Available online at www.rajournals.in

RA JOURNAL OF APPLIED RESEARCH
ISSN: 2394-6709
DOI:10.31142/rajar/v5i2.01
Volume: 05 Issue: 02 February -2019

International Open Access
ICV- 74.25
Impact Factor :5.985
Page no.- 2269-2273

A Case Report of the Largest Right Ventricular Hémangioma

Y. Kettani[1], O. Louizi[1], S. Hallab[1], N. Doghmi[1], M. Cherti[1] A. Saoud[2], A. Bakkari[2], Y. Elfakir[2]

[1.] Cardiology B service, Ibn Sina hospital
[2.] Radiology service, Maternity hospital, UH of Ibn Sina

ARTICLE INFO

Published Online:
03 February 2019

Corresponding Author:
Y. Kettani

ABSTRACT

Background: Primary cardiac tumors are rare and often asymptomatic or present with unspecific symptoms. Benign cardiac tumors of vascular origin are especially rare, with only few existing data in the literature.

Case presentation: A 26-year-old woman presented cardiac symptoms like palpitations four month earlier; he started complaining of fatigue and mild exercise dyspnea. His cardiologist discovered a systolic murmur. Echocardiography showed a large right ventricular mass, which engaged in the tricuspid valve in hoes generating a transtricuspidien gradient of 15 mmHg, Our patient then underwent cardiovascular magnetic resonance (CMR) imaging and a thoracoabdominopelvic CT scan the day after admission, which revealed that there isn't enormous than this mass. The surgeons were preparing to operate the patient but the patient's biological assessment was disrupted, and 2 days later a cardiogenic shock state requiring administration of dobutamine without improvement.
The evolution was marked by the installation of a severe bradycardia with impaired consciousness with a GCS 3 having motivated the family to take agonizing to his parental home, the patient unfortunately died on his arrival home.

Conclusion: We conclude that MRI is advantageous over a combination of TTE and TEE for the detection and complete morphological and functional evaluation (hemodynamic effects) of cardiac masses.

