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CASE REPORT

Primary cardiac congenital yolk sac tumor - case report and literature review

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Abstract

Intracardiac tumors are rare, especially in the neonatal period. We present an infant of 1-year and 11 months old with a diagnosis of an intracardiac antenatal tumor, along with literary review on the topic. The referred tumor evolved rapidly with metastases at the time of diagnosis. A biopsy of a pulmonary metastatic lesion was performed, which showed germinative cells tumor (GCT). The patient underwent intensive care treatment, and is now in good condition, following the proposed treatment with chemotherapy for TCG.

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INTRODUCTION

Cardiac neoplasms are rare, with prevalence rates of 0.001 to 0.3% in autopsy series[1]. Intracardiac tumors have been known since medieval times, remaining a diagnostic and therapeutic challenge[2]. There are no large studies on the subject, most being case reports, particularly in malignant tumors[2].

Most cardiac tumors are benign (75%), followed by metastatic involvement[2,3]. In the pediatric age group, the most common primary cardiac tumors are rhabdomyomas, unlike in adults, where atrial myxoma is predominant[1,2]. The incidence of primary cardiac neoplasm in pediatrics has increased annually[3].

The clinical picture depends on the location of the tumor and its relationship with the dimensions of the cardiac chambers and adjacent structures. Patients can range from asymptomatic to overt heart failure with arrhythmias and thromboembolism[1].

Primary and secondary malignant neoplasms of the heart have a poor prognosis, requiring a multidisciplinary approach[1].

Given the rarity and unique presentation of this form of tumor, we present a case in order to enrich the discussion of the topic. We describe the clinical, radiological and laboratory findings in the investigation of congenital cardiac tumors. For a literature review, a search was performed in the Pubmed database in January 2019. The search for "congenital cardiac tumor" yielded 1778 articles, for " malignant primary pediatric cardiac tumor" resulted in 69 studies, and for " primary cardiac yolk sac tumor" others 14. We have refined the search to " best matches" and latest articles.

CASE REPORT

Primigesta, at the 28th gestational week, underwent obstetric ultrasound in prenatal care at the Hospital das Clínicas, Faculty of Medicine, University of São Paulo (HC-FMUSP). Fetal examination revealed the presence of a cardiac mass measuring approximately 2.5x2.4cm (5.35cm³) in the interventricular septum, which was initially diagnosed as a cardiac rhabdomyoma.

After birth, seeing an asymptomatic infant and clinical examination with a slight murmur (2+/6+), conservative treatment was chosen, without surgical intervention, with periodic consultations and imaging tests for follow-up. A beta-blocker was started, as the mass invaded both ventricles, reducing their volume, but without outflow tract obstruction. At around 1 year of age, an echocardiogram (Figure 1) performed in the emergency room due to dyspnea showed an increase in intracardiac tumors (5.7x5.4x5.1cm), as well as obstruction of the inflow and outflow tracts of the right ventricle. Computed tomography of the chest was performed (Figure 2) showing the appearance of multiple lesions, most with signs of necrosis, located in the lungs and mediastinum. A percutaneous biopsy of a pulmonary nodule was performed.

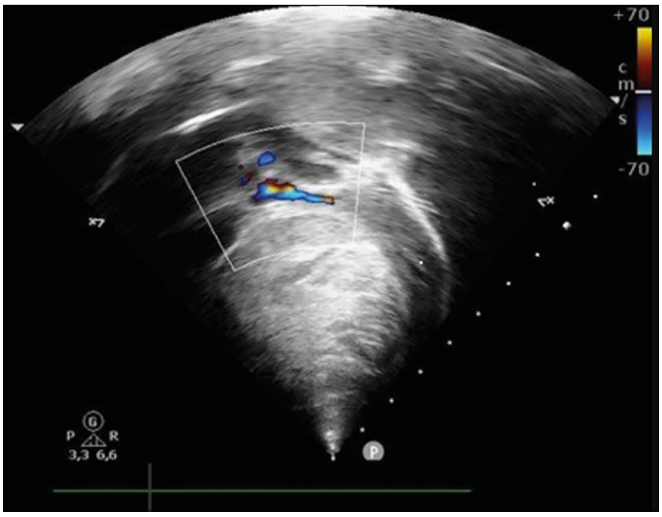


Figure 1. Echocardiogram Caption: Presence of a mass in the ventricular septum of heterogeneous content . Note obstruction to ventricular outflow.

