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EC CARDIOLOGY
Case Report

Cardiac Angiosarcoma Causing a Massive Pericardial Effusion and Obstruction of the Inferior Vena Cava: Case Report

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Abstract

Primary cardiac malignant neoplasms are rare and occur less commonly than metastatic disease of the heart. However, angiosarcoma represent the most frequent malignant tumor of the heart and 75% of them are in the right heart especially in the right atrium which can provoke many serious complications. Due to the extreme rarity of it there is no great experience in its management. We report a case of a women who was hospitalized in our department for acute dyspnea to whom we discover in the echocardiography a cardiac mass diagnosed later as angiosarcoma and treated with cardiac surgery.

Keywords: *Angiosarcoma; Malignancy; Primary Cardiac Tumors; Cardiac Surgery*

Introduction

Primary tumors of the heart, except for atrial myxomas, are very rare (a prevalence of 0.001 - 0.28%) [1] and most of them are benign. Metastases to or direct invasion of the heart are far more common. Sarcomas are the largest group of primary cardiac malignant neoplasms and have a mesenchymal origin. The most common cardiac sarcomas are angiosarcomas, rhabdomyosarcomas, malignant mesotheliomas and fibrosarcomas [1,2]. Leiomyosarcomas are extremely rare. Unfortunately, most of these tumors have already metastasized to the lung, liver and brain at the time of presentation [3] and each clinical case of this tumor requires documentation of the diagnosis, treatment efficacy, and a comparison with the literature data.

Case Presentation

39 years old female without history of cardiovascular diseases, who was admitted with recent onset of shortness of breath, on examination BP was 110\80 mm hg, regular heart rate at 118 beats\min, there was no abnormal findings on pulmonary auscultation, but heart beats were not audible. Chest-x-ray didn't show any special anomalies and electrocardiogram was normal except for sinus tachycardia at 120 beats\min.

In the echocardiography there was a mass with an heterogenous texture multilobulated that measure 6.5 cm in his high axis which occupies the right atrium and diving in the right ventricle to block the tricuspid valve. There was also a massive pericardial effusion without signs of tamponade (Figure 1-3).

