

¹⁸F-FDG PET/CT imaging of malignant PEComa originating in the pericardium: A case report

Abstract

Primary perivascular epithelioid cell tumors (PEComas) originating in the pericardium are extremely rare. We present a rare case of a malignant PEComa arising from the pericardium, with abnormal fluorine-18-fluorodeoxyglucose (¹⁸F-FDG) uptake maximum standardized uptake value (SUVmax 23.1) on positron emission tomography/computed tomography (PET/CT) scan, indicating its highly aggressive nature. Imaging revealed a soft tissue mass in the left atrial appendage with poorly defined borders to adjacent structures. The patient underwent CT-guided biopsy, and histopathological examination confirmed the diagnosis of malignant PEComa. This case underscores the atypical imaging features of a malignant pericardial PEComa, which, due to its elevated ¹⁸F-FDG uptake, should be considered in the differential diagnosis of cardiac masses with similar PET/CT characteristics, as well as for distinguishing between benign and malignant lesions.

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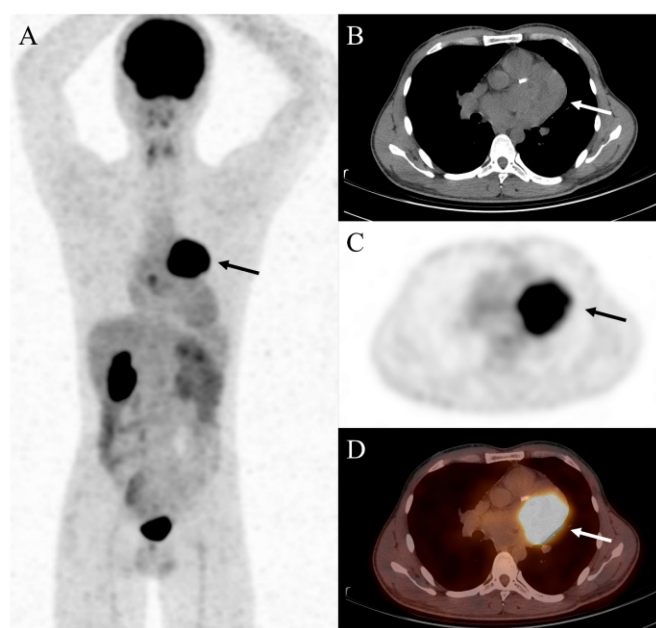


Figure 1. A 30-year-old male patient presented with sudden onset of left shoulder and upper arm pain 6+ months ago, without any obvious triggers. This was followed by chest pain in the precordial region during sleep, described as persistent, crushing pain. After admission to a local hospital, the patient lost consciousness and was resuscitated. Acute myocardial infarction was considered, and emergency coronary angiography and stent implantation were performed. Cardiac computed tomography (CT) suggested the presence of a cardiac mass. Subsequently, the patient did not experience further chest pain, chest tightness, fever, visual disturbances, dizziness, or other discomforts. To clarify the nature of the mass, the patient was referred to our hospital and instructed to maintain a 6-hour fasting state prior to undergoing the positron emission tomography (PET)/CT scan. Maximum intensity projection (MIP) image (A), axial CT (B), PET (C) and fusion images (D) revealed a soft tissue mass in the upper outer region of the left atrial appendage, with heterogeneous density. The largest cross-sectional area measured 66×52mm, and there was an unclear boundary with the pulmonary artery trunk and the left ventricular apex. Fluorine-18-FDG uptake was abnormally increased, with a maximum standardized uptake value (SUVmax) of 23.01. The imaging features were suggestive of a neoplastic lesion. The patient subsequently underwent "CT-guided cardiac mass biopsy," and pathology results confirmed a soft tissue tumor. The tumor cells were spindle-shaped, with some cytoplasm showing red staining and other cytoplasm appearing clear. Immunohistochemistry showed that the tumor cells were SMA (+), HMB45 (-), desmin (-), A103 (-), MITF (-), PCK (-), EMA (-), PAX-8 (-), TFE3 (-), CD63 (+), and the Ki67 proliferation index was 15%. The diagnosis was consistent with perivascular epithelioid cell tumors (PEComa). Perivascular epithelioid cell family tumors are classified based on biological behavior into benign, borderline, and malignant categories. The cells in this focal region appeared relatively mild; however, several high-risk factors were present: the tumor was large, some areas showed increased cytological atypia, nuclear division was active, pathological mitosis was observed, and necrosis was present. Considering these factors, a final diagnosis of malignant PEComa was made. Perivascular epithelioid cell tumor is a rare mesenchymal tumor, typically originating from the gastrointestinal tract, retroperitoneum, uterus, or soft tissues of the body wall [1, 2]. However, there are few reports of tumors originating in the pericardium. To the authors' knowledge, the only report on the PET/CT imaging features of a pericardial PEComa was by Li et al. in 2023, describing a case of a benign perivascular epithelioid cell tumor in the pericardium in a 32-year-old female [3]. In Li's case, the fluorine-18-fluorodeoxyglucose (¹⁸F-FDG) uptake of the pericardial PEComa was mildly elevated, similar to myocardial uptake, with a SUVmax of only 3.1. Both the imaging and pathological results indicated a benign tumor. In contrast, our case shows significantly elevated ¹⁸F-FDG uptake (SUVmax of 23.1), and pathology suggests a malignant PEComa. This case highlights the rare ¹⁸F-FDG PET/CT imaging findings of a malignant PEComa originating in the pericardium, which contributes to the understanding of this potentially rare tumor. Positron emission tomography/CT imaging can be helpful in differentiating between benign and malignant lesions, staging, and monitoring disease progression.

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