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Case Report
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Acute Myocardial Infarction Caused by Coronary Artery Compression by Perivascular Epithelioid Cell Tumor: A Case Report

*Qiubing Zhang[1#], Fang Gou[2#], Zhitao Yan[1], Yuanjun He[1] and Ping Shi[2]**

[1]Department of Cardiology, Guangyuan Central Hospital, China

[2]Guangyuan Central Hospital, China

[#]These authors contributed equally to this work

Abstract

A 30-year-old male underwent percutaneous coronary intervention for acute myocardial infarction. The patient developed fever after the operation, and CT revealed a 6.4 cm × 4.8 cm soft tissue mass around the proximal segment of the right coronary artery, between the left main pulmonary artery and the left heart. The density of the mass was not uniform; the boundary was unclear, with invasion of the pericardium, left coronary artery, some branches of the left pulmonary artery and vein, and the left heart. The pathological examination at the superior hospital suggested that the mass was mainly composed of spindle cells with slightly transparent cytoplasm. Immunohistochemistry showed HMB-45 (+), EMA (±), SMA (+), MDM2 (+), and CD63 (partial +). No MDM2 gene amplification was found by Fluorescence *in situ* Hybridization (FISH). This is the first reported case of acute myocardial infarction caused by coronary artery compression by myocardial Perivascular Epithelioid cell tumor (PEComa), which differs from previously reported cases of pericardial/myocardial PEComas.

Keywords: Perivascular Epithelioid Cell tumor (PEComa); Coronary artery; Acute myocardial infarction

Introduction

Cardiac tumors are very rare in clinical practice, but affect patients of all ages. Benign tumors account for 80%, mainly including myxoma, papillary elastic fibroma and rhabdomyoma, of which left atrial myxoma is more common. Malignant tumors account for 20%, mainly including sarcomas, which were most common in right atrial and metastases. In 2015, the fourth edition of World Health Organization (WHO) classification of lung, thymus and heart tumors published by the International Agency for Research on Cancer (IARC) added unknown biological behavior tumors and germ cell tumors. Unknown biological behavior tumors include inflammatory myofibroblastoma and ganglioma, while germ cell tumors include teratoma and yolk sac tumor [1]. According to the WHO definition of soft tissue tumor in 2020, Perivascular Epithelioid cell tumor (PEComa) belongs to the category of tumors with uncertain differentiation [2]. PEComa is a mesenchymal tumor family composed of Angiomyolipoma (AML), Lymphangioleiomyomatosis (LAM), renal capsule tumor, clear cell myomelanoma and lung clear cell "sugar" tumor, which co-express melanocytes and smooth muscle markers, and have the characteristics of perivascular distribution. PEComa can occur in viscera (especially gastrointestinal tract and uterus), retroperitoneal and abdominal pelvic region, and some soft tissues and skin. It is most common in women, and the incidence rate of male to female is 1:6 [3]. Herein, we reported the case of a 30-year-old male with acute myocardial infarction caused by tumor compression of the coronary artery.

Case Presentation

A 30-year-old man reported no history of hypertension, diabetes, heart disease, autoimmune disease and other diseases. He denied recent history of upper respiratory tract infection and diarrhea as well as any family history of stroke. The patient reported smoking for 10+ years, 10 cigarettes per day, occasional drinking, and long-term staying up late and other detrimental living habits. Three days before admission, the patient had no obvious inducement for shoulder and back pain, and no obvious symptoms such as chest tightness and chest pain. The patient self-administered ibuprofen,

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Correspondence:
Ping Shi, Guangyuan Central Hospital, Guangyuan 628000, Sichuan, China, E-mail: zqbgf715@163.com

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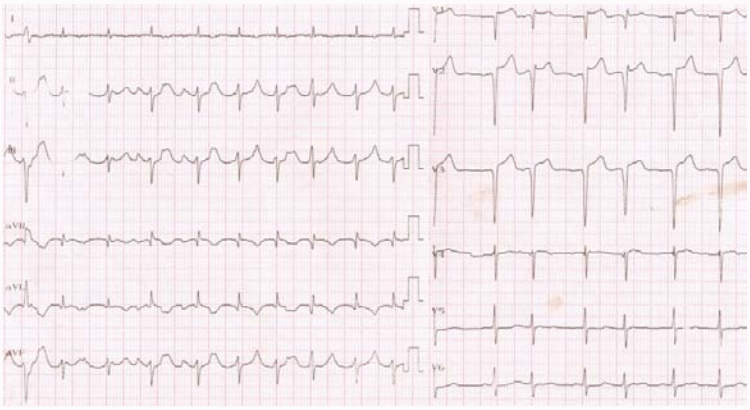

Figure 1: The electrocardiogram showed avR and V1-V4 ST segment elevation.

