Extrapleural pneumonectomy for scimitar syndrome

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Scimitar syndrome is a rare congenital anomaly characterized by partial or total anomalous pulmonary venous drainage of the right lung into the inferior vena cava. The description “scimitar” comes from the classic appearance resembling the Turkish sword of a crescent vascular density at the lower right heart border on the chest radiograph. Scimitar syndrome may appear as early as infancy but has been discovered as late as the eighth decade of life. Treatment options are close observation, repair with an intra-atrial baffle, direct reimplantation of the anomalous drainage to the left atrium, or pneumonectomy. This case presentation concerns a woman who had repair of scimitar syndrome at age 3 and who at age 34 years required right extrapleural pneumonectomy after multiple pulmonary infections. She may be the oldest patient documented in whom pneumonectomy was performed for scimitar syndrome and the only one in whom extrapleural pneumonectomy was carried out.

Clinical Summary

Our patient initially was brought for repair of scimitar syndrome at 3 years of age. The anomalous common right pulmonary vein drained into the inferior vena cava immediately above the diaphragm with no other anomalous blood. An intra-atrial diversion of the anomalous venous drainage to the left atrium was performed along with an atrial septectomy. For 10 years she did well, but a quantitative perfusion scan performed at this point revealed no perfusion to the right lung. She was presumed to have venous occlusion.

When the patient was first seen in our facility, she was having increasingly frequent respiratory infections with at least a dozen right-sided pneumonias necessitating hospitalization. A quantitative perfusion scan confirmed no perfusion to the right lung. A chest radiograph showed a mildly hypoplastic lung. A magnetic resonance angiogram demonstrated an atretic pulmonary artery with drainage to the left atrium via small collateral veins (Figure 1). Echocardiogram showed no evidence of a septal defect. Cardiac catheterization confirmed normal pulmonary artery pressures. Pulmonary function tests demonstrated a forced expiratory volume in 1 second of 1.52 L (49% of predicted normal); lung volumes showed moderate restriction and air trapping with a total lung capacity of 67% of predicted normal with air trapping.

Right pneumonectomy was recommended and accepted. A right anterolateral thoracotomy was performed through the fifth intercostal space. Complete pleural symphysis was immediately evident. An extrapleural dissection was carried out, working laterally to medially. The lung was dissected off the diaphragm in an extrapleural plane and the pericardium was preserved medially. The pulmonary artery was patent, approximately 1 cm in size, and was divided with a vascular stapler. Only small collateral vessels were identified and ligated in the region of the pulmonary vein. The airway was divided and covered with a pericardial fat pad. The patient did well and was discharged home on the fifth postoperative day. At early follow-up she is doing well with no recurrent respiratory symptoms.

Discussion

Scimitar syndrome typically appears early in life with symptoms of heart failure or respiratory distress. Symptoms at this stage are generally severe and must be addressed immediately. Repair focuses on returning the pulmonary drainage to the left side of the heart, although the repair may fail either early or over time. Pneumonectomy is carried out in the setting of lung agenesis or failed repair. Presentation is variable in adulthood.
and ranges from an incidental finding to severe symptoms—typically, recurrent respiratory symptoms in the affected lung or dyspnea owing to pulmonary hypertension from long-standing shunting. Indications for repair are (1) pulmonary hypertension owing to left-to-right shunt with a shunt fraction greater than 50% as determined by cardiac catheterization and (2) recurrent pulmonary infections.

This case demonstrates the technical difficulty of pneumonectomy after failed repair of scimitar syndrome in the setting of recurrent infection. It points out that surgical resection of a lung should not be delayed when a propensity to become infected is evident. Inflammation can lead to dense pleural adhesions. Although an extrapleural approach may be radical, it potentially reduces the risk of postpneumonectomy space infection in patients with loculated fluid collections or ongoing parenchymal infection. Additionally, it may reduce postoperative bleeding from collateral blood supply. Preoperative imaging with magnetic resonance angiography of the vascular anatomy is critical to avoid intraoperative surprises. Pneumonectomy is unlikely to adversely affect pulmonary function and may actually improve it.

References

Successful perioperative management of a middle mediastinal paraganglioma

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Middle mediastinal paragangliomas are very rare, slow-growing tumors, but almost all of them are very hypervascular tumors. Complete surgical resection is difficult to achieve because of their proximity to the heart, great vessels, and trachea. We report successful complete resection incorporating preoperative embolization and a clamshell bilateral thoracotomy.

Clinical Summary
A 52-year-old woman was found to have an abnormal shadow on a chest radiograph for a medical checkup. Chest computed tomography at another hospital revealed a large well-enhanced mass with a cystic lesion located between the superior vena cava, aortic arch, right pulmonary artery, left atrium, and trachea (Figures 1 and 2). A video-assisted thoracoscopic biopsy was performed at another hospital to make the diagnosis, but massive bleeding occurred during the procedure, and the biopsy was abandoned. The patient was then referred to our department. We strongly suspected a paraganglioma of the middle mediastinum.

The serum noradrenaline level was slightly increased, and an iodine 123–meta-iodobenzylguanidine scan showed uptake in the tumor. An angiographic study showed many feeding arteries, and the main feeders were 3 thick bronchial arteries. Preoperative embolization with Gelfoam (Pfizer, Ann Arbor, Mich) and several microcoils was performed the day before the operation. At operation, we were ready to perform cardiopulmonary bypass; however, complete resection of the tumor without cardiopulmonary bypass was performed via the clamshell approach. Finally, intraoperative blood loss was reduced to 1070 mL. Histologic diagnosis was reported as a typical paraganglioma, 7 cm in size. There was no sign of recurrence on a computed tomography scan 1 year after surgery.

Comment
Aorticopulmonary paragangliomas are rare neoplasms; 79 anterior and middle mediastinal paragangliomas, which represent a surgical challenge, were reviewed. Because of their location close to the great vessels and trachea, complete resection is very difficult. Paragangliomas are locally invasive and have a high local recur-