

Primary Cardiac Angiosarcoma with Extensive Visceral Metastases: Utility of ¹⁸F-Fluorodeoxyglucose Positron Emission Tomography-Computed Tomography in Response Assessment to Sorafenib

Abstract

Primary cardiac angiosarcomas although rare neoplasms remain the most common primary malignant neoplasms affecting the heart. We discuss the sequential positron-emission tomography-computed tomography findings of a 50-year-old man who was diagnosed with the metastatic cardiac angiosarcoma at the outset and developed progressive disease despite sorafenib therapy.

Keywords: Cardiac angiosarcoma, metastases, positron-emission tomography-computed tomography, sorafenib

A 50-year-old man diagnosed with primary cardiac angiosarcoma underwent ¹⁸F fluorodeoxyglucose positron-emission tomography-computed tomography (FDG PET-CT) for initial staging and metastatic work-up. The scan findings revealed an enlarged right atrium with increased FDG uptake in the right atrial appendage [Figure 1a solid black arrow, c and d]. FDG avid metastases were seen in the bilateral lungs [Figure 2a], adrenals [Figure 2c], and spleen [Figures 1a dashed-red line and 2c]. He was started on sorafenib therapy and a followup of PET-CT scan was done after 3 months that revealed progressive metastatic disease in bilateral lungs [Figures 1b and 2b], liver [Figures 1b solid purple arrow and 2d], spleen [Figures 1b solid red arrow and 2d], abdominal lymph nodes, and bones [Figures 1b solid blue arrows and 2f]. Angiosarcoma accounts for <1% among sarcomas and primary angiosarcoma of the heart and the great vessels accounts for only 3% of all the angiosarcomas^[1] with an autopsy prevalence of 0.001%–0.28%.^[2] Most of these tumors primarily arise in the right atrium, involving the lateral wall of

the right atrium and mostly sparing the septum which results in delay in the initial diagnosis.^[3] Frequent sites of extra-cardiac metastases include lung, liver, brain, bone, lymph nodes, adrenal glands, spleen, and intestine.^[4-6] Our case had metastatic disease involving the lungs, adrenals, spleen, liver, and bones with all of them progressing on follow-up PET-CT scan. Utility of 18-F-FDG PET-CT in initial staging as a part of extra-cardiac metastatic work-up and response assessment to chemo or radiotherapy; although, scarce has been reported in the literature by Hod *et al.*, Tan *et al.*, Jain *et al.*, and Dhull *et al.*, respectively.^[7-10] Due to aggressive and rapidly progressive behavior of the primary and metastatic disease, we advocate extensive use of 18-F-FDG PET-CT as a “one-stop shop” for immaculate assessment of the disease burden during the initial staging and restaging purposes.

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Conflicts of interest

There are no conflicts of interest.

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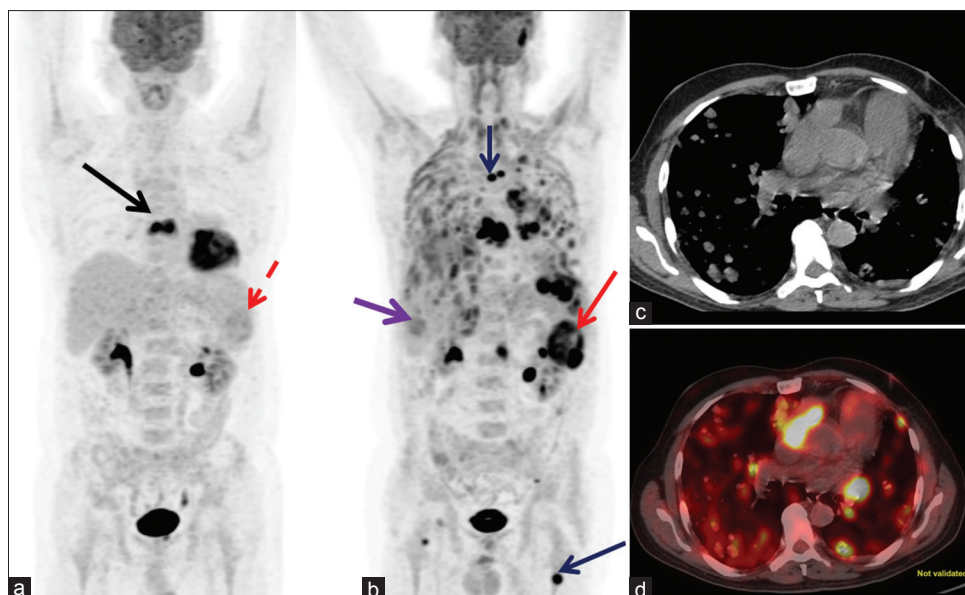


