We describe a case of 63-year-old woman with pulmonary artery sarcoma successfully treated with chemotherapy. She developed acute shortness of breath, and left chest and shoulder pain. Although a diagnosis of acute pulmonary embolism was made at a local hospital and she received anticoagulation and thrombolysis therapy, no improvement was achieved. Thereafter, she underwent a pulmonary thromboectomy in our hospital, and the histological diagnosis was intimal sarcoma of the pulmonary artery. Since post-operative computed tomography (CT) scans of the chest showed obvious persistence of an intraluminal hypodense area in the left main pulmonary artery, the patient was treated with four cycles of a doublet chemotherapy consisting of ifosfamide (2.5 g/m²/day) on days 1–5 and epirubicin (45 mg/m²/day) on days 2 and 3. CT scans of the chest after four cycles showed marked regression of the intraluminal hypodense area in the left main pulmonary artery. This is the first case of pulmonary artery sarcoma responding to chemotherapy. Surgical resection is currently the most hopeful treatment for pulmonary artery sarcoma. However, intensive chemotherapy is worth trying in unresectable patients.

Key words: intimal sarcoma – chemotherapy – pulmonary artery

INTRODUCTION

Pulmonary artery sarcoma is a rare tumor that frequently is misdiagnosed as pulmonary embolism (1–7). According to previous reports, only surgical resection offers a chance of prolonged survival (5). The role of chemotherapy and radiotherapy is still undefined. We describe a rare case of pulmonary artery sarcoma undergoing successful treatment by a two-drug combination chemotherapy consisting of ifosfamide and epirubicin.

CASE REPORT

A 63-year-old woman visited a local hospital with complaints of acute shortness of breath, and left chest and shoulder pain on October 2, 2003. Physical examination on admission revealed low grade fever (37.5°C) and tachycardia (110/min). Respiratory sounds were markedly diminished in the left lower chest. Laboratory findings demonstrated leukocytosis (10.6 × 10³/cm) and elevation of C-reactive protein (18.7 mg/dl). Analysis of arterial blood gas showed hypoxia (PaO₂ 74.1 mmHg) and hypocapnia (PaCO₂ 30.4 mmHg). Computed tomography (CT) scans of the chest disclosed a large filling defect arising from the main pulmonary trunk and extending into the left upper lobe and interlobar arteries (Fig. 1). She was given a diagnosis of acute pulmonary embolism and treated with insertion of an inferior vena cava filter, anticoagulation therapy (heparin and warfarin) and thrombolysis therapy (tissue plasminogen activator). Follow-up CT scans of the chest on October 6, 2003 revealed no improvement of the filling defect. The patient was therefore transferred to our hospital for further evaluation and surgical resection of the saddle pulmonary embolus.

She was asymptomatic on admission to our hospital. She underwent perfusion scintigraphy, which showed no perfusion in the left lung. Venography of the lower extremities detected no deep vein thrombosis. Thereafter, she underwent a pulmonary thromboectomy on October 27, 2003. The resected specimen consisted of mucoid regions and an endothelial blood clot. Histological findings showed proliferation of spindle cells with hyperchromatic and pleomorphic nuclei in a myxoid and collagenized background, which was consistent with intimal sarcoma of the pulmonary artery (Fig. 2A and B). Post-operative CT scans of the chest showed obvious persistence of an intraluminal hypodense area in the left main pulmonary artery.
From December 8, 2003, the patient was treated with four cycles of doublet chemotherapy consisting of ifosfamide (2.5 g/m²/day) given as a continuous intravenous infusion on days 1–5 and epirubicin (45 mg/m²/day) given as a continuous intravenous infusion on days 2 and 3. This was repeated every 4 weeks until March 14, 2004. She achieved the maximum response, and CT scans of the chest on March 26, 2004 showed marked regression of the intraluminal hypoattenuated area in the left main pulmonary artery (Fig. 3B). Adverse events were a grade 4 (National Cancer Institute common toxicity criteria version 2.0) neutropenia in all cycles, febrile neutropenia in the third and fourth cycles, grade 3 thrombocytopenia in the fourth cycle and grade 4 anemia in the second to fourth cycles. Hematological recovery was rapid. She tolerated this chemotherapy well without severe non-hematological toxicities. She was discharged on June 11, 2004 and is followed monthly at the out-patient clinic.

