
CONVERGENCE AND A CASE OF PULMONARY ANGIOSARCOMA

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INTRODUCTION

Case reports are a time-honored way for physicians to share knowledge in cases that are especially rare, interesting, or instructive. A case report typically involves only the care of one patient and therefore often appears to represent an isolated endeavor. Most cases, however, are not isolated events but rather a convergence of multiple forces. To illustrate this, I have received permission from a patient to identify him in this case discussion.

"Convergence" is defined in *The American Heritage Dictionary of the English Language* as "the act, condition or quality of converging or approaching the same point from different directions; tend towards a meeting or intersection." This case report is a story of extraordinary convergence, of people and places, of hope and happiness.

THE INITIAL DIAGNOSIS

In October 2005, I received a call from a former resident now on staff at M.D. Anderson Cancer Center, Dr. David Rice. He had received information and studies on a patient in New Zealand dying from a pulmonary artery sarcoma that had been deemed unresectable in both New Zealand and Australia. David asked a simple question: "What can we do?"

The patient, Bryan Hodder, was at the time a 74-year-old retired farmer in New Zealand. Having spent a lifetime working hard so that others could eat, Bryan and his wife Lois were looking forward to travel and family. In June 2004, he noticed his first symptoms of wheezing when he lay on his left side in bed. He had also developed a slight but persistent cough. Bryan and Lois were to leave soon on a long-awaited eight-week overseas vacation. Before leaving, Bryan visited his family physician, who diagnosed asthma and gave him an inhaler to take with him on his trip. He noticed increasing fatigue leading up to his trip but attributed this to doing too much in preparation for leaving. By the second week of their vacation tour, despite having been a very vigorous man all his life, Bryan was having trouble keeping up. By week four in Copenhagen, his cough was much more insistent and his wheeze clearly audible. A call to the hotel

doctor resulted in a phone diagnosis of asthma and a prescription for inhalers and cough medicine. A week later in Dover, he was seen by a physician who wrote more prescriptions and asked to see him the following day. Because the tour was leaving, however, this was not possible, and Bryan soldiered through the end of the tour with increasing fatigue. The day after his return, his physician ordered a chest X-ray, and the result was unremarkable: although he had never smoked, and despite several bronchoscopies, no one could explain the shadow in his lung. In fact, he became known at the Christchurch Hospital as "the mystery man."

Consultation was arranged with Mr. Singh, the local cardiothoracic surgeon, who finally provided him with a diagnosis - a sarcoma of his left pulmonary artery. Mr. Singh told him this sarcoma was unresectable, and follow-up opinions from experts in Auckland and Australia concurred. Radiation therapy was started as palliation for the tumor, which had occluded the blood supply to his left lung and threatened the blood supply of his right lung. Told that his tumor was incurable, his family braced itself for what appeared to be the inevitable end.

Eventually, the Hodders received a fax from a niece who had searched the Internet and found some information on cardiac tumor work being

done at the M.D. Anderson Cancer Center and The Methodist DeBakey Heart & Vascular Center, both in Houston's Texas Medical Center. Bryan initially chose not to pursue this as he feared another disappointment and had already prepared himself for the worst. Then his family physician, Dr. John Cook, told him, "You have nothing to lose; there's nothing else here in New Zealand or Australia. Let's give it a try." Bryan agreed, and the call was made to Houston.

A CASE OF EXTRAORDINARY CONVERGENCE

Dr. David Rice emigrated from Ireland to pursue a career as a thoracic surgeon specializing in thoracic oncology. He planned to do a surgery residency at The Mayo Clinic. At the time, I served as program director of the Baylor College of Medicine Cardiothoracic Surgery residency and had worked with Dr. Jack Roch, chief of thoracic surgery at M.D. Anderson Cancer Center, to create a "general thoracic" spot within the Baylor cardiothoracic residency - an approach that was subsequently embraced by the thoracic surgery residency review committee and led to formalized "thoracic" and "cardiac" tracks within cardiothoracic residencies. Offering Dr. Rice a spot at Baylor with additional time at M.D. Anderson, Jack

and I lured him away from Memorial Sloan Kettering and other institutions where he interviewed. Dr. Rice worked with me at the Methodist DeBakey Heart & Vascular Center and completed his cardiothoracic residency at Baylor, then joined the thoracic surgery faculty of the M.D. Anderson Cancer Center, where he is now recognized as a national leader in thoracic oncologic surgery.

I have been fortunate to maintain contact with my former resident and current colleague and have been privileged to operate with him on several challenging cardiac tumor cases. When Dr. Rice received studies on a New Zealand patient and a phone call asking if he could help, David seized the opportunity and made another call - this one across the street.

