Cardiomyopathy Induced by Pulmonary Sequestration in a 50-Year-Old Man

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A 50-year-old black man presented at the emergency department with midsternal, non-radiating chest pressure and chronic dyspnea on exertion. Four years before the current admission, he had been diagnosed with nonischemic cardiomyopathy at another facility. After our complete evaluation, we suspected that his symptoms arose from left-to-left shunting in association with pulmonary sequestration, a congenital malformation. Our preliminary diagnosis of secondary dilated cardiomyopathy was confirmed by normalization of the patient's ventricular size and function after lobectomy. To our knowledge, this patient is the oldest on record to present with cardiomyopathy consequent to pulmonary sequestration. His case is highly unusual because of his age and the rapid resolution of his symptoms after lobectomy. We believe that pulmonary sequestration should be included in the differential diagnosis of dilated cardiomyopathy. (*Tex Heart Inst J* 2015;42(1):63-5)

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Pulmonary sequestration (also known as bronchopulmonary sequestration) is a lesion comprising 0.15% to 6.4% of all congenital pulmonary malformations. When the lesion is located within a normal lobe and lacks its own viscera, the anomaly is called intralobar sequestration. This particular malformation has no connection with the bronchial system. Most often, intralobar sequestration is present in the left lower lobe, has a systemic arterial supply that typically arises from the descending thoracic aorta,¹ and drains through the pulmonary venous system.² This congenital anomaly most often presents with pulmonary symptoms in infancy or childhood and is rarely found in the adult population or with isolated cardiac manifestations.¹ We submit here a highly unusual (perhaps unique) late presentation of this formation in an adult who presented with nonischemic cardiomyopathy.

Case Report

In December of 2012, a 50-year-old black man with nonischemic cardiomyopathy presented at the emergency department for investigation of midsternal, nonradiating chest pressure. This atypical angina, rated 7/10 on a pain scale, was constant, was not relieved by nitroglycerin, and was transiently relieved by morphine. During the taking of his initial history, the patient stated that he “walks everywhere” at a slow pace but had chronic dyspnea upon any additional exertion.

Four years before the current admission, the patient had received a diagnosis of nonischemic cardiomyopathy from another facility. During that earlier admission, the patient had displayed signs of heart failure (HF) and a reduced left ventricular (LV) ejection fraction (documented by echocardiography). His coronary arteries had appeared to be normal upon cardiac catheterization. A β -blocker and an angiotensin-converting enzyme (ACE) inhibitor, started at that time, had been continued to the time of the present admission, with no recurrent HF admissions.

An electrocardiogram (ECG) administered in the emergency room showed sinus tachycardia with LV hypertrophy and a left-axis deviation. An initial chest radiograph was interpreted by radiology as showing a small focal infiltrate in the airspace at the base of the left lung (Fig. 1). Because of the patient's ongoing chest pain, negative troponin assay, and ECG results that were not indicative of ischemia, our emergency department requested a computed tomographic angiogram, a cardiology consultation, and a transthoracic echocardiogram. Vital signs at the time of the cardiology con-

sultation included a blood pressure of 124/89 mmHg and a pulse rate of 89 beats/min. The patient's height was 5 ft 11 in, and his weight was 280 lb. The results of his physical examination were noted to be normal for an obese man who had no noted cardiac, pulmonary, or lower-extremity abnormalities (there was, for example, an absence of murmur over his anterior lung or chest). When the cardiology consultant examined the patient's computed tomographic angiogram, however, he found an anomalous arterial blood supply to the left lower lobe, consistent with intralobar pulmonary sequestration (Fig. 2). Moreover, the transthoracic echocardiogram showed a dilated LV with an end-diastolic dimension of 59 mm and an ejection fraction estimated at 0.35 to 0.40. There was grade 2 diastolic dysfunction, with mild right ventricular enlargement and an estimated pulmonary arterial pressure of 30 mmHg.

Fig. 1 Chest radiograph from day of presentation suggests left lower-lobe infiltrate.

Fig. 2 Computed tomographic angiogram (3-dimensional volume rendering) reveals an anomalous origin of the pulmonary arterial supply (arrow) from the descending aorta.

