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CASE REPORT

A rare case of rapidly growing isolated right ventricular phyllodes tumor leading to death

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ABSTRACT

An 82-year-old woman with a known chronic small "clot" in the heart presented with rapidly progressive dyspnea. A [^{18}F]fluoro-2-deoxy-D-glucose ([^{18}F]FDG) and cardiac-MRI demonstrated rapid enlargement of the right ventricular presumed clot now expanding and obliterating the right ventricular cavity. Patient continued to deteriorate despite urgent cardiac surgery to remove the mass and died 8 days later. Cardiac explant pathology demonstrated phyllodes tumor.

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CASE PRESENTATION

An 82-year-old woman presented with rapidly progressing shortness of breath over a one-month period. She had a known small cardiac right ventricular mass, which was favored to be a thrombus on multiple prior imaging studies. A chest computed tomography (CT) performed at outside hospital demonstrated a large right ventricular mass and she was referred to our hospital for further management. A [^{18}F]FDG PET/CT was performed which demonstrated a 7.4 x 5.4 cm partly calcified mass occupying and expanding the right ventricular cavity (Figure 1). It had intense heterogeneous rim of increased FDG uptake with central photopenic/necrotic areas. The right ventricular cavity was expanded and almost completely obliterated by the mass with deviation of septum towards the left. There was enlargement of the right atrium and IVC due to backpressure changes. In addition, there were mild pericardial and bilateral pleural effusions. A cardiac-MRI performed the next day corroborated the findings of the [^{18}F]FDG PET/CT with a large right ventricular mass expanding and obliterating the right ventricle with backstream enlargement of the right atrium and IVC (Figure 2). On a prior MRI 6 months prior and a CT 3 months back, this mass measured approximately 2.8cm x 2.0 cm.

Detailed history revealed that she had a history of high-grade right breast phyllodes tumor with pulmonary metastases treated with mastectomy and adjuvant radiation, 3 years ago. A working diagnosis of either a primary or a metastatic cardiac tumor was considered. Given the history, a possibility of an isolated cardiac metastasis from phyllodes tumor was also kept. Her condition continued to deteriorate during the hospital admission, requiring vasopressor and inotropic support. A decision was made to proceed with urgent surgical resection of mass to restore the right ventricular cavity. A 7.4 x 5.4 cm large mass was removed from the right ventricle during surgery and sent for histopathologic evaluation. She continued to be on supportive care post-operatively and could not be weaned off and died 4 days after surgery. Cardiac explant pathology revealed a diagnosis of phyllodes tumor.

