Primary Leiomyosarcoma of the Pulmonary Artery: A Case of a 20-Month Survivor after Incomplete Surgical Resection

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Abstract

We report a 53-year-old man who presented with dyspnea and low-grade fever. Cardiac ultrasound showed pulmonary hypertension and an ill-defined echogenic mass within the pulmonary trunk. Computed tomography scan revealed an inhomogeneous mass which filled the main pulmonary trunk with near-total occlusion, and extended into both pulmonary arteries. Anticoagulant therapy was administered based on a presumptive diagnosis of pulmonary thromboembolism. Positron-emission tomography scan was useful for differentiating the mass, which was determined as a pulmonary artery sarcoma by surgical resection. Although complete resection was impossible, the patient survived for 20 months with adjuvant chemotherapy and medical treatment.

Key words: pulmonary sarcoma, prognosis, pulmonary embolism


Introduction

Pulmonary artery sarcoma is a rare disease with a poor prognosis (1). Diagnosis is difficult as the clinical presentation is similar to that of pulmonary thromboembolism. Aggressive surgical treatment is considered the only strategy for prolonged survival (1-4). Herein, we present a patient with a pulmonary artery sarcoma who survived for 20 months with adjuvant chemotherapy and medical treatment following incomplete tumor resection.

Case Report

A 53-year-old man was admitted to our hospital with shortness of breath on exertion and elevation of fever. He had a sustained low-grade fever of unknown origin for over a 6-month period. Although he was treated with prednisolone due to suspected autoimmune disease, his symptoms had gradually worsened. Physical examination revealed distention of the neck vein, but no edema of his face or legs. There was no palpable lymphadenopathy. A Levine grade 3/6 systolic murmur and a widely split second heart sound was heard in the 4th right and 2nd left intercostal spaces, respectively. His C-reactive protein was 8.65 mg/dL and brain natriuretic peptide was 638 pg/mL. Arterial blood gas analysis revealed a slightly low PaO2 of 76.9 mmHg in room air. Chest radiography revealed cardiomegaly (Fig. 1A), and his electrocardiogram showed sinus rhythm and incomplete right bundle branch block. Cardiac ultrasound showed right ventricular (RV) and right atrial (RA) dilatation suggesting severe pulmonary hypertension, and an ill-defined echogenic mass within the pulmonary trunk (Fig. 2A).

We suspected acute or chronic pulmonary thromboembolism, and performed a computed tomography (CT) angiogram. Contrast-enhanced CT scan revealed an inhomogeneous mass filling the main pulmonary trunk with near-total occlusion and extending into both pulmonary arteries (Fig. 1B). A presumptive diagnosis of pulmonary thromboembolism was made, and the patient was treated with...
960,000 units of urokinase within a week, as well as unfractionated heparin. Despite anticoagulant therapy, however, his symptoms failed to resolve (New York Heart Association functional class III/IV), and the mass remained unchanged. Positron-emission tomography scan (PET) showed accumulation of \( ^{18} \text{F-fluorodeoxy glucose} \) in the mass within the lumen of the pulmonary artery, which suggested the presence of malignant neoplasm (Fig. 1C). The mass was ultimately determined as a pulmonary artery sarcoma by surgical resection, the diagnosis of primary leiomyosarcoma of the pulmonary artery was made.

Although a follow-up cardiac ultrasound two weeks later showed no RV and RA dilatation (Fig. 2B) and no pulmonary hypertension, CT scan revealed the residual tumor in both pulmonary arteries (Fig. 1D). The patient subsequently received postoperative chemotherapy consisting of doxorubicin (100 mg twice per month, total 200 mg). The patient was treated as an outpatient for nine months. However, he was re-admitted with shortness of breath and elevation of fever 14 months after surgery. A chest CT scan revealed the massive tumor occupying both pulmonary arteries (Fig. 1E), with worsening of pulmonary hypertension. Although the patient was treated with chemotherapy consisting of pirarubicin (70 mg) and cyclophosphamide (1,300 mg) on day 1 and 21, there was no change in CT scan findings. Prostaglandin I2, (360 μg/day) and sildenafil citrate (60 mg/day) transiently improved the patients' symptoms of pulmonary hypertension. However, due to RV failure resulting in uncontrollable worsening of anasarca with pleural effusion and ascites, the patient died 20 months after surgery.
Autopsy results showed leiomyosarcoma of the pulmonary trunk and right main pulmonary artery, compatible with recurrence of leiomyosarcoma. Tumor embolism and thromboembolism were observed in the right pulmonary artery, bilateral peripheral pulmonary arteries with dissemination to the pulmonary valve, and in the left lower pulmonary artery. Metastases were only observed in the stomach, and these were less than 3 cm in diameter. There were several signs of right heart failure, with myocardial hypertrophy of the right ventricle, systemic edema, rupture of the penile skin, translucent ascites (7,500 mL), and left pleural effusion (200 mL).

