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Compression of the Pulmonary Artery and Coronary Artery Caused by Pulmonary Epithelioid Hemangioendothelioma: A Case Report

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ABSTRACT

A 63-year-old woman with a five-month history of pulmonary epithelioid hemangioendothelioma (PEH) presented to the emergency department, due to worsening dyspnea and chest pain. The electrocardiography showed a pattern of ST-segment elevation in leads I, AVL, and poor R-wave progression consistent with anterolateral ischemia. Emergent coronary angiography revealed severe stenosis of the left main coronary artery. Then, contrast-enhanced computed tomography scan indicated the right pulmonary artery and left main coronary artery narrowing by compression of metastasized PEH. Finally, the patient died of deteriorated multi-organ failure.

INTRODUCTION

Pulmonary artery stenosis (PAS) is commonly congenital and involves narrowing of vessels, most often in conjunction with congenital cardiac defects, and can form anywhere within the pulmonary artery tree. It occurs in 2%-3% of all patients with congenital heart disease [Bacha 2001]. PAS also can result from an acquired lesion, such as tumor compression, but it is rare [Armstrong 2013]. Acute myocardial infarction (AMI) is characterized by an occlusive coronary artery, which mostly is caused by plaque rupture and subsequent thrombosis and sometimes caused by non-atherosclerotic diseases, such as coronary embolism, aortic dissection, coronary artery anomalies, arteritis, and other diseases [Thygesen 2018; Fabris 2016]. Tumor compression or invasion of the coronary artery leading to occlusion has been reported but is rare, especially involving the pulmonary artery and coronary artery simultaneously [Lu 2015; Al-Hawwas 2018].

Pulmonary epithelioid hemangioendothelioma (PEH) is a rare low-to-intermediate malignant vascular tumor that commonly affects noncardiac organs in previous reports [Rosenberg 2018]. Here, we present a case that showed PEH, resulting in severe stenosis of the right pulmonary artery and left main coronary artery and AMI, due to external compression.

CASE REPORT

A 63-year-old female patient was referred to our hospital, due to progressive dyspnea with swelling of lower limbs for three months and worsening chest pain for one day. She was diagnosed and treated as heart failure for three months prior. When she came into the emergency department, she presented dysphoria, orthopnea, and diaphoresis. Her vital signs on admission were as follows: body temperature 36.1°C, heart rate 140bpm, respiratory rate 30bpm, blood pressure 129/90mmHg, and oxygen saturation of 90% on room air. Electrocardiography (ECG) findings revealed ST-segment elevation in leads I, aVL, and poor R wave progression. (Figure 1) Troponin T rose from 25.9ng/L (relative index 0-14ng/L) initially to 576.7ng/L 2 hours later. Emergent coronary angiography revealed 90% distal occlusion of the left main coronary artery (LMCA), 90% ostium occlusion of the left anterior descending coronary artery (LAD), and suspected compression of aortic root. (Figure 2) Then contrast-enhanced computed tomography scan immediately was performed, which showed the compression of soft tissues on the LMCA and the right pulmonary artery, resulting in severe stenosis of these arteries. (Figure 3) Bedside transthoracic echocardiography showed severe biventricular dysfunction. The left ventricular ejection fraction (LVEF) was 25%, and fractional area change (FAC) was 20%. Further illness history inquiry showed she received left pulmonary nodule resection five months prior without the following treatment. She denied a history of coronary heart disease. Postoperative pathologic findings indicated PEH (ERK (+), CD31 (+), CD34 (±), CAMTA-1(+), TFE-3(-), CK (-), TTF-1(-), P63 (-), Ki67 (+, ~10%)). So, we thought the soft tissues in the CT findings were metastasized PEH. Considering the poor prognosis, the patient refused further intervention treatment. Finally, the patient died of deteriorated multi-organ failure.

