CASE REPORTS

Postoperative Course in 7 Cases of Primary Sarcoma of the Lung

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Unlike lung metastases of extrapulmonary sarcomas, primary sarcoma of the lung is very rare. We analyzed 7 primary sarcomas treated surgically in Hospital Universitario La Paz, Madrid, Spain between 1985 and 2001. Preoperative histologic diagnosis was correct for 2 patients. Surgery was performed in all 7 patients. Resection was extended to the left atrium in 1 patient, to the chest wall in another, and the parietal pleura were removed from 2 others. Surgical resection was considered complete in 6 cases. Histology revealed 4 cases of malignant fibrous histiocytoma, 1 angiosarcoma, 1 osteogenic sarcoma, and 1 undifferentiated sarcoma. Enlarged lymph nodes removed during surgery were tumor free.

Three patients received complementary treatment. The 3 longest-surviving patients were treated with surgery alone; at the end of the study, these patients remained alive 16, 9, and 4 years after surgery. One patient has a recurrent lymph node tumor in a single lung. Three patients died within 1 year and another died 24 months after surgery.
In conclusion, surgical treatment of primary sarcoma of the lung can achieve good survival.

Key words: *Lung sarcoma. Malignant fibrous histiocytoma. Angiosarcoma.*

Evolución posquirúrgica de 7 sarcomas pulmonares primitivos

Los sarcomas primitivos pulmonares son muy raros, pero no los metastásicos. Se analizaron 7 sarcomas primarios operados en nuestro servicio entre 1985 y 2001. El diagnóstico histológico preoperatorio fue correcto en 2 pacientes. Se realizó cirugía en todos. En uno se amplió la resección a aurícula izquierda, en otro a pared torácica y en otros 2 se extirpó pleura parietal. Se consideró cirugía completa en 6 casos. En el estudio histológico 4 fueron fibrohistiocitomas malignos, uno angiosarcoma, uno sarcoma osteogénico y uno sarcoma indiferenciado. Las adenopatías extirpadas estaban libres de tumor.

Se hizo tratamiento complementario en 3 pacientes. En los 3 de mayor supervivencia se realizó sólo cirugía, con un seguimiento de 16, 9 y 4 años, respectivamente. Uno tiene actualmente una recidiva tumoral nodular en pulmón único. En cuanto a los fallecidos, 3 murieron antes de un año y otro después de 24 meses de evolución.

En conclusión, la cirugía en los sarcomas pulmonares primitivos puede conseguir una elevada supervivencia.

Palabras clave: *Sarcomas pulmonares. Fibrohistiocitoma maligno. Angiosarcoma.*

Introduction

Primary sarcomas are estimated to represent less than 0.5% of all malignant tumors of the lung.[1,2] Experience with this rare type of tumor is necessarily limited and it is, therefore, difficult to define the most common histologic types. The presence of a sarcoma at any other site must be ruled out before diagnosing the tumor as a primary tumor of the lung. However, sarcomas occurring outside the lung rarely remain occult and their lung metastases frequently occur at multiple sites.

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This report presents 7 cases of primary sarcoma of the lung that were treated in our hospital over a period of 16 years and analyzes both histologic characteristics and survival.

Case Descriptions

Between 1985 and 2001, 7 patients with primary sarcoma of the lung, 6 of whom were men, were treated surgically in the thoracic surgery department of Hospital Universitario La Paz, Madrid. Patient age ranged from 42 to 66 years (mean, 55 years). Respiratory symptoms were present in 5 patients, 2 of whom also presented general malaise, weakness, and weight loss. In 2 cases, the sarcoma was discovered by chance following chest radiography performed for other reasons.

Chest radiographs (Figure 1) revealed tumor masses that were generally well circumscribed and ranged in size from 5 to 20 cm; the tumor was accompanied by atelectasis in 2

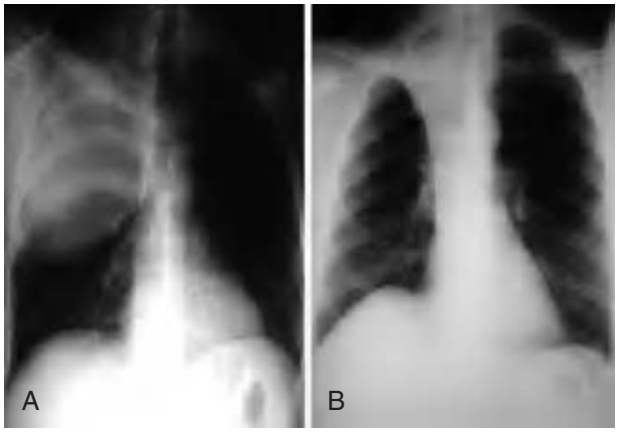

Figure 1. Chest radiograph showing a 15 cm mass (A) and radiograph of a tumor accompanied by atelectasia (B).

