Case Report

Primary Pulmonary Sarcomatoid Carcinoma

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ABSTRACT

Objective: To describe the characteristics and the result of surgical treatment in a series of patients with primary pulmonary sarcomatoid carcinoma (PSC).

Methodology: A descriptive study of 11 patients with primary PSC who were treated by the Thoracic Surgery Department at the Hospital Universitario 12 de Octubre in Madrid (Spain) between 2005 and 2009. We analyzed age, gender, histologic type, pathological stage, type of surgery and survival (in months).

Results: Ten patients were male and 11 were smokers; mean age of was 55. The pathologic stages were: 4 stage IIA, 3 stage IIIB, 2 stage IB and 2 stage IA. The most frequent histologic type was pleomorphic carcinoma, which was found in 5 cases. Complete resection was performed in 10 cases, and 7 received adjuvant therapy. Seven are disease-free after a mean follow-up period of 49 months.

Conclusions: Complete surgery in the initial stages of primary PSC can improve survival.

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Carcinomas sarcomatoides pulmonares primarios

RESUMEN

Objetivo: Describir las características y el resultado del tratamiento quirúrgico de una serie de pacientes con carcinomas sarcomatoides pulmonares primarios (CSPP).


Resultados: Diez pacientes eran varones y 11 eran fumadores, con una edad media de 55 años. Los estadios patológicos fueron 4 estadios IIA, 3 estadios IIIB, 2 estadios IB y 2 estadios IA. El tipo histológico más frecuente fue el carcinoma pleomórfico, con 5 casos. Se realizó resección completa en 10 casos, y 7 recibieron terapia adyuvante. Siete de ellos se encuentran libres de enfermedad en un periodo de seguimiento con una media de 49 meses.

Conclusiones: La cirugía completa en estadios iniciales de los CSPP puede mejorar la supervivencia.

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Introduction

Sarcomatoid carcinomas of the lung and pleura are rare tumors with a complex differential diagnosis. Primary pulmonary sarcomatoid carcinomas (PSC) must be differentiated from sarcomas and metastatic carcinoma, and account for 0.3% of all malignant neoplasms of the lung. Since 2004, they have been classified according to the WHO grading system as pleomorph carcinoma, fusiform cell carcinoma, giant cell carcinoma, carcinosarcomas and blastomas.1 The histological diagnosis may be suspected by the analysis of small samples, but definitive diagnosis is only possible with the complete tumor resection, due to its histological heterogeneity and pleomorphism.1

This disease generally occurs in older adults, with a mean age of 60–70 years, mainly males and smokers. The prognosis for these patients is poor, with a mean survival of 9–12 months in patients with complete tumor resection and 5-year survival of approximately 20%.2

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**Methodology**

This was a retrospective, descriptive study in which 488 lung resections for lung cancer carried out in the Thoracic Surgery Department of Hospital Universitario 12 de Octubre de Madrid between 2005 and 2009 were reviewed; data were obtained from the clinical records of the hospital’s Commission on Cancer Registry. Patients with a histological diagnosis of primary pulmonary sarcomatoid carcinoma were selected and analyzed. In all cases, contrast computed tomography of the lower cervical spine, chest, and abdomen was performed, and a bronchoscopy was carried out. Age, gender, histological type, disease stage, type of surgery (complete with free surgical margins), survival in months, adjuvant treatment (chemotherapy or radiation therapy), tumor recurrence, and disease-free survival were analyzed in all patients.

**Results**

Of the total of 488 patients with lung resections, 11 had a histological diagnosis of primary pulmonary sarcomatoid carcinoma. Of these, 10 were male and all had a history of smoking: 6 ex-smokers and 5 active smokers. They were aged between 46 and 80 years with a mean age of 55 years. Eight had tumors confined to the upper lobe and 4 had an endobronchial lesion (Table 1).

The preoperative diagnosis obtained from bronchoscopy was as follows: 6 cases of large cell carcinoma, 2 cases of epidermoid carcinoma, one case of adenocarcinoma and 2 undiagnosed cases.

