

Case Report

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Pericardial Synovial Sarcoma: A Case Report and Review of Literature



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Abstract

Pericardial Synovial Sarcoma (PSS) is a rare malignant soft-tissue tumor. Early diagnosis and multimodal management could be improving patient outcome. PSS diagnose is difficult due to localization but an aggressive disease. Using all the available medical methods is important to avoid progression and recurrence of this kind of tumors. Multidisciplinary treatment is necessary, and radiotherapy is a good and safe option. We present our experience of a forty-year-old man with this tumor.

Keywords: Pericardial; Sarcoma; Treatment

Abbreviations: PSS: Pericardial Synovial Sarcoma; IORT: Intraoperative Radiation Therapy; RT: Radiotherapy; CT: Chemotherapy

Introduction

Pericardial synovial sarcoma is a type of sarcoma uncommon [1]. It is usually diagnosed in young adults. Patient's present unspecific symptoms, like fever, cough, pain chest and fatigue. In other cases, patients present pericardial effusion and cardiac tamponade. Only 30 cases of synovial sarcoma of the pericardium have been reported in the English language literature, 20 included follow-ups [2,3].

The purpose of this report is to describe the case of a patient with pericardial synovial sarcoma whose initial presentation was severe pericardial effusion. Despite treatment, the patient died in one year. Then, we reviewed the medical literature for the epidemiology, clinical picture, and relevance of complementary imaging tests, therapeutic conduct and prognosis of this neoplasm.

Case Report

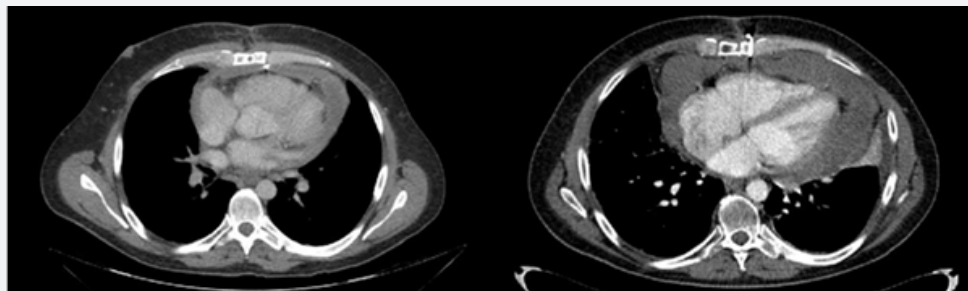


Figure 1: CT with pericardial mass.

40-year-old man visited our Hospital in June 2014 due to chest pain. The physical examination had no abnormal findings. After admission, the chest X-ray indicated cardiomegaly and echocardiography showed dates of pericardial effusion. The patient received treatment and was discharged from hospital after one week with pericarditis diagnosis. Four months later, he started with similar symptoms and echocardiography showed

massive pericardial effusion with a tumorous region in the pericardial space. Echocardiography showed a big mass with compression of right cavities without stenosis, gradient valve tricuspid of 3,7mmHg. The chest CT, as well as confirming the effusion, revealed a pericardial cystic mass with pericardial thickening (Figure 1). Is suggested of neoplasm like mesothelioma or sarcoma. Pericardiocentesis was performed and 1000 mL of

pericardial fluid was drained. According to these findings, surgical resection through pericardiectomy with median sternotomy was performed on October 2014 with pathology study. The tumor was in the pericardial space, with irregular morphology, size of 14x5.7x6 cm and 1500mL serohematic pericardial fluid. Histological analysis showed spindle cells-type sarcoma, necrosis and 18 mytosis. Immunohistochemical analysis showed that the tumor cells were positive for bcl-2, CD99, vimentin and the Ki-67 proliferative index was 50%; however, staining for S100 proteins, CD₃₄, CK AE₁/AE₃, EMA, enolasa, muscle-specific actin (HHF35) and desmin was negative. On the other hand, fluorescence in situ hybridization revealed a moderate prevalence (64%) of cells with interruption of the SYT gene (18q11.2). Final diagnosis was monophasic synovial sarcoma with poorly differentiated areas with unspecified margins. Patient had a good clinical evolution but the control CT scan