Discussion

Pulmonary artery sarcoma is a rare tumor of the cardiovascular system; diagnosis is rarely made preoperatively as the tumor is commonly misdiagnosed as pulmonary thromboembolism. Anderson et al reported six cases of primary pulmonary sarcoma from 150 pulmonary thromboendarterectomy operations (5). Histologically, the most frequent diagnosis in that study was undifferentiated sarcoma (one-third of the histological diagnoses of the reviewed cases), followed by leiomyosarcoma and fibrosarcoma or rhabdomyosarcoma (6). Nevertheless, histopathological classification is
generally not useful clinically or prognostically (7). Most pulmonary artery sarcomas arise within the pulmonary trunk or pulmonary valve lesion, which is consistent with the present case.

Although the present case exhibited a low-grade fever of unknown origin over a 6-month period, the patient did not complain of a dyspnea for several months. Early definitive diagnosis is often difficult as symptoms resulting from pulmonary hypertension do not generally appear until over half of the pulmonary vasculature has been occluded. In addition, the differential diagnosis between thromboembolism and sarcoma is difficult, as in most cases the diagnosis is determined by examining resected or autopsy specimens (6). Nevertheless, a reliable diagnosis of pulmonary artery sarcoma has been reported using enhanced CT or transesophageal echocardiography (8, 9). PET is also useful for differentiating the tumor from a thromboembolism (10), which shows increased uptake in the area of the tumor, especially in patients without extramural invasion such as the present case. Early definitive diagnosis of pulmonary artery sarcoma may also be possible by blood aspiration cytology using an endovascular catheter (4).

Early radical surgical resection with cardiopulmonary bypass is the only strategy for potential cure (3). With this strategy, it is important to obtain a tumor-free margin, thus as much as possible of the pulmonary artery and the tumor is resected, and reconstruction with a cryopreserved pulmonary artery allograft and/or pneumonectomy can also be undertaken if necessary (4, 11). However, in the present case it was impossible to completely resect the tumor because the mass extended too far into both pulmonary arteries and it adhered to the intima and the posterior leaflet of the pulmonary valve.

The prognosis of patients with primary sarcomas of the pulmonary artery is extremely poor (12). In a review of 93 cases, Krüger et al reported a median survival time of 1.5 months, while the median survival time of patients with surgical tumor excision could be prolonged to 10 months (1). In seven cases of pulmonary artery sarcoma reported by Mayer et al, three patients were alive at 21, 35, and 62 months after complete resection, while four patients died 7, 9, 18, and 19 months (3). In a further six cases of pulmonary artery sarcoma reported by Anderson et al, a patient receiving antitumor chemotherapy alone died at 11 months, while a patient with limited tumor resection died at five months (5). The present case seemed to have a favorable survival time despite incomplete surgical resection.

Chemotherapy using doxorubicin is a potential strategy in pulmonary artery sarcoma cases where the surgical tumor resection is incomplete. The two most active agents in the treatment of soft tissue sarcoma are doxorubicin and ifosfamide. Doxorubicin is most active at doses of ≥75 mg/m², with single-agent response rates of approximately 20 to 35% (13), while two-drug combination chemotherapy consisting of ifosfamide and epirubicin has also been used successfully (14). In addition, selective vasodilators such as prostaglandin I₂ and phosphodiesterase-5 inhibitors have been reported to be partially useful for patients with residual tumor or pulmonary hypertension.

In summary, the present case suggests that PET is useful for the diagnosis of pulmonary artery sarcoma, while surgical treatment with adjuvant chemotherapy is an effective strategy for improving of the survival duration of patients even in cases where complete resection is impossible.

The authors state that they have no Conflict of Interest (COI).

References


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