Case Reports

Concurrent Aortic Valvular Disease and Pulmonary Sequestration:

Clinical Implications

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Pulmonary sequestration refers to segmental lung tissue that has no connection with the bronchial tree or pulmonary arteries. In adults, the clinical sequelae are usually related to infection. Patients are typically referred for sequestrectomy even when they are asymptomatic. There are no guidelines for treating patients who have pulmonary sequestration and coexisting cardiac valvular disease, in which case the venous drainage patterns of sequestra pose the additional risks of infective endocarditis and volume overload.

We present the cases of 2 adult patients—one symptomatic and one asymptomatic—who had concurrent aortic valvular disease and pulmonary sequestration, and we discuss the factors involved in our evaluation of their cardiac risk and our treatment decisions. In view of the sparse data to predict cardiac risks, we think that pulmonary sequestrectomy in adult patients with concurrent valvular conditions should be considered on a case-by-case basis. ***(Tex Heart Inst J 2014;41(6):649-52)***

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Pulmonary sequestration is a nonfunctioning mass of normal lung tissue that lacks normal communication with the tracheobronchial tree. The arterial supply of the anomalous lung segment is usually systemic and arises from a lower thoracic or abdominal aortic branch. However, the venous drainage can be to the right or left cardiac chambers. This poses risks of infective endocarditis (IE) and volume overload, which might be exacerbated by existing cardiac valvular disease.

Indications for pulmonary sequestrectomy include malignancy, recurrent infections, the risk that the normal pulmonary parenchyma will be destroyed, and an unclear diagnosis of a posterior mediastinal mass. In general, authors have considered surgical removal to be mandatory upon the actual or suspected diagnosis of any pulmonary sequestrum.[1,2] However, others have claimed that conservative management is an option in asymptomatic adult patients.[3]

We found only 2 published case reports of native-valve IE associated with pulmonary sequestration.[4,5] Given the rarity of these coexisting conditions, few data are available to guide management. We describe the cases of 2 adult patients with aortic valve disease and pulmonary sequestration, and we discuss some principles that might help to guide management and surgical decisions.

Case Reports

Patient 1

In December 2007, a 52-year-old man presented at our institution with chest pain, exertional dyspnea, and orthopnea. An echocardiogram revealed critical aortic stenosis, with an aortic valve area of 0.7 cm^2 and a mean aortic valve gradient of 45 mmHg. The patient's medical history included appropriately treated latent tuberculosis and a remote history of pneumonia. Preoperative chest radiographs suggested subsegmental atelectasis of the right lower lobe. The patient underwent aortic valve replacement through a 3rd-interspace right minithoracotomy, with use of a 23-mm bioprosthetic Biocor™ Stented Valve System (St. Jude Medical, Inc.; St. Paul, Minn). In September 2010, the 55-year-old patient presented with recurrent pneumonia. Computed tomograms of the chest showed a 5 × 7-cm mass in the right lower lobe (Fig. 1). An aberrant arterial supply that arose from the infradiaphragmatic abdominal aorta was consistent with the diagnosis of an intralobar pulmonary sequestrum (Fig. 2).

There was no clinical or echocardiographic evidence of endocarditis. However, the long-term risk of bioprosthetic IE was a concern, so we performed a right lower lobectomy. The patient's postoperative course was uneventful, and he had experienced no IE or further episodes of pneumonia as of March 2014.

Patient 2

In June 2012, a 48-year-old man underwent routine echocardiographic examination of his known bicuspid aortic valve (BAV), from which he was asymptomatic. His medical history included the human immunodeficiency virus (HIV), treated for many years with antiretroviral medication; and remote episodes of cytomegalovirus retinitis, *Pneumocystis carinii* pneumonia, and *Mycobacterium avium* complex pneumonia. He reported no use of tobacco, alcohol, or intravenous drugs. His CD4 count was 867 cells/μL.