DISCUSSION

PAS, most often seen in children with congenital heart diseases, is caused by obstructive lesions in the pulmonary artery and its branches [Kim 2020]. PAS may present either as an

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isolated lesion or in conjunction with cardiac malformations. Congenital PAS is observed in congenital heart diseases, such as tetralogy of Fallot, pulmonary valve stenosis, ventricular septal defect, and patent ductus arteriosus, as well as in genetic diseases, such as Williams, Alagille, Ehlers-Danlos, and Noonan's syndromes [Bacha 2001]. Acquired PAS commonly is observed in patients following cardiac surgery but also is seen in rare conditions, such as fibrosing mediastinitis or mediastinal tumors [Yang 2021]. Patients with PAS may present dyspnea, fatigue, tachycardia, and swelling of the lower limbs. These symptoms progressively can worsen over time. If untreated, the condition may progress to right ventricle dysfunction and refractory heart failure. In our case, the patient presented progressive dyspnea over the past month, most likely due to PAS.

The most common cause of AMI is atherosclerosis within the coronary arteries. About 4%-7% of patients with AMI may have no evidence of atherosclerotic lesions demonstrated by coronary angiography or necropsy [Thygesen 2018; Waller 1996]. The nonatherosclerotic causes include coronary embolism, dissection, vasospasm, and congenital coronary artery anomalies. A metastatic tumor also may cause coronary artery stenosis by external compression and result in myocardial ischemia and even AMI, as the case presented [Waller 1996; Al-Hawwas 2018; Waller 1996]. These conditions can be confirmed by invasive angiography and contrast-enhanced CT angiography.

Epithelioid hemangioendothelioma (EHE) is classified as a locally aggressive tumor with metastatic potential by

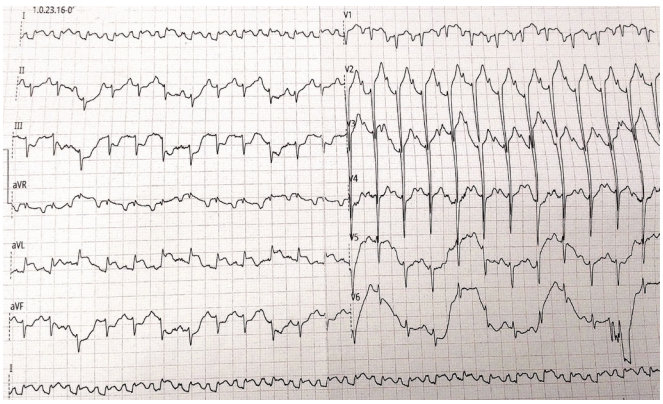

Figure 1. Electrocardiogram on admission showed ST-segment elevation in leads I, aVL, and poor R wave progression.

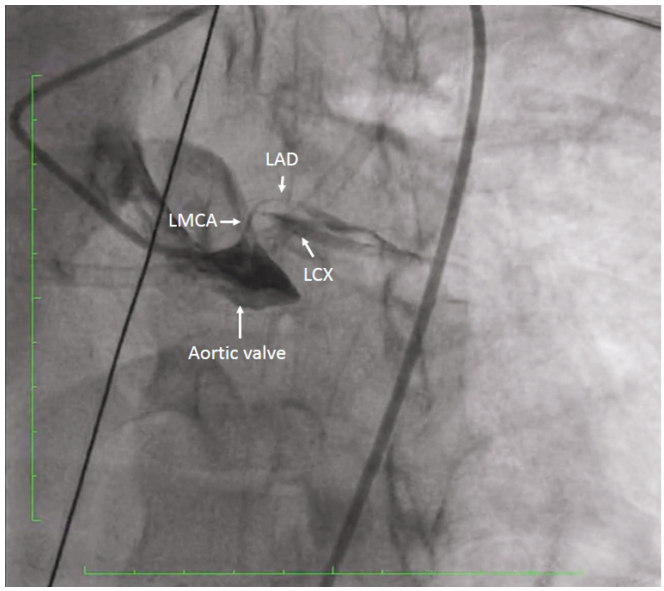

Figure 2. Emergent coronary angiography revealed 90% distal occlusion of LMCA, 90% ostium occlusion of LAD and suspected compression of aortic root (A and B). LAD, left anterior descending coronary artery; LCX, left circumflex coronary artery; LMCA, left main coronary artery