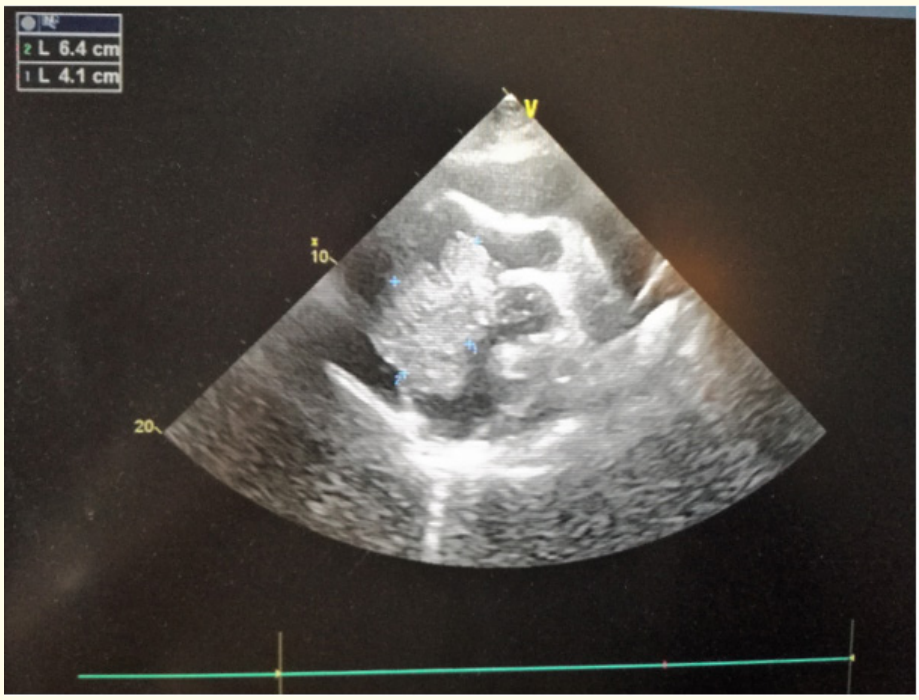

Figure 1

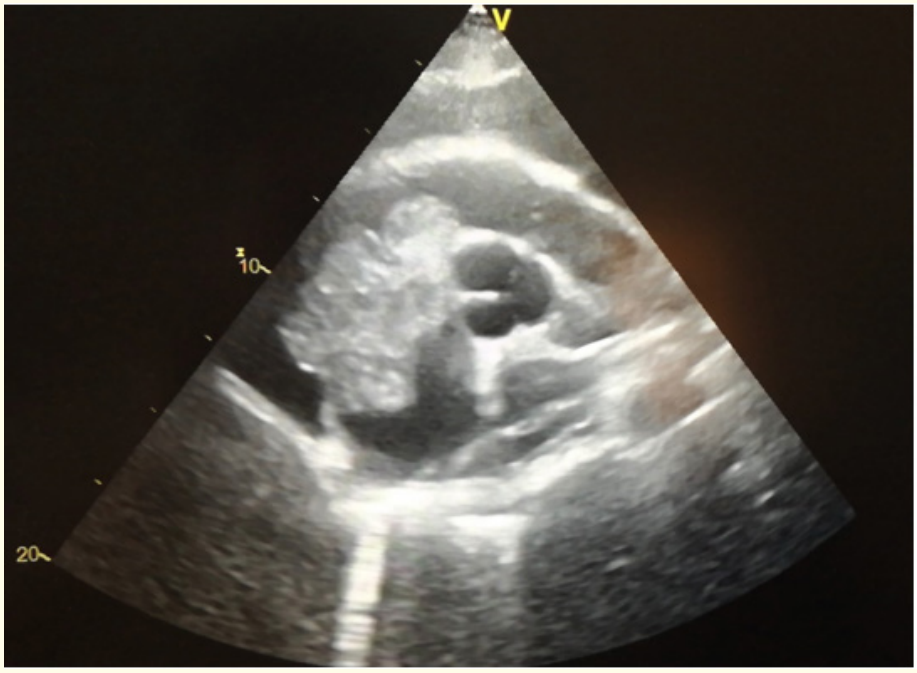

Figure 2

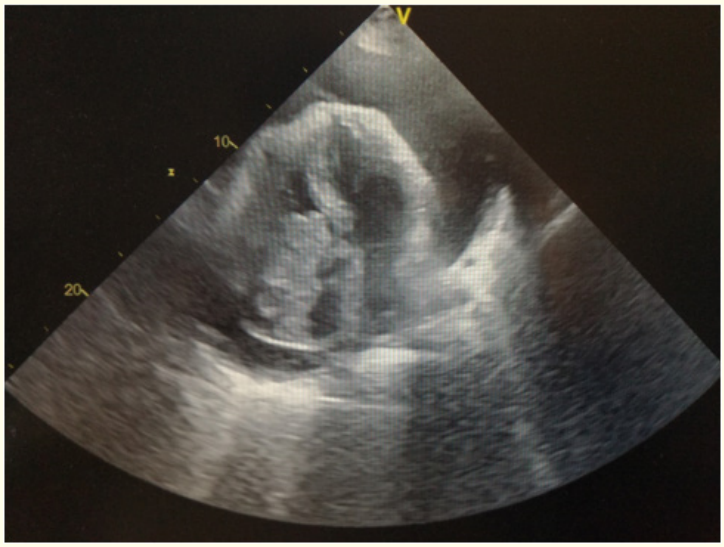

Figure 3

Figure 1-3: *Echocardiographic short axis and apical views visualizing a 6.5 cm multilobulated mass, which occupies the right atrium and dives into the right ventricle (and probably penetrating its free wall). Associate to an important pericardial effusion.*

A CT thoracic scan was performed for further diagnosis has confirmed the findings of the echocardiography which was a mass located in the right atrium extended to the inferior vena cava but did not occlude it (Figure 4 and 5).

