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**Intimal sarcoma of the pulmonary artery with multiple lung metastases: Long-term survival case**

García-Cabezas S *et al*. Long-term survival case of metastatic PAIS

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**Abstract**

Pulmonary artery intimal sarcoma (PAIS) is a rare tumor with a very poor prognosis. Clinical and radiological findings usually mimic thromboembolic disease, leading to diagnostic delays. The treatment of choice is surgery, and adjuvant chemotherapy and radiotherapy have limited results. We report the case of a 48-year-old male patient, initially suspected with pulmonary thromboembolism. The angio-CT revealed a filling defect in the pulmonary artery trunk. The patient underwent surgery, resulting in with complete resection of the mass with a diagnosis of PAIS. The tumor progressed rapidly in the lung, requiring surgery of multiple lung metastases. The patient was treated with stereotactic body radiation therapy (SBRT) on two occasions for new pulmonary lesions. In the last follow‑up (4 years after initial diagnosis), the patient was disease-free. In conclusion, SBRT proved to be an alternative treatment to metastasectomy, allowing palliative chemotherapy to be delayed or omitted, which may result in improved quality of life.

**Key words:** Intimal sarcoma of the pulmonary artery; Lung metastases; Treatment; Metastasectomy; Stereotactic body radiation therapy

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**Core tip:** Intimal sarcoma of the pulmonary artery is a rare tumor with a very poor prognosis. It has been described in a limited number of reports. This case is a uncommon patient with long-term survival despite having rapid metastatic progression, who maintains a complete remission after initial surgical treatment, completed after occurrence of progression with stereotaxic body radiotherapy.

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**INTRODUCTION**

Pulmonary artery intimal sarcoma (PAIS) is a rare tumor first described by Mandelstamm in 1923[1]. Since then, about 300 cases have been reported in the literature[2,3]. The prognosis is generally poor, with a median overall survival of approximately 17 mo[2-4]. Clinical and radiological findings usually mimic thromboembolic disease, leading to diagnostic delays[5]. Surgical resection of the primary tumor is the best therapeutic option to prolong survival and adjuvant chemotherapy and radiotherapy have limited results[2,3]. The treatment of choice is surgery, as adjuvant chemotherapy and radiotherapy have limited results. When metastases occur, they may be resected in specific patients[6,7]. Otherwise, treatment is generally systemic and palliative in nature. In recent years, stereotactic body radiation therapy (SBRT) for lung metastases, a high-precision external radiotherapy technique, alternative to metastasectomy, has undergone significant development. Prospective phase I/II studies have shown that SBRT is safe and effective as treatment of lung metastases in oligometastatic patients who are not candidates for surgery[8,9]. SBRT of inoperable lung metastases is today considered routine in many centers. We report the case of a rapidly metastatic PAIS, with sustained complete remission following surgical resection and SBRT.

**CASE REPORT**

A 48-year-old male patient presenting with sudden-onset symptoms of sweating, dizziness and falling to the ground, with loss of consciousness, and spontaneous recovery. After observing electrocardiographic changes, he was hospitalized with suspected acute coronary syndrome. An angio-CT was performed, resulting in a diagnosis of pulmonary thromboembolism, with no improvement after anticoagulant therapy. The patient was transferred to our hospital, where a repeat angio-CT was performed, revealing a filling defect in the pulmonary artery trunk, extending from the subvalvular area to the origin of the right pulmonary artery, with no change in size with respect to the previous angio-CT (Figure 1). The patient was operated on for a suspected primary tumor of the pulmonary artery, resulting in with complete resection of the mass, whose pathological result was an intermediate-grade malignant tumor suggestive of PAIS (Figure 2). Following an extension study, CT showed only a nonspecific pulmonary nodule of 4 mm in diameter in the right upper lobe. No adjuvant treatment was given. Three months later, a positron emission tomography-computed tomography (PET-CT) found that the previously mentioned nodule measured 6 mm, suggestive of metastasis. Another 4 mm de novo nodule was found and two more of 2-3 mm in size, possibly granulomas, all without an increase in metabolic activity. The patient underwent surgery where 4 bilateral pulmonary lesions, compatible with metastasis, were resected. Fifteen month later, a new PET-CT reveals a subpleural nodule in the left upper lobe, again suspicious of metastasis (Figure 3A), as well as several millimeter-size nodules reported in the previous CT, not metabolically characterizable. After discussion in a multidisciplinary committee, the patient was given treatment with SBRT (12 Gy × 5 fractions), with excellent control. Six months later, growth of the two new pulmonary lesions noted in the previous CT was observed (Figures 3B and C). Chemotherapy was prescribed, which was rejected by the patient, and a second course of SBRT was given on both pulmonary lesions. Nine months after SBRT, the patient is disease-free by PET-CT.

