Endovascular occlusion of right to left arteriovenous shunt associated with persistent left superior vena cava

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Left-sided superior vena cava (SVC) as the result of persistence of the left superior cardinal vein in postnatal life is a rare congenital anomaly, is usually associated with other cardiac defects, and can cause symptoms of right to left shunt. We report the case of a 58-year-old Asian man with a history of end-stage renal disease and Ebstein anomaly that was corrected surgically who presented with progressively worsening disabling dyspnea. An echocardiogram with concomitant intravenous saline injection raised the suspicion of right to left shunt, a finding that was confirmed with contrast injection of the left SVC that rapidly filled the left heart chambers and subsequently the aortic arch. To treat this anomaly, we accessed the left basilic vein under ultrasound guidance and inserted a 14F sheath into the left subclavian vein. A covered stent was then prepared at the back table with three Prolene 4-0 sutures that were wrapped around the middle portion of the graft to achieve a controlled area of stenosis after deployment. The stent graft was placed along the proximal innominate vein and the contiguous part of the left SVC. Coil embolization was then performed with coils that were positioned at the stenotic area of the covered stent. An immediate venogram demonstrated residual flow into the left SVC; however, a delayed venogram 2 weeks after the procedure showed occlusion of the left SVC and the development of collaterals to the right innominate vein that was draining to a normal right SVC. The patient remained marginally hypotensive after surgery, but he soon noted a substantial improvement in his symptoms. A repeat echocardiogram with intravenous saline injection confirmed the correction of the right to left shunt. Endovascular repair of persistent left SVC is feasible and safe and can be performed with minimal morbidity. (J Vasc Surg 2006;44:875-8.)

The persistence of the left superior cardinal vein in postnatal life is a rare anomaly associated with the development of a persistent left superior vena cava (SVC). In this syndrome, right SVC may or may not be present. Other congenital heart anomalies may also coexist. A persistent left SVC may be the cause of venous blood shunting directly into the left atrium and causing dyspnea and cyanosis. Several techniques for open repair have been described in the literature. In this article, we report the successful correction of this anomaly by using the endovascular approach.

CASE REPORT

A 58-year-old man of Asian origin who had recently migrated to the United States was seen by our service originally for placement of hemodialysis access. He reported at that time a long history of dyspnea on minimal exertion. Subsequent workup demonstrated that he had Ebstein anomaly, which manifested as severe tricuspid valve regurgitation and an associated atrium secundum septal defect. He underwent placement of a prosthetic valve and repair of the atrial defect. During surgery, a left-sided SVC was noted, a finding that was subsequently confirmed by a venogram performed in the context of dialysis access site revision. His postoperative course was complicated by the presence of pericardial tamponade that was successfully treated with the creation of a pericardial window.

Over the course of the next several months, he developed left upper extremity swelling secondary to stenosis along the left subclavian vein that responded temporarily to balloon dilation (Fig 1, *A*). Symptomatically, dyspnea continued to be his major complaint. He remained oxygen dependent, with severe activity and lifestyle limitations. He was therefore re-evaluated with transesophageal echocardiography and concomitant use of intravenous saline injection, which acts as a sonographic contrast agent and improves the quality and sensitivity of the study. Mild residual tricuspid valve regurgitation was again noted, whereas the calculated ejection fraction was 50% to 55%. It is interesting to note that saline flowed from the venous system directly to the left heart, thus raising the question of a severe right to left shunt. The computed tomographic angiography that followed demonstrated filling of the left SVC and the left heart before any opacification of the right heart chambers. This prompted a more detailed venogram via a left brachial approach under ultrasound guidance. A catheter was advanced to the proximal left-sided SVC, and contrast injection demonstrated filling of the left heart chambers and rapid filling of the aortic arch, thus confirming the suspected SVC drainage to the left atrium and a right to left shunt (Fig 1, *B*).