Cardiology, internal medicine, pulmonology, and thoracic surgery physicians all reviewed the case. Given the patient's ongoing chest pain, we arrived at a differential diagnosis; infection, pulmonary embolus in the sequestered lung, and cardiac ischemia were under consideration for cause. When review of the images showed that turbulent blood flow in the anomalous vessel was the cause of incomplete opacification, thrombus was ruled out, along with infection. Enoxaparin was discontinued, as well as the antibiotic agents. After a vasodilator nuclear stress test yielded results that were negative for ischemia, the patient experienced an unexplained resolution of his atypical chest pain. We then turned our attention to the differential diagnosis for a source of the dilated cardiomyopathy.

A multidisciplinary discussion led us to the possibility that a left-to-left shunt could account for our patient's dilated cardiomyopathy. When we presented our findings to him, he requested continued investigation into the relationship of these findings to his chronic condition. An invasive preoperative evaluation with cardiac catheterization revealed normal coronary arteries and normal right-sided pressures. The interventional cardiologist injected the anomalous vessel with contrast solution, and fluoroscopy showed that arterial flow from the aorta entered the left lower-lung field and drained through the pulmonary veins into the left atrium. There was no evidence of left-to-right shunting; however, left-to-left shunting was clearly present. In consultation with thoracic surgery, we discussed options for removing the aberrant vessel and the sequestered pulmonary segment.

Intraoperatively, the patient was found to have a 2-cm anomalous vessel arising from an enlarged 3-cm descending thoracic aorta (Fig. 3). The left inferior pul-

Fig. 3 Computed tomographic angiogram (maximum intensity projection) displays the origin of the pulmonary arterial supply (arrow), its size (compared with that of the enlarged descending aorta), and incomplete opacification of the anomalous vessel.

monary vein was gigantic, at approximately 8 cm in diameter. A left lower lobectomy enabled removal of the anomalous vessel.

Postoperative echocardiography, performed 7 days after the initial imaging, showed a modest improvement in LV end-diastolic dimension but notable improvement in LV ejection fraction, which was estimated at 0.55 to 0.60. Clinically, the patient was ambulatory soon after surgery and already had begun to note improvements in his symptoms. Within days after the thoracotomy, he reported absence of the dyspnea that he had experienced earlier at any pace above a slow walk.

After discharge, this patient was lost to follow-up. Our electronic record shows that he has had no further contact with our health system.

Discussion

Intralobar pulmonary sequestration with cardiac involvement is typically discovered in infancy, and rarely in early adulthood.² The usual findings that lead to a diagnosis of pulmonary sequestration are in fact pulmonary: some combination of cough, infection, dyspnea, or hemoptysis.³ Cardiovascular symptoms, when caused by pulmonary sequestration in the absence of associated intracardiac abnormalities, are rare. The most often reported is HF, which usually appears in the neonatal period or in childhood. In adults, the documented appearance of cardiovascular complications ascribed to sequestration has been limited to a few case reports, which are discussed below.

Cardiac complications are caused by the high cardiac output that results from volume overload to the LV and left-to-left shunting. The magnitude of the shunt can play a role in the timeline of symptoms. Solit and colleagues⁴ described the case of a 23-year-old patient with intralobar pulmonary sequestration in whom the preoperative hemodynamic study showed a high cardiac output with no sign of heart failure; the diagnosis of sequestration was made after routine chest radiography. Fabre and colleagues⁵ described the case of a 25-year-old patient who had an undiagnosed pulmonary sequestrum associated with aortic stenosis, cardiomegaly, and HF. The patient's condition improved after removal of the sequestrum.

Our patient did have a previous diagnosis of cardiomyopathy, but otherwise had no structural cardiac abnormality. It can be hypothesized in his case that the shunt was relatively small earlier in life and had increased in size and severity over time. This could explain why the sequestrum was overlooked well into adulthood—until the patient's presentation with HF.

After our patient's initial admission for HF at another hospital, he was treated with ACE inhibitors and β -blockers, which appeared to stabilize his symptoms and did indeed prevent recurrent hospital admissions for

heart failure. Complete evaluation at our facility led us to suspect that the symptoms were due to left-to-left shunting through the sequestrum. Our preliminary diagnosis of secondary dilated cardiomyopathy was confirmed by normalization of the patient's ventricular size and function after lobectomy.

To our knowledge, this patient is the oldest on record to make an initial presentation with cardiomyopathy consequent to pulmonary sequestration. The case is also highly unusual because of the rapid resolution of symptoms after lobectomy. We believe that pulmonary sequestration should be included in the differential diagnosis of dilated cardiomyopathy.

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