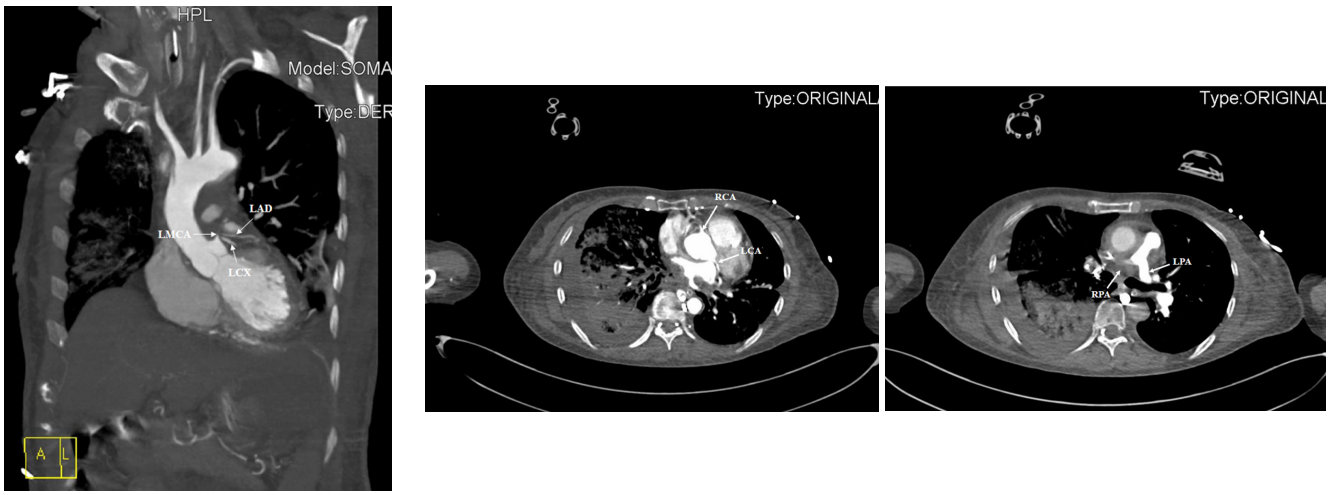

Figure 3. Contrast-enhanced CT scan showed the compression of soft tissues on the LMCA (A and B), as well as the right pulmonary artery (C), resulting in severe stenosis of these arteries. LAD, left anterior descending coronary artery; LCA, left coronary artery; LCX, left circumflex coronary artery; LMCA, left main coronary artery; LPA, left pulmonary artery; RCA, right coronary artery; RPA, right pilmonary artery

the World Health Organization [Choi 2021]. Any site in the body can be affected by EHE, but the most commonly affected organs are the liver, lung and bones, while heart or epicardial tissues rarely are affected. Patients with pulmonary involvement or metastases have poorer prognosis [Lau 2011; Wu 2019; Rosenberg 2018]. Previous reports showed that approximately 30% of cases present as PEH [Rosenberg 2018; Sardaro 2014]. PEH is a rare malignant vascular neoplasm described as intermediate between hemangioma and angiosarcoma, with 60% of 5-year survival probability [Bagan 2006]. The clinical symptoms and signs are nonspecific, such as chest pain, cough, dyspnea, hemoptysis, etc. Some asymptomatic patients are discovered accidentally. PEH presents distant hematogenous metastases mainly in the liver but also in the bone, skin, serosa, spleen, and retroperitoneum [Sardaro 2014]. Metastasis in the mediastinum is possible as well.

There are limited clinical data to guide standard treatment strategies, due to the rarity of PEH cases. In asymptomatic patients, watchful waiting is sometimes a reasonable strategy [Kitaichi 1998]. Surgery resection remains the first choice for the cases of unilateral single or multiple nodules. For metastatic disease, the treatment of choice includes cytotoxic chemotherapy, immune therapy, and targeted therapy. But these are mainly reported by case reports and small-sample case series [Rosenberg 2018]. For the patient in our case, balloon angioplasty and stent implantation of the stenosed arteries seemed reasonable treatment options. Relief of stenosis can improve the patient's hemodynamics, clinical symptoms, and short-term prognosis [Kim 2020; Han 2019]. The challenge is defining the goals of therapy and balancing the risks of treatment. Unfortunately, given the complexity of the patient's condition and poor prognosis, the patient refused further intervention treatment.

