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CASE REPORT

# Volumetric Modulated Arc Radiotherapy Efficacy after Double Recurrences of Cardiac Sarcoma

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**Abstract:**

***Background:***

Volumetric Modulated Arc Therapy (VMAT) has recently become a pivotal treatment of oncological diseases due to the high-precise delineation of target volume contours with sparing organs at risk. This procedure requires a high level of experience and precision and is achievable only with advanced diagnostic support. Magnetic Resonance (MRI) and multimodality imaging, such as 18F-fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT), are fundamental in implementing radiotherapy guidance.

***Case Report:***

A 54-year-old patient underwent surgery twice to remove primitive and recurrent cardiac sarcomas of the left atrium. The appearance of a further relapse required radiotherapy as the only possible treatment. Cardiac MRI was then performed to define the degree of atrial mass invasiveness, and 18F-FDG PET/CT was performed to assess the activity and staging of the cardiac lesion. It revealed high 18F-FDG uptake not only in the left atrium lesion but also in a pancreatic lesion with elevated 18F-FDG uptake (SUV max 5.5). The pancreatic biopsy performed a few days later confirmed the myxoid sarcoma metastasis, and surgeons defined it as not operable due to the patient's clinical condition. Radiotherapy was then urgently performed with the VMAT technique. After 40 days, a cardiac MRI showed a reduction in the cardiac mass with improvement in the respiratory and cardiac symptoms; then, the patient started chemotherapy. One year after diagnosis, the patient is still alive and is receiving chemotherapy with gemcitabine and docetaxel with good compliance.

***Conclusion:***

The correct and timely management of a patient suffering from a rare oncological disease has allowed a better and longer survival, especially due to VMAT, a sophisticated procedure that requires high expertise. This case also demonstrates that cardiac MRI and whole-body imaging procedures, such as 18FDG PET/CT, can be useful in staging patients with oncological diseases.

**Keywords:** VMAT, Cardiac sarcoma, Therapy strategies, MRI, 18F-FDG PET/CT, Case report.

| Article History | Received: September 20, 2022 | Revised: January 13, 2023 | Accepted: January 26, 2023 |
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## 1. INTRODUCTION

Primitive heart tumors are rare and have variable incidence, ranging from 0.001% to 0.03% [1]. Most of them are benign, and among malignancies, metastatic cardiac tumors are more common than primary neoplasms [2].

Cardiac malignancies are still characterized by poor prognosis as complete surgical resection is achieved in less than 50% of patients, and local recurrences and metastasis occur even within 1 year [2].

The diagnosis of these malignancies is still complex and challenging due to non-specific symptoms and the need for sophisticated diagnostic investigations that allow the identification of lesions but also the discrimination with benign

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pathologies [3].

Moreover, treatment protocols are not well defined; beyond surgery, radiotherapy and chemotherapy are considered, but all these procedures are sometimes not even possible in a few patients as they are associated with severe side effects [4, 5].

This report presents the case of a patient with double recurrence of a rare primitive cardiac sarcoma to whom the most advanced radiotherapy planning performed with Volumetric Modulated Arc Therapy (VMAT) was decisive for his management and prognosis.

## 2. CASE REPORT

A 54-year-old patient presented with worsening dyspnoea and chest pain. Due to the smoking habit, he performed a chest computed tomography (CT) scan before and after contrast medium injection, which showed a large (5.5 x 6 cm) irregularly rounded hypodense tissue in the left atrium size; furthermore, signs of post-capillary pulmonary hypertension and bilateral pleural effusion were also detected.

After a few weeks, the patient underwent thoracotomy and resection of the endocavitary portion of the left atrial mass. The histological examination concluded mesenchymal neoformation with fused cells and myxoid stroma with frequent atypical mitosis, proliferative index of Ki67=25%, positive immunohistochemical reactions for CD34 (++) and negative for CK pool and CD31.

Only 2 months later, the patient presented again with dyspnoea and chest pain. Another chest CT scan was performed that showed a recurrence of the atrial sarcoma with diameters of 4.2x3.9 cm, which partially occupied the mitral orifice.

Even this new lesion was surgically removed, and the histological examination suggested the recurrence of a myxoid sarcoma.

Unfortunately, one month later, the follow-up chest CT showed a further recurrence of 4.3x3.7 cm. The patient was defined as inoperable by cardiac surgeons and not amenable to chemotherapy by medical oncologists because of his precarious clinical condition, mainly related to cardiac function.

Radiotherapy was then proposed as the unique possible treatment.

Cardiac Magnetic Resonance (MRI) was then performed to define the degree of atrial mass invasiveness to the posterior wall and left pulmonary veins and to plan the radiotherapy treatment (Fig. **1**).