**DISCUSSION**

PAIS is characterized by insidious growth, causing extensive local invasion and hematogeneous metastases. Because it is a rare tumor, only case reports and small case series have been published, most of them focused on the histopathological findings and surgical aspects of its management[3,4,10-12]. Few patients achieve long-term survival and they are those without disease dissemination. The largest analysis of outcomes of this tumor reported better median survival of patients who received multimodality treatment with respect those who had single treatment (median survival of 24.7 and 8.0 mo, respectively). However, single treatment was defined as either surgery, chemotherapy or radiotherapy alone, instead of surgery without postoperative treatment[2]. Mussot *et al*[3] described a surgical series of 31 patients. They concluded that there appeared to be no statistical survival benefit in those who received adjuvant treatment compared to those who did not. A recent study[4] analyzed 20 patients diagnosed with PAIS obtaining a median overall survival of 17 mo: patients who received postoperative chemo and radiotherapy showed a trend towards better survival compared to those who had surgery alone (24 mo *vs* 8 mo, *P* = 0.3417). Successful cases reported in metastatic patients are anecdotal. Thus, Said *et al*[13] reported a case of pulmonary artery angiosarcoma, with a follow-up of 5 years and multiple repeat lung metastasectomies, which has a disease-free interval of 1 year. Choi *et al*[14] published a case of PAIS with metastases in the thyroid and adrenal glands, 4.7 and 6.3 years, respectively, after initial surgery. Both metastases were surgically resected, with an unusual survival of 12.5 years up to the last follow-up. In our case, dissemination occurred much earlier, as the patient was operated on for lung metastases 5 mo after the initial surgery, though it is likely that patient was already metastatic at diagnosis. He is currently disease-free, 4 years after diagnosis. To our knowledge, this is the first published case of metastatic PAIS with long-term survival treated with surgery and SBRT.

In conclusion, SBRT proved to be an alternative treatment to metastasectomy, allowing palliative chemotherapy to be delayed or omitted, which may result in improved quality of life.

**COMMENTS**

***Case characteristics***

A 48-year-old male presenting sudden-onset symptoms of sweating, dizziness and fall, with momentary loss of consciousness.

***Clinical diagnosis***

Acute coronary syndrome.

***Differential diagnosis***

Thromboembolic disease.

***Imaging diagnosis***

Angio-CT: Filling defect in the pulmonary artery trunk, extending from the subvalvular area to the origin of the right pulmonary artery.

***Pathological diagnosis***

Intermediate-grade malignant tumor suggestive of pulmonary artery intimal sarcoma.

***Treatment***

The patient underwent surgery. The tumor progressed rapidly in the lung, requiring surgery of multiple lung metastases. Finally, he was treated with stereotactic body radiation therapy on two occasions for new pulmonary lesions.

***Experiences and lessons***

This case report describes a uncommon patient with exceptional long-term survival despite having rapid metastatic progression. This case teaches us that SBRT is an alternative treatment to metastasectomy, allowing palliative chemotherapy to be delayed or omitted.

***Peer-review***

The article presents an unusual case of intimal sarcoma of the pulmonary artery. There are a small number of cases reported. This is a rare sarcoma with very good response to treatment with radiotherapy.