As a native Houstonian, I was fortunate to be accepted into Baylor College of Medicine for my medical school education. There the convergence of two extraordinary opportunities - working with great surgical talent gathered by Dr. Michael E. DeBakey and working at The Methodist Hospital - influenced me to pursue a career in surgery. I was accepted into Baylor's general surgery residency, where I worked with teachers including Dr. DeBakey, Dr. Stanley Crawford, Dr. Jimmy Howell, and Dr. George Noon, who inspired me to seek cardiovascular surgery training. Because of my strong interest in congenital heart surgery, I was accepted into a position at The Texas Heart Institute under Dr. Denton Cooley. In 1984, I was fortunate to operate with Dr. Cooley on a young man from Italy who had what was thought in Italy to be an unresectable left atrial cardiac tumor. To everyone's surprise, Dr. Cooley excised the entire heart, excised the tumor, and reimplanted the heart. We lost this patient despite this dramatic and innovative approach to an otherwise unsolvable problem, but I was infected with a lifelong interest in cardiac tumors and their surgical treatment. We have with time established one of the largest experiences with malignant primary

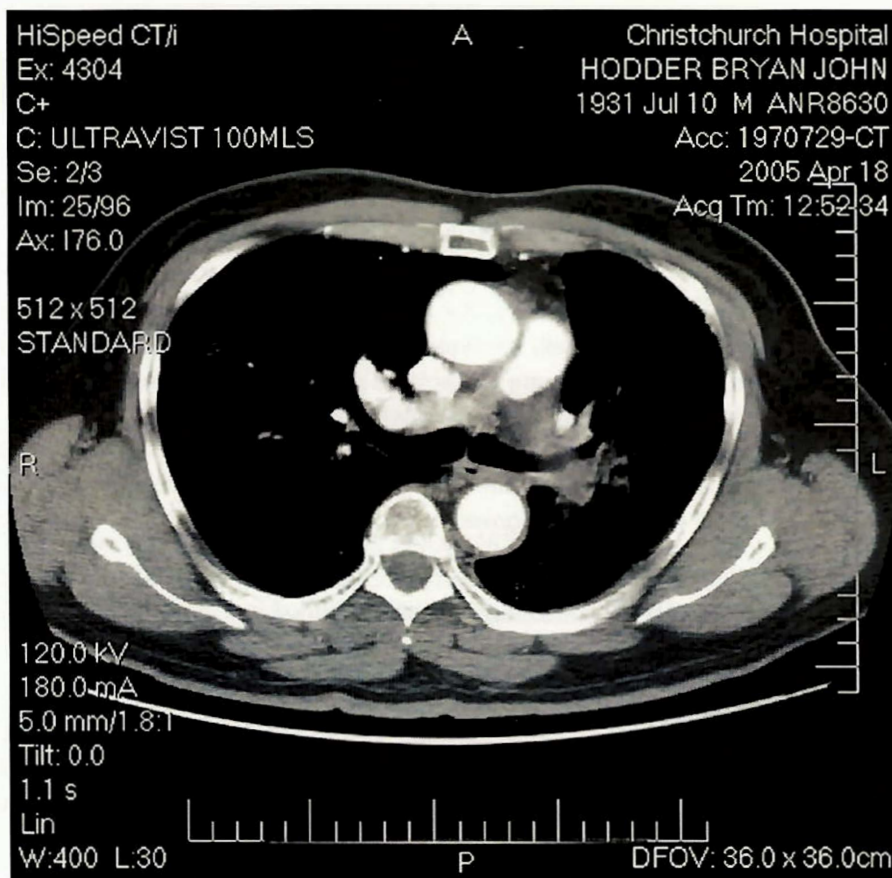


Figure 1. CT scan of an intraluminal mass within the pulmonary artery.

cardiac tumors in the world within the Methodist DeBakey Heart & Vascular Center and often share this experience with our colleagues at M.D. Anderson Cancer Center and elsewhere. One day I received a call from my former resident, Dr. Rice, regarding a patient in New Zealand. Dr. Rice posed the question, "Can we do anything about this?" I thought we could.

With all the appropriate people contacted and in agreement that something could be done, the discussion turned to "How?" Our normal approach is to bring patients into our hospital for care, but New Zealand has a national health service that covers medical expenses, and they calculated that it would likely cost more to send the patient to America than to bring our team to New Zealand. Dr. Rice and I were asked if we would be willing to come to New Zealand. I had never been to New Zealand and so got out my maps. Arrangements were made to

go halfway around the world - not just to a different time zone but to a different day due to crossing the international date line - to care for a man we had yet to meet. Thus, an unusual convergence of lives in Christchurch, New Zealand leads to our case report.