The disease stages were as follows: 4 cases of stage IIA, 3 stage IIB, 2 stage IB and 2 stage IA.

The most frequent histological type was pleomorphic carcinoma, which was found in 5 cases, followed by 3 carcinosarcomas, 2 giant cell variants and one fusiform cell carcinoma.

Surgical resection was performed in all 11 cases (7 lobectomies and 4 pneumonectomies); in one of the cases, the resection was incomplete due to infiltration of the bronchial resection margin. Eight patients received adjuvant therapy with chemotherapy: 4 with stage IIA, 3 with stage IIB and one patient with stage IB. Two of the latter also received radiation therapy.

Of the 10 patients with complete resection, 7 were disease-free for a follow-up period of between 25 and 72 months (mean=49 months). Three patients died 2, 3 and 14 months after surgery, and two had distant recurrence after 2 and 8 months. These latter patients received chemotherapy and radiation therapy.

Of the 4 patients who died, 3 had pleomorphic carcinoma.

**Discussion**

In this case series, all the patients had a history of smoking and the majority were men, which corresponds with the characteristics of other published series.\(^3\)\(^4\) Moreover, endobronchial lesions are common in this type of neoplasm.\(^5\)

All patients with complete resection who died had the pleomorphic carcinoma histological type, which appears to be the most common and aggressive variant, with a survival of between 5 and 35 months.\(^6\)\(^-\)\(^8\)

In this series, 70% of the patients who had complete surgical resection had a mean disease-free survival of 49 months, which contrasts with other published series that report short periods of survival.\(^3\)\(^-\)\(^8\)\(^10\) This is probably because most of these patients underwent surgery in the early stages of the disease and no pathological lymph nodes were found in any of the cases.\(^11\)

Very few data are available in the literature on adjuvant chemotherapy and radiation therapy in these types of tumors. This approach has been proposed in some publications, but as the series in question were very small, the results are inconclusive. Moreover, preoperative histological diagnosis is rarely available in these patients, due to the need for large samples to obtain the diagnosis.\(^12\)

However, many authors recommend adjuvant treatment according to the disease stage.\(^11\) In our patients, chemotherapy was administered to patients with clinical stage IIA or higher and radiation therapy was reserved only for cases of tumor relapse.

In conclusion, pulmonary sarcomatoid carcinomas are a very rare group of neoplasms with a poor prognosis. Nevertheless, complete surgery in the early stages of the disease substantially improves survival.

<table>
<thead>
<tr>
<th>Histological type</th>
<th>Disease stage</th>
<th>Age</th>
<th>Gender</th>
<th>Smoker</th>
<th>Location</th>
<th>Resection</th>
<th>Follow-up</th>
<th>Adjuvant treatment/recurrence</th>
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<tbody>
<tr>
<td>Pleomorphic carcinoma</td>
<td>IIA</td>
<td>76</td>
<td>Male</td>
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<td>Right upper lobe</td>
<td>Lobectomy</td>
<td>72 months</td>
<td>Chemotherapy</td>
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<td>IIB</td>
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<td>Male</td>
<td>Ex-smoker</td>
<td>Middle lobe</td>
<td>Lobectomy</td>
<td>72 months</td>
<td>Chemotherapy</td>
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<td>IIA</td>
<td>54</td>
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<td>Ex-smoker</td>
<td>Left upper lobe</td>
<td>Pneumonectomy</td>
<td>2 months (died)</td>
<td>Chemotherapy radiation therapy on recurrence</td>
</tr>
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<td>Fusiform cell variant</td>
<td>IIB</td>
<td>46</td>
<td>Male</td>
<td>Yes</td>
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<td>Ex-smoker</td>
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<td>Right lower lobe</td>
<td>Lobectomy</td>
<td>30 months</td>
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<tr>
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<td>Left lower lobe</td>
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<td>25 months</td>
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<td>Lobectomy</td>
<td>8 months (died)</td>
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References