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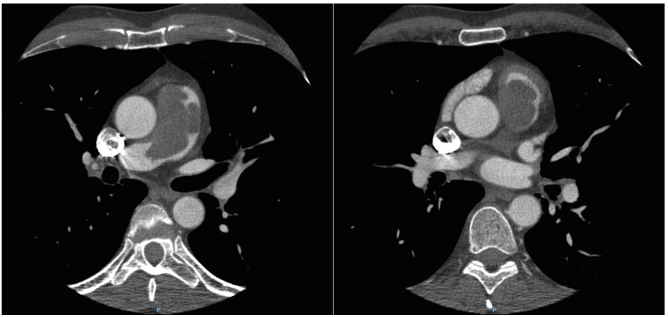
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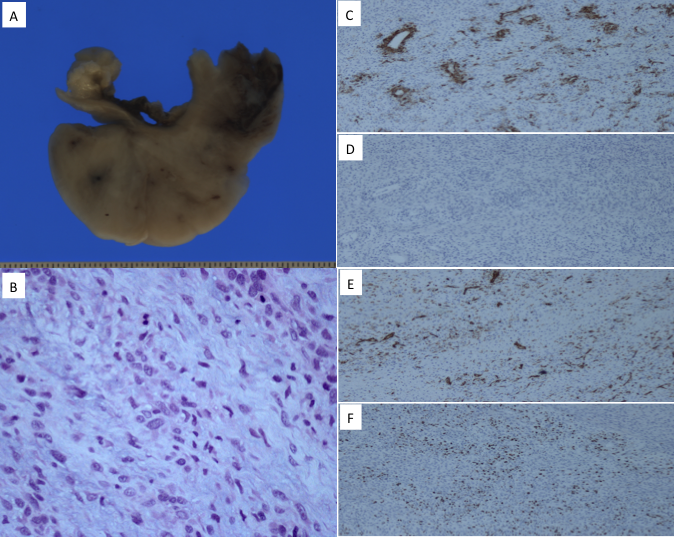
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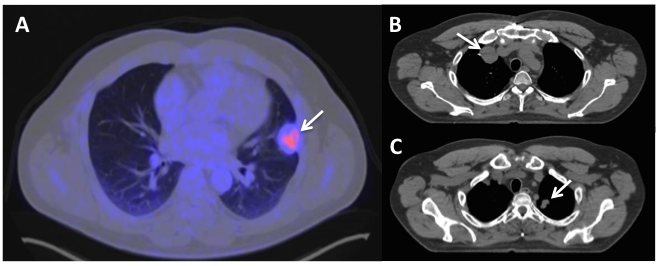
Grade E (Poor): 0



**Figure 1 Angio-CT: Filling defect in pulmonary artery trunk measuring 58 mm × 32 mm × 44 mm, extending from the subvalvular area to the origin of the right pulmonary artery.**



**Figure 2 The patient was operated on for a suspected primary tumor of the pulmonary artery, resulting in with complete resection of the mass, whose pathological result was an intermediate-grade malignant tumor suggestive of Pulmonary artery intimal sarcoma.** A: Macro: View of the pulmonary artery transversal section with infiltrating sarcoma on the lumen; B: High power of the tumor. Note the variable atypia 200 ×. Immunohistochemical stainings 100 ×; C: Smooth muscle actin: Focal tumor cell reaction 100 ×; D: Desmine: Negative 100 ×; E: CD31: Reaction of the endothelium surrounded by negative tumor cells 100 ×; F: Ki-67: Variable proliferation index in tumor cells 100 ×.



**Figure 3 Positron emission tomography–computed tomography, three months later.** A: Subpleural hypermetabolic nodular lesion in left upper lobe (SUV max 3.8) measuring 27 mm × 30 mm, suggestive of tumor activity; B, C: computed tomography, a 38-mm nodule in right upper lobe and another 20-mm nodule in left upper lobe, compatible with metastasis.