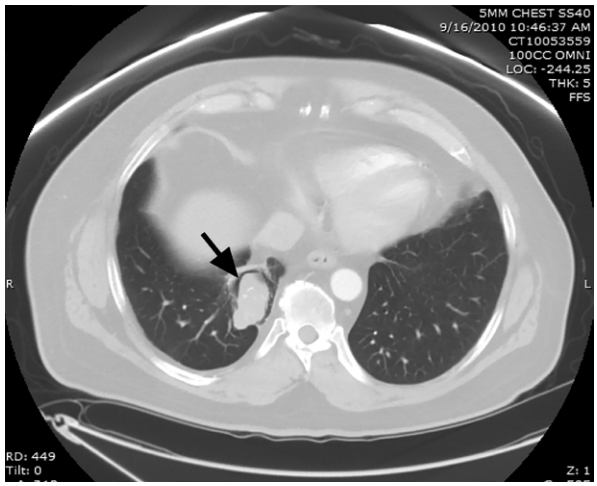

Fig. 1 *Patient 1. Computed tomogram shows an intralobar pulmonary sequestrum in the right lower lobe (arrow).*

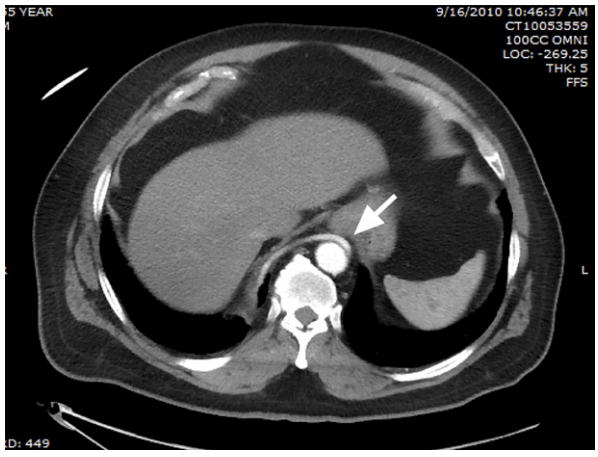

Fig. 2 *Patient 1. Computed tomographic angiogram shows an arterial branch (arrow) that arises from the infradiaphragmatic aorta and supplies the intralobar sequestral segment.*

A transthoracic echocardiogram showed a mildly calcified BAV without significant aortic stenosis, a mean aortic valve gradient of 9 mmHg, an aortic valve area (measured by means of the continuity equation) of 2 cm^2, an aortic root diameter of 3.9 cm, and an ascending aortic diameter of 3.5 cm. Computed tomograms showed an ill-defined, 3 × 1.3-cm area of ground-glass opacity and focal air-trapping in the posteromedial left lower lobe (Fig. 3). The area received arterial blood supply from the descending thoracic aorta (Fig. 4). The venous drainage was into both ipsilateral pulmonary and hemiazygos veins, consistent with an extralobar pulmonary sequestrum (Fig. 5). Because the patient had no symptoms related to valvular disease or pulmonary sequestration, we recommended conservative management consisting of serial clinical and echocardiographic

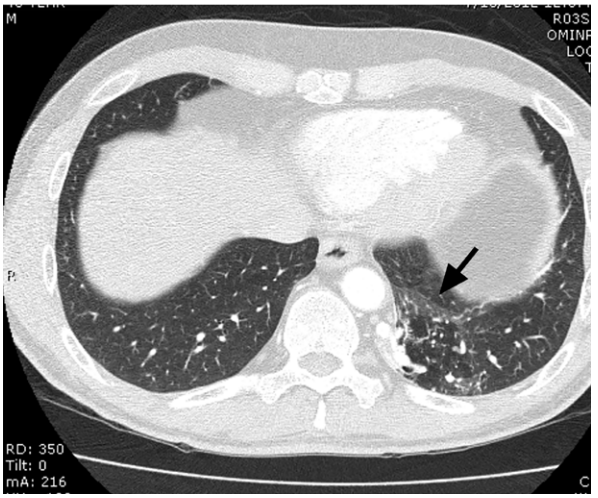

Fig. 3 *Patient 2. Computed tomogram shows an extralobar pulmonary sequestrum in the left lower lobe (arrow).*