three months later, showed tumor persistence, 7.5x11x2.8cm with compression in right ventricle. The tumor grew from the inner surface of the pericardium near the origin of the pulmonary artery and ascending aorta with invasion of left atrium and right coronary. The study was completed with angiography scan. The case was evaluated in multidisciplinary committee. The specialist decided re-intervention, but complete resection was not possible because of the proximity and attachment to the great vessels. In CT scan after surgery, the mass was reduced around 50%. In multidisciplinary committee, we discussed the possibility of adjuvant radiation therapy. Finally, patient received radiation treatment with prescription dose of 45Gy, 1.8Gy daily (Figure 2). He didn't had any toxicity. After three months, patient had a new recurrence. In committee we decided not surgery, patient received chemotherapy. Finally, patient had a new recurrence and died in 12 months after diagnosis (Figure 3).

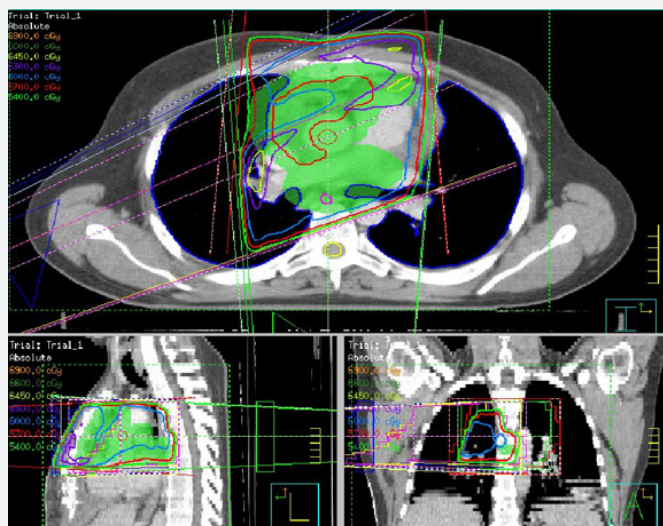


Figure 2: Radiotherapy planning for pericardial sarcoma.

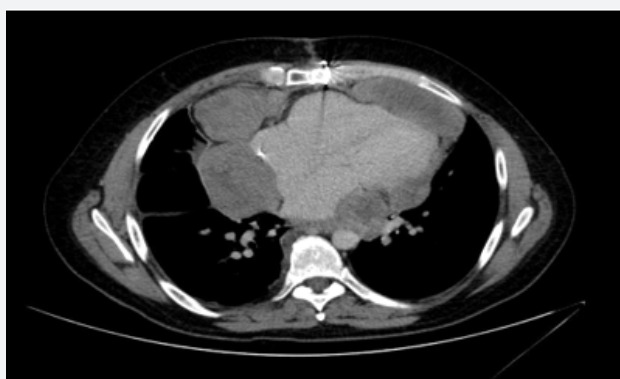


Figure 3: Tumor recurrence.

Discussion

Pericardial Synovial Sarcoma (PSS) is a rare primary malignant tumor of the heart and has an unclear prognosis. Primary cardiac tumours are uncommon, occurring less frequently than metastatic tumours with an incidence in autopsy

series ranging only from 0.001-0.03% [4]. Most of the primary cardiac tumours are benign and approximately 25% of them are malignant, of which the majority are sarcomas but originating from the pericardium are exceedingly rare accounts for approximately 4.2% of primary cardiac sarcomas. In descending order of frequency, these are angiosarcoma, fibrosarcoma, rhabdomyosarcoma, and malignant fibrous histiocytoma [5]. It is more frequent in young adults between 15 and 35 years of age [6]. PSS was first described in 1978 [7]. The signs and symptoms caused by the primary cardiac sarcoma are extremely variable and nonspecific. Clinical manifestations can be divided into four categories: systemic; embolic; cardiac; and secondary to metastasis.