CLINICAL SUMMARY

The patient is a 74-year-old male non-smoker who was in previous good health when he developed a cough and wheezing. An initial diagnosis of asthma was established and bronchodilator therapy initiated. Despite treatment, the patient developed worsening cough and wheezing with increasing fatigue. A chest X-ray showed a left hilar mass, and a follow-up chest CT scan revealed an intraluminal mass within the pulmonary artery that completely obstructed the left pulmonary artery and compromised the right pulmonary artery (Figure 1). A presumptive diagnosis of pulmonary artery sarcoma was made

that was initially deemed unresectable, and five courses of radiotherapy were given. The patient remained symptomatic despite radiotherapy, and further consultation was requested from The Methodist DeBakey Heart & Vascular Center and the M.D. Anderson Cancer Center. Surgical resection was recommended, and the patient had a preoperative coronary arteriogram that revealed triple vessel coronary artery obstructive disease.

The patient underwent a median sternotomy incision that allowed complete mobilization of the left lung. The inferior pulmonary ligament and both pulmonary veins were divided using gentle retraction on the heart for exposure. The left main bronchus was exposed at its origin from the trachea between the ascending aorta and the superior vena cava allowing staple closure and division. The left main pulmonary artery was fully mobilized, and the patient was given heparin and placed on cardiopulmonary bypass. While on complete bypass but without cardiac arrest, the main pulmonary artery was opened to explore the extent of involvement there. The main pulmonary artery could be divided just distal to the pulmonary valve and just distal to the take off of the right main pulmonary artery to achieve complete resection. This allowed completion of the left pneumonectomy with resection of the main pulmonary artery and partial resection of the right pulmonary artery. The pulmonary artery was reconstructed with pulmonary allograft, with the proximal anastomosis just distal to the pulmonary valve, end-to-end anastomosis between the allograft right pulmonary artery and the patient's right pulmonary artery, and oversewing of the allograft left pulmonary artery. Subsequent three-vessel coronary artery bypass was completed with left internal mammary artery to the left anterior descending coronary artery and reversed autologous saphenous vein graft to the right and obtuse marginal coronary arteries without cardiac arrest.

The patient was extubated several hours after surgery and had an uneventful post-operative course. Final pathology revealed pulmonary artery angiosarcoma with margins free of tumor. At 18 months of follow-up, the patient is doing well and free of known disease.

DISCUSSION

Cardiac tumors occur both as primary cardiac tumors arising from the heart and secondary cardiac tumors that have spread to the heart. Primary cardiac tumors are quite rare, occurring with an autopsy incidence of about 0.003%.¹ Of the primary cardiac tumors, about 75% are benign and 25% are malignant, and 75% of the malignant tumors are sarcomas.² Primary pulmonary artery sarcomas are a subset of primary cardiac tumors and are even more uncommon. The first case was reported in 1923 from an autopsy by Mandelstamm,³ and since then less than 200 patients with primary PA sarcoma have been described.⁴ For surgical case series, there are less than four patients in the largest collection in date. These tumors are subclassified as rhabdomyosarcoma, osteogenic sarcoma, angiosarcoma, or unclassified leiomyosarcomas.

Patients often present with dyspnea, cough, hemoptysis, or chest pain with lung involvement being present in most cases.⁵ The usual duration of symptoms prior to diagnosis is from three to 12 months. Findings upon clinical evaluation include right ventricular dysfunction, pulmonary hypertension, and pulmonary insufficiency. Because these symptoms sometimes mimic pulmonary embolus, these patients are often initially treated with anticoagulation therapy and later found to have a refractory intraluminal mass.

The results of chemotherapy management alone are suboptimal,⁶ and there is no established treatment regimen. Survival for this disease is often reported in weeks instead of months or years. Radiation therapy can be used for pulmonary artery sarcoma because of its ability to avoid large doses to the

cardiac muscle.⁷

To achieve complete resection, the entire right ventricular outflow tract or main PA trunk may require excision. Because of the extensive surgery required to achieve a complete resection, the lack of clinical information indicating a benefit, and the lack of preoperative diagnoses, most institutions have not performed such radical procedures. We have performed eight radical resections for primary pulmonary artery sarcoma, often with homograft replacement and always with cardiopulmonary bypass, with a current mean survival of 39 months - double the current survival in the literature (in press).

Surgical resection combined with neoadjuvant chemotherapy remains the mainstay approach for pulmonary artery sarcomas and we believe gives the best chance for survival. With resection alone, the mean survival ranges from 2 to 12 months in the current literature.⁶⁻⁸ The results of adjuvant chemotherapy have been inconsistent with positive^{8,10} and negative¹¹ reports. When a preoperative diagnosis can be made, we and others believe that there is a clinical benefit to neoadjuvant chemotherapy.⁹ Additionally, chemotherapy regimens for angiosarcoma have improved in the current era. Due to the rarity of this type and location of tumor, there are no prospective trials to evaluate these treatment regimens.