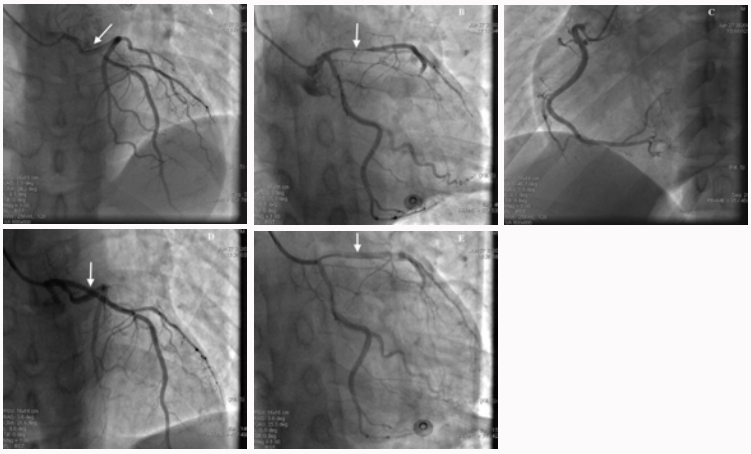

Figure 2: The percutaneous coronary intervention in the emergency ward showed no obvious stenosis in the left main coronary artery, about 99% stenosis in the proximal segment of the left anterior descending artery, and no obvious stenosis in the left circumflex artery and the right coronary artery. A 3.0 mm × 30 mm BuMA stent was implanted into the proximal segment of the left anterior descending artery after 2.5 mm × 15 mm balloon pre-dilation. Angiography showed that the stenosis had disappeared.

which was not effective. Seven hours before admission, the patient felt persistent worsening of shoulder and back pain, accompanied by precordial pain, without other related symptoms. Physical examination indicated Blood pressure 93/69 mmHg, and no other obvious abnormality. Laboratory examination showed leukocytes 15.57×10^9/L, neutrophils 13.43×10^9/L, neutrophil percentage 86.2%, C-reactive protein 102.36 mg/l, cTnI 4.4 ng/ml, CK-MB >60 ng/ml, and myoglobin 428.4 ng/ml. No obvious abnormality was found in other biochemical examinations. The electrocardiogram showed avR and V1-V4 ST segment elevation (Figure 1), which was diagnosed as acute anterior wall ST segment elevation myocardial infarction. The patient was administered aspirin 300 mg, ticagrelor 180 mg and rosuvastatin 10 mg. Thereafter, percutaneous coronary intervention in the emergency ward showed no obvious stenosis in the left main coronary artery, about 99% stenosis in the proximal segment of the left anterior descending artery, and no obvious stenosis in the left circumflex artery and the right coronary artery. A 3.0 mm × 30 mm BuMA stent was implanted into the proximal segment of the left anterior descending artery after 2.5 mm × 15 mm balloon pre-dilation. Angiography showed that the stenosis had disappeared (Figure 2).

The patient developed recurrent low grade fever of 38°C from the second day after operation. After ruling out infections of digestive and urinary systems, chest CT was performed to clarify the cause of fever. Chest CT showed that stent shadow was seen in the left anterior descending artery, and about a 6.4 cm × 4.8 cm soft tissue mass was seen in the left pulmonary artery and left heart, with uneven density, unclear boundary, suggestive of neoplastic lesions or hematoma. The enhanced CT further revealed an irregular mass around the proximal right coronary artery and between the left main pulmonary artery and left heart (Figure 3); suggestive of lesions invading the pericardium, left coronary artery and partial branches of left pulmonary artery and vein, and left heart. The transthoracic echocardiography showed EF 30%, a solid mass of about 6.3 cm × 5 cm × 6 cm near the left atrial appendage and the main pulmonary artery, with blood vessels inside, local invasion of left atrium and left ventricular wall, unclear boundary with myocardium, and abnormal segmental movement of left ventricular wall (Figure 4). According to the results of the patient's CT and echocardiography, we reexamined the patient's angiographic images (Figure 5), which showed a space-occupying lesion in the left heart, blood vessels in the left anterior descending artery and the left circumflex artery supplying to the space-occupying lesion, and some vessels disappeared after stent implantation. In order to clarify the diagnosis and treatment, the patient was referred to the superior hospital.