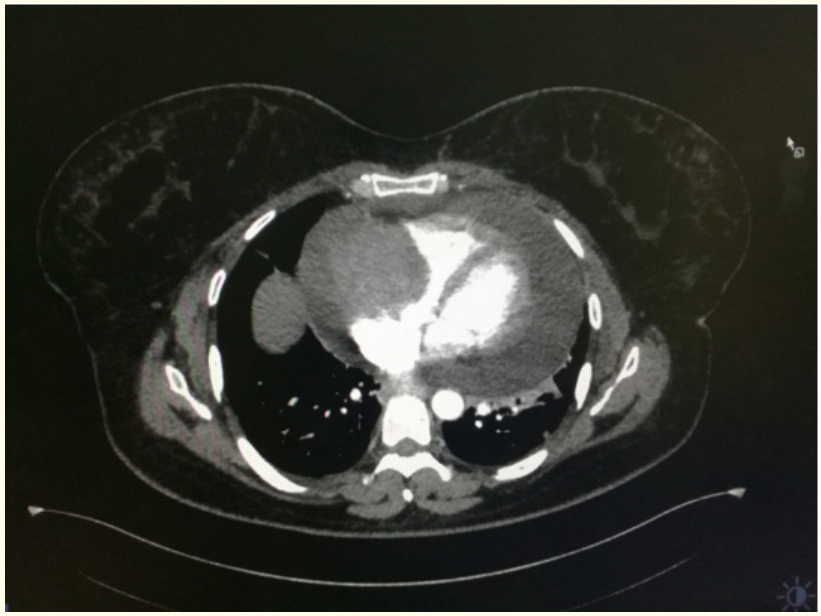

Figure 4

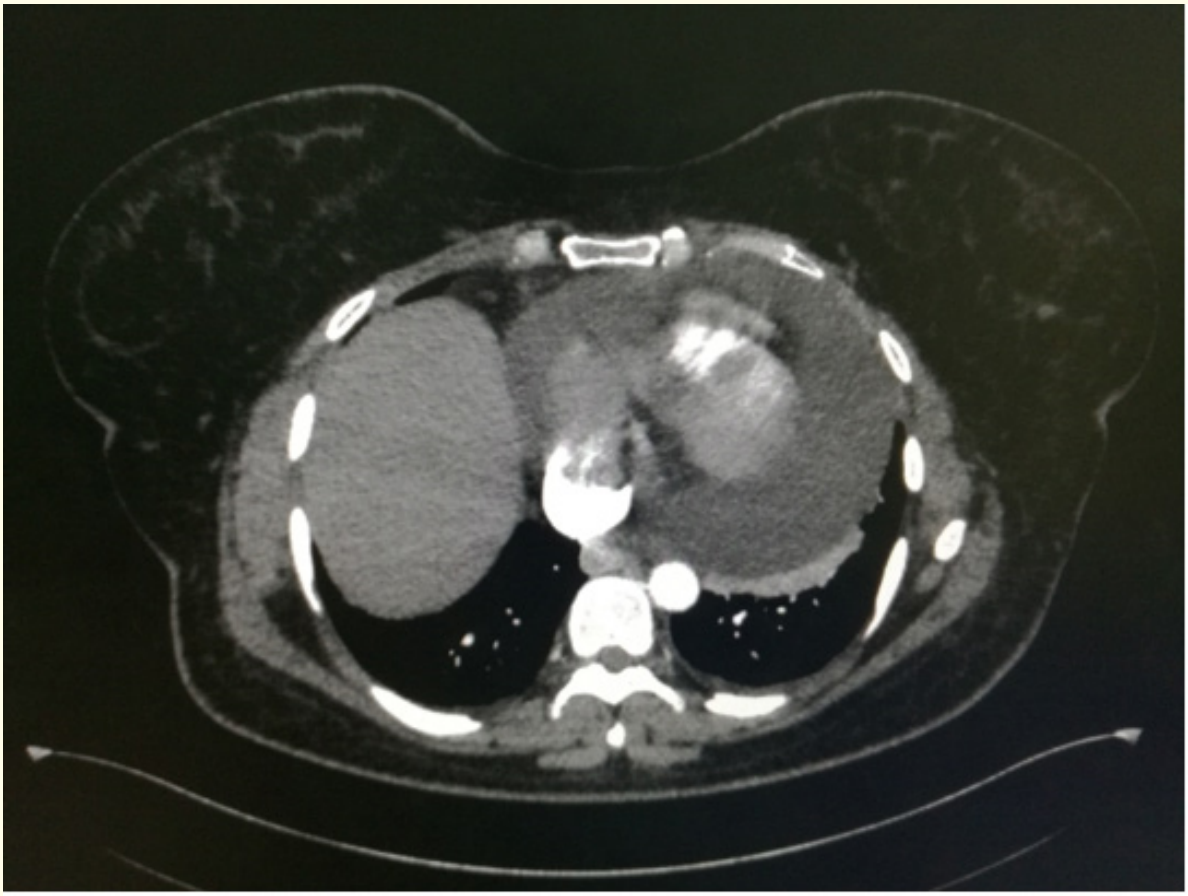

Figure 5

Figure 4 and 5: *Transversal CT scan views reveals the right atrium mass and the partial occlusion of the inferior vena cava.*

Patient was transferred to cardiac surgery department the same day where she was operated. At the time of surgery, the right atrial appendage was noted to be very congested and the right atrial cavity was found to be replaced by a friable tumor which had "fronds like" appearance more over right ventricle 's free wall was infiltrated but tricuspid valve was free of the tumor. Then, the entire free wall of right atrium and the tumor were excised extending from and to the origin of vena cava. A sample of the tumor was subjected to frozen section examination which suggested the diagnosis of sarcoma. Then, the right atrium was reconstructed using autologous pericardium (Figure 6-8).

Subsequently; Macro and microscopic study has revealed the diagnosis of mild differentiated primitive cardiac angiosarcoma.

After recovery from surgery the patient was submitted to chemotherapy and tumor surveillance plan involving serial echocardiograms is planned.

