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Commentary: Opportunity is like a sunrise. If you wait too long, you miss it

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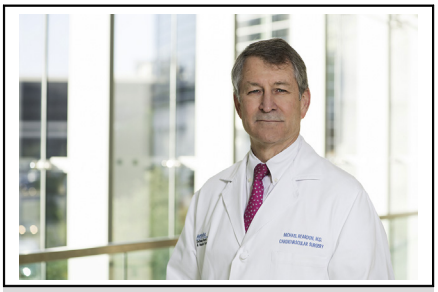

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CENTRAL MESSAGE

Pulmonary artery sarcoma is a rare and deadly disease. Treatment by an experienced, multidisciplinary team is optimal.

Our recent 42-year review of the Surveillance, Epidemiology, and End Results database yielded 442 cases of primary cardiac sarcoma in the United States.[1] This disease is both rare and deadly, with a median survival of 7 months and 1-, 3-, and 5-year survivals of 40.7%, 15.6%, and 9.8%.[1] In this large review, both surgery and chemotherapy were associated with improved survival. Given the very uncommon occurrence of primary cardiac sarcoma and the much more common presentation of benign cardiac tumors, it is not unusual for the proper diagnosis and treatment of primary cardiac sarcoma to be delayed.[2] This is further exacerbated for pulmonary artery sarcoma by the much more common occurrence, and physician recognition of, pulmonary emboli.

In this issue of the *Journal*, Malik and colleagues[3] present a case of a pulmonary artery mass that was diagnosed as a pulmonary embolus and treated with oral anticoagulation for 6 months with progression of the mass before the patient was referred to their center for treatment. The initial computed tomography (CT) scan of the chest showed a large pulmonary mass that appeared to expand the right main pulmonary artery. Follow-up CT of the chest at the initial institution showed an increase in size of the mass. We unfortunately do not know from the report if this CT of chest was done at 6 months or if there were interval diagnostic imaging before this. At this point, the patient was referred to the Western University group, where a multidisciplinary evaluation suggested malignancy and operative resection recommended. The authors nicely describe and beautifully show in their video their technical approach to this resection and reconstruction. The patient did well and at 6 months had no detectable tumor. Postoperative adjuvant chemotherapy was given.

The Western University group are to be congratulated on both their approach to this difficult disease as well as their masterful surgery. Besides this surgical tour de force, there are a number of lessons for surgeons and teams interested in treating pulmonary artery sarcoma. The first lesion is to maintain a high index of suspicion for cases that are not straightforward. Did this case present any clinical reasons to suspect pulmonary embolus, such as deep vein thrombosis or reasons for hypercoagulopathy? The initial CT scan of the chest suggested an expansion of the right main pulmonary artery, which is common with sarcoma but uncommon with clot. We have created imaging approaches both to separate benign from malignant cardiac tumors[4] as well as specific imaging characteristics of pulmonary sarcoma.[5]

The next lesson is having a multidisciplinary evaluation to plan treatment. Our group uses a multidisciplinary cardiac tumor team for all cases suspected of malignancy to plan treatment approaches.[6] Our surgical approach is similar to the authors', and we have presented this in detail previously.[7] We differ in several areas. When pulmonary root resection and replacement is needed, we divide the main pulmonary artery to allow us to reach the posterior root and dissect down to right ventricular muscle much like we do when doing a Ross procedure.[8] We replace the root with allograft but use artificial graft to replacement

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the main pulmonary arteries going out to the first major branch.[7] When pneumonectomy is required, the surgeons must assess the density of adhesions to the chest wall, as the authors did, before proceeding with pneumonectomy. The need to divide extensive adhesions can lead to significant bleeding into the pneumonectomy space after a cardiac resection and pump run. This can lead to multiple transfusions and eventual unilateral pulmonary edema in the remaining lung and death. In these cases, we resection the cardiac component and disconnect the pulmonary veins. The main stem bronchus is left intact for bronchial flow. The chest is closed and a standard pneumonectomy is then done using a thoracotomy approach after 24 to 48 hours, when the coagulopathy from the cardiopulmonary bypass has abated. We refer to this as the Texas Two Step[9] and have had no deaths in 7 cases since instituting this approach. We have recently reviewed our experience with the surgical resection of 20 consecutive cases of pulmonary artery sarcoma.[10] This is a young patient group with a median age of 52.5 years. The pulmonary valve is often involved, and complete pulmonary root resection and replacement was needed in 80% of the cases. Similar to the case presented, pneumonectomy is common and needed in 35% of our cases. Unlike the current case presented, most of our cases had previous surgery elsewhere and were referred to our site when there was a recurrence. The 10-year survival from the initial diagnosis or from our surgery was 16.4% and 8.4%, respectively.

We again congratulate the authors on their approach and excellent outcome in this difficult case. Missing the diagnosis at initial presentation is a missed opportunity to institute appropriate care. Fortunately, the Western University group was able to correct this and give their patient additional sunrises to enjoy.

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