Figure 1: (a) Pretherapy ¹⁸F fluorodeoxyglucose positron-emission tomography-computed tomography maximum intensity projection image showing focal increased fluorodeoxyglucose uptake in the thorax region (solid black arrow) suggestive of cardiac angiosarcoma. Dashed-red arrows show increased fluorodeoxyglucose uptake in the liver, spleen, thorax, and pelvis suggestive of metastases. (b) Posttherapy ¹⁸F fluorodeoxyglucose positron-emission tomography-computed tomography maximum intensity projection image showing increased fluorodeoxyglucose uptake in the liver, spleen, thorax, and pelvis suggestive of metastases. (c) Trans-axial computed tomography image showing soft-tissue lesion in the right atrial appendage (d) Fused trans-axial positron-emission tomography-computed tomography image showing increased fluorodeoxyglucose uptake in the right atrial appendage suggestive of cardiac angiosarcoma

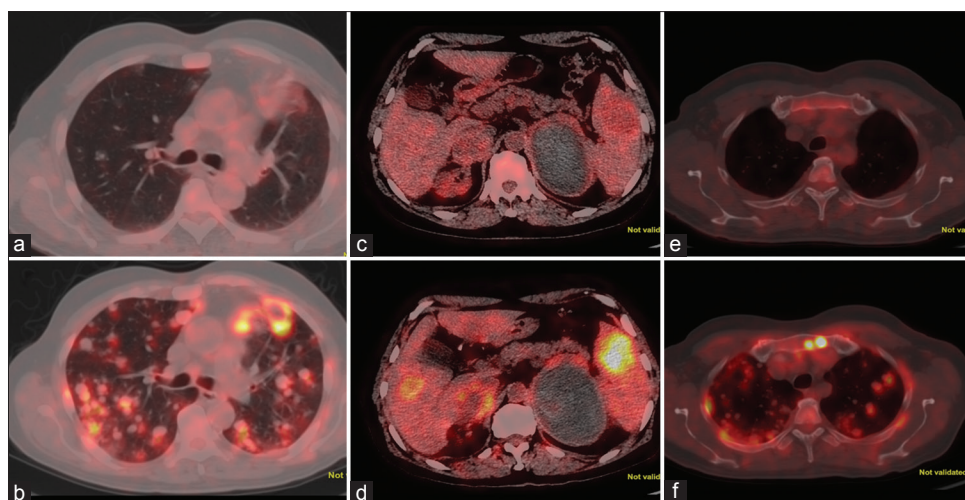


Figure 2: Fused axial pre-therapy positron-emission tomography-computed tomography image showing nodules in the right lung (a) which increased in size and fluorodeoxyglucose uptake in post-therapy scan. (b) Fused axial pre-therapy image (c) showing mildly fluorodeoxyglucose avid hypodense lesion in the spleen which increases in size and fluorodeoxyglucose avidity in post-therapy scan. (d) However, no significant change in Fluorodeoxyglucose avid liver lesion in segment V is seen. No significant fluorodeoxyglucose uptake is seen in the sternum (e) whereas post-therapy scan (f) shows increased fluorodeoxyglucose uptake in the sternum with no cortical changes suggestive of marrow metastases

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