Furthermore, 18F-fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) was performed to assess the metabolic activity of the cardiac lesion and complete the staging; an area of intense uptake corresponding to left atrium sarcoma (SUV max 6.8) extended to pulmonary vessels (Figs. **2B** and **2C**) and a pancreatic lesion with high 18F-FDG uptake (SUV max 5.5) suggestive for relapse was described (Figs. **2D** and **2E**). 18F-FDG uptake was also evident along the surgical wound for the 2 thoracotomies (Fig. **2A**).

The pancreatic biopsy performed a few days later confirmed the myxoid sarcoma metastasis, and surgeons defined it as inoperable due to the patient's clinical condition.

Radiotherapy with the VMAT technique was then urgently performed. The patient underwent to CT scan centring, using the wing board for the arms and chest and the Combi fix for the legs as immobilization systems. Three radiopaque markers were placed on the intermammillary line. The chest scan was conducted from the lung apices to the lung bases with a 3 mm slice thickness. An image fusion was carried out with the images of the centring CT and those of magnetic resonance imaging for a greater definition of the processing volumes.

The Planning Target Volume (PTV) included the myxoid sarcoma mass with an expansion of 0.5 mm, and as organs at risk (OAR), the heart, oesophagus, left and right lung, and spinal cord. With the VMAT imaging technique (IGRT), a total dose (TD) of 45 Gy was delivered in 25 fractions of 1.8 Gy/day (Fig. **3**). Before each treatment, Cone Beam CT images were acquired, and the sessions were performed under electrocardiographic monitoring to exclude the onset of malignant cardiac arrhythmias.

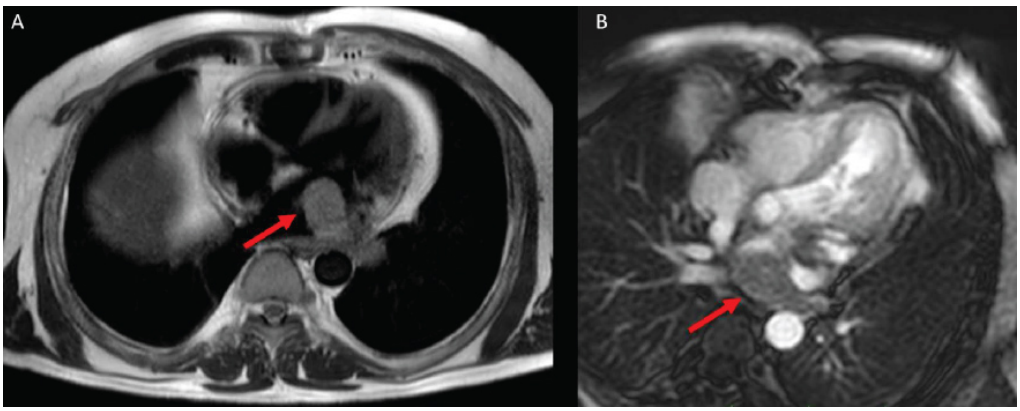


**Fig. (1).** Pre-treatment MRI imaging (**A**) T1w axial showed left atrial involvement from sarcoma. (**B**) T1wCE+ showed involvement left pulmonary vein.

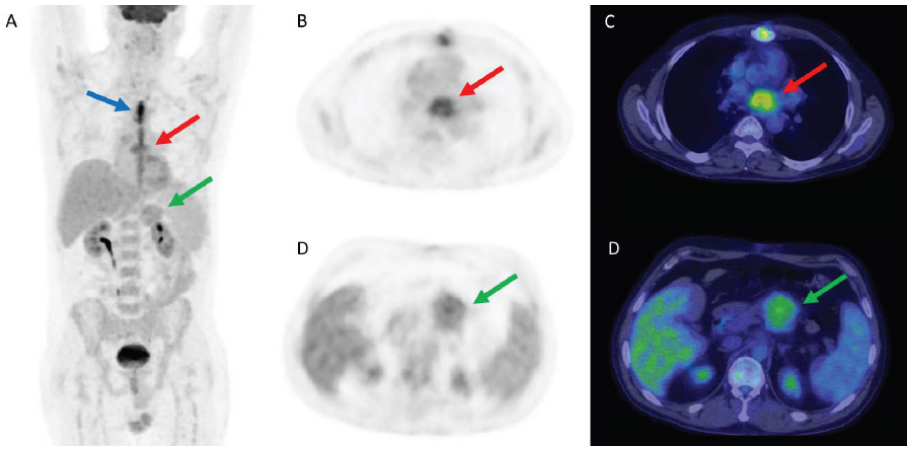


**Fig. (2).** 18F-FDG PET/CT MIP image (**A**) showed longitudinal 18F-FDG uptake in the middle of the thorax due to the recent sternotomies (blue arrow); also, in transaxial PET and fused images, mass in the left atrium was evident (**B**, **C**) and characterized by high 18F-FDG uptake (SUV max 6.8) corresponding to the sarcoma (red arrows) and a lesion with intense 18F-FDG uptake (SUVmax 5.5) in the mesogastric region (**D**, **E**) compatible with pancreatic metastasis (green arrows).

