Cardiac Angiosarcoma - Importance of Imaging in Early Diagnosis

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Introduction

This is the case of angiosarcoma resection in young male patient with no evidence of metastasis on routine radiological tests. On admission, his clinical condition was heart failure (HF) functional class (FC) III (NYHA - New York Heart Association). Transthoracic echocardiography (TTECHO) revealed mass with compression of the right chambers and moderate pericardial effusion (PE). PE puncture with laboratory analysis was little enlightening and the final diagnosis was only possible by clinical pathology (CP) and immunohistochemistry (IHC). Although there is no formal clinical indication for computed tomography (CT) of the heart and coronary arteries, it proved crucial for surgical planning. In chemotherapy regimen with Paclitaxel, 12 months after the surgery, the patient progressed to HF FC I (NYHA) and presented normal imaging tests.

Case Report

L.R.O, 17, black, born in São Paulo, with unimportant medical history, admitted to the emergency room with respiratory distress for two years, which worsened in the past 60 days. During that period, the patient underwent many treatments to respiratory tract infections, which turned out unsuccessful. The patient was admitted to another hospital via emergency room 45 days before, undergoing an echocardiography test that revealed moderate PE. He had pericardial puncture followed by drainage and biopsy, which proved to be nonspecific to laboratory and CP tests.

After two weeks of hospital discharge, the symptoms got worse and were associated with fever peaks (38 ºC). The patient was then referred to the ER of our service. The patient was lucid, conscious, pallor (+/3+), no fever, tachypnea (28 ipm), jugular vein distension at 45 degrees (++/3+), mild edema of the lower limbs. Cardiac auscultation was normal except for tachycardia (108 bpm), and lung auscultation revealed diminished breath sounds on the right base. Abdomen with hepatomegaly 3+/4+, painful on palpation in the right hypochondrium.

Keywords

Hemangiosarcoma/surgery; Heart Failure; Echocardiography; Pericardial Fluid/pathology.

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Chest X-ray in the anteroposterior projection showed significant increase in the cardiac area related to the right heart chambers and ECG revealed low QRS voltage in peripheral leads.

Transthoracic echocardiogram (Figure 1) showed an image suggestive of hyperechoic mass, heterogeneous, rounded shape, fixed, with intrapericardial location, measuring 10.3 x 11.5 cm, related to the right atrium and right ventricle, with compressive signs. There were signs of restricted right ventricular filling. Mild pericardial effusion.

Cardiac magnetic resonance imaging (CMRI) (*General Electric* 1.5T) showed a mass originating in the lateral wall of the right atrium extending to the right ventricle, measuring 12.0 x 11.0 x 10.0 cm, causing significant compression of these chambers (Figure 2). It was closely related to the superior vena cava, inferior vena cava, aortic root and right pulmonary veins. Sequences in cine-MRI suggested areas of necrosis and hemorrhage. Gadolinium infusion revealed severe diffuse perfusion (suggesting rich vascularization), however, heterogeneous and with fibrotic areas.

Computed tomography of the coronary arteries conducted on a multi-slice GE scanner with 64 detectors showed rounded heterogeneous hypervascular mass measuring 11.0 x 9.8 x 9.7 cm, determining a compressive effect in

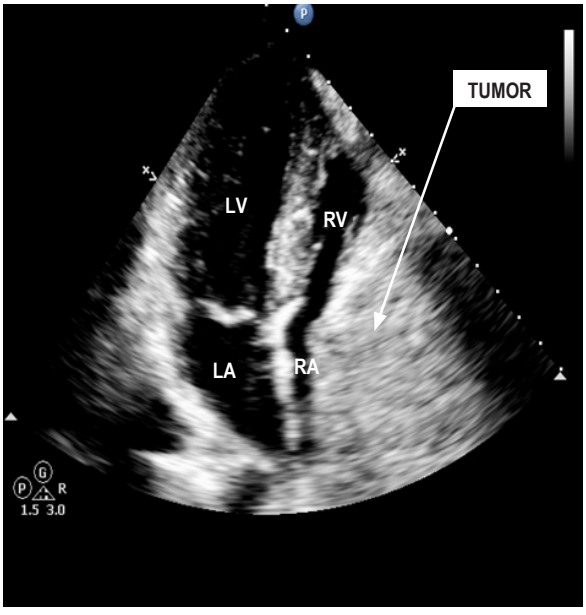

Figure 1 – *TTECHO (apical 4-chamber). LA: left atrium; RA: right atrium; LV: left ventricle; RV: right ventricle.*

the right heart chambers. There was close contact with the pericardium of the right ventricular free wall with no obvious sign of invasion or impairment of the pericardium or coronary artery throughout its path. It was not possible to determine a cleavage plane between the mass and the right atrial wall or the aortic root (Figure 3).