KEYWORDS: CMR, Cardiac Tumor, Echocardiography, Intracardiac angioma

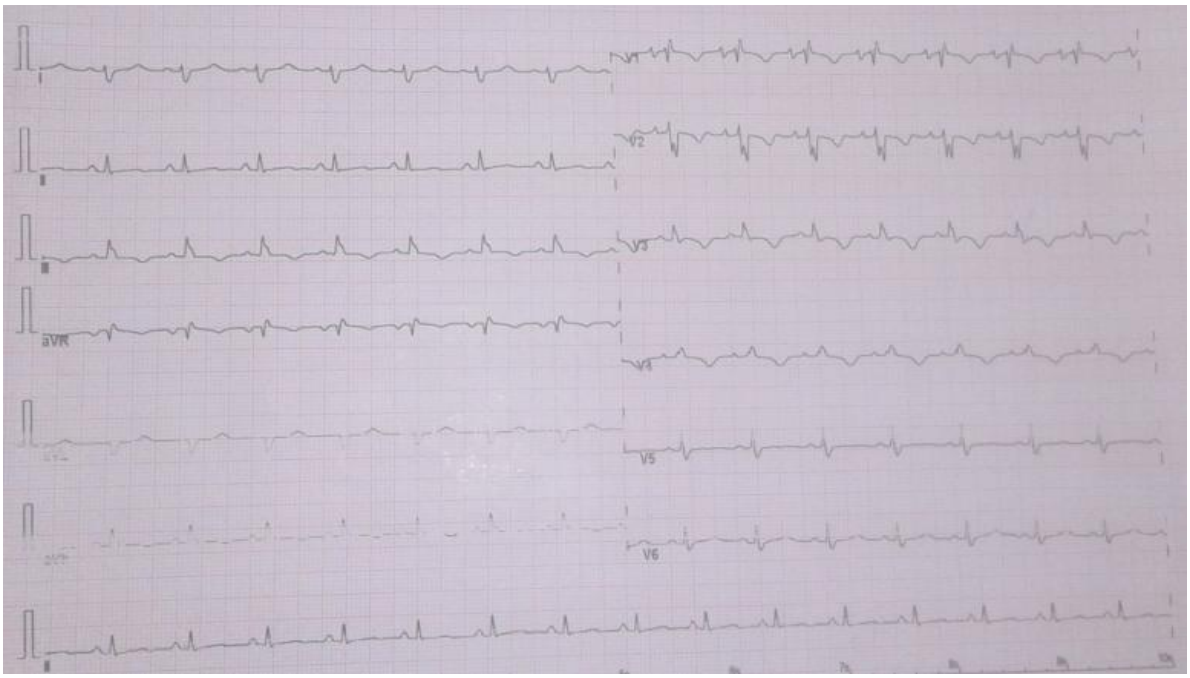
Background

Primary cardiac tumors are rare and their incidence ranges from 0.0017 to 0.28 % as reported in autopsy studies [1]. the most common benign tumors of the heart are myxomas, followed by lipoma, papillary fibroelastoma, Angioma, fibroma, hemangioma, rhabdomyoma, and teratoma. Only about 5 % of all benign cardiac tumors are angiomas [2], and diagnosing them is often difficult. Many of the primary cardiac tumors are asymptomatic and are detected postmortem. If these tumors are symptomatic, embolization, obstruction, and arrhythmogenesis are the major modes of presentation [2]. Large cardiac tumor obliterating the right ventricular cavity was not commonly described. Therefore we present the largest Right Ventricular Hémangioma with spleen localization.

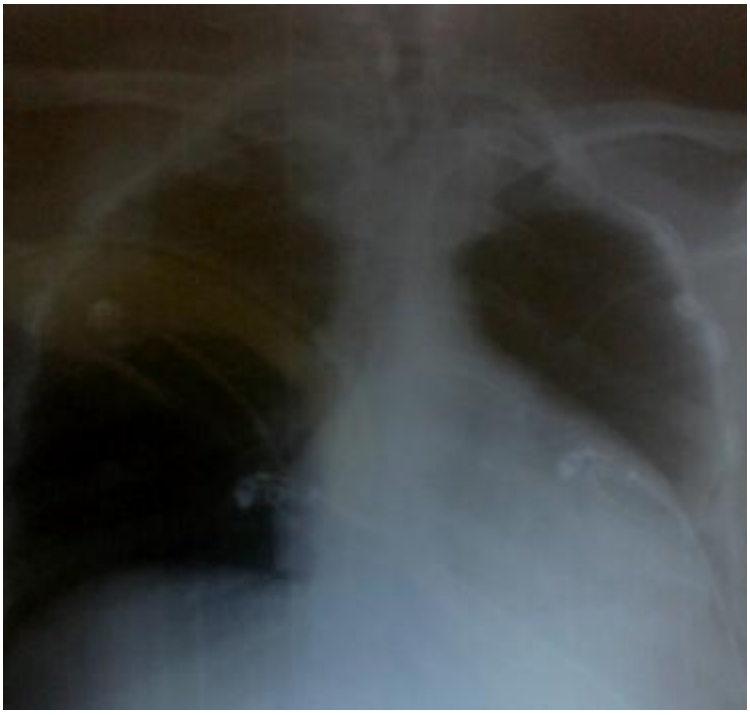
Case presentation

A 26-year-old woman presented cardiac symptoms like palpitations four month earlier; he started complaining of fatigue and mild exercise dyspnea. His cardiologist discovered a systolic murmur. Echocardiography showed a large right ventricular mass.

On physical exam, the patient had stable vital signs. Neck veins were not distended. Breath sounds were decreased in bilateral lung bases. She had a prominent left parasternal heave, and her apex beat was displaced to the sixth intercostals space (ICS) at the left anterior axillary line. Heart sounds were distinct with a regular rhythm. There was a grade 2/6 mid-to-late systolic murmur at the second ICS of the left parasternal border (LPSB) and a grade 3/6 holosystolic murmur at the fourth to fifth ICS of the LPSB. A fluid wave was elicited on the abdominal exam. She hadn't cyanosis or clubbing of the extremities. There were no noted skin lesions or abnormal pigmentation.
A 12-lead electrocardiogram showed T-wave inversion in the inferior and precordial leads and signs of right ventricular hypertrophy (Fig 1).