FOLLOW UP

Dr. Rice and I performed the surgery with Mr. Hodder's local cardiothoracic surgeon, Mr. Harsh Singh. The family later sent me a newspaper clipping discussing this case as a "first" for New Zealand (Figure 2). In this interesting convergence of lives, Dr. Rice and I were fortunate to care for a wonderful man and meet his loving family, make many new friends in the Christchurch medical community, visit the South Island wine country, and "go cramping" (climbing) in their Southern Alps - an experience I will carry with me to the end of my professional career.

and beyond with fond remembrance. I recently received a letter from Mr. Hodder's daughter, Josie, which stated, "My daughter, Jazmynn, and myself have had an extra 18 months - so far, that is - of treasured times with our dad and granddad still in our lives." Mr. Hodder is currently doing well (Figure 3), and I have received a recent letter from Bryan and Lois saying, "We are traveling to Brisbane, Australia this week to see our grandson and great grandson for two weeks."

The convergence allowing the care of Mr. Hodder is rooted deep within The Methodist DeBakey Heart & Vascular Center. In the broader sense, it represents the culmination of the life work of many people - beginning with Dr. DeBakey and including our hospital staff, nurses, administrators and physicians, all of whom paved the way for our work in cardiac malignancy that eventually led us to Mr. Hodder. I hope you all revel in his recovery and his family's joy - the Hodders greatly appreciate your efforts.

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Christchurch Heart Cancer Operation a 'First' for New Zealand

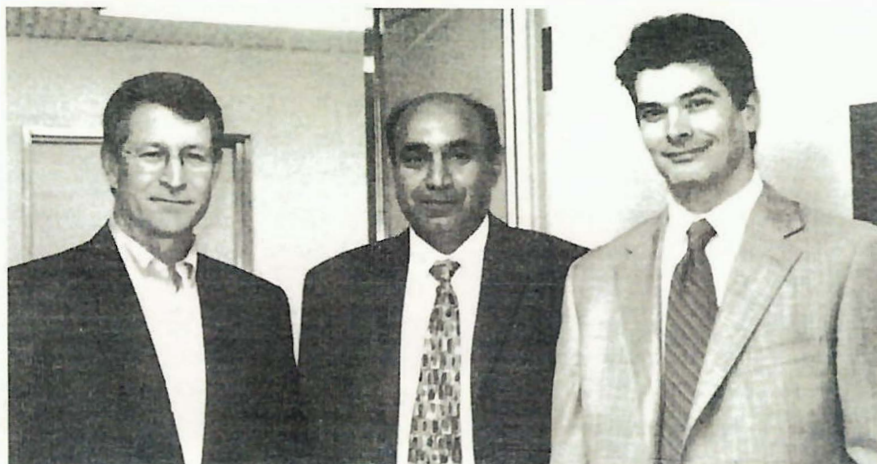


Figure 2. Mr. Hodder's heart surgery makes headlines as a New Zealand first. Pictured from left to right are Dr. Michael Reardon, Mr. Harsh P. Singh and Dr. David Rice.

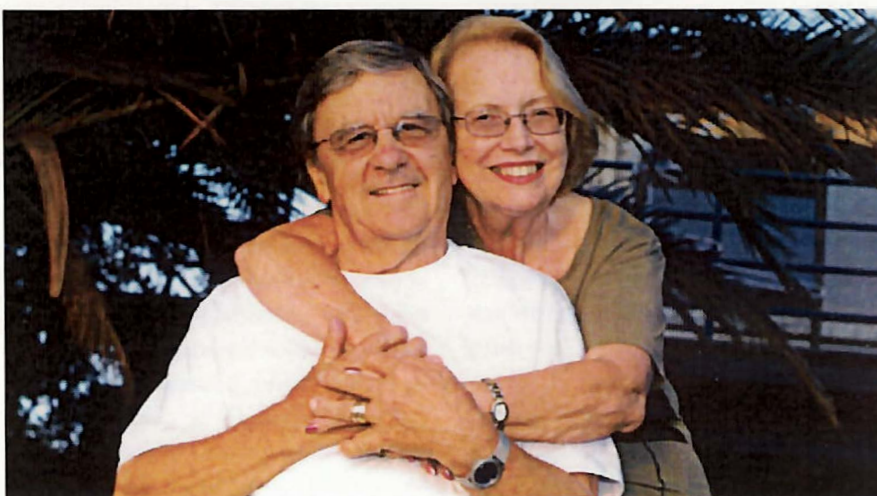


Figure 3. Bryan and Lois Holder on their Golden Wedding Anniversary, two years post surgery.