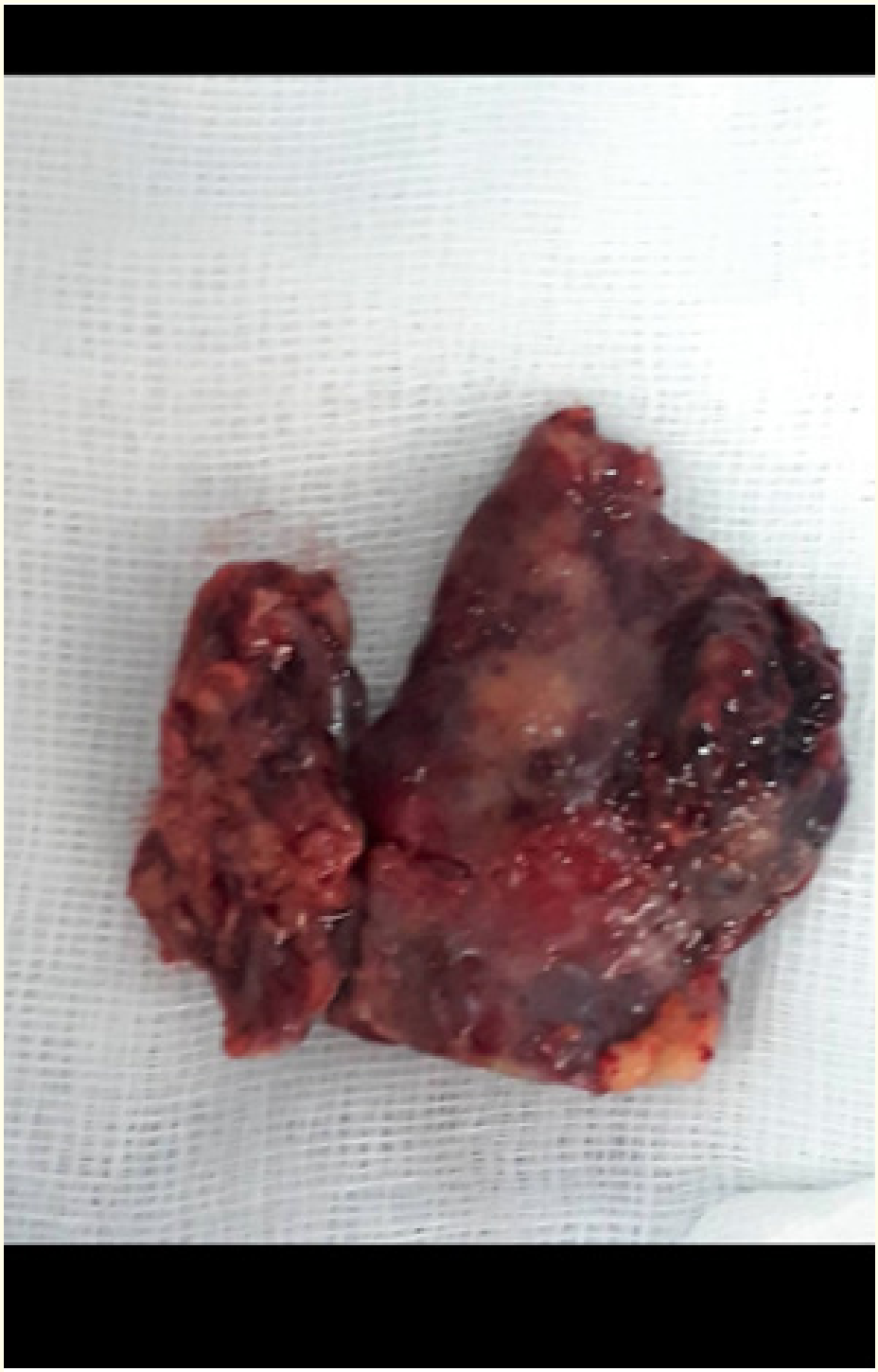

Figure 6

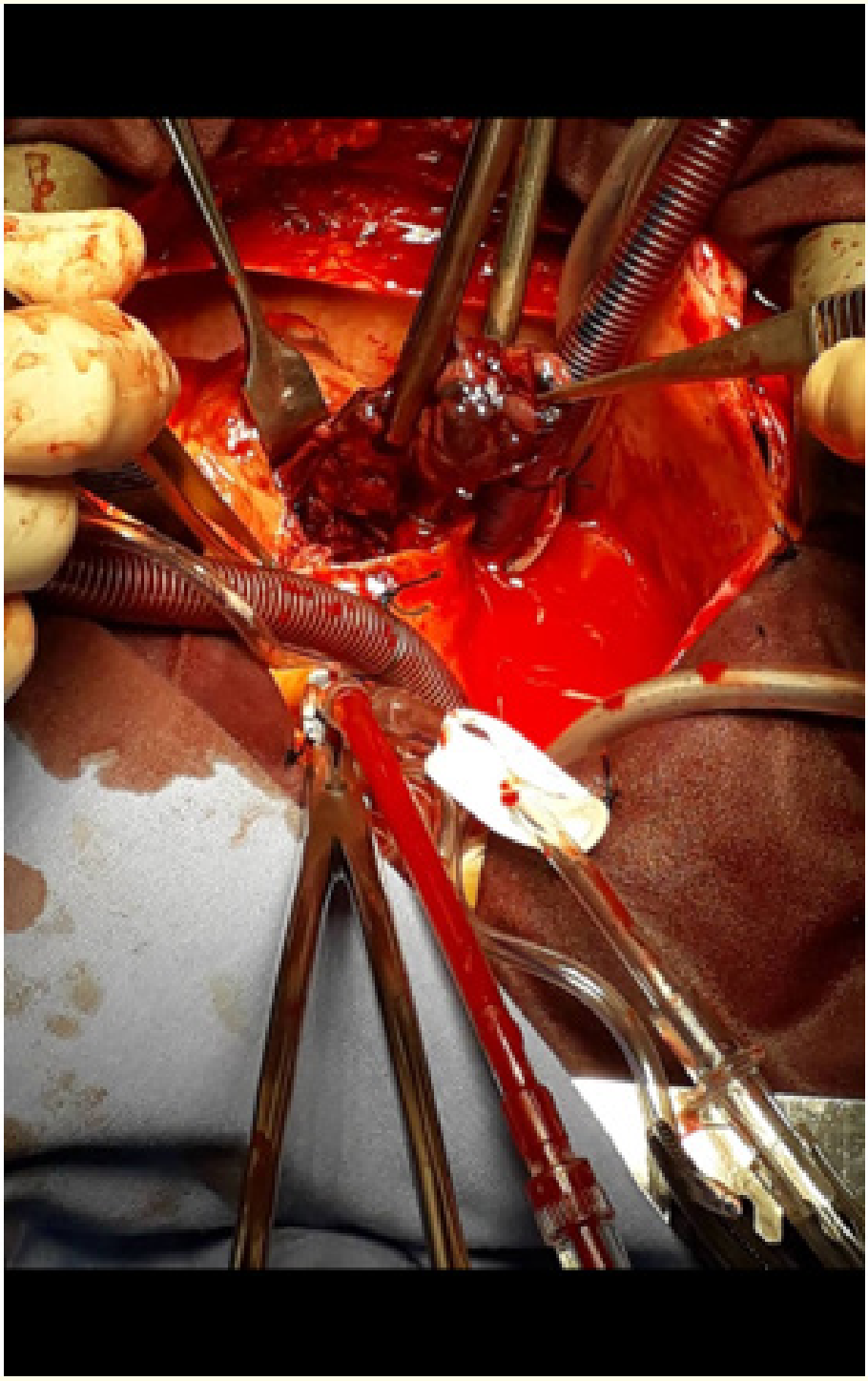

Figure 7

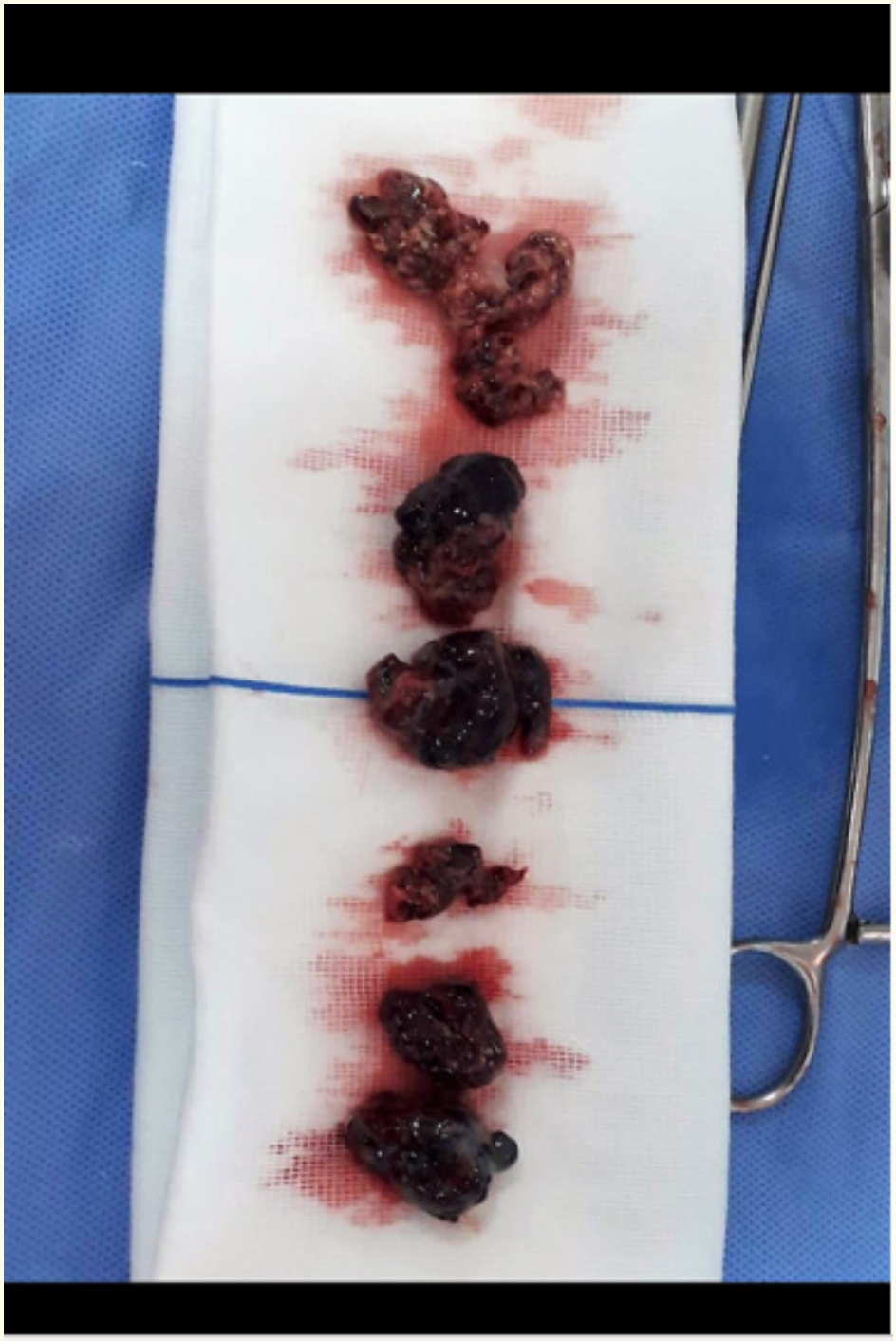

Figure 8

Figure 6-8: *Real life mass during and after extraction.*

Discussion

Soft tissue sarcoma is the most common malignant tumor of the heart, pericardium, and great vessels. It is a rare tumor and its presentation is nonspecific and subtle [2,4].

Primary cardiac angiosarcoma is the most common histological subtype and constitutes 30% of those cases [4]. Males are usually affected more than females in a 2-3/1 ratio [8] which is not the case here.

In a review of 366 cases of angiosarcoma, only 3% were in the heart or great vessels. The most common locations were skin (33%), soft tissue (24%), breast (8%), bone (6%), spleen (4%) and miscellaneous sites [5].

Primary cardiac angiosarcoma is an endothelial cell tumor. Nearly 90% of tumors occur in the right atrium as a multicentric mass. It is characterized by an aggressive and permeating growth within the surrounding myocardial wall but can project into or fill the atrial chamber and invade the vena cava and tricuspid valve [6]. Less than 5% occur in the left atrium or ventricles [4]. Some rare cases of cardiac angiosarcomas are originating in the right coronary artery [7].