Although the pleural effusions on the chest radiograph make it difficult to delineate the cardiac silhouette, the lack of prominent vascular markings taken with the seemingly rounded apex and prominent right cardiac border point to a predominantly right-sided pathology. (Figure2)



Transthoracic echocardiography revealed a large heterogeneous mass in her slightly dilated right ventricle (Figure 3, 4, 5), with a large implantation base on the free wall of the right ventricle and which engaged in the tricuspid valve in hoes generating a transtricuspidien gradient of 15 mmHg(Figure 6), Her cardiac valves were normal without stenosis or regurgitation, and the size and function of her left ventricle were normal. A small, not significant pericardial effusion was also detected. The mass was easily visible on the parasternal long and short axis (level of aortic valve) views, measuring 6,4 x 6,0 cm (Figure 7), prolapsing or plopping motion of the mass through the PV and into the main pulmonary artery (MPA) during systole and into the right atrium. Also consistent with the previous TTE, the right-sided chambers were dilated (transverse diameters of 52 mm for the RA and 42 mm for the RV), with evidence of depressed RV function.

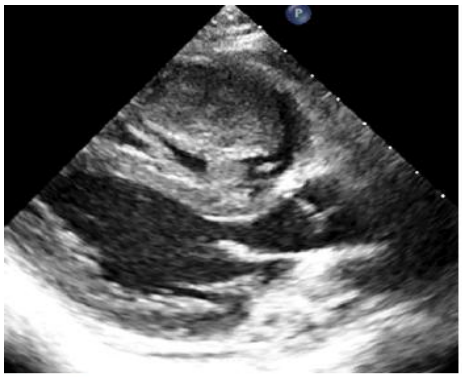


Fig.3

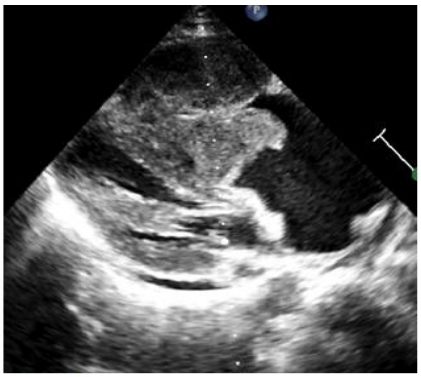


Fig.4

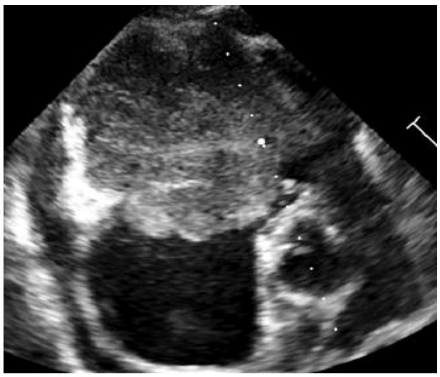


Fig.5

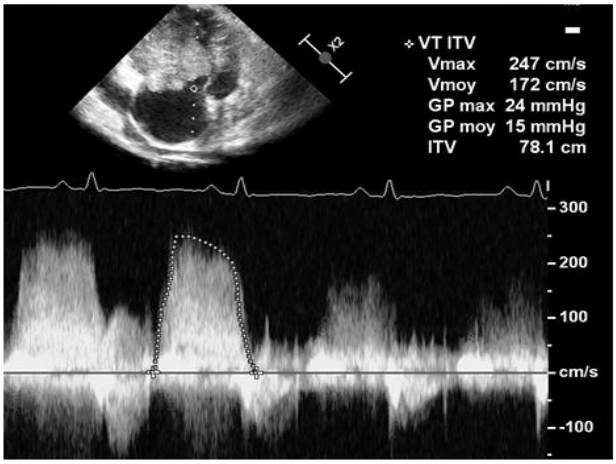


Fig.6

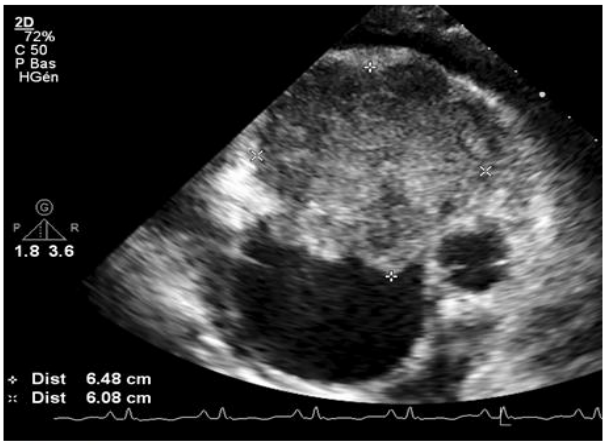


Fig.7

Our patient then underwent cardiovascular magnetic resonance (CMR) imaging (Figure 8, 9) and a thoracoabdominopelvic CT scan (Figure 10, 11) the day after admission, which revealed a 64 × 62 mm right ventricular mass with an important implantation base at the floor and a transtricuspidien extension in the right atrium with heterogeneous enhancement usual found in hemangiomas.