In conclusion, our case suggested that early therapy and close follow up are necessary for patients with PEH. PEH can simultaneously cause extrinsic compression of the pulmonary artery and coronary artery. Active intravascular intervention may improve the symptoms and short-term prognosis.

REFERENCES

Al-Hawwas M, Tsitlakidou D, Gupta N, Iliescu C, Cilingiroglu M, Marmagkiolis K. 2018. Acute Coronary Syndrome Management in Cancer Patients. Curr Oncol Rep. 20(10): 78.

Armstrong MA, Pollock GF. 2013. Pericarditis and Pulmonary Artery Stenosis Due to an Extragonadal Non-seminomatous Germ Cell Tumor: Case Report and Review of the Literature. The Journal of Emergency Medicine. 45(5): e157-e160.

Bacha EA, Kreutzer J. 2001. Comprehensive management of branch pulmonary artery stenosis. J Interv Cardiol. 14(3): 367-375.

Bagan P, Hassan M, Barthes FLP, Peyrard S, Souilamas R, Danel C, et al. 2006. Prognostic Factors and Surgical Indications of Pulmonary Epithelioid Hemangioendothelioma: A Review of the Literature. The Annals of Thoracic Surgery. 82(6): 2010-2013.

Choi JH, Ro JY. 2021. The 2020 WHO Classification of Tumors of Bone: An Updated Review. Adv Anat Pathol. 28(3): 119-138.

Fabris E, Kilic ID, Caiazzo G, Serdoz R, Foin N, Sinagra G, et al. 2016. Nonatherosclerotic Coronary Artery Narrowing. JACC: Cardiovascular Imaging. 9(3): 317-320.

Han XJ, Li JQ, Khannanova Z, Li Y. 2019. Optimal management of coronary artery disease in cancer patients. Chronic Diseases and Translational Medicine. 5(4): 221-233.

Kim CW, Aronow WS, Dutta T, Spevack DM, Frishman WH. 2020. Treatment of Peripheral Pulmonary Artery Stenosis. Cardiology in Review. 29(3): 115-119.

Kitaichi M, Nagai S, Nishimura K, Itoh H, Asamoto H, Izumi T, et al. 1998. Pulmonary epithelioid haemangioendothelioma in 21 patients, including three with partial spontaneous regression. Eur Respir J. 12(1): 89-96.

Lau K, Massad M, Pollak C, Rubin C, Yeh J, Wang J, et al. 2011. Clinical Patterns and Outcome in Epithelioid Hemangioendothelioma With or Without Pulmonary Involvement. Chest. 140(5): 1312-1318.

Lu DY, Yu WC, Chen CK, Sung SH. 2015. Tumor Invasion of Myocardium Presented with Acute Coronary Syndrome. Acta Cardiol Sin. 31(3): 257-260.

Rosenberg A, Agulnik M. 2018. Epithelioid Hemangioendothelioma: Update on Diagnosis and Treatment. Current Treatment Options in Oncology. 19(4).

Sardaro A, Bardoscia L, Petruzzelli MF, Portaluri M. 2014. Epithelioid hemangioendothelioma: an overview and update on a rare vascular tumor. Oncology Reviews.

Thygesen K, Alpert JS, Jaffe AS, Chaitman BR, Bax JJ, Morrow DA, et al. 2018. Fourth Universal Definition of Myocardial Infarction (2018). Journal of the American College of Cardiology. 72(18): 2231-2264.

Waller BF, Fry ET, Hermiller JB, Peters T, Slack JD. 1996. Nonatherosclerotic causes of coronary artery narrowing--Part I. Clin Cardiol. 19(6): 509-512.

Waller BF, Fry ET, Hermiller JB, Peters T, Slack JD. 1996. Nonatherosclerotic causes of coronary artery narrowing--Part III. Clin Cardiol. 19(8): 656-661.

Wu X, Li B, Zheng C, Hong T, He X. 2019. Clinical characteristics of epithelioid hemangioendothelioma: a single-center retrospective study. European Journal of Medical Research. 24(1).

Yang S, Wang J, Li J, Huang K,Yang Y. 2021. Refractory pleural effusion as a rare complication of pulmonary vascular stenosis induced by fibrosing mediastinitis: a case report and literature review. Journal of International Medical Research. 49(5): 3000605211010073.