The most common systemic manifestations are fever, chills, fatigue, night sweats and weight loss. Embolic manifestations occur mainly in sarcomas invading the cardiac cavities, causing pulmonary or systemic embolism. Cardiac manifestations depend on the location of the tumor: predominantly pericardial, intramural or intracavitary. In the case of pericardial sarcoma,

the most common initial clinical picture results from massive hemorrhagic pericardial effusion and cardiac tamponade [4]. The organs most often affected by primary cardiac sarcoma metastases are the lungs, brain and bones. The tumors are classified into three histologic subtypes: monophasic, biphasic and poorly differentiated [6]. The histologic type was judged to be monophasic in the current case. Biphasic PSS tumor composed of spindled and epithelioid areas or as a monophasic tumor, characterized by the spindle cell component only. Although immunological characteristics, such as positivity for bcl-2 and CD99, are not exclusive to synovial sarcoma, a definitive diagnosis can be made based on the presence of the chromosomal change t(X;18) (p11;q11), the cytogenetic hallmark of synovial sarcoma. The SS18 gene on 18q and either the SXX1 or SXX2 gene on Xp undergo reciprocal translocation, forming a fusion gene, SS18-SSX, with the subsequent production of fusion proteins; more

than 90% of synovial sarcomas present with this characteristic molecular signature. In addition, biphasic synovial sarcoma has a SYT-SSX1 fusion transcript, whereas SYT-SSX2-positive tumors are monophasic synovial sarcomas [6]. It is most frequent presenting symptom is signs of cardiac tamponade; subsequent diagnostic imaging, including echocardiography or chest CT, reveals a pericardial mass requiring biopsy for diagnosis [8].

An important survival factor is complete resection, but it is rare. Radiotherapy and chemotherapy are indicated in incomplete resection to prevent locoregional recurrence. While broad surgical resection is the first choice for treatment, chemotherapy and radiotherapy are carried out when surgical resection is unsuccessful; however, the prognosis of this disease is poor [9]. Median survival was around 15-25 months in some series.

Table 1: Cases reports with follow-up.

Reference	Age	Gender	Clinical	Surgery	CT (type)	RT (dose/Gy)	Immunohistochemically positive antigens and molecular analysis	Outcome
Muramatsu et al. [8]	57	M	Cardiomegaly	Incomplete	Ifosfamide, adriamycin	50Gy	CK, CD99 and bcl-2 Fusion SYT/SSX negative.	Died at 3y
Phatak et al. [5]	27	M	Dyspnea, cough	Incomplete	Ifosfamide and MESNA	Yes	BCL-2, CD 99, vimentin, FLI1, enolase. EMA. SYT gene: rearrangement	Recurrence at 9m. Second line CT. Stable at 18m
Bezerra et al. [2]	29	F	Dyspnea, chest pain	Incomplete	2-cycles	No	Spindle cells No analysis	Died before second cycle of CT
Cheng et al. [9]	31	M	Fever, cough	Biopsy	No (refused)	No	EMS, CK7, 19, AE1/AE3 SYT gene: rearrangement	Alive at the the 12m follow up
	31	M	Fever, night sweats, chest pain	Incomplete	Doxorubicin, ifosfamide, diamminedichloroplatinum	No	CD99, calpoein, bcl-2, vimentin	Died 27-m after presentation
							SYT gene: reaarrement	
	38	M	Chest distress, shortness of breath	Biopsy			Vimentin, bcl-2, calponin, CD99. SYT gene: rearrangement	Awaiting heart transplantation
Akerström et al. [12]	54	F	Malaise, fever, chest pain	Incomplete	Ifosphamide	No	EMA, vimentin, VER EP4, bcl2, CK AE1/AE3	Died after 26-m
							SYT-SSX fusion gene	
Moorjani et al. [13]	61	M	Dysnea	Yes	Yes	No	EMA, CK AE3	Alive after 2-m of surgery
Katakura et al. [14]	70	F	Cough, dysnea	Complete	No	No	SYT-SSX fusion gene	Died 79-d after operation
Korula et al. [15]	49	M	Dyspnea, chest pain	Biopsy	Yes	No	EMA, CK(AE1/AE3), bcl2	Palliative care
Zhang et al. [16]	29	M	malaise	Incomplete	Yes	No	-	Alive at 13-m
De Zwaan et al. [17]	19	M	Dyspnea	Biopsy	-	-	EMA, CD34, CD99, vimentin and MNF116. FISH (X; 18) translocation	Palliative care
Hing et al. [18]	15	M	Cough, tight chest	Incomplete	Cisplatin, doxorubicin, cyclophosphamide	No	EMA, vimentin, bcl2	Died 31-m after presentation