Treatment options were at that point discussed with the patient, who was interested in the least invasive approach. Repeat sternotomy was considered hazardous by the cardiac surgeons because of a dilated thin-walled right ventricle. Ligating the axillary vein was an option; however, collaterals from chest wall would

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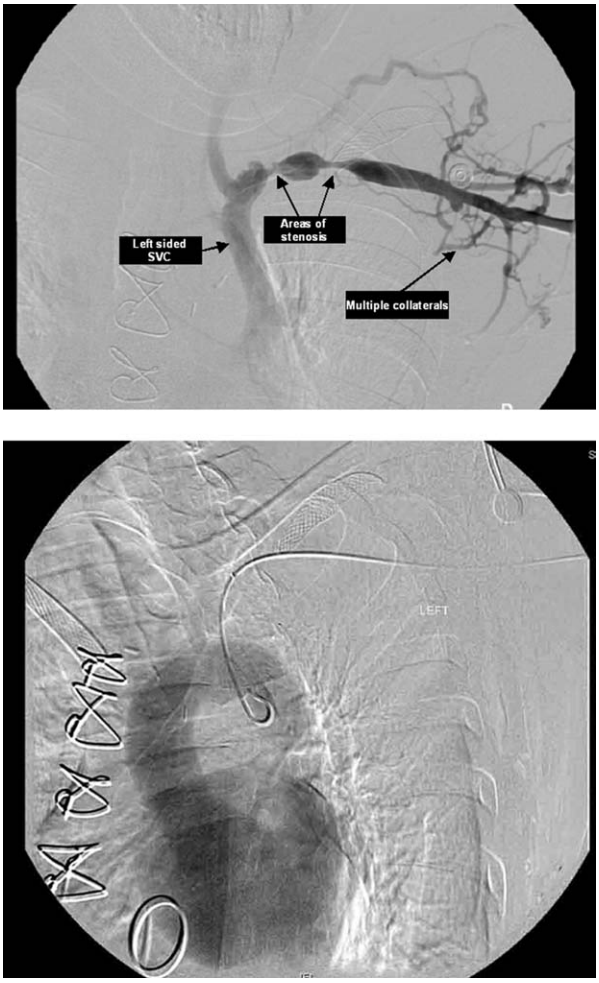

Fig 1. A, Venogram demonstrating areas of stenosis of the left subclavian vein, multiple collaterals, an occluded stent placed in the proximal left cephalic vein, and the left-sided superior vena cava *(SVC)*. **B,** Contrast injection via a catheter placed in the proximal left-sided SVC was followed by rapid filling of the left heart chambers, followed by visualization of the aortic arch, thus confirming the presence of right to left arteriovenous shunt.

continue to fill the left-sided SVC, thus maintaining the right-to-left shunt. An off-label use of the currently available endograft devices seemed to offer a reasonable alternative means of occluding the flow through the persistent SVC.

The patient was taken to the operating room, where a cavogram was performed percutaneously via a left basilic vein approach. An Amplatz wire was then inserted and directed through the left heart to the right inferior pulmonary vein. The patient was systemically heparinized at that point with heparin 100 U/kg. The left-sided SVC measured 14 mm in diameter; therefore, we planned to use a 16-mm device to occlude the SVC before its confluence with the left inferior pulmonary vein. An initial attempt was made to place an Occluder device (Cook Inc, Bloomington, Ind) in the left-sided SVC superior to the left inferior pulmonary vein. Because of the large profile of this device (18F system),