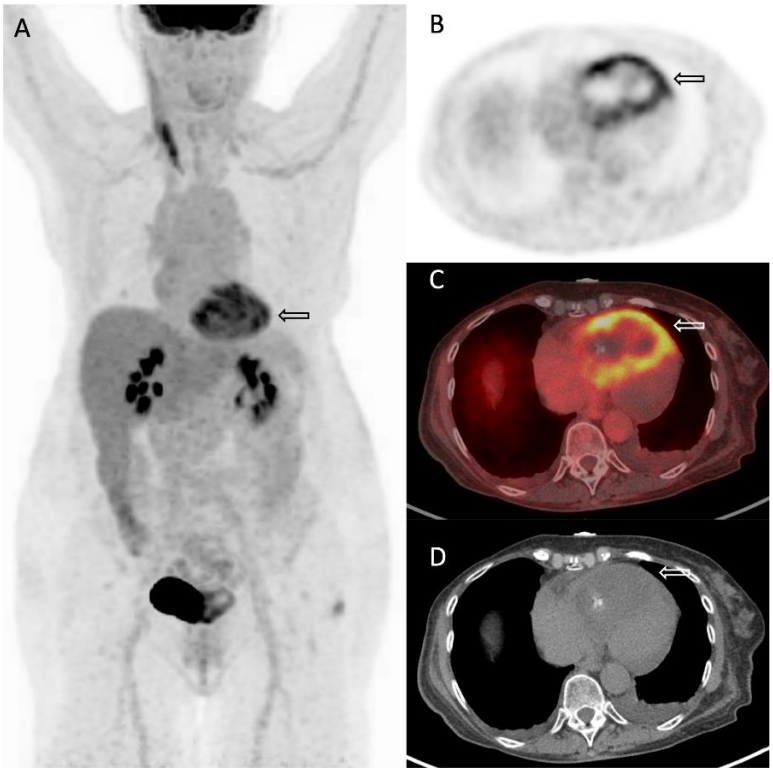

Figure 1. A: [^{18}F]FDG PET/CT whole body maximum intensity projection (MIP) image demonstrates heterogeneous FDG uptake in the region of the cardiac ventricles (hollow arrow). B: Axial attenuation corrected [^{18}F]FDG PET image demonstrates a peripheral rim-like FDG uptake within the right ventricular cavity (hollow arrow). C and D: Axial fused [^{18}F]FDG PET/CT (C) and CT (D) images demonstrating a large heterogeneous mass with some internal calcifications expanding the right ventricular cavity (hollow arrows). It shows peripheral rim-like FDG uptake and central photopenia, likely suggesting central necrosis. There is enlargement of the right atrium and small pericardial and pleural effusions

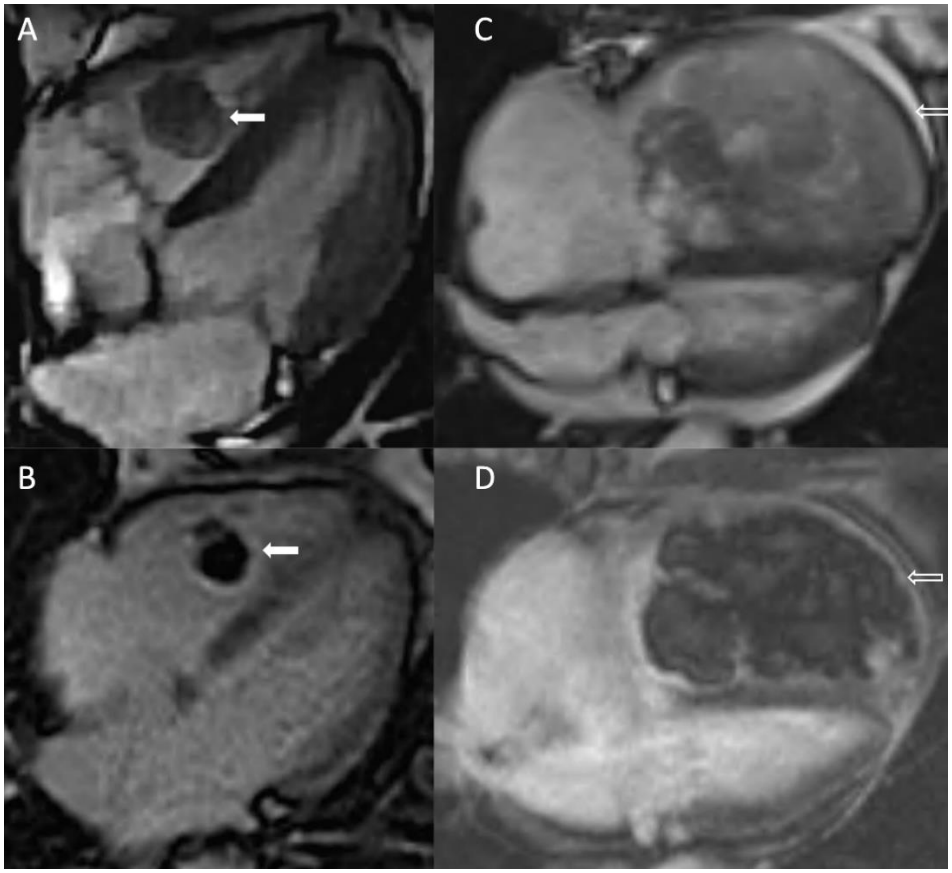

Figure 2. A and B: Cardiac-MRI True Fast Imaging with Steady-state Precession (TrueFISP) (A) and Contrast Enhanced Inversion Recovery (B) images, 6 months prior demonstrates a small round homogeneous non-enhancing mass in the right ventricular cavity, presumed to be a thrombus (solid arrows). C and D: MRI TrueFISP (C) and Contrast Enhanced Inversion Recovery (D) images at the time of presentation demonstrates marked interval enlargement of the right ventricular mass, now expanding and obliterating the entire right ventricular cavity and showing only a few areas of internal heterogeneous enhancement (hollow arrows)

