LETTERS TO THE EDITOR

Leiomyosarcoma of the pulmonary veins extending into the left atrium or left atrial leiomyosarcoma: Multimodality therapy

To the Editor:

We read with interest the article by Shimono and associates (J Thorac Cardiovasc Surg 1998;115:469-1) titled "Pulmonary Leiomyosarcoma Extending Into Left Atrium or Pulmonary Trunk: Complete Resection With Cardiopulmonary Bypass." Although it is true that primary pulmonary leiomyosarcoma is a rare tumor, we think that the second case reported consisted in a left atrial leiomyosarcoma originating in the atrial wall and extending into the pulmonary veins. The most important series in the world concerning this tumor is the experience of the Armed Forces Institute of Pathology (AFIP),[1] with 12 cases reported and 6 cases surgically resected. Controversy still exists concerning their origin. The AFIP clearly demonstrated that the majority of cardiac leiomyosarcomas arise in the left atrium; at surgery, some of these appear to arise from a pulmonary vein. For these reasons, the cell origin of cardiac leiomyosarcoma may reside in the smooth muscle media of pulmonary veins. However, it is likely that some leiomyosarcomas originate within the left atrium itself, because the left atrium is a preferred site of origin of most cardiac sarcomas, and because the subendocardial lining of the atrium normally contains bundles of smooth muscle cells. Their large size at the time of diagnosis makes assessment of the precise site of origin impossible in most cases. To the best of our knowledge, 20 cases of left atrial leiomyosarcomas have been previously reported in the literature (Table I). The mean postoperative survival in those patients is 6.8 months.[1,2] In our opinion, 7 months is insufficient follow-up time to evaluate whether surgery without adjuvant therapy has been successful. Shimono and associates stated that their case was the first report of the resection of a left atrial tumor by thoracotomy through the rib bed, and we understand why the majority of cardiac surgeons relating their experience preferred the sternotomy approach. Surgery is indicated to obtain a definitive histologic diagnosis and to evaluate the prognosis, and as much of the tumor as possible must be removed during the operation. Visualization of tumors in the left atrium appears to be more difficult, and detection of unresected portions of tumor after the operation can be extremely difficult if not impossible.[3] This, evidently, can be extremely difficult in tumors of the left atrium with involvement of the pulmonary veins[4] by the thoracotomy approach.

Krüger and associates[5] have analyzed 19 cases of leiomyosarcoma of the pulmonary artery. After surgery, the median survival time of the patients was prolonged to 10 months. The surgical technique (pneumonectomy compared with mere "excision" of the tumor from the vascular bed) did not have any effect on the survival time ($P = .43$). Resection involving pneumonectomy may lead to recurrence of the tumor in the remaining lung owing to the proximal location of the tumor. We have treated 1 patient with leiomyosarcoma of the pulmonary artery trunk. The tumor was successfully resected without pneumonectomy with a convenient follow-up of 1 year.

Regarding adjuvant chemotherapy, which was not indicated in the 2 cases reported by Shimono and associates, recent reports seemed to demonstrate the benefit of a chemotherapy regimen in those type of tumors.[4-6] Doxorubicin-based postoperative adjuvant therapy has been used on the basis of its effectiveness in treating soft tissue sarcomas in other locations.[6] Because of the poor prognosis of patients with leiomyosarcomas of the heart and the great vessels, the recommended therapy is surgery followed by chemotherapy and radiation.[2,4] Some authors[4,5] have demonstrated that radio-

Table I. *Primary leiomyosarcoma of the left atrial cavity*

References	*No. of cases*	*Treatment*	*Follow-up*
Sande MA, Am Rev Respir Dis 1970;102:258-63	1	Autopsy	
Hardin NJ, Johns Hopkins Med J 1974;134:141-55	1	Autopsy	
Karasawa T, Acta Pathol Jpn 1981;31:195-202	1	Autopsy	
Leiba VI, Arkh Patol 1981;43:71-3	1	Autopsy	
Donovan VM, Arch Intern Med 1982;142:1923-5	1	Resection	Operative death
Reece IJ, J Thorac Cardiovasc Surg 1984;88:439-46	1	Resection	8 mo
Chaloupka JC, Cardiovasc Intervent Radiol 1986;9:132-5	1	Autopsy	
Miki H, Kokyu to Junkan 1987;35:203-7	1	Resection	1 mo
Closas J, Rev Esp Cardiol 1988;41:302-5	1	Autopsy	
James CL, Pathology 1989;21:308-13	1	Resection	1 mo
Fyfe AI, Can J Cardiol 1991;7:193-6	1	Resection	7 mo
Antunes MJ, Ann Thorac Surg 1991;51:999-1001	1	Resection	23 mo
Takamizawa S, Intern Med 1992;31:265-8	1	Resection	6 mo
Ghyra SA, Ann Thorac Surg 1996;61:1840-1	1	Resection	Alive (1 y)
Burke A, Atlas Tumor Pathol 1996;16:127-69	6	Resection	6.8 mo