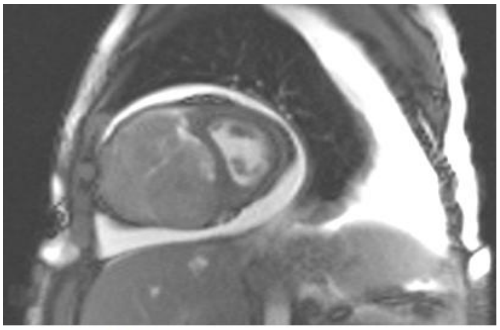


Fig. 8: CMR

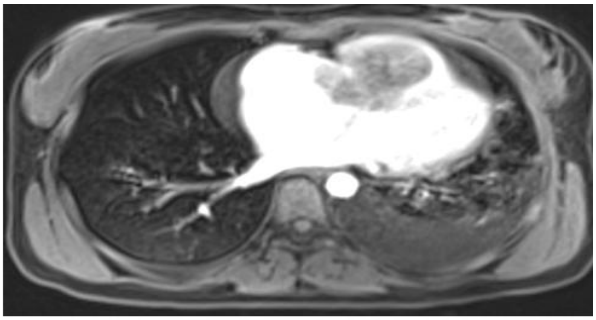


Fig. 9: CMR

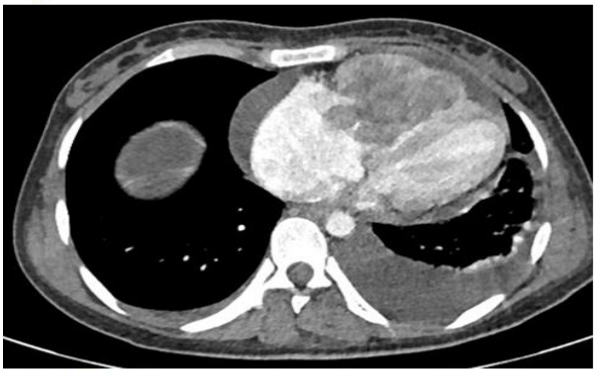


Fig 10: CT scan

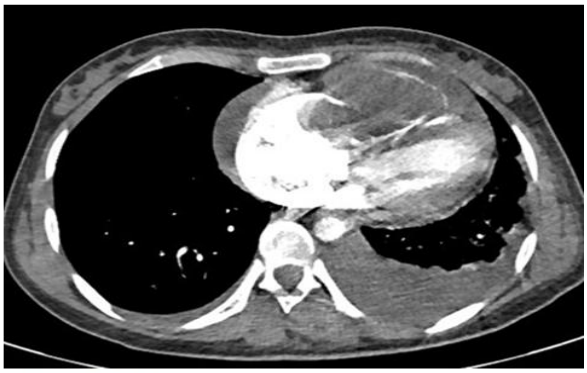
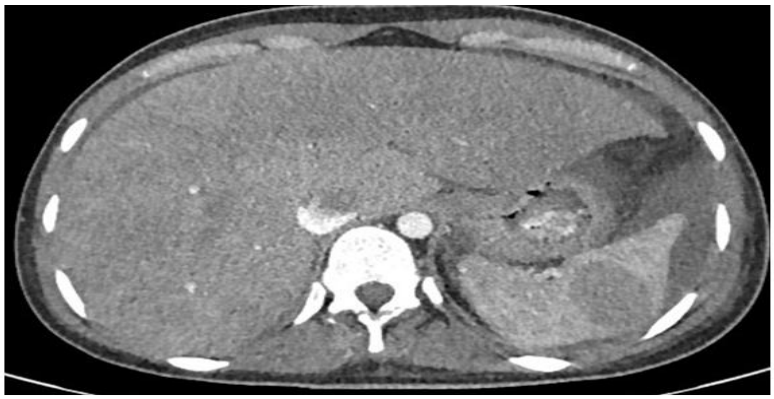
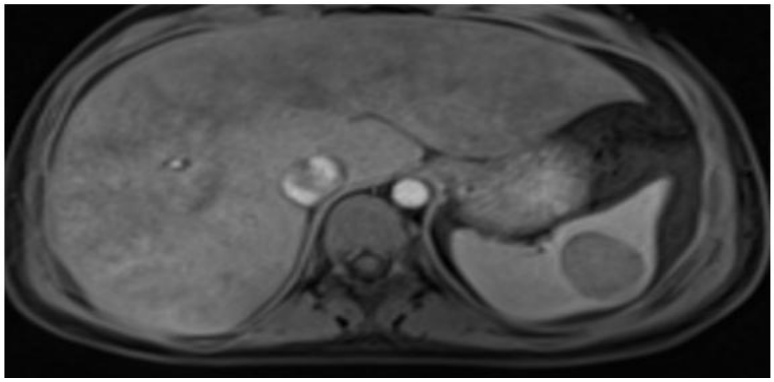