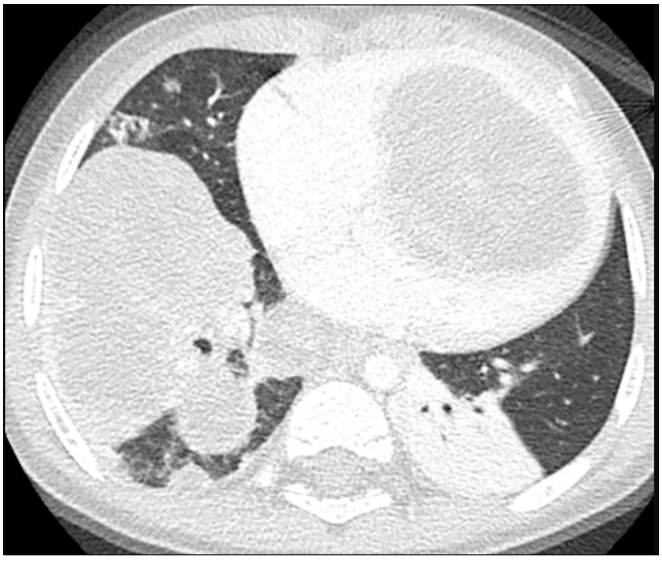


Figure 2. Chest CT Caption: Cardiac lesion best seen in LV, with pulmonary masses on the right; also note atelectasis in left lower lobe.

Histopathological analysis with hematoxylin and eosin staining revealed a neoplasm composed of tubulopapillary structures and anastomosed cords against a background of loose myxoid stroma . The cells had eosinophilic cytoplasm and enlarged nuclei, with frequent mitotic figures. Immunohistochemical stains were diffusely positive for SALL-4, high- fetoprotein and placental alkaline phosphatase. Neoplastic cells were also focally positive for OCT4, CDX2, and low molecular weight cytokeratins (CAM 5.2). Stains for S-100, CD30, CD117 (C-KIT), WT-1, TTF-1 and calretinin were negative. Subsequent clinical investigation revealed a serum alpha-fetoprotein level of 22,470 ng /ml. The diagnosis of congenital cardiac primary yolk sac tumor was performed, starting at this time protocol treatment with chemotherapy (Ifosfamide , cisplatin and etoposide) for GCT (Germ Cell Tumors – GALLOP 2017 GCT protocol)[1].

At the present time, the patient is still under treatment, in good general condition and excellent evolution, with a significant reduction in cardiac tumor (4x3 cm without outflow tract obstruction) and alpha-fetoprotein (16.1 ng /ml).

DISCUSSION

Primary cardiac tumors are rare, especially in the neonatal period, and most cases correspond to rhabdomyomas, teratomas and fibromas[4,5]. Mediastinal tumors, particularly GCTs (Germ Cell Tumors), are extremely rare and are usually located in the anterior mediastinum[1]. They correspond to 6 to 18% of tumors in this location. They present peaks of incidence in childhood and after 10 years of age, with a predominance of males in adolescence[1].

Congenital primary cardiac tumors are mostly benign, corresponding to rhabdomyomas[6]. When malignant, they are mostly represented by sarcomas[4]. The present case is a congenital embryonic germ cell tumor (yolk sac), which makes it relevant due to its histological rarity and location of presentation. It is important to emphasize that the detection of arrhythmia in the antenatal period makes fetal echocardiography mandatory in an attempt to make an early diagnosis of cardiac abnormalities[1]. In this case, the tumor was a routine examination finding. Most pediatric tumors may show volume reduction with advancing age, as they are benign[3]; This fact justifies and scientifically supports the conservative conduct in the reported initial care.

Clinically, such lesions may present with nonspecific symptoms such as fever, weight loss, cough and night sweats. Depending on their location, they can cause pericardial effusion (more common in lymphoma), vena cava syndrome, or more severely heart failure, arrhythmias (due to infiltration in the neural circuit or myocardium) and embolisms. There have been reports of intracardiac endodermal sinus tumors with syncope and epileptic manifestations[1,2,3,7]. Furthermore, cardiac tumors may be associated with genetic syndromes such as Carney syndrome , neurofibromatosis , tuberous sclerosis (particularly rhabdomyomas)[1], Gorlin-Goltz and Beckwith-Wiedemann syndrome[1].

The echocardiographic examination is the first step in obtaining the diagnosis, followed by computed tomography or cardiac resonance. PET CT showed a sensitivity of 90% in differentiating benign from malignant tumors in the literature. Angiography is important to rule out coronary stenoses[2].

Cardiac tumors such as rhabdomyomas generally do not require resection given their spontaneous involution; however, the best therapeutic regimen for cardiac tumors has not yet been established, due to the low number of cases. Surgical procedures with simple or complex resection (assisted by Fontan circulation, when the right side of the heart is affected), the use of an artificial heart and even heart transplantation may be indicated. Benign tumors can be treated with surgical resection, but interdisciplinary cooperation between oncologists, cardiologists, surgeons and radiologists must be joint, especially for malignant tumors[2].

Surgical results for benign tumors show a good prognosis, with low recurrence rates, while malignant tumors remain poor, with death from metastasis[2]. A recent Chinese study showed a median survival of 9 months for malignant tumors from diagnosis[2,3]. In malignant GCT-type tumors, however, the overall survival is 60 to 88%, demonstrating that mediastinal GCTs have a favorable prognosis and do not differ from germ cell tumors at other sites[1].

CONCLUSION

The uniqueness of the case is presented not only by the rarity of the tumor, GCT, intracardiac , in an infant, but also by the difficult clinical management and multidisciplinary approaches of both Oncology and the Intensive team. We hope that this study can also be discussed and evaluated by colleagues who will find similar cases, considering this hypothesis as a cardiac tumor.

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