

Case Report

A Case of Pulmonary Artery Intimal Sarcoma Presenting as a Lung Tumor

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- Pulmonary artery intimal sarcoma
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- Proton therapy
- Surgery

Abstract

We report a very rare case of pulmonary artery intimal sarcoma presenting as a lung tumor. A 30s-year-old female was referred to our hospital with continuous cough. Chest X-P and contrast-enhanced CT showed a large oval mass in right lung, and FDG-PET/ CT revealed increased uptake of FDG in the right lung tumor, mediastinum, and right ventricle. She underwent a surgical treatment, and the pathological finding was pulmonary artery intimal sarcoma. Proton-beam radiotherapy was applied to treat a local recurrence which was considered to be radioresistant, and the effect was excellent.

INTRODUCTION

Pulmonary artery intimal sarcoma (PAS) is very rare disease, and the prognosis is generally poor, with death resulting in most patients within a few months from diagnosis [1,2]. PAS are often misdiagnosed as pulmonary thromboembolism because of their similar clinical symptoms [3,4]. The tumor mostly originated in the pulmonary trunk, and both the right and left branches are likely involved [1,5]. Surgery is considered to be a best choice for treating PAS. On the other hands, response to chemotherapy and radiotherapy is poor, and these treatments still remains controversial. Here, we report a very rare case of PAS that presented as a lung tumor.

CASE PRESENTATION

A female patient in her 30s was referred to our hospital with a complaint of continuous cough. The patient had no smoking history and no remarkable medical or familiar history. Chest X-ray radiography showed a large oval mass and peripheral infiltrative shadow in right middle lung field (Figure 1). Chest enhanced computed tomography (CT) showed a defect in the right pulmonary artery and large enhanced mass in right S6 (Figure 2). Ventilation-perfusion lung scintigraphy showed

a defect of blood flow in the right lung. Fluorodeoxyglucose-positron emission computed tomography (FDG-PET / CT) was performed in the differential diagnosis between malignant tumors and benign disorders. FDG-PET / CT showed an intense uptake in the right lung mass as well as in the mediastinum and right ventricle (Figure 3). Bronchoscopic examination revealed edematous and obstructing changes in right B6. The patient underwent the right pneumonectomy and pulmonary artery angioplasty. The tumor replaced as a S6 tumor and an obstructive mass in right main trunk of pulmonary artery, and extended into right ventricle. Histological examination of the resected specimen showed spindle and anaplastic cells with nuclear pleomorphism, and diagnosed as pulmonary artery intimal sarcoma. Twelve months after the surgery, Magnetic Resonance Imaging (MRI) revealed local recurrence in right ventricular outflow tract (Figure 4). Proton-beam therapy was selected for treating local recurrence of PAS, because PAS was thought to be radioresistant for achieving local control. The patient was admitted to Proton Therapy Center of Tsukuba University to undergo proton-beam radiotherapy with a total dose of 62.7 GyE in 19 fractions after 10 Gy of conventional radiotherapy in our hospital. Fifteen months after proton-beam radiotherapy, CT showed complete response of recurrent tumor.



Figure 1 Chest radiograph showing a lung tumor in the right middle lung field.

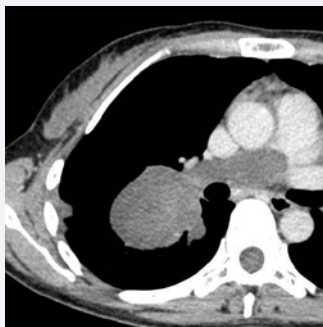


Figure 2 Chest enhanced CT showing a defect in the right pulmonary artery and large enhanced mass in right S6.

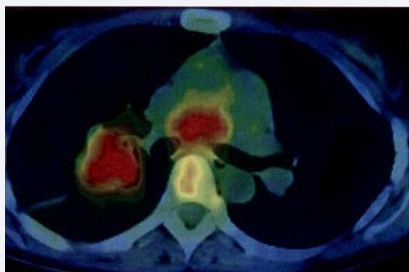


Figure 3 FDG-PET/ CT revealing massive FDG accumulation in the right lung tumor and in the lesion in the mediastinum.



Figure 4 Chest MRI demonstrating a local recurrence in right ventricular outflow tract.

DISCUSSION

PAS is a very rare disease with about a few hundred cases reported to date [3, 6], and aggressive malignancy that leads to heart failure and can metastasize to the bone, lung, and visceral organs. Intimal sarcoma is malignant mesenchymal sarcoma arising in a large artery [5]. The defining features of these tumors are intraluminal growth with obstruction of the lumen and seeding of emboli. Retrograde extension of the intimal sarcoma to the pulmononic valve and the right ventricle is reported to be a very rare condition [3]. Clinical diagnosis of PAS is considered to be difficult, and PASs are often misdiagnosed, and initially diagnosed as pulmonary embolisms. This mistaken diagnosis may lead to inappropriate therapy for PAS patients. PET/ CT is reported to be useful in the differentiation of PAS and pulmonary embolism based on the SUV max value [7, 8]. Lung sarcoma was suspected before surgery in our case because of PET/ CT and CT findings. However the tumor was diagnosed with PAS presenting as a lung tumor postoperatively. Mori reported the pulmonary artery sarcoma presenting as an isolated lung mass is very rare [9]. Inoue reported that significant extension of PAS into lung was also extremely rare [6]. Our case has the right lung tumor and intraventricle tumor in chest X-ray and CT findings. Some papers reported that prognosis of PAS is from 6 months to 2 years [1,7]. Wong described median overall survival was 17 months [10]. Surgical resection of the tumor offers the best choice of prolonged survival. Radical surgery is considered the first-line treatment when tumor localizes unilaterally. Wong reported that surgery provided significant symptomatic improvement and long-term survival [10]. Blackmon described patients underwent curative resection have longer overall survival compared to those with incomplete resection [1]. On the other hands, some reports recommended multimodal therapy combined chemotherapy and / or radiotherapy [1]. However the response of chemotherapy or radiotherapy to PAS is generally poor. The efficacy of chemotherapy or radiotherapy for PAS still remains controversial [4,6]. In this case, we selected proton-beam radiotherapy for achieving local control. Some reports described particle-beams radiotherapy was effective for several sarcomas. Kamada reported carbon-ion radiotherapy produced excellent results for bone and soft-tissue sarcoma [11]. Ishigami reported that proton therapy was especially beneficial for local recurrence of PAS because proton-beam radiotherapy offered a good physical dose distribution [2]. Our case showed good response to proton-beam radiotherapy.

CONCLUSIONS

We reported a very rare case of pulmonary artery intimal sarcoma presenting as a lung tumor. Proton-beam radiotherapy was effective for recurrent tumor of PAS.

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