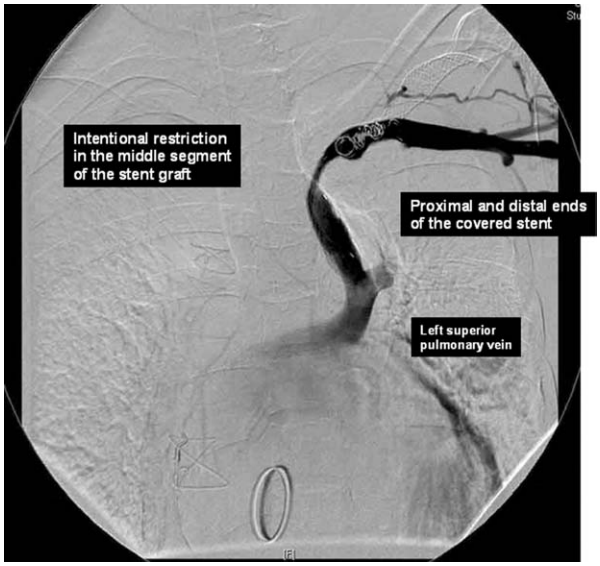

Fig 2. After coil embolization, the flow of contrast through the stent graft slowed down substantially. The left superior pulmonary vein was again shown to join the superior vena cava proximal to the landing zone of the stent graft.

advancement of the delivery sheath was not possible. As a result, this approach was aborted. Over the wire, a 14F sheath was inserted and positioned at the mid left subclavian vein. At the back table, we modified a 16 mm × 9.5-cm Excluder (W.L. Gore, Sunnyvale, Calif) stent graft by suturing three 4-0 polypropylene sutures—placed 5 mm apart—around its middle portion to create a controlled area of stenosis in the graft, approximately 6 mm in diameter. We then positioned the stent graft with its distal end above the left superior pulmonary vein, whereas the middle stenotic region created by the polypropylene suture portion was placed just distal to the confluence of the left subclavian and internal jugular veins. The graft was deployed in the usual fashion and the desired configuration; a constriction at its mid portion was confirmed with fluoroscopy and performance of a new venogram.

A number of coils were then packed at the area of stenosis, initially 9 and then 7 and 3 mm in diameter. A completion venogram confirmed good placement of the coils and some residual flow in the graft (Fig 2). The procedure was terminated at that point. The introducer sheath was removed, and a cutdown was performed on the basilic vein to repair the venotomy. It is interesting to note that the patient developed persistent hypotension after surgery that required the administration of vasopressors at a very low rate for 5 days. This could have been at least in part due to increased blood flow through the right heart chambers in a patient with a dilated right ventricle and recent tricuspid valve repair. Alternatively, the temporary hypotensive episode could have been explained by a temporary decrease in the preload after the right-sided SVC was occluded and while chest wall collaterals were developing. After that point, the patient progressively improved and was able to ambulate without any supplemental oxygen over the next 5 days. His Pao_2 was now 72 mm Hg on room air, with a corresponding alveolar-arterial gradient of 36 mm Hg. This repre-

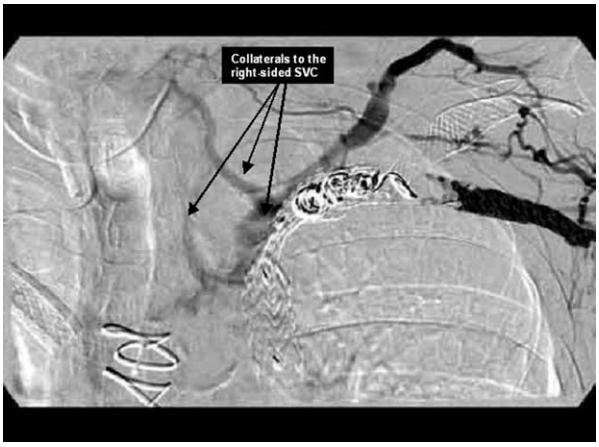

Fig 3. Two weeks after the initial coil embolization, a new venogram confirms occlusion of the left-sided superior vena cava *(SVC)* and collateral blood flow to the right-sided SVC.

sented a marked improvement over a preoperative Pao_2 of 52 mm Hg and alveolar-arterial gradient of 190 mm Hg. There was no evidence of venous congestion of his left upper extremity at any point after the procedure. A repeat cavogram 2 weeks after the coil embolization demonstrated occlusion of the stent graft, with the development of large collaterals that were draining blood from the left upper extremity to the right-sided SVC (Fig 3). His left basilic vein, which had been used as an access vessel for the endograft placement, was patent. A follow-up transthoracic echocardiogram with concomitant intravenous saline injection confirmed cessation of the right to left shunt.