Fig 11: CT scan

Magnetic resonance (CMR) imaging and thoracoabdominopelvic CT scan (Figure 12, 13) shows second spleen localization. A hepatomegaly of the turgor type secondary to an infusion disorder related to the hyperpression of the right cavities.



(Figure 12)



(Figure 13)

The surgeons were preparing to operate the patient but the patient's biological assessment was disrupted, with hepatic cytolysis (26X normal) and a very impaired crase assessment (TP = 10%).

Surgery is referred to until its condition is stabilized, but the patient presented 2 days later a cardiogenic shock state requiring administration of dobutamine but without improvement.

The evolution was marked by the installation of a severe bradycardia with impaired consciousness with a GCS 3 having motivated the family to take agonizing to his parental home, the patient unfortunately died on his arrival home.

Discussion

Hemangiomas are vascular tumors composed of blood vessels that can be either capillaries (capillary hemangiomas) or large cavernous vascular channels (cavernous hemangiomas). Any organ can be involved by these tumors, but most frequently they are localized to the skin and subcutaneous tissues [3]. Their localization to the heart is extremely rare. Twenty-three cases of cardiac hemangioma have been described in the literature [4-15]. Cases diagnosed at autopsy are not included. There were 16 female and 7 male patients. Ages ranged from 3 to 63 years (mean age, 23 years). Nine patients (40%) were under the age of 15 years [4, 7, 8,9,10, 11, 12, 13, 14].The most frequent clinical presentation was dyspnea on exertion (10123, 43%); less frequently patients presented for arrhythmias (423, 17%), pseudoangina (3123, 13%), signs of right heart failure (3/23, 13%), pericarditis or pericardial effusion (2/23, 8%), and failure to thrive (2/23, 8%). Two patients (2/23, 8%) were totally asymptomatic. Systolic murmur mimicking pulmonary stenosis was noted in 5 cases (21%). Our patient presented for dyspnea on exertion and hepatalgia. Another interesting clinical setting in which cardiac hemangioma may be observed is the Kasabach-Merritt syndrome, which is characterized by multiple systemic hemangiomas associated with recurrent thrombocytopenia and consumptive coagulopathy. Cardiac involvement in this syndrome has been recently reported [24]. Diagnosis of cardiac hemangiomas was always difficult to establish. Whereas in 17 cases (17/23,74%) an abnormal chest roentgenogram was noted at the initial work-up, the diagnosis of cardiac tumor and more particularly of cardiac hemangiomas was consistently delayed and required further multiple investigations

Echocardiography was performed in 11 patients (11/23, 48%). This examination showed a cardiac tumor in 9 of 11 (81%). In 2 patients (18%) Myxomas, the most common cardiac tumor in adults, are usually located in the left atrium (75%). Only 5.0% are located in the ventricles, but there have been several case reports of large myxomas leading to RVOT obstruction.2,5,7,8,9,19 Fibromas, while more common in children compared to adults, are typically located in the ventricles and can also lead to RVOT obstruction.11,12 Cardiac hemangiomas are rare,

comprising only approximately 2.8% of the already limited subset of primary cardiac tumors. They are comprised of benign proliferations of endothelial cells, and are histologically similar to hemangiomas found in the skin, subcutaneous tissues, and other parts of the body.14 Specific morphology can vary, including capillary, cavernous, and arteriovenous forms, with most hemangiomas containing a mix of these subtypes.15 In a review of 23 cases, Brizard et al.16 noted that the most frequent clinical presentation was dyspnea on exertion (43%). Signs of right heart failure and a pulmonary stenosis murmur were only encountered in 13% and 21%, respectively.