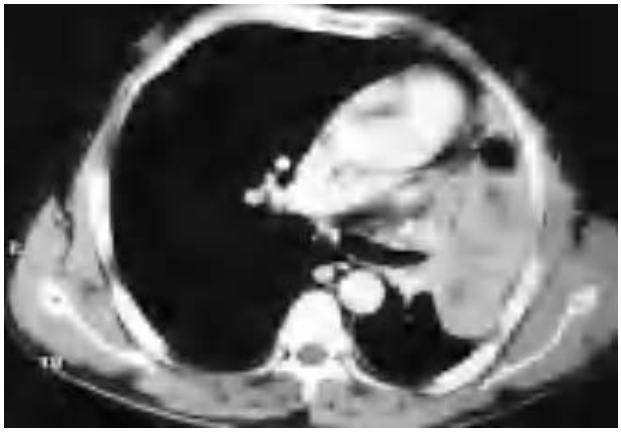

Figure 2. Computed tomography scan corresponding to case 4 showing central excavation of the tumor.

cases. Computed tomography (CT)—as shown in Figure 2—allowed tumor extension to be assessed and in 1 case revealed invasion of the chest wall with destruction of the rib; diseased mediastinal lymph nodes with a diameter of greater than 1 cm were observed in 2 patients.

Fiberoptic bronchoscopy was performed in all patients and on 3 occasions revealed a visible tumor; however, biopsy only yielded a correct diagnosis in 2 cases. Transthoracic fine needle aspiration was performed in 2 cases and examination of the aspirates yielded an erroneous diagnosis of carcinoma. Analysis of tumor extension (abdominal CT, bone scintigraphy, head CT, and abdominal ultrasound) was undertaken in 3 patients but did not reveal evidence of tumors at other sites. Mediastinoscopy was performed in 2 patients with suspected lymph node tumors based on the results of CT; the results were negative in both cases. Table 1 shows the sex, age, and clinical characteristics of the patients along with the diagnostic methods used.

Resection was performed in all 7 patients: 3 pneumonectomies and 4 upper lobectomies. Surgery was extended to the left atrium in 1 patient, to the chest wall in another, and to the parietal pleura in 2 others.

All of the resected tissues were subsequently analyzed to determine the histologic characteristics of the sarcomas.

Table 2 shows the type of surgery performed, the TNM classification, the definitive diagnosis, and the survival time for each patient.

TABLE 1
Distribution of Patients According to Sex, Age, Symptoms, and Diagnostic Procedures*

Case	Sex	Age, Years	Symptoms	Radiography	Bronchoscopy	Peoperative Diagnosis
1	W	68	Cough, pain	6 cm mass	Visible	Reticulohistiocytoma
2	M	43	Cough, dyspnea, weight loss	5 cm mass	Not visible	Carcinoma
3	M	42	Pain, cough, general malaise	15 cm mass with chest wall involvement	Not visible	Adenocarcinoma (FNA)
4	M	64	Dyspnea, cough, hemoptysis	20 cm mass	Visible	Undifferentiated carcinoma (FNA)
5	M	66	Asymptomatic	10 cm mass	Visible	Sarcoma (FB)
6	M	49	Catarrhal symptoms	6.5 cm mass	Not visible	Carcinoma (FNA)
7	M	56	Cough, weight loss	Atelectasia with 6 cm mass	Not visible	Suspected carcinoma (FNA)

*W indicates woman; M, man; FNA, fine needle aspiration; FB, fiberoptic bronchoscopy.