DISCUSSION

The patient had a history of right breast high-grade phyllodes tumor with pulmonary metastases, treated with right mastectomy and radiation. There was an untreated chronic small right ventricular mass, which showed minimal growth over time and was considered to be a chronic thrombus on multiple prior imaging. It underwent rapid enlargement in size leading to rapid deterioration of clinical condition and death despite urgent surgery. This is an unusual presentation of a usually benign tumor with a devastating outcome.

Phyllodes tumor is a type of fibroepithelial neoplasm of breast comprising less than 1% of all breast neoplasms [1]. Pathologically they can be categorized as Benign, Borderline or Malignant [2]. Ramakant et al. retrospectively studied 150 patients with phyllodes tumor from 2003 to 2013 and found that 51.33% were benign, 16% were borderline and 32.67% were malignant [3]. Benign hyllodes tumors can be visually and radiologically indistinguishable from benign breast fibroadenomas. Malignant tumors tend to present at a later age, are often larger and show fast growth and metastases [3]. In some patients, a chronic stable or slow growing lesion for many years may present with a sudden rapid increase in size [4]. After local resection, phyllodes tumor tends to have a high local recurrence rate from 30-65%, which is higher in malignant subtype [5]. A retrospective analysis of 295 patients between 1952 and 2010 demonstrated the most common sites of metastases were lungs (75.7 %), bone (18.9 %), brain (10.8 %), and liver (5.4 %), whereas there were no cardiac metastases. Metastases occurred on overage 21 months (2–57) after surgery [6].

Cardiac metastases have been rarely reported in phyllodes tumor. To the best of our knowledge, there are seven reported cases of cardiac metastases and one case report of pulmonary artery involvement from breast phyllodes tumors [7–10]. In 1998, Myojin et al. from Japan described a case of right ventricular mass in a 47-year-old women presenting with severe hemodynamic shock. They were able to resect a large multilobular phyllodes tumor and perform tricuspid valvuloplasty, but the patient

succumbed to multiorgan failure 15 days later [7]. In 2011, Garg et al. described a similar case in a 35-year-old woman who died 8 days after removal of a right ventricular phyllodes tumor metastasis [11]. In 2014, Mačák et al. described a case of a right ventricular metastatic phyllodes tumor with osteosarcomatous differentiation in a 74-year-old woman which lead to her death from cardiac failure shortly after presentation [12]. In 2014, Bhambhani et al. described a case of phyllodes tumor metastasis to the left atrium in a 50-year-old women presenting with tachycardia, dyspnea and atrial fibrillation. Cardiac surgery removed a large mass from the left atrium. Subsequent whole body [^{18}F]FDG PET/CT imaging revealed an FDG-avid mass abutting the left superior pulmonary vein with access into cardiac chamber. Additional small FDG-avid nodules presumed to be hematogenous metastases were found in ascending aorta and abdominal wall [10]. In 2009, Nakatsu et al. described a right ventricular outflow tract and main pulmonary artery mass in a 65-year-old women presenting with progressive chest pain and dyspnea. A lifesaving surgery was performed. She, however, succumbed to her tracheal metastases 77 days after the cardiac surgery [9]. All of these patients had prior history of a prior breast mass resection with or without radiotherapy.

Several pathways of metastatic involvement of the heart have been described, including tumor embolization and hematogenous spread, lymphomatous spread, and direct contiguous extension [13]. Studies have shown a high degree of hematogenous spread in malignant phyllodes tumors at 20% [14–16]. Hematogenous dissemination in phyllodes tumor has been associated with a very poor prognosis with most patients dying in a short time after diagnosis [17]. A high incidence of local recurrences and delayed distant metastases warrant close surveillance of these patients.

CONCLUSION

This case report describes the salient [^{18}F]FDG PET/CT imaging features of cardiac metastases of phyllodes tumor. This case also highlights the importance of keeping this possibility in differential diagnoses in patients who have a prior history of phyllodes tumors to ensure early diagnosis and treatment of such rare tumors.

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