The epicardium is the most common location for these tumors, but they can also be located in the myocardium and endocardium.[11] In terms of the involved cardiac chamber, Kojima et al. determined that majority (36%) are found in the right ventricle,[12] but only a few cases in the world literature had significant RVOT obstruction.[11, 12, 13, 14, 15]

While echocardiography is still the diagnostic modality of choice to screen for intracardiac tumors, other tests may provide additional useful information. A cardiac MRI sometimes demonstrates the light bulb sign on T2-weighted images, which is due to a higher signal coming from the sluggish blood flow within the vascular tumor.[15]

Complete resection is the treatment of choice, although some hemangiomas may be deemed no resecable due to anatomical considerations such as extensive involvement of the coronaries. There was one reported case of spontaneous resolution of the mass.

Conclusion

We conclude that MRI has a definite advantage over a combination of TTE and TEE in the imaging evaluation of cardiac masses. It is better suited for suggesting the etiology, delineating the extent, the relationship to adjacent structures, and the presence of any hemodynamic effects or associated extra cardiac lesions.

Conflicts of interest and funding

All authors have none to report. The authors have not received any funding or benefits from industry or elsewhere to conduct this study.

Reference

1 Reynen K. Frequency of primary tumors of the heart. Am J Cardiol. 1996;77:107.

2 Shapiro LM. Cardiac tumors: diagnosis and management. Heart. 2001;85:218

3 Yan XG, Jia B, Zhu BX, Hu XH. Successful resection of a primary cardiac fibroma in a neonate: report of a case. Surg Today. 2014 Sep;44(9):1768-70.

4 Gopal AS, Stathopoulos JA, Arora N, Banerjee S, Messineo F. Differential diagnosis of intracavitary tumors obstructing the right ventricular outflow tract. J Am Soc Echocardiogr. 2001 Sep;14(9):937-40.

5 Reiss N, Theissen P, Feaux de Lacroix W. Right-ventricular hemangioma causing serious outflow-tract obstruction. Thorac Cardiovasc Surg. 1991 Aug;39(4):234-6.

6 Iba Y, Watanabe S, Akimoto T, Abe K, Koyanagi H. Pedicled cardiac hemangioma with right ventricular outflow tract obstruction. Jpn J Thorac Cardiovasc Surg. 2005 May;53(5):269-71.

7 Ainsworth CD, Salehian O, Nair V, Whitlock RP. A bloody mass: rare cardiac tumor as a cause of symptomatic ventricular arrhythmias. Circulation. 2012 Oct 9;126(15):1923-31.

8 El Fawal S, El Metwally EA. Report of a right ventricular outflow tract myxoma in a young adult. J Cardiovasc Magn Reson. 2015; 17(Suppl 1): P374.

9 Gribaa R, Slim M, Kortas C, Kacem S, Ben Salem H, Ouali S, Neffati E, Remadi F, Boughzela E. Right ventricular myxoma obstructing the right ventricular outflow tract: a case report. J Med Case Rep. 2014 Dec 17;8:435.

10 McAllister H. Tumors of the heart and pericardium. In: Silver MD, editor. Cardiovascular pathology. New York: Churchill Livingstone; 1983. p. 909e43.

11 Sharma J, Hsu D, Weinstein S. Obstructive right ventricular outflow tract hemangioma in an adolescent. Congenit Heart Dis. 2011 Nov-Dec;6(6):657-60.

12 Botha J, Ihlberg L, Elhenawy A, Abbott M, Butany J, Paul N, Brister SJ. A giant cavernous hemangioma of the heart. Ann Thorac Surg. 2010 Jul;90(1):293-5.

13 Moreira D, Rodrigues B, Delgado A, Nascimento C, Nunes L, Nobre A, Cravino J, Santos O. Hemangioma of the right ventricular outflow tract, clinical report. Rev Port Cir Cardiotorac Vasc. 2012 Apr-Jun;19(2):79-81.

14 Soberman MS, Plauth WH. Hemangioma of the right ventricle causing outflow tract obstruction. J Thorac Cardiovasc Surg 1988.