TABLE 2
Type of Surgery, TNM Classification, Definitive Diagnosis, and Survival

Case	Surgery	TNM	Histologic Diagnosis	Survival
1	Right pneumonectomy and atrial resection (1997)	T3N0	Undifferentiated sarcoma	2 years
2	Left pneumonectomy (1985)	T2N0	Osteogenic sarcoma	3 months (lost to follow-up)
3	Right upper lobectomy and incomplete rib resection (1985)	T3N0. Positive surgical margins	Angiosarcoma	2 months
4	Right upper lobectomy (1999)	T2N0	Malignant fibrous histiocytoma	6 months
5	Right upper lobectomy (1995)	T2N0	Malignant fibrous histiocytoma	9 years
6	Right upper lobectomy (2000)	T2N0	Malignant fibrous histiocytoma	4 years
7	Right upper lobectomy (1988)	T2N0	Malignant fibrous histiocytoma	16 years

With the exception of one patient who was lost at 3 months after surgery, follow-up was complete in all cases. Two patients died within the first year due to brain and bone metastases. One patient survived for 2 years before dying from widespread metastasis. Currently, 3 patients survive 16, 9, and 4 years after surgery. One of these patients has lymph node recurrence in a single lung with poor functional reserves that preclude further surgical intervention. This recurrence occurred after 7 years of disease-free survival. Adjuvant treatment with chemotherapy and radiotherapy was employed in 3 patients based on histologic characteristics, tumor size, and involvement of neighboring structures. The patients who are currently surviving were treated with surgery alone.

Discussion

Sarcomas are tumors of mesenchymal origin that on rare occasions are found as primary tumors in the lung. Despite the fact that extrapulmonary sarcomas rarely remain occult, the presence of a sarcoma of different origin must be ruled out prior to diagnosis as a primary tumor of the lung. These tumors arise from stromal elements of the bronchial wall, the blood vessels, or the lung interstitium.

Primary sarcomas of the lung form a heterogeneous group that contains different histologic varieties. The majority are soft-tissue sarcomas. However, additional varieties of vascular origin or, less commonly, arising from bone or cartilage (osteosarcoma, chondrosarcoma, and osteogenic sarcoma) have also been reported.[3,4]

The rarity of this type of tumor, which represents less than 0.5% of all malignant tumors of the lung, has meant that the patient series presented in the literature contain only small numbers of cases[4-6]; furthermore, publications prior to 1975 included lymphoproliferative tumors within this type of sarcoma.[1]

The most frequent type of sarcoma found in the patient series presented here was malignant fibrous histiocytoma (4 cases). This type, first described by O´Brien and Stout, was for years incorrectly included within the fibrosarcomas and leiomyosarcomas.[7] A review of this type of tumor published by Halyard et al[8] in 1996 described 6 subtypes; the most common subtype in the lung is the storiform variety.

Sarcomas arising in bone are a genuine exception and are only described in isolated cases.[3] An osteogenic sarcoma that displayed an unfavorable course was identified retrospectively in our patient series.

Primary sarcomas of the lung grow through the parenchyma and in this way can reach a large size (20 cm in one of the cases presented here). Although their invasive tendency means that they can spread into the chest wall, the mediastinum, or the cardiac chambers, the symptoms are usually limited. They are tumors that rarely metastasize via the lymphatic system—in the literature only Reynard et al[9] have reported lymph node tumors in 25% of a series. Metastasis occurs more commonly through the blood, the most frequently affected organs being the brain, lung, and skeleton.

Although most of the cases reported here occurred in men (6 men and 1 woman), both sexes are equally affected. In general, these tumors can appear at any age—the mean age of presentation in our patient series was 55 years. However, leiomyosarcomas and rhabdomyosarcomas are described as occurring more frequently in young patients.

Primary sarcomas of the lung commonly present in radiographs as tumor masses. These masses can sometimes reach a substantial size and invade neighboring structures, and can also undergo necrosis.

Assessment of the literature reveals that fiberoptic bronchoscopy is of limited use for diagnosis because tumors only occasionally invade the bronchial wall,[8] meaning that percutaneous fine needle aspiration of the tumor is necessary in order to obtain a histologic diagnosis. However, the cytology can be unreliable and histologic differentiation requires immunohistochemistry, electron microscopy, or analysis of cell structure.[10] Mediastinoscopy is justified in those cases in which invasion of the lymph nodes or mediastinal tumors are suspected.[6,9]

Surgery is the best treatment and the literature reports surgical interventions extended to the cardiac chambers and wall,[11,12] even including cardiopulmonary bypass, given that resection is the best method of eradicating these tumors.[13] In our patient series, resection of the left atrium was performed in 1 patient because of venous thrombosis not caused by the tumor.

