Extended right pneumonectomy with partial left atrial resection for primary leiomyosarcoma of the mediastinum

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Primary leiomyosarcoma of the mediastinum is a rare malignancy that usually occurs either in the mesenchymal cells of the soft tissue of the visceral mediastinum or in the smooth muscle of the great vessels. The available data on this condition are minimal at best, but it seems that the most significant factor affecting survival is the ability to completely resect the mediastinal sarcoma.

We report on a case of leiomyosarcoma of the mediastinum invading the left atrium and the right lung that was completely resected via a right pneumonectomy with en bloc partial resection of the left atrium.

Clinical Summary

A 62-year-old man was referred to our attention because a chest radiograph taken after 15 days of antibiotic therapy showed a persistent right lower pulmonary lobe infiltrate. This is an occasional finding during cardiovascular examinations performed to study recent single events of transient ischemic attack.

Workup included a full-body computed tomographic scan, which revealed a mediastinal mass of 6 cm in diameter extending to the right lower pulmonary lobe and to the inferior pulmonary vein, with extrinsic infiltration of the left atrium wall. The tumor presented an intravascular extension into the left atrial cavity via the inferior pulmonary vein and a floating neoplastic polyp. No suspicious mediastinal lymph nodes were observed.

Extrathoracic staging was negative for metastatic disease. An F-18 fluorodeoxyglucose positron emission tomographic scan of the tumor was positive, with no other suspicious uptake. We performed 2 transesophageal needle biopsies of the mass by using a flexible bronchoscope to obtain a preoperative histologic sample; but this failed to yield a diagnosis. We then used a rigid bronchoscope, and the samples so obtained led to a preoperative histologic diagnosis of undifferentiated necrotic neoplasm. A more specific diagnosis was not possible because of the necrotic aspect of the available tissue.

The functional evaluation included routine blood tests, electrocardiogram, spirometry, and a perfusion lung scan, and there was no evidence of concomitant cardiac or pulmonary disease. The spirometry showed a forced expiratory volume in 1 second of 3.15 L (97.4% of the predicted value) and a predicted carbon monoxide diffusing capacity of 153.6%. The perfusion lung scan showed a right lung perfusion of 48.3%.

The case was discussed multidisciplinarily, and on the basis of these discussions, we recommended proceeding first with surgical resection of the tumor by right pneumonectomy with partial resection of the left atrium (because of the intracardiac floating polyp and its subsequent high risk for future thromboembolic complications and consequent cerebral damage).

We used a right hemiclamshell approach. Because the tumor had developed in the posterior mediastinum and involved the right pulmonary lower lobe and the left atrium at the origin of both right pulmonary veins, we chose first to separate the right pulmonary artery and the right main stem bronchus. Cardiopulmonary bypass was instituted without cardiac arrest, and the left atrium was opened along the origin of both pulmonary veins. The tumor was revealed to involve the posterior left atrial wall, with almost complete obstruction of the right inferior pulmonary vein's ostium, and presented with an intracardiac polypoid growth. We resected the left atrium along the orifice of the right superior and inferior pulmonary veins, en bloc with the tumor, the right lung, and the pericardium. We then reconstructed the left atrium. A radical ipsilateral mediastinal lymph node dissection was also performed. We then weaned the patient from cardiopulmonary bypass and reconstructed the right pericardium with a polytetrafluoroethylene prosthetic patch to avoid cardiac herniation.

Pathologic examination indicated a medium-grade leiomyosarcoma arising from the posterior mediastinum (outside the heart), which involved the pericardium, the left atrium wall at the origin of the pulmonary veins, and the lower pulmonary lobe (Figure 1). Immunohistochemical stains were strongly positive for both smooth muscle actin and myosin but were negative for desmins, S-100 protein, cytokeratin, CD34, CD31, and CD117. All 14 lymph nodes examined were negative. The postoperative outcome was normal, and the patient was discharged 10 days after the

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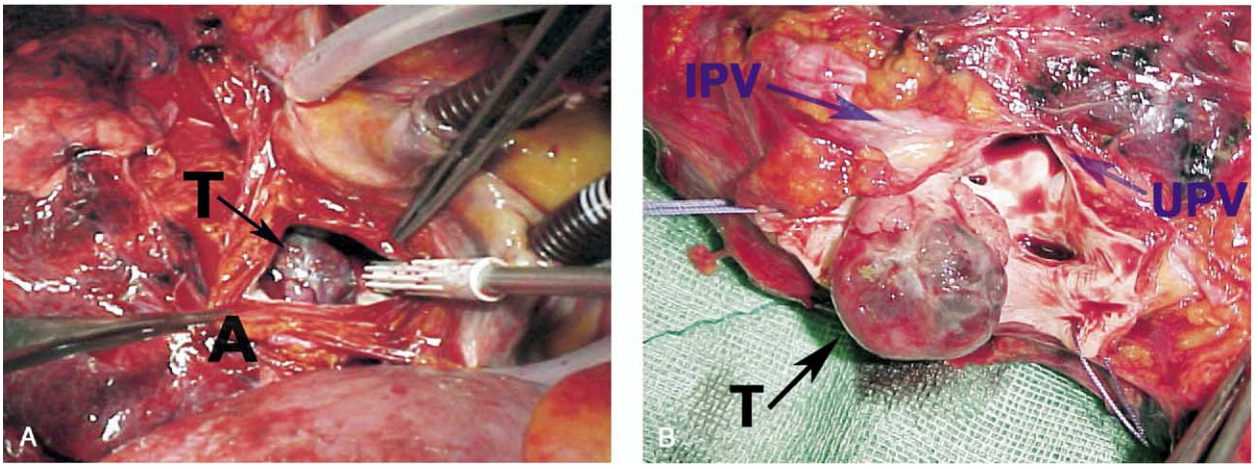

Figure 1. A, Operative view showing the intracardiac polypoid tumor *(T)* growth resected in extracorporeal circulation without cardiac arrest. *A,* Right atrium. B, Macroscopic view of the resected specimen illustrating the intra-atrial mass *(T)* originating from the inferior pulmonary vein *(IPV). UPV,* Upper pulmonary vein.

operation. No adjuvant therapy was proposed, and the patient is still alive at 6 months, without evidence of disease at a recent chest computed tomography and F-18 fluorodeoxyglucose positron emission tomography scan.

Discussion

Primary leiomyosarcoma of the mediastinum develops either in the mesenchymal cells of the soft tissue of the visceral mediastinum or in the smooth muscle of the great vessels. In the literature, a few sporadic cases[1-4] are reported, and there are 2 retrospective cohort studies. In 1994, Moran and colleagues[5] reported on a series of 10 patients with primary leiomyosarcoma of the mediastinum, and in 1998, Burt and colleagues[6] reported on their experience with 47 patients with primary sarcoma of the mediastinum. Both studies concluded that the overwhelming factor in determining survival was the ability to completely resect the tumor.

In the case presented here, with the goal of completely resecting the tumor, we used a very aggressive surgical approach, performing an extended right pneumonectomy with partial resection of the left atrium by using cardiopulmonary bypass. Our experience confirmed that of Conner and associates,[7] who reported on a similar case of performing a resection with cardiopulmonary bypass.

The discussion is still open regarding the role of adjuvant therapy after complete operation, and the lack of sufficient data makes the decision difficult. We chose not to administer any adjuvant therapy, because we considered radiotherapy of the heart potentially dangerous and unjustified after a pathologically proven complete resection. Adjuvant chemotherapy was also ruled out because of the medium grade of the leiomyosarcoma and the impossibility of evaluating any potential response to the therapy in the absence of any detectable disease.

Long-term survival in primary leiomyosarcoma of the mediastinum depends on the ability to completely resect the tumor. In select potentially resectable cases, we strongly advocate complete resection of the tumor, even if a cardiopulmonary bypass is required. Also, although we await new data on this condition, we recommend determining the usefulness of any possible adjuvant therapy on a case-by-case basis.

References

1. Shields TW, Robinson PG. Mesenchymal tumors of the mediastinum. In: Shields TW, LoCicero J III, Ponn RB, editors. General thoracic surgery. Philadelphia: Lippincott Williams & Wilkins; 2000. p. 2371-2.
2. Rasaretnam R, Panabokke RG. Leiomyosarcoma of the mediastinum. *Br J Dis Chest.* 1975;69:63-9.
3. Sunderrajan EV, Luger AM, Rosenholtz MJ, Maltby JD. Leiomyosarcoma in the mediastinum presenting as superior vena cava syndrome. *Cancer.* 1984;53:2553-6.
4. Steen BC, Florez MS, Fernandez FL, Garcia TJ, Jareno EJ, Ancochea BJ. Mediastinal leiomyosarcoma. *An Med Interna.* 1993;10:83-5.
5. Moran CA, Suster S, Perino G, Kaneko M, Koss MN. Malignant smooth muscle tumors presenting as mediastinal soft tissue masses: a clinicopathologic study of 10 cases. *Cancer.* 1994;74:2251-60.
6. Burt M, Ihde JK, Hajdu SI, Smith JW, Bains MS, Downey R, et al. Primary sarcomas of the mediastinum: results of therapy. *J Thorac Cardiovasc Surg.* 1998;115:671-80.
7. Conner WC, Fink GW, McGinnis KM, Alfieris GM. Surgical management of leiomyosarcoma of the mediastinum. *Ann Thorac Surg.* 2004; 77:334-6.