							SYT-SSX fusion gene	
Van der Mieren et al. [19]	26	M	Cardiac tamponade	Yes	Doxorubicin, hydrochloride	Yes	EMA, vimentin, CK	Alive >5 years
							SYT-SSX transcript	
Yano et al. [20]	22	F	Shortness of breath, cough	Yes	Yes	Yes	EMA, vimentin	No recurrence at 12-m
							SYT-SSX2 expression	
Anand et al. [21]	29	M	Acute breathlessness	Incomplete	Yes	Yes	EMA, CD99	Alive at 13-m
Val-Bernal et al. [22]	76	F	Unspecific symptoms	Autopsy	No	No	EMA, cytokeratin 5,6,7, thrombomodulin	Died
Dos Santos et al. [23]	37	M	Fever	Incomplete	No (refused)	No	-	Died 15-d
Oizumi et al. [24]	19	F	Dyspnea, fatigue	Complete	-	-	Vimentin	Died 7-m
							SYT-SSX transcript	
Al-Rajhi et al. [25]	19	M	Shortness of breath	Incomplete	No	Yes	SYT-SSX fusion gene	No evidence of disease at 12m

Due to its extreme rarity, the most effective therapeutic strategy for treating pericardial synovial sarcoma could not be determined, and the complete or near total excision of the tumor was performed in only five of the reported cases [10-13]. The pericardial tumor observed in the present case was also considered to be unresectable, due to its strong attachment to the visceral pericardium [6]. Management of synovial sarcoma of pericardial origin is challenging and needs a multimodal approach. It is an aggressive tumor with a poor prognosis. Most patients have a short survival time. One exception is the case report of a patient whose survival exceeded 14 years and involved recurrences and multiple surgical resections. Surgical resection can lead to curative therapy. Most often, due to the site of origin, tumors are adherent to adjacent vital organs and are non-resectable at the time of diagnosis. Therefore, EBRT and less commonly neo-adjuvant chemotherapy are used to make the tumor resectable [14-18]. In our patient, we could not attempt complete surgical resection. Therefore, after the initial biopsy and pericardial window, neo-adjuvant chemotherapy was done with six cycles of high dose ifosfamide (2000mg/m²) and mesna. Due to the rarity of these tumors, it is very difficult to establish an optimal regimen for chemotherapy. In general, synovial sarcomas are chemo-sensitive to ifosfamide and show a good response. A case series of 13 patients with synovial sarcoma treated with high-dose ifosfamide showed a response in all patients, with 4 clinical remissions achieved. Combining doxorubicin with ifosfamide may achieve a higher response rate (58%) but patient tolerability and side effects need to be considered [19-21]. The initial favorable response to neoadjuvant high dose ifosfamide in our patient made the tumor resectable and may be a viable alternative to EBRT alone in non-resectable tumors [5].

EBRT is a very useful modality of treatment post-surgery when the margins of resection are positive. In our patient, EBRT

was initiated one month after surgery. Previous reports have shown similar favorable results. To the authors' knowledge only EBRT has been employed in previous cases. Intraoperative Radiation Therapy (IORT) with a radiation dose of 15-45 Gray appeared to be useful for local control and to be more effective in patients without distant metastases at the first visit in a study that included 24 patients with soft tissue sarcomas (1 case of synovial sarcoma) [22-24]. Close monitoring for recurrence is very crucial in management of this tumor. CT or magnetic resonance imaging should be performed every 6 months to detect subclinical recurrences. We performed CT imaging every three months along with regular clinical follow-up or symptoms. Repeat imaging is essential to be able to diagnose subclinical recurrence and initiate repeat broad surgical resection and follow-up chemotherapy if needed. We used the anti-mitotic agent phosphamide for treating the recurrence [5].

Cardiac transplant had been an alternative in some cases with more survival, but it is controversial due to tumoral etiology.

This report described one case of pericardial synovial sarcoma, an extremely rare primary cardiac tumor, despite proper treatment, has an unclear prognosis [25] (Table 1).

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