Prognostic factors include tumor size, valid for some patients[4,5,14] but of limited significance in others[5]; degree of histologic malignancy[4,6,9,12]; and endobronchial localization[2,4]; however, diagnosis can be considered to be obtained earlier in this type of tumor because the symptoms are more florid. A number of opinions exist regarding histologic type; while some authors suggest that the best prognosis is shown by malignant fibrous histiocytoma,[8,12] others find no relationship between prognosis and histologic type.[4,6] In summary, the low frequency of occurrence of these tumors and the variation in the published cases makes it difficult to establish genuine prognostic factors.

Adjuvant treatment with radiotherapy and chemotherapy does not lead to a significant improvement in survival[13] but may be indicated in incomplete resections, in patients for whom surgery is not indicated, and in those patients with positive lymph nodes or highly malignant tumors. Patients showing long survival in our group had been treated by surgery alone. All had malignant fibrous histiocytomas, were classified as T2 N0, and had a tumor diameter of less than 12 cm.

In conclusion, sarcomas of the lung are very rare as primary tumors and should only be considered as such following exclusion of other sites. Preoperative diagnosis is difficult since the tumor is rarely visible by endoscopy and needle aspiration is unreliable. Surgical resection is the best form of treatment and should be radical in order to prevent recurrence. There appears to be little justification for adjuvant treatment following complete surgery.

REFERENCES

1. Martini N, Hadju SI, Beattie EJ Jr. Primary sarcoma of the lung. J Thorac Cardiovasc Surg. 1971;61:33-8.
2. Guecion JG, Rosen SH. Bronchopulmonary leiomyosarcoma and fibrosarcoma. A study of 32 cases and review of the literature. Cancer. 1972;30:836-47.
3. Robison PG, Shields TW. Uncommon primary malignant tumors of the lung. In: Shields TW, Locicero J III, Ponn RB, editors. General thoracic surgery. 5th ed. Philadelphia: Lippincott Williams & Wilkins; p. 1533-53.
4. Nascimento AG, Unni KK, Bernatz PE. Sarcomas of the lung. Mayo Clin Proc. 1982;57:355-9.
5. McCormack PM, Martini N. Primary sarcomas and lymphomas of lung. In: Martini N, Nvogt-Moykopf I, editors. International trends in general thoracic surgery. Vol. 5. St. Louis: Mosby; 1988.
6. Janssen JP, Mulder JS, Wagenaar S, et al. Primary sarcoma of the lung: a clinical study with long-term follow-up. Ann Thorac Surg. 1994;58:1151-5.
7. Lee JT, Shelburne JD, Linder J. Primary malignant fibrous histiocytoma of the lung. A clinicopathologic and ultrastructural study of five cases. Cancer. 1984;53:1124-30.
8. Halyard MY, Camoriano JK, Culligan JA, Weiland LH, Allen MD, Pluth JR, et al. Malignant fibrous histiocytoma of the lung. Cancer. 1996;78:2492-7.
9. Régnard JF, Icard P, Guibert L, de Montpreville VT, Magdeleinat P, Levasseur P. Prognostic factors and result after surgical treatment of primary sarcomas of the lung. Ann Thorac Surg. 1999;68:227-31.
10. Attanoos RL, Appleton MACA, Gibbs AR. Primary sarcomas of the lung: a clinicopathological and immunohistochemical study of 15 cases. Histopathology. 1996;29:29-36.
11. Porte HL, Metois DG, Leroy X, Conti M, Gosselin B, Wurtz A. Surgical treatment of primary sarcoma of the lung. Eur J Cardiothorac Surg. 2000;18:136-42.
12. Bacha EA, Wright CD, Grillo HC, Wain JC Moncure A Keel SB, et al. Surgical treatment of primary pulmonary sarcomas. Eur J Cardiothorac Surg. 1999;15:456-60.
13. Ríos Zambundio A, Roca Calvo MJ, Polo García LA. Fibrohistiocitoma maligno de pulmón variedad estoriforme. Arch Bronconeumol. 2003;39:431-2.
14. Burt M, Zakowski M. Rare primary malignant neoplasms. In: Pearson FG, Deslauriers J, Ginsberg RJ, Hiebert CA, McKneally MF, Urschel HC Jr, editors. Thoracic surgery. p. 807-26.