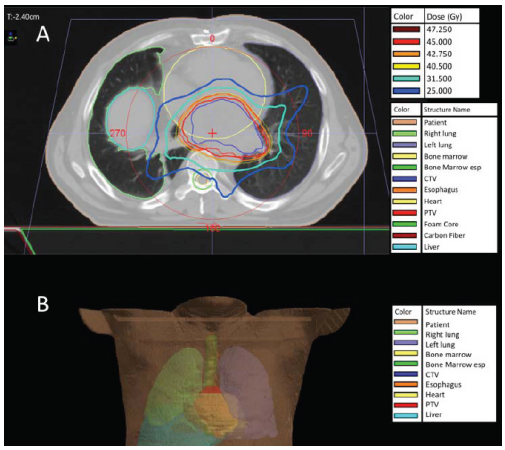


**Fig. (3).** Images of RT treatment plan; (**A**) Target volume and OARs delineation with image registration between the planning CT scan and T1wCE Cardiac MRI Dose distribution: red, 100% of Total Dose (TD); orange, 95% TD; green, 30% TD; blue, 5% TD; (**B**) Three-dimensional visualization of the OARs and target volume for RT planning.

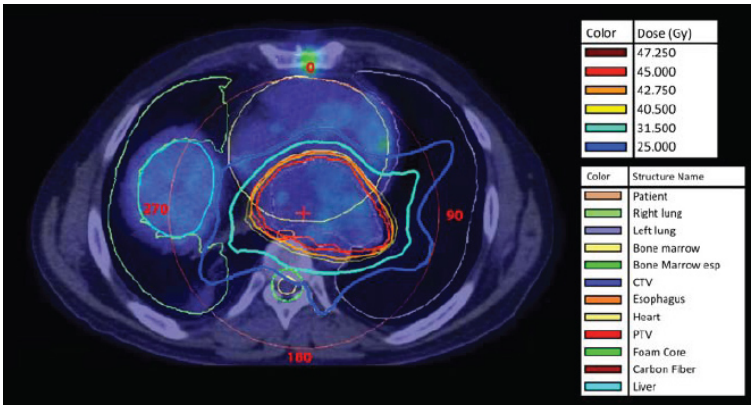


**Fig. (4).** 18F-FDG PET/CT transaxial fused image was used to overlap the isodose curves of the radiotherapy plan as a further comparison evaluation of the irradiation distribution and showed matching of target volumes.

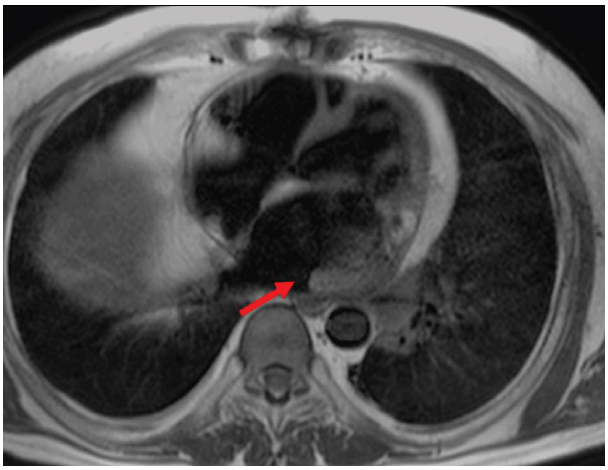

**Fig. (5).** Post-treatment MRI imaging T1w showed volume reduction of sarcoma without pulmonary vein involvement.

During treatment, the patient underwent weekly echocardiography to detect acute complications even though treatment continued and ended without significant side effects.

In addition, 18F-FDG PET/CT fusion images were used as a comparative evaluation of the radiotherapy plan, reporting the isodose curves on them and showing the correspondence of target volumes (Fig. **4**).

After 40 days, a cardiac MRI showed a reduction in the cardiac mass (Fig. **5**).

After the improvement of respiratory and cardiac symptoms, the patient began chemotherapy treatment.

A year after diagnosis, the patient is still alive and is undergoing chemotherapy with gemcitabine and docetaxel with promising results and good compliance.

## 3. DISCUSSION

Surgery is considered the treatment of choice for cardiac malignancies; despite the current technological advances in surgical removal of the left atrium lesions, it is associated with a high incidence of major hospital complications and mortality. Complete surgical removal of cardiac sarcomas is often problematic and may require resection of large wall portions; unfortunately, due to the massive local diffusion and the frequent presence of distant metastasis, complete resection is not possible in all patients. Furthermore, for rapidly growing local relapses, short-term cardiac surgery re-operations are often required, associated with aggressive chemotherapy treatments [6 - 8].

