The appearance of the superior vena cava obstruction progressively disappeared, and the baby experienced an uneventful postoperative course. He was extubated on postoperative day one and discharged from the hospital on postoperative day five. The baby was subsequently followed in the surgical and cardiology clinic at 3 months and 1 year after the procedure, and there was no evidence of recurrence. The echocardiogram performed 3 months and 1 year after surgical excision revealed good ejection fraction, no pericardial mass, and no presence of effusion (Fig. 5).

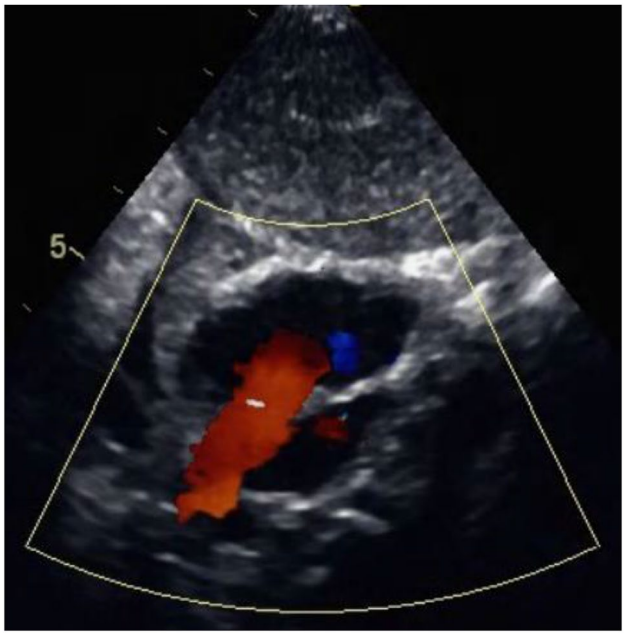

Fig. 5 The two-dimensional color Doppler echocardiography presented that the blood flow through the superior vena cava can smoothly pass through the atrial defect

Discussion and conclusion

Superior vena cava obstruction is characterized by edema of the head, neck, and upper extremities because of increased venous pressure in the upper body. Obstruction can be caused by external compression of the SVC or thrombosis of blood within the SVC [5]. Almost 75–80% of cases of SVC obstruction in published modern series are due to lung cancer, with bronchogenic carcinoma being the most common cause, followed by nonhodgkin lymphoma [6]. The rare causes of this syndrome include mediastinal goiter, colon carcinoma and metastatic hepatomegaly [7–9]. Almost all cases of SVC obstruction have been reported in adults, rarely in infants. We report an unusual case of superior vena cava obstruction caused by an intrapericardial teratoma in an infant [6].

Intrapericardial teratoma is a rare form of primary cardiac tumor that occurs in 5 to 6 out of 10,000 children [10]. Most clinical findings of intrapericardial teratomas are either due to large pericardial effusion or due to the mass effect of the tumor. Inrapericardial teratomas are usually right-sided masses located close to the aorta, superior vena cava, right atrium, or pulmonary artery. They are typically attached to one of the great vessels via a pedicle [11]. Macroscopically, masses are generally polycystic with areas of liquid and solid tissue. The size of the masses ranges from a few millimetres to 15 centimetres [12]. In our patient, this mass was located between the superior vena cava and the aorta and originated from the anterolateral portion of the ascending aorta. This mass is polycystic with areas of more liquid tissue and floats by massive pericardial effusion so that it escapes attention because of weakened cardiac compression and is not displayed on ultrasound at the first time, except for pericardial effusion. After diuretic therapy, this mass drops following effusion reduction and compresses the superior vena cava immediately so that puffiness of the face presents unexpectedly. This mass is incidentally revealed by echocardiography and thoracic computed tomography angiography. Echocardiography may not be able to demonstrate the exact relationship of the tumor with vascular structures. However, it is often the first investigation in the diagnostic work-up but needs to be followed by thoracic computed tomography angiography. On thoracic computed tomography angiography, the mass location, extent, and relationship with the vascular structures, to which they are commonly adherent, are very well depicted [13]. In our case, echocardiography helped us investigate this mass first while superior vena cava obstruction appeared. Thoracic computed tomography angiography revealed that this mass was located between the superior vena cava and the aorta and originated from the anterolateral portion of the ascending aorta, which is useful for performing surgical intervention.

The majority of these tumors are benign, usually without any associated cardiac malformation. There is general agreement that, soon after discovery, tumor removal is needed immediately for hemodynamic reasons [2]. Incomplete resection can lead to pericardial effusion with hemodynamic consequences that can (in some cases) be life threatening [14]. A median sternotomy is the procedure of choice to achieve complete resection. The surgical procedure for removal is usually uncomplicated. Although the literature supports resection without the use of cardiopulmonary bypass, in some cases, it has been used. Because the vascular supply is derived from the adventitial vessels of the aorta [15], there is a small risk of massive hemorrhage from the aorta during dissection. The outcome of intrapericardial teratomas is favourable after complete surgical resection. In our patient, the tumor was removed completely via median sternotomy without the use of cardiopulmonary bypass. A portion of the adventitia of the aorta was resected along with the mass to avoid incomplete resection. No evidence of recurrence was found 1 year after the procedure.

Abbreviations

SVC Superior Vena Cava

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Author contributions

WH-concept and figure preparation, GK-manuscript writing and data collection. All authors read and approved the fnal manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
This study was approved by the Ethics Committee of Children's Hospital Affiliated Shandong University (Jinan Children's Hospital)(approval number: SDFE-IRB/T-2024058).

Consent for publication
The patient's parent provided written consent to use clinical information for scientifc publications.

Competing interests
The authors declare no competing interests.

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