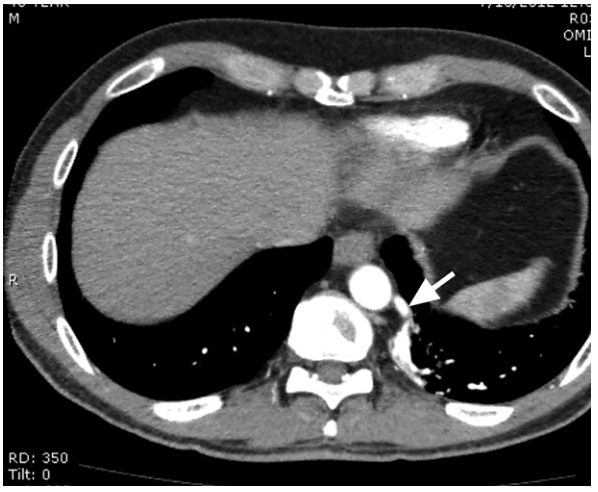

Fig. 4 *Patient 2. Computed tomographic angiogram shows that the arterial supply (arrow) of the extralobar pulmonary sequestrum arises from the descending thoracic aorta.*

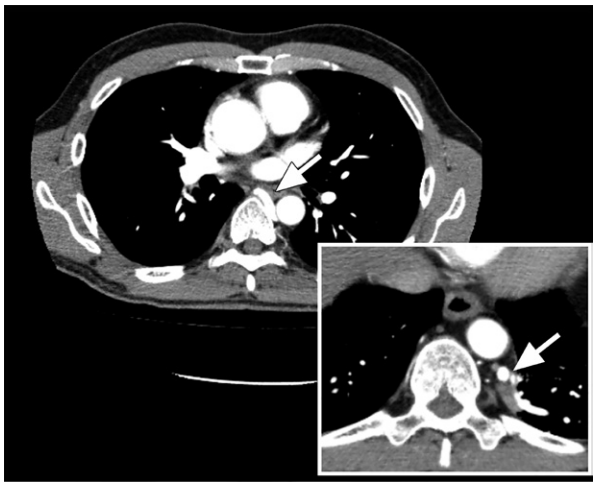

Fig. 5 *Patient 2. Computed tomogram shows drainage of the extralobar pulmonary sequestrum through a hemiazygos vein (arrow). The inset shows pulmonary venous drainage (arrow) at the level of the sequestrum.*

evaluations. As of July 2014, no IE had been detected, and the patient had experienced no further episodes of pneumonia.

Discussion

Pulmonary sequestrectomy is typically successful in adult and pediatric patients. Some authors recommend that all pulmonary sequestra be treated operatively.[1,2] Conversely, Dimitriou and colleagues[3] monitored 5 asymptomatic or mildly symptomatic adult patients for a median period of 7.8 years and observed unchanged lesion size in 4 of 5 patients, no major infection, and resolution of mild infection by means of oral antibiotic therapy. The authors concluded that a conservative approach is a legitimate option in such patients.

The cardiac implications of pulmonary sequestration are related to the venous drainage patterns of the anomalous lung segment. Sequestra can drain to the right atrium through the systemic veins (hemiazygos or azygos) or to the left atrium through the pulmonary veins. Thus, infected sequestra can provide a direct hematogenous connection to either side of the heart (Fig. 6) and a consequent risk of IE.

The true risk of infection from pulmonary sequestration is difficult to discern, because sequestra can remain undetected until recurrent pulmonary infection occurs. In case series of pulmonary sequestration, infection has been found to be the presenting symptom in 27% to 70% of adults.[6,7] In a review involving 10 patients older than 50 years of age, 3 (30%) were asymptomatic, perhaps indicating a greater likelihood that older patients undergo chest imaging for unrelated reasons.[8] The reported infectious organisms have included *Staphylococcus*, *Streptococcus*, *Enterococcus*,[9] *Pseudomonas*, *Mycobacterium* species, and *Aspergillus*.