The results of CT and echocardiography at the superior hospital were consistent with those at our hospital. PET-CT Figure 6 showed a mass with increased glucose metabolism in the outer upper area of

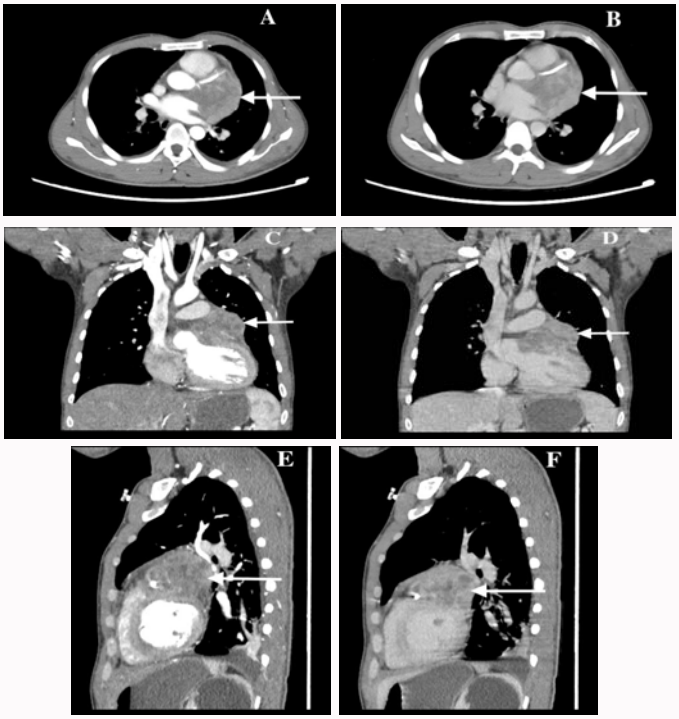

Figure 3: The enhanced CT revealed a 6.4 cm × 4.8 cm soft tissue irregular mass around the proximal right coronary artery and between the left main pulmonary artery and left heart, with invasion of left coronary artery and partial branches of left pulmonary artery and vein, and left heart.

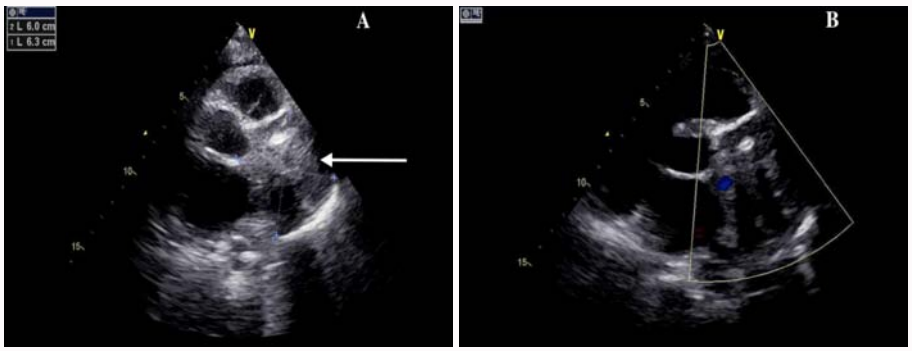

Figure 4: The transthoracic echocardiography showed a solid mass of about 6.3 cm × 5 cm × 6 cm (Figure 4A) near the left atrial appendage and the main pulmonary artery, with blood vessels inside (Figure 4B), local invasion of left atrium and left ventricular wall, unclear boundary with myocardium, and abnormal segmental movement of left ventricular wall.

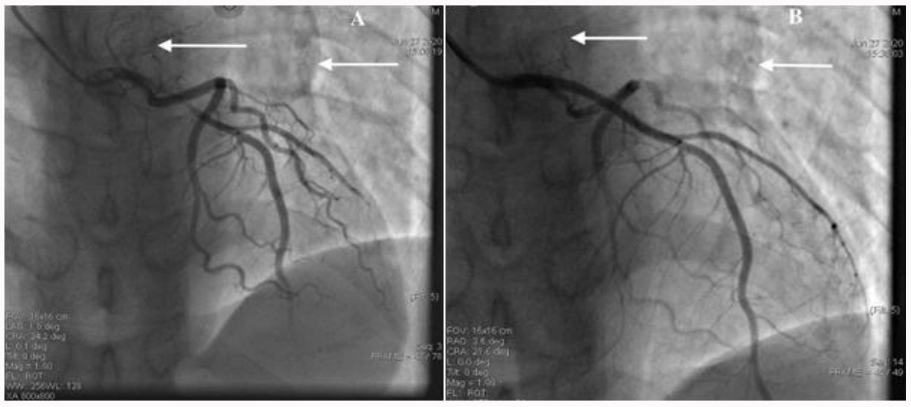

Figure 5: Reexamination of the angiographic image showed a space-occupying lesion in the left heart, blood vessels in the left anterior descending artery and the left circumflex artery supplying to the space-occupying lesion (Figure 5A), and some blood vessels disappeared after stent implantation (Figure 5B).