Local relapses also occurred in the patient, who underwent surgery twice; the third cardiac surgery after the second relapse was not possible due to the previous resections of the cardiac wall.

Transthoracic and/or transesophageal ultrasonography are the first diagnostic examinations due to their versatility and routine use in cardiopathic patients, but they are limited in the comprehensive evaluation of the cardiac chambers [9, 10]. CT imaging can be used to evaluate the heart and the surrounding mediastinum, providing better soft-tissue contrast [11].

MRI is considered a powerful reference technique for characterizing suspected cardiac mass; it describes tissue morphology, size, extension, perfusion and characterization using late-gadolinium enhancement, orienting toward the histopathological diagnosis [12]. Furthermore, MRI represents the non-invasive technique able to evaluate the functional impact of cardiac lesions, treatment planning and post-treatment follow-up [13].

Unfortunately, claustrophobia and the presence of devices incompatible with magnetic fields may limit its use, and images might be influenced by artifacts from respiratory movements [10].

18F-FDG PET/CT is an established whole-body technique considered a powerful tool with a great impact on the diagnosis and treatment of oncological diseases [14].

18F-FDG PET/CT is a multimodality imaging that allows metabolic evaluation also through the collection of semiquantitative parameters, such as the Standardized Uptake Value (SUV) and the merging with morphological images.

The heart evaluation is difficult because of the inhomogeneities of myocardial 18F-FDG uptake mainly in the lateral and anterior regions and lesser in the septal region; nevertheless, literature reports a sensitivity of 100% and specificity of about 86% using a SUVmax cutoff of 3.5 in detecting cardiac malignancies and post-surgical recurrences [2, 15].

Recently, literature reports that 18F-FDG PET/CT is a valid support in radiotherapy planning, and the merging with MRI images further implements this use due to the help in delivering the radiation directly to the mass and trying to avoid damage to healthy cardiac tissue [10].

Also, in our patient, the multimodality approach with MRI and 18F-FDG PET/CT was fundamental for exactly characterizing the heart lesion morphologically, functionally and metabolically and for the patient's whole body staging.

VMAT has become a pivotal treatment of oncological diseases due to its high-precision delineation of target volume contours and organs at risk. This procedure requires a high

level of expertise and precision and is achievable only with advanced diagnostic support. MRI and multimodality imaging, such as 18F-FDG PET/CT, are strategic in implementing radiotherapy guidance [16].

In External Beam Radiotherapy, advances in rotational, intensity-modulated delivery techniques with VMAT have allowed individualizing the radiation dose distribution to the cardiac volume while sparing the surrounding normal tissues and organs, thus optimizing treatment efficacy and minimizing surrounding organs toxicity [17].

Few case reports of cardiac sarcoma are described in the literature, and to our knowledge, none of them have been treated with VMAT.

The case we reported is an example of how modern radiotherapy techniques can be used as a non-invasive therapeutic treatment; in particular, VMAT reduces toxicity and increases efficacy.

The goal of the treatment would have been the eradication of the lesion, but the reduction in size with consequent improvement in cardiovascular function could be considered an excellent result. Therefore, this improvement made it possible for the patient to undergo chemotherapy for the residual heart lesion and pancreatic metastasis.

## CONCLUSION

This is a virtuous example of the multimodal approach in both therapy and diagnosis. The correct and timely management of a patient with a rare oncological disease has allowed him a better and longer survival, especially due to VMAT, a sophisticated procedure that requires high expertise.

This case also demonstrated that cardiac MRI and whole-body imaging procedures, such as 18FDG PET/CT, help stage all patients with cardiac oncological diseases.

## LIST OF ABBREVIATIONS

| | | |
|---|---|---|
| **VMAT** | = | Volumetric Modulated Arc Therapy |
| **CT** | = | Computed Tomography |
| **MRI** | = | Magnetic Resonance |
| **18F-FDG PET/CT** | = | 18F-fluorodeoxyglucose Positron Emission Tomography/Computed Tomography |
| **SUV** | = | Standardized Uptake Value |
| **PTV** | = | Planning Target Volume |
| **OAR** | = | Organs at Risk |
| **IGRT** | = | Imaging Guided RadioTherapy |
| **TD** | = | Total Dose |

## ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Not applicable.

## HUMAN AND ANIMAL RIGHTS

Not applicable.

## CONSENT FOR PUBLICATION

A written informed consent was obtained from the patient for the publication of this report and any accompanying images.

## STANDARDS OF REPORTING

CARE guidelines were followed in this case study.

## FUNDING

None.

## CONFLICT OF INTEREST

Dr. Corinna Altini is the editorial advisory board member for the journal Current Medical Imaging.

## ACKNOWLEDGEMENTS

Declared none.

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