Transthoracic biopsy with thick needle was performed to collect material and analyze the pericardial fluid, and pericardial biopsy was also conducted and proved typical of spindle cell carcinoma. Total abdominal tomography and positron emission tomography (PET) ruled out impairments from a distance.

Due to developments with clinical worsening, exploitative thoracotomy surgery turned out to be imperative. Performed with total circulatory arrest, the tumor that confirmed the CT findings was resected. It presented a wine color, fibro-elastic consistency and no defined cleavage planes (Figure 4). Resection of the right atrial free wall was necessary. Reconstruction with bovine pericardial patch was conducted.

Postoperatively, the patient developed anasarca and hypoalbuminemia, and responded to diuretics. Control pre-hospital discharge TTECHO was normal.

CP showed surgical margins, visceral and parietal pericardium infiltrated by cancer. Based on the tumor CP and IHC results (Table 1), chemotherapy regimen was started with Paclitaxel 150 mg once a week for seven months with good tolerance. Reevaluated four, six, eight, twelve and eighteen months after the surgery, the patient progressed well and without clinical signs of CHF or echocardiographic abnormalities.

Discussion

In primary cardiac tumors, there is a predominance of benign tumors. Histological types and clinical presentations depend on the age group. Only 25% of primary tumors are malignant. Sarcomas are the most prevalent types, accounting for about 50% to 75% of the cases, followed by primary lymphomas of the heart. The most common primary malignant tumors of the heart include angiosarcoma, leiomyosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, undifferentiated sarcoma, fibrosarcoma and malignant lymphoma.[1]

Angiosarcoma is the most common one among primary cardiac sarcomas in adults and accounts for 30% to 37% of the cases. They occur in the age group of 30 to 50, but can occur in any age group. Its general presentation are irregular nodular invasive masses of low attenuation on CT scan, presenting heterogeneous signal intensity and hypervascularization on

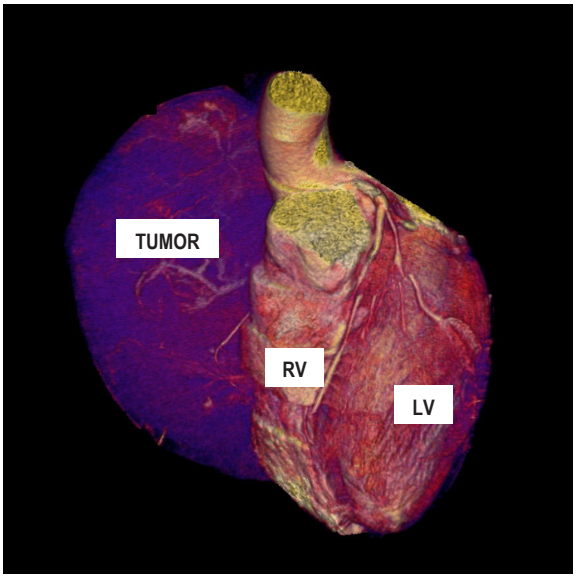

Figure 3 – *CT of the heart and coronary arteries displaying vascularized mass (arrow) and in close contact with the right chambers. PT: pulmonary trunk; LV: left ventricle; RV: right ventricle.*

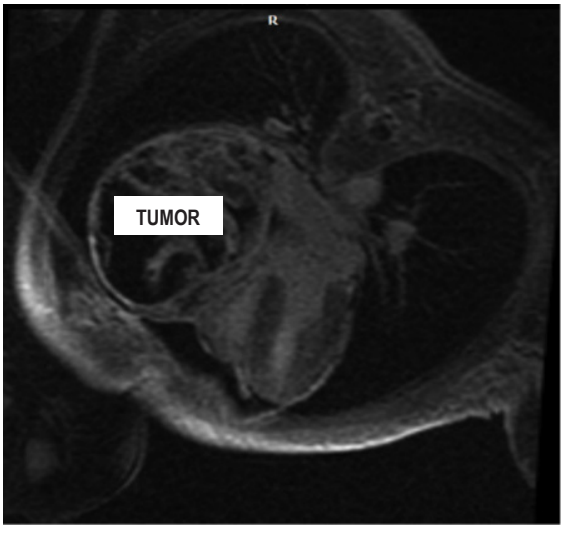

Figure 2 – *Delayed enhancement series in RMC, displaying extensive heterogeneous areas of fibrosis.*

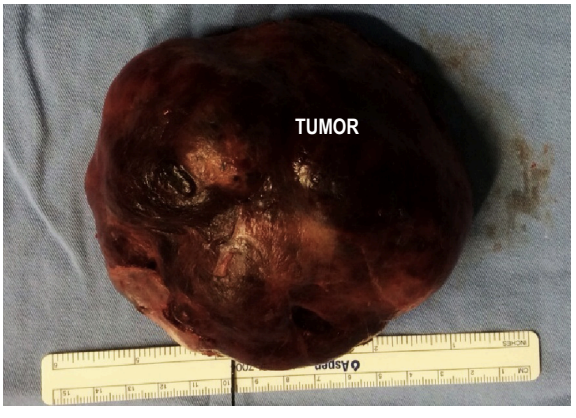

Figure 4 – *Macroscopic aspect of the tumor in surgical sample.*

Table 1 – Immunohistochemistry profile

Antibody	Clone	Interpretation
EMA	E29	Negative in neoplastic cells
EMA	E29	Negative in neoplastic cells
ENOLASE (NSE)	E27	Negative in neoplastic cells
ENOLASE (NSE)	E27	Negative in neoplastic cells
CD99		Negative in neoplastic cells
CD34	QBEnd-10	Positive in neoplastic cells
CD34	QBEnd-10	Positive in neoplastic cells
S100	Policional	Negative in neoplastic cells
S100	Policional	Negative in neoplastic cells
AML	HUC1-1	Negative in neoplastic cells
AML	HUC1-1	Negative in neoplastic cells
HHV8	13B10	Negative in neoplastic cells
HHV8	13B10	Negative in neoplastic cells
CD31	JC70A	Positive in neoplastic cells
CD31	JC70A	Positive in neoplastic cells
MYOD1	5.8A	Negative in neoplastic cells
MYOGENIN	POLYCLONAL	Negative in neoplastic cells
DESMIN	D33	Negative in neoplastic cells
AE1AE3	AE1AE3	Negative in neoplastic cells
KI 67	(30-9)	Positive in 60% of neoplastic cells

resonance, as well as fibrosis and hemorrhage.[1] Invasive methods such as transvenous cardiac biopsy guided by echocardiography may be helpful, but a negative result does not rule out the possibility of angiosarcoma.[2] These generally have distant metastases in 75% of patients at diagnosis, especially for the lungs, thoracic lymph nodes, mediastinum and spine. Average survival after onset of symptoms is 6 to 12 months.[3]

Treatment is based on the tumor resection, which may be partial or total, where possible. When limited to the free atrial wall, atrial septum or a small portion of the ventricle or heart valve, complete resection should be done in an attempt to alleviate symptoms and increase postoperative survival.[4]

In a case report by Tomasa Centella et al.[5], a patient with no evidence of metastases was subjected to left atrial mass resection receiving six cycles of chemotherapy with ifosfamide, Adriamycin and methotrexate. After two years, she had local recurrence and underwent a new excision followed by six cycles of chemotherapy. Death occurred two years after the second operation, and survival of four years.[5] In a case report by Shao-wei Chen et al.[6], a 33-year-old patient with cardiac tamponade and radiological evidence of mass with active bleeding underwent excision surgery and atrial reconstruction using bovine pericardium. Adjuvant therapy with docetaxel and gemcitabine was performed early in the postoperative period. After 14 months, the patient was alive, but with brain and lung metastasis.[6]

Treatment combining many therapeutic types has been reported by Baay et al.[7]. In that report, the patient underwent chemotherapy with doxorubicin, dacarbazine, ifosfamide and mesna, supplemented by radiotherapy, and then transplanted. Two months later, he received two additional courses of chemotherapy with the same drugs and was maintained with long-term cyclosporine and prednisone. The clinical outcome was favorable and metastases were not detected up to 33 months after surgery, showing that aggressive treatment can provide longer remission of cardiac sarcoma.[7]

Early diagnosis of cardiac angiosarcoma in the absence of metastases seems to have been instrumental for the good outcome of the case presented here. The imaging methods proved to be important from the characterization of the tumor to the surgical planning. The presence of coronary involvement by the tumor is always a challenge for the surgeon and CT angiography of the coronary arteries was an indispensable resource in the surgical strategy. The literature provides no consensus on the choice of the chemotherapeutic agent for the present case and, as it seems, the protocol will depend on the individual experience of each service.

Conclusion

This report suggests that early diagnosis of cardiac angiosarcoma associated with chemotherapy may offer additional survival to that found in the current literature and

cardiac angiography is a method to be considered when we need to assess involvement of the coronary arteries.

Authors' contributions

Research creation and design: Rached HRS; Data acquisition: Rached HRS, Soares AA, Aitta R, Nery Jr R, Succi JE, Villiger LEO; Data analysis and interpretation: Rached HRS; Manuscript drafting: Rached HRS, Soares AA; Critical revision of the manuscript as for important intellectual content: Rached HRS, Nery Jr R.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

This study is not associated with any graduate program.

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