Pulmonary sequestration can also lead to congestive heart failure consequent to increased venous return to the heart. Whereas the anomalous drainage is usually from the sequestrum itself, the entire ipsilateral lung can be involved.[10] In one case,[11] drainage from the sequestrum to the left atrium led to left-to-left shunting. This resulted in an initial misdiagnosis of residual aortic stenosis after aortic valvuloplasty. Subsequent lobectomy resulted in the regression of cardiac chamber size and the resolution of heart failure in that patient. In another case,[12] a large-caliber arterial vessel fed a pulmonary sequestrum and functioned as an arteriovenous malformation, with resultant left ventricular enlargement and mitral regurgitation. After lobectomy alone, the patient's cardiac dilation and mitral regurgitation resolved. In short, the hemodynamic consequences of shunting might exacerbate or mask underlying cardiac valvular disease, or lead to an erroneous diagnosis of a primary cardiac valvular disease. These diagnostic uncertainties might be particularly confounding in cases of extralobar sequestration, which is often associated with congenital heart disease.

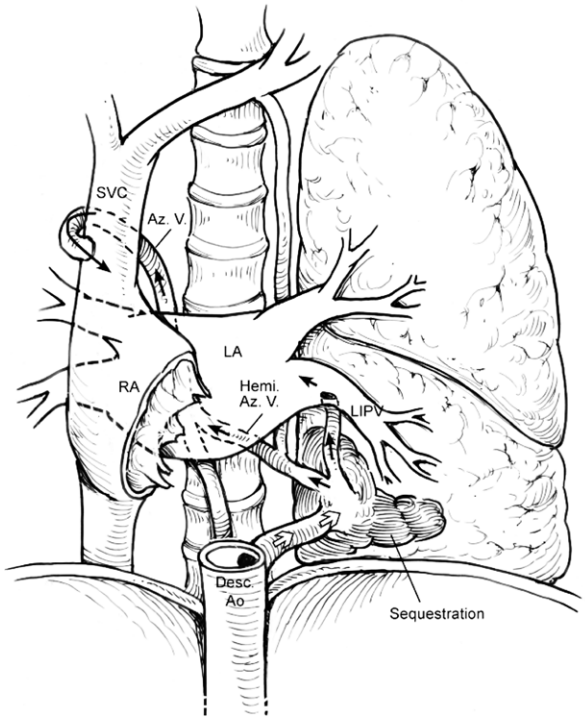

Fig. 6 *Drawing shows an example of extralobar pulmonary sequestration with anomalous drainage to both ipsilateral pulmonary veins and the azygos system.*

Az. V. = azygos vein; Desc Ao = descending aorta; Hemi Az. V. = hemiazygos vein; LA = left atrium; LIPV = left inferior pulmonary vein; RA = right atrium; SVC = superior vena cava

Another factor in concurrent valvular disease and pulmonary sequestration is the contribution of native valvular pathologic conditions to the risk of infection. It is generally accepted that abnormal valves are prone to endocarditis; however, the relative risk of a bicuspid configuration is not known. Investigators in a multicenter observational study of IE patients[9] found that, in comparison with trileaflet valves, bicuspid configuration was an independent predictor of aortic root abscess but not of in-hospital or 5-year mortality rates. The authors also noted that, although BAV is common in cases of aortic valve IE, the overall risk of IE in BAV is low.

Finally, an additional consideration is an immunocompromised state, as in our 2nd patient. His current CD4 count was above the threshold for acquired immunodeficiency syndrome (AIDS), but his medical history included AIDS-defining illnesses. In the absence of intravenous drug abuse, IE in HIV patients is rare, and HIV-1 alone is not considered to be a significant risk factor for IE. Nonintravenous-drug-abusing HIV-1 patients with IE appear to have mortality rates similar to those of non-HIV-1 patients with IE, independent of medical therapy or surgical treatment.[13] To our knowledge, no association of infected pulmonary sequestration and HIV has been reported.

Conclusion

Concurrent cardiac valvular disease and pulmonary sequestration present diagnostic and management challenges. Patients should optimally undergo lung resection if they have pulmonary sequestration with malignancy or recurrent infections, or if a posterior mediastinal mass has an uncertain nature. However, investigators have challenged the need for routine resection in asymptomatic adult patients with isolated pulmonary sequestration. Associated cardiac valvular disease raises specific concerns of infectious risk and hemodynamic changes. In view of the sparse data to predict cardiac risks, we think that pulmonary sequestrectomy in adult patients with concurrent cardiac valvular conditions should be considered on a case-by-case basis.

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