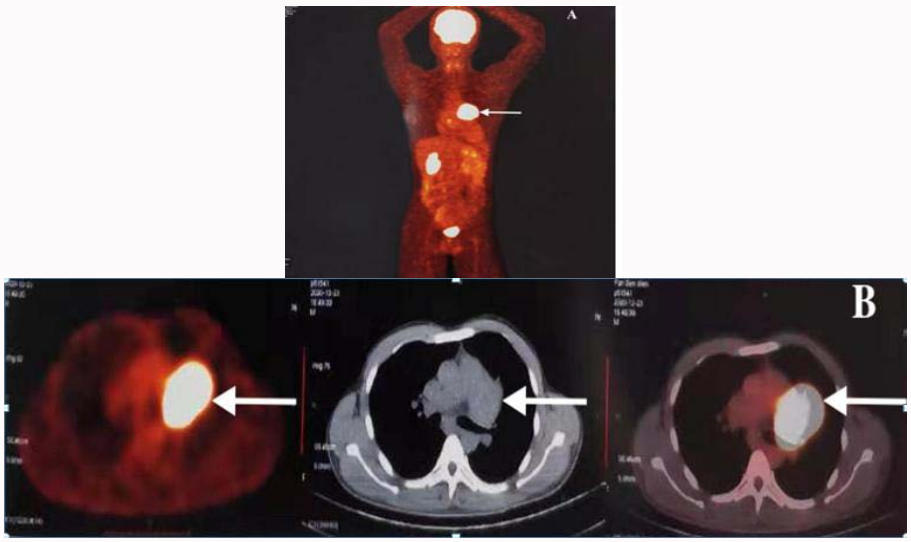

Figure 6: The PET-CT showed a mass with increased glucose metabolism in the outer upper area of the left atrial appendage, with a maximum cross-sectional area of 66 mm × 52 mm, increased glucose metabolism in the left part of the left atrial appendage, but the nature of the mass was unknown.

the left atrial appendage, with a maximum cross-sectional area of 66 mm × 52 mm, increased glucose metabolism in the left part of the left atrial appendage, the nature of the mass was unknown. Histological biopsy of the heart revealed that the tumor had short spindle shape and slightly transparent cytoplasm. Immunohistochemistry showed that the tumor cells were HMB45 (+), EMA (+), SMA (+), MDM2 (+), and CD63 (partial +). Fluorescence *in situ* Hybridization (FISH) showed no amplification of MDM2 gene. However, the histopathological sections of the patient could not be obtained from the superior hospital. Therefore, the microscopic findings of the tumor could not be provided in this case report.

Because the tumor invaded the left heart and the boundary between the tumor mass and the myocardium was unclear, effective tumor resection could not be performed. The patient has been following-up for 15 months. The patient has recurrent symptoms of dyspnea and has been repeatedly hospitalized. He is currently waiting for heart transplantation.

Discussion

Cardiac tumors, both benign and malignant are very rare especially PEComa of myocardium or pericardium. The clinical manifestations of cardiac tumors are diverse and lack specificity. Their symptoms and signs are dependent on the growth site, growth rate, size, invasion force, activity, tumor exfoliation, bleeding, and necrosis. Previously reported cases of myocardial or pericardial PEComa were identified during medical treatment after embolization events, hemodynamic obstruction, and other systemic symptoms, but no cases of acute ST-segment elevation myocardial infarction caused by myocardial or pericardial PEComa compression on coronary arteries have been reported. This case was a 30-year-old male with clinical manifestations of acute chest pain, and shoulder and back pain. The electrocardiogram showed acute myocardial infarction and percutaneous coronary intervention was performed in the emergency ward. Cardiac space-occupying lesions were found by chest CT because of recurrent fever, and confirmed by histopathological biopsy as PEComa. This is the first reported case of acute ST-segment elevation myocardial infarction caused by PEComa compression of the coronary artery.