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therapy and/or chemotherapy administered in the postoperative period was followed by a significantly prolonged survival time ($P = .023$). Pulmonary metastases are present in up to 60%[4] of patients. Distal microembolization is common; therefore adjuvant therapy, even in the absence of pulmonary nodules, would seem appropriate. In conclusion, total surgical resection (which was best performed with the aid of cardiopulmonary bypass through a median sternotomy), with the addition of chemotherapy, radiotherapy, or both, should offer these patients significant palliation and an opportunity for increased length of survival.

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Reply to the Editor:

We thank Babatasi, Massetti, Galateau, and Khayat for their interest in our article. We shall first respond to their comment that the tumor in the second case originated in the atrial wall and extended into the pulmonary vein. Because of space limitations in our article, we did not show the photographs of the operative specimen and histologic findings in detail. On the macroscopic findings, the tumor filled the left lower pulmonary vein and was strongly attached to the wall of the pulmonary vein. Most of the intra-atrial tumor was free from the atrial wall and weakly attached to the atrial wall around the orifice of the pulmonary vein (Fig 1). Therefore, the tumor, the left lung, and the left side of the left atrial wall were excised en bloc.

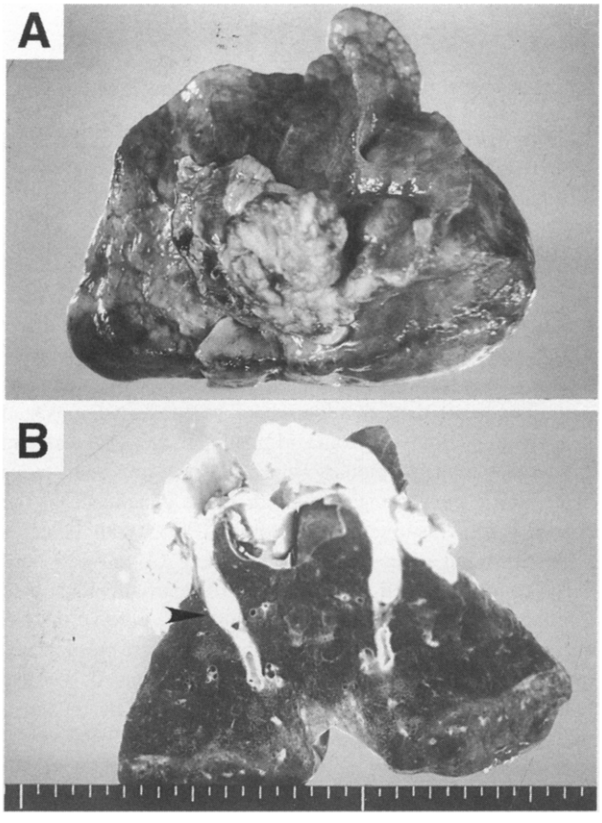

Fig 1. A, Left pneumonectomy specimen with the tumor mass and the left atrial wall. **B,** Macroscopic finding of the left lower pulmonary vein and the tumor. The tumor is strongly attached to the wall of the pulmonary vein *(arrow)*.

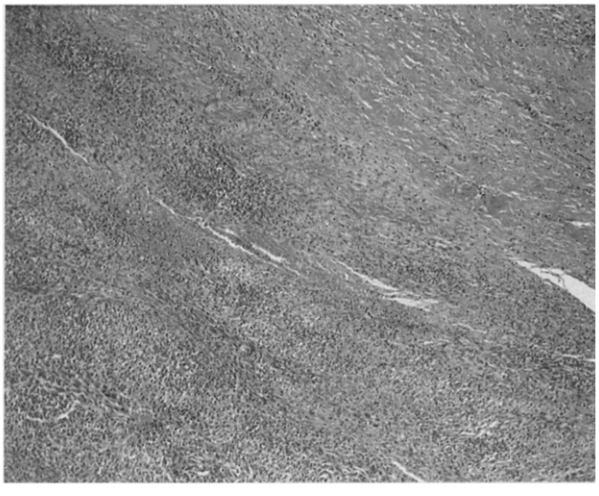

Fig 2. Histologic findings at the portion of the tumor strongly attached to the vein wall.

Histologic examination revealed that the tumor arose from the media of the pulmonary vein at the point where it was strongly attached (Fig 2).