DISCUSSION

Pulmonary artery sarcoma is a rare tumor. Approximately 120 cases have been reported in the literature (1,2). A review of these cases revealed that ifosfamide and epirubicin are effective treatment options.

Figure 1. CT scan of the chest on October 24, 2003 shows a large filling defect arising from the main pulmonary trunk and extending into the left upper lobe and interlobar arteries.

Figure 2. (A) A section of the pulmonary artery shows an intraluminal sarcoma with thrombosis (hematoxylin and eosin stain 40x). Histological findings showed proliferation of spindle cells with hyperchromatic and pleomorphic nuclei in a myxoid and collagenized background, which was consistent with intimal sarcoma of the pulmonary artery. (B) The sarcoma consists of spindle cell proliferation (hematoxylin and eosin stain 400x).

Figure 3. (A) Post-operative CT scan of the chest showed obvious persistence of an intraluminal hypoattenuated area in the left main pulmonary artery. (B) CT scan of the chest on March 26, 2004 showed marked regression of the intraluminal hypoattenuated area in the left main pulmonary artery.
of 110 cases of pulmonary artery sarcoma showed a slight female predominance (1:1.3). Patients ranged in age from 22 to 80 years with a mean age of 48 years (2). Pulmonary artery sarcomas cause symptoms suggestive of recurrent pulmonary emboli such as dyspnea, chest pain and shortness of breath (1). Therefore, pulmonary artery sarcomas are very often confused with acute or chronic thromboembolic disease. This patient also visited a local hospital with complaints of acute shortness of breath, and left chest and shoulder pain, and she was given a diagnosis of acute pulmonary embolism. The absence of improvement despite anticoagulation therapy and atypical distribution of filling defects by CT scan, magnetic resonance imaging (MRI) or pulmonary angiography usually suggests a misdiagnosis of pulmonary embolism (3). The diagnosis can be strongly suspected when a CT scan or MRI shows a lobulated and heterogeneous hilar mass that originates from the pulmonary artery trunk or main pulmonary artery and expands into the peripheral artery. In this case, CT scans of the chest showed a large filling defect arising from the main pulmonary trunk and extending into the left upper lobe and interlobar arteries, and anticoagulation and thrombolysis therapy were ineffective. She was given a definite diagnosis at surgery. A definite diagnosis of pulmonary artery sarcoma is difficult. According to a review of 110 cases of pulmonary artery sarcoma, pre-operative or ante-mortem diagnosis without an exploratory thoracotomy was made in seven cases. In the majority of cases, the diagnosis was obtained at autopsy (2).

The role of chemotherapy and radiation therapy for pulmonary artery sarcoma is still undefined. Only complete surgical resection offers the chance of prolonged survival (5). Therefore, surgical treatment should be attempted in any case, even if cure seems unlikely. To our knowledge, chemotherapy has been tried in 16 patients including our case (2,6–10). However a definite effect (objective response) from chemotherapy has not been reported. This is the first case achieving a durable response by chemotherapy. Reichardt et al. reported the feasibility and usefulness of dose-intensive chemotherapy with ifosfamide and epirubicin in a phase II study for adult patients with metastatic or locally advanced soft tissue sarcoma, which resulted in a response rate of 52%, with a complete remission rate of 22% (11). We expect that marked regression of the pulmonary artery sarcoma by this chemotherapy in this patient may lead to survival prolongation.

In conclusion, surgical resection is currently the most hopeful treatment for pulmonary artery sarcoma. However, intensive chemotherapy is worth trying in unresectable patients.

References