PEComa mostly occurs before the age of 40 years, and consists of epithelioid cells and/or spindle cells that continuously express HMB-45. To date, less than 10 cases of PEComa of myocardium or pericardium have been reported, with an average age of 24.5 years (2 to 48 years) and no gender preponderance. These patients have corresponding signs and symptoms. The most common symptoms are dyspnea and arrhythmia. At the time of diagnosis and autopsy, it was found that the tumors were at least 6 cm in size. Tumors have been found in the pericardial cavity and/or closely related to the large vessels in the posterior wall of the heart [4,5].

With the continuous development of medical technology, the application of transthoracic or transesophageal echocardiography, multi-slice spiral CT, Magnetic Resonance Imaging (MRI), coronary angiography and PET-CT have become very important in the diagnosis of cardiac space-occupying lesions or tumor. These methods can quickly obtain real-time cardiac images, with high time and spatial resolution, and tumor tissue characteristics. However, the final diagnosis depends on histopathological examination. The main functions of coronary angiography in patients with cardiac tumors are: (1) to show the vessels that can be ligated during the operation; (2) If there is a visible blood supply, it may not be an intracardiac thrombus; (3) Myxoma can be found earlier than echocardiography; (4) Some myxomas can bleed into the right or left atrium, which can be seen in coronary angiography [6]. In this case report, the patient underwent coronary angiography after acute myocardial infarction, which showed severe stenosis of the left anterior descending coronary artery, but the presence of tumor was not detected. The patient underwent chest CT due to recurrent fever, which showed cardiac space-occupying lesions. Reexamination of the coronary angiography showed that the coronary artery had blood vessels supplying to the tumor, and some blood vessels of the tumor were lost after stent implantation. Subsequently, the patient underwent transthoracic echocardiography, enhanced CT, PET-CT and other examinations to further confirm the existence of the tumor, the size and shape of the tumor and its relationship with surrounding tissues. The histological sections showed that the tumor cells were short spindle-shaped and the cytoplasm was slightly transparent. The immunohistochemical results were HMB45 (+), EMA (±), SMA (+), MDM2 (+) and CD63

(partial +), which confirmed that the tumor was PEComa. However, the histopathological sections of the patient could not be obtained from the superior hospital. Therefore, the microscopic findings of the tumor could not be provided in this case report.

Surgery or conservative treatment should be selected according to the patient's condition. However, PEComa is very rare and no optimal treatment has been established. Studies have shown that chemotherapy and radiotherapy have no significant benefit for patients with PEComa, and surgical resection is the only treatment for the primary lesion, local recurrence and metastasis [3,7]. Among the reported patients with PEComa, seven patients underwent surgical treatment, of which two died after surgery, and the remaining five patients showed no tumor recurrence during short-term follow-up, but long-term follow-up results were not provided [5]. In this case, chest echocardiography and enhanced CT examination indicated that PEComa had invaded the myocardium, and surgical resection may not have the desired effect. For patients with this condition, multidisciplinary discussions are needed to develop a treatment plan that is most appropriate for the patient.

References

1. Burke A, Tavora F. The 2015 WHO classification of tumors of the heart and pericardium. J Thorac Oncol. 2016;11(4):441-52.
2. Sbaraglia M, Bellan E, Dei Tos AP. The 2020 WHO classification of soft tissue tumors: News and perspectives. Pathologica. 2021;113(2):70-84.
3. Hornick JL, Fletcher CDM. PEComa: What do we know so far? Histopathology. 2006;48(1):75-82.
4. Vang R, Kempson RL. Perivascular epithelioid cell tumor ('PEComa') of the uterus: A subset of HMB-45-positive epithelioid mesenchymal neoplasms with an uncertain relationship to pure smooth muscle tumors. Am J Surg Pathol. 2002;26(1):1-13.
5. Zhang L, Carpenter D, Dehner LP. Ruptured pericardial perivascular epithelioid cell tumor (PEComa) leading to sudden death: An autopsy case report and review of the literature. Cardiovasc Pathol. 2016;25(1):63-6.
6. Gupta PN, Sagar N, Ramachandran R, Rajeshekharan VR. How does knowledge of the blood supply to an intracardiac tumor help? BMJ Case Rep. 2019;12(2):e225900.
7. Armah HB, Parwani AV. Perivascular epithelioid cell tumor. Arch Pathol Lab Med. 2009;133(4):648-54.