DISCUSSION

A persistent left SVC is a rare anomaly found in 0.3% to 3% of the general population and up to 10% of patients with heart disease.[1] A persistent left SVC that drains into the left atrium is an even more rare defect,[2] usually associated with cardiac malformations such as common atrium and endocardial cushion defects.[3,4] Symptoms vary and include cyanosis that can become severe, clubbing, polycythemia, and, more rarely, cerebral emboli that may cause cerebral abscesses.[5] Therefore, intervention is necessary to eliminate symptoms and to prevent associated complications. Ebstein anomaly is an uncommon congenital cardiac malformation of the tricuspid valve and right ventricle and is characterized by a downward displacement of the septal and often the posterior tricuspid valve leaflets into the right ventricle. Other congenital anomalies, such as atrial septal defect, ventricular septal defect, pulmonary outflow obstruction, coarctation of the aorta, and patent ductus arteriosus, may coexist.[6,7] To date, to our knowledge, SVC malformations have been described in conjunction with this anomaly.

Traditionally, the treatment of a persistent left SVC has been open surgical repair. Several options are available. Division of the SVC and oversewing of the atrial side is an option if there is a patent innominate vein, which is not always the case. Transposition of the SVC to the right atrium is also possible if the anomalous SVC is of adequate length.[8,9] Intra-atrial partitioning to direct the blood from the SVC to the right atrium by unroofing the coronary sinus,[10] as well as an extracardiac method of transposition of the left SVC to the left pulmonary artery,[11] has been described.

To our knowledge, there has been only one previous report of a similar treatment for symptomatic right to left shunt resulting from persistent left SVC, in which Troost et al[12] treated a patient with left-sided SVC, right to left shunt, and frontal cerebral abscess by using an Amplatzer (AGA, Golden Valley, Minn) occluder placed in the SVC. The minimally invasive nature of this technique makes it extremely appealing for high-risk patients or for those who have had a previous median sternotomy, as was the case with our patient.

Several technical aspects are worth mentioning. A commercially available stent graft that has been routinely used for endovascular repair of abdominal aortic aneurysms in our practice was used. Because endovascular ligation is not yet possible, we took the innovative approach to deploy a stent graft to cover the branches draining into the left SVC. The graft was bow-shaped with the precise placement of sutures that predictably narrowed its middle segment. The purpose of this maneuver was twofold. First, it allowed for secure placement of coils that were confined at the narrowest segment of the graft to achieve cessation of flow without risking an accidental distal embolization in the arterial circulation with potentially disastrous consequences. In addition, it allowed for preservation of oxygenated blood flow from the left pulmonary vein that was emptying into the distal left SVC just above the left atrium. Similar results can theoretically be achieved with devices functioning as vascular plugs, such as the Cook Occluder or the Amplatzer occluder. Whereas the former requires a large access vessel because of the 18F profile, the Amplatzer is a rather attractive option because it comes in a substantially smaller platform in which an 8F sheath can be used to deliver a 16-mm plug. Despite a theoretical concern, acute upper extremity edema was not seen in our patient, likely because he already had central venous stenoses and well-developed collaterals. In the venogram obtained 2 weeks after the stent graft placement and the coil embolization, prominent collaterals were seen to drain the left upper extremity, thus confirming this hypothesis. Furthermore, the increased preload to the right ventricle, which could potentially pose a stress on the tricuspid valve, did not seem to be an issue on the postoperative echocardiogram.

In summary, we successfully used an endovascular approach to treat a patient with symptomatic left persistent SVC and right to left shunt. Advances in technology and improvements in the stent graft design are likely to make this approach more feasible in the future.

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