The right atrium as site for this type of tumor is rare and some specific presentation could be occurred such as pulmonary embolism, right sided cardiac failure on the account of tricuspid valve blockage, pericardial effusions (tamponade) and paroxysmal atrial arrythmias [9].

Symptoms are nonspecific and subtle which the most common complaint is dyspnea or shortness of breath. More specific symptoms begin usually when the tumor has extended to neighboring tissues and structures and in this phase of his natural history, the tumor was already at the metastatic stage. For that, early diagnosis is the key for curative treatment and good prognosis, but it remains elusive.

According to Mayer., *et al.* [14] half the patients with cardiac sarcomas, are presented with high grade tumor and distant metastases.

Echocardiographic is the most sensitive imaging technique, findings should be interpreted not only by detection of a mass but also by hemodynamic consequences, mobility, consistency and base of the tumor. Transesophageal echocardiography has 97% sensitivity in detecting cardiac masses [10].

The location, shape, size, attachment, and mobility of a tumor can be found via transthoracic echocardiography, whereas transesophageal echocardiography provides information about the insertion point of the tumor [10,11]. However, soft-tissue characterization remains limited compared with that achieved with computed tomography (CT) and magnetic resonance (MR) imaging, and myocardial disease such as tumor infiltration is not clearly depicted [12-14].

No evidence-based treatment guidelines are available due to the rarity of the disease. Early diagnosis and radical excision are essential in the potential achievement of long-term survival for cardiac sarcoma, but this is rarely accomplished due to the locally aggressive nature of the tumor and the presence of early metastases, therefore, the overall results are very poor [16].

Although it appears that most angiosarcomas are resistant to chemotherapy and radiation, aggressive treatment does seem to contribute to reducing tumor size.

The primary site of the tumor may influence treatment planning. Right-sided tumors appear to be bulkier and more infiltrative, with greater likelihood for metastases for which a neoadjuvant chemotherapy is needed [17].

Operative mortality has been reported to be high at 8.3% with an overall actuarial survival of 14% at 24 months after resection [12,15].

Angiosarcomas are rare but lethal tumors; in urgent situations such as tamponade or tricuspid valve blockade, echocardiography should be the first exam to consider so can surgery be started as quick as possible. Outside these cases, a careful clinical exam followed by electrocardiogram, a chest X ray, echocardiography and then CT scans for further investigation should be undertaken.

Conclusion

It should be recognized that currently there are no effective ways to influence the development of cardiac angiosarcomas. The treatment is still controversial and palliative in most cases. The most effective method is surgical resection, but its potential for prolongation of survival is based rather on the natural course of the disease than on the possibility of radical tumor resection.

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