Endarterectomy for pulmonary artery sarcoma: Too much, too little, or just right?

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Central Message

Pulmonary artery sarcoma is rare and aggressive. Complete resection is needed for local control.

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Primary cardiac sarcoma is a highly unusual tumor with a limited survival without surgical resection. A 17-year review of sarcomas identified with the Surveillance, Epidemiology and End results database identified 210 primary cardiac sarcomas and 24,404 soft-tissue sarcomas, showing the rarity of primary cardiac sarcoma.¹ The overall median survival of primary cardiac sarcoma in this series was 6 months and that of soft-tissue sarcoma was 93 months. These highly aggressive primary cardiac sarcomas had a median survival of 12 months in those who had surgical resection and 1 month in those that did not.

Our group has a long-standing interest in primary cardiac sarcoma and has accumulated a large surgical experience with our partners at MD Anderson Cancer Center.² We have shown that neoadjuvant chemotherapy improves survival for right heart sarcomas.³ We believe, based on our large experience, that neoadjuvant therapy would likely also be beneficial in pulmonary artery sarcoma. Pulmonary artery sarcoma generally presents in an advanced symptomatic stage requiring resection to alleviate heart failure, and getting a tissue biopsy is more complex than when dealing with right heart sarcomas, making a tissue diagnosis more difficult before surgery. We have previously published our results with pulmonary artery sarcoma showing an improved survival versus published data at that time.⁴ This led us to read with great interest the article in this issue of the *Journal* by Yin and colleagues.⁵

These authors present 12 cases of pulmonary artery sarcoma collected over an 11-year period at a busy cardiovascular center in China. Ten patients underwent surgical resection, and 2 had exploratory thoracotomy only. For the 10 patients having resection, all were treated with endarterectomy, with 3 requiring an addition of pneumonectomy. Interestingly 9 (75%) of these cases had pulmonary trunk involvement, and 6 (50%) were initially misdiagnosed as pulmonary embolus, which is consistent with our own experience. There was 1 operative mortality

(8.3%). Median survival for the entire group was 18 months. The median survival was significantly longer for those that had adjuvant chemotherapy and radiation versus those that did not (28 months vs 8 months). The result that caught our attention was that all 8 late deaths were related to recurrence of the malignancy. Although recurrence is not uncommon in cardiac sarcoma, it is generally metastatic disease after surgical resection, which has been our Achilles heel. In this series, there were 5 recurrences at the distal pulmonary artery and 1 in the pulmonary trunk, and only 2 deaths were due to distant spread. The 75% local recurrence rate seen in this series shows inadequate local control. Our primary role as surgeons is to achieve complete resection for local control and, failing this, we must evaluate our choice to operate, our surgical technique used, or both.

After reading this interesting article, we reviewed our cardiac sarcoma experience. Our group has now operated on 115 primary cardiac sarcomas, of which 15 were primary pulmonary artery sarcomas. We have found that pulmonary artery sarcoma generally begins in the pulmonary root as Yin and colleagues found and about 25% of our cases had initial endarterectomy elsewhere and were referred to us for resection after local recurrence. We have found that the sarcoma expands the pulmonary arteries as it grows distally but does not penetrate the artery itself to involve mediastinal tissue outside of the artery. This allows complete resection of the pulmonary trunk and main pulmonary arteries out to the upper lobe artery take off without difficulty. This is our standard approach, believing this represents a better cancer resection and our previous experience would appear to support this

both for local recurrence and survival.⁴ We agree that extension beyond the upper lobe branch will require either endarterectomy or pneumonectomy for removal and do use endarterectomy in this circumstance when the lung can be saved.

We congratulate Yin and colleagues on their series of pulmonary artery sarcoma. We will never have enough cases to compare endarterectomy with complete resection but suggest that when possible, complete resection should be considered as a more acceptable malignancy operation. This excellent series will